The Sri Lanka Journal of Neurology (SLJN) is a forum for debate, education and entertainment for health professionals interested in Clinical Neurology, Neurosurgery and Neurosciences. The Journal is aimed at practicing Neurologists, Neurosurgeons and Neuroscientists with commitments in Sri Lanka and has relevance to all those working in the health sector. The Journal’s prime responsibility is to the members of the Association of Sri Lankan Neurologists (ASN) and its objective is to promote good clinical practice and influence policy making across the medical world through publication of original research and peer reviewed articles on current issues and to foster responsible and balanced debate on issues that affect medicine and health care in Sri Lanka. Contributions to the SLJN reflect its national and multidisciplinary readership and include current thinking across a range of medical specialties and the Journal assists the ASN in its continuing medical education programme.

While members of the ASN receive the SLJN as one of the benefits of membership, it will be available to other health professionals on paying a subscription fee. The Journal has full editorial independence.
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The formation of the Association of Sri Lankan Neurologists in 2007 was a landmark event in the professional and academic calendar of medical professionals in the country. Over the last seven years it has made a remarkable progress and today stands very strong in comparison to other neurology associations world over. Though still young and with a small membership, it has made friends with other neurological associations, is affiliated to the World Federation of Neurology and plays a significant role in the activities of the WFN. Members regularly attend and present their work in these international meetings. World Stroke Congress, International League against Epilepsy and the Movement Disorders Society are other major organizations where our members play an important role. It is expected that the contributions we make to these organizations will improve further with the young and energetic increasing membership of the ASN. The ASN journal is published annually and this is the third issue. We need to increase the number of issues per year and I am confident that we can achieve this with the help of the young members and also attracting papers from the region.

There are neurologists now working in the north and east of the country, parts of Sri Lanka affected by the now defunct war. The epidemiology of neurological disorders may be different in those regions and they are likely to see hitherto unseen problems and unusual excesses of some disorders and may face unexpected difficulties in managing their patients. The ASN had been regularly conducting monthly meetings, the ASN Forum where clinical cases are presented and an update lecture is given. These are at present held in Colombo but we have been able to transmit these sessions live via skype to centers in Jaffna and Galle and are very well received by the post graduates in these faraway places. The ASN council meets bimonthly in the evenings on a Sunday for its council meeting and this ends up with another CME meeting based on the Continuum series of the AAN which we receive courtesy WFN.

Our annual academic meetings have gained a high reputation for its contents, faculty and fellowship. They are attended by delegates from the region and the faculty include world authorities and have included higher officials of the WFN and the ABN. We also have cohosted the Asia Pacific Stroke Conference in 2011 with great success and this year we cohost the annual congress of the Asian Society against Dementia with the participation of many overseas delegates and a large faculty from Australia, India, UK, Taiwan, Singapore, Thailand, Japan, USA, Hong Kong, Philippines and Indonesia. The association has its own office in Colombo with a full time administrator and is open on all week days. And this year we are proud to announce that we now have our own academic gown and ASN tie designed. At the Annual Academic meeting in November the council will be wearing their new gowns and ties and the ASN is thankful to the current president Dr. Arjuna Fernando and the committee who took the initiative to accomplish this exciting task. So a look at the past makes us proud of our achievements and we look forward to a future of the ASN with more international recognition and representation.

Saman B Gunatilake
Editor in Chief
The burden of epilepsy: data from an epilepsy clinic

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Sri Lanka Journal of Neurology, 2014, 3, 2-4

Abstract

Introduction: Over half of the 50 million people with epilepsy worldwide are estimated to live in Asia, but information about the recognition of the burden created by the disease is scarce. Studies in the region and in Sri Lanka focusing on the burden of epilepsy are limited.

Design/Setting: A retrospective hospital-based study was conducted to determine educational, marital, and employment opportunities of patients with epilepsy attending the epilepsy clinic at the National Hospital of Sri Lanka for one year. An interviewer based questionnaire was used to collect data.

Results: Data was gathered from 500 patients. Male to female ratio was 1.08:0.92. Two hundred and twelve patients (42.4%) belonged to the 16 to 30 age group while 140 (28%) were from the 31 to 45 age group. Most of the patients in the aforesaid age groups (n=352) were unemployed (n=211, 59.94%) and only 89 (25.3%) were employed. Two hundred and sixty four patients (75%) were unmarried while 7 (1.9%) were divorced. The number of patients who received primary and secondary education were 44 (24.4%) and 93 (51.6%) respectively. Two (1.1%) patients went to university. Forty one (22.8%) had no schooling.

Interpretation: Many epilepsy patients have fewer educational, marital, and employment opportunities which contribute towards poor interpersonal skills, social withdrawal and low self-esteem. This emphasises that epilepsy is a chronic disabling neurological disorder which causes many psychosocial tribulations that need to be addressed by healthcare professionals.

Index words: Burden of epilepsy in Sri Lanka. Psychosocial consequences of epilepsy. Educational, marital, and employment opportunities of patients with epilepsy

Introduction

Epilepsy is a neurological condition that knows no geographic or social boundaries, occurring in men and women and affecting people of all ages. It has been estimated that at least 50 million people worldwide have epilepsy¹,². More than 80% of people with epilepsy live in developing countries, where effective treatment of this condition is scarce². Over half of the 50 million people with epilepsy worldwide are estimated to live in Asia which is a heterogeneous and resource-constrained continent³. Although much research is done in Asia, information about the recognition of the burden created by epilepsy is scarce. There has been a long standing concern that, although progress continues to be made in relation to medical management of epilepsy, including the development of a number of new antiepileptic drugs, attention to the social adjustment of individuals with the condition is still limited⁴. People with epilepsy in developing regions carry a heavy burden of stigma with associated poor social and economic status⁵. Studies in the region and in Sri Lanka focusing on the burden of epilepsy are limited.

The present study was conducted at the epilepsy clinic, National Hospital of Sri Lanka to assess the educational, marital, and employment opportunities of patients with epilepsy.

Design/Setting

A retrospective hospital-based study was conducted to determine educational, marital, and employment opportunities of patients with epilepsy attending the epilepsy clinic at the National Hospital of Sri Lanka. An interviewer based questionnaire was used to collect data on educational, marital, and employment opportunities. Data was collected from patients attending the above clinic from January 2002 to January 2003. Statistical analysis was done through SPSS version¹⁰. Informed consent was obtained from the study population.

Results

Data was gathered from 500 patients who attended the clinic during the stipulated study period. Of these patients 260 (52%) were males. Two hundred and twelve
patients (42.4%) belonged to the 16 to 30 age group while the second highest number, 140 (28%) were between 31 and 45 years. Forty patients (8%) were over 45 years of age while 54 (10.8%) patients were between the ages of 11 and 15. Twenty two (4.4%) were from the 6 to 10 age group and 32 (6.4%) were 5 years and below. Most of the patients between 16 and 45 (n=352) were unemployed (n=211, 59.94%) and only a minority were employed (n=89, 25.3%). Fifty two (14.7%) were certified unemployable. Two hundred and sixty four patients (75%) in the aforesaid age group were unmarried while 7 (1.9%) were divorced. The remaining 81 were married (23.01%).

While the majority of patients who were above 30 years (n=180) had received secondary education (n=93, 51.6%) a significant proportion of patients (n=44, 24.4%) received only primary education. Only 2 (1.1%) patients went to university. Forty one (22.8%) had no schooling.

Discussion

People with disabilities are among the most vulnerable in any society. The psychosocial and economic consequences of epilepsy in less developed countries are generally acknowledged to contribute substantially to the burden of this disease as it is associated with social stigma. Stigmatization leads to discrimination, and people with epilepsy have been the target of prejudicial behaviour in many spheres of life, over many centuries and in many cultures. Stigma has a severe impact on individuals and their families, as well as on the effectiveness of public health programmes. Despite enormous cultural diversity across the world, the areas of life affected are remarkably similar. They include marriage, interpersonal relationships, employment, education, mobility, leisure activities and attendance at social and religious functions. Discrimination against people with epilepsy in the workplace and with respect to access to education is not uncommon. In a study done in Hong Kong 94.1% of respondents thought that people with epilepsy could be married, but only 67.8% would allow their child to marry a person with epilepsy. Thus people with epilepsy may have fewer educational, marital, and employment opportunities. The findings of this study may lend support to the supposition that people with epilepsy have to face. As mentioned in other studies social withdrawal and isolation may contribute towards these problems. Fewer educational, marital, and employment opportunities in turn worsens social withdrawal, thus a vicious cycles ensues.

Conclusion

WHO defines health as “a state of complete physical, mental, and social well-being and not merely the absence of disease”. Many patients in the above population have fewer educational, marital, and employment opportunities which contribute towards poor interpersonal skills, social withdrawal and low self-esteem which negatively affect their capacity for health. This emphasizes that epilepsy is a chronic disabling neurological disorder which causes many psychosocial tribulations that need to be addressed by healthcare professionals.

Strategies to improve the well-being of people with epilepsy need to be multi-modal in nature, addressing stigma-mediated disadvantage through education and advocacy of influential groups, promoting empowerment among people with epilepsy, expanding opportunities for education and employment.

While this study represents institutional data and may have an element of referral bias it highlights psychosocial tribulations in epilepsy patients and the need for intervention. However further community based epidemiological studies are necessary to assess the burden of epilepsy in the community.

References

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Non motor symptoms in patients with Parkinson disease: experience from a tertiary care center in Sri Lanka

S B Gunatilake¹, N D Perera¹, K M Bandara¹, A U Pathirana¹, A G S Asiri², M T M Riffsy²


Abstract
Non motor symptoms (NMS) of Parkinson’s disease (PD) lead to morbidity, impaired quality of life and frequent hospitalization. NMS are under-recognized and under-reported. NMSQuest is a validated screening tool adopted to identify these symptoms in patients with PD. We conducted a descriptive cross sectional study during March 2012 – September 2012 in a tertiary care hospital in Sri Lanka to describe the non-motor symptoms of PD and to determine the percentage of NMS that remain undeclared to the healthcare professionals. 103 patients, age 64.55 (SD 9.62, range 43-86) years, 58 (56.3%) males and 45 (43.7%) females completed the study. Mean duration of illness was 3.16 yrs. 98.1% of patients had at least one NMS. The mean total NMSQ-T was 9.08 +/- 5.087(SD). Memory loss was the commonest NMS. NMSQ-T increased with the severity of PD (p<0.001) but no association was found between NMSQ-T and age, sex or duration of illness. Loss of weight, difficulty in swallowing and falls were significantly commoner among males than females. (p <0.05) A significant proportion of NMS (Mean 76.1 +/- 17.7, range 36.7% to 100%) were undeclared to the health care personnel. Incomplete bowel evacuation, sweating, diplopia and anxiety were the least declared symptoms. In conclusion, NMS are present in the majority of patients with PD increasing with the severity of disease. A significant proportion of NMS are undeclared to the healthcare personnel highlighting the importance of using screening tools to identify these symptoms during health visits.

Index words: Parkinson’s disease, non motor symptoms

Introduction
Parkinson’s disease (PD) is the second commonest neurodegenerative disorder affecting people from all cultures and races around the world. Although the motor symptoms of Parkinson’s disease are well defined, the non-motor features of this disorder are under-recognized and consequently, under treated. However, James Parkinson, who first described the disease in 1817, described the non-motor symptoms, sleep disturbance, constipation, dysarthria, dysphonia, dysphagia, sialorrhoea, urinary incontinence and ‘constant sleepiness with slight delirium’ in his essay titled “The Shaking Palsy”¹.

There has been growing interest in research on non-motor symptoms (NMS) of PD and recent studies reveal that these symptoms could manifest prior to the onset of motor symptoms (pre motor PD) inevitably merging with disease progression²,³. As disease advances, non-motor symptoms dominate the clinical picture causing additional morbidity, impaired quality of life and shortened life expectancy. National Institute for Clinical Excellence in England has found under-reporting and under-recognition of non-motor symptoms an important unmet need in PD⁴. Additionally, non-motor symptoms are a frequent cause of hospitalization and institutionalization, which increases the cost of care of patients with PD⁴.

Some of these symptoms are undeclared to the health care providers, further delaying recognition. A recent international survey has shown that up to 62% of symptoms of Parkinson’s disease remain undeclared to the health care professionals because they are unaware that the symptoms are linked to Parkinson’s disease or are embarrassed to talk about them¹⁰. Early identification would result in better outcome for the patients by symptom directed therapy.

Because of the range of non-motor symptoms, questionnaires and tools have been developed to help identify NMS in PD patients. NMSQuest is a self-administered 30 item questionnaire, validated as a screening tool identifying nine NMS domains. (Appendix 1) Chaudhri and colleagues described the presence of approximately 10-12 NMS in PD patients with the use of NMSQuest⁶.

There is paucity of studies on non-motor symptoms of PD from developing countries and no published data in Sri Lanka. We conducted a cross sectional study in a tertiary care institution in Sri Lanka to identify the presence of NMS in patients diagnosed with PD and to identify the proportion of patients with undeclared symptoms.
Objective

To describe the non-motor symptoms of Parkinson’s disease and to determine the percentage of non-motor symptoms that remains undeclared to the healthcare professionals.

Methods

This was a descriptive cross sectional study conducted at Colombo South Teaching Hospital, a tertiary care center in the city of Colombo. Study was conducted during March 2012 – September 2012. We recruited 103 patients diagnosed as PD (according to diagnostic criteria of the United Kingdom Parkinson’s Disease Society Brain Bank) attending neurology and medical clinics. Exclusion criteria were employed to exclude patients with an alternative explanation as the cause for Parkinsonism (Table 1).

Table 1. Exclusion criteria

<table>
<thead>
<tr>
<th>Features considered as exclusion criteria in patients with parkinsonism</th>
</tr>
</thead>
<tbody>
<tr>
<td>• History of repeated strokes with stepwise progression of parkinsonism features</td>
</tr>
<tr>
<td>• History of repeated head injury</td>
</tr>
<tr>
<td>• History of definite encephalitis</td>
</tr>
<tr>
<td>• Neuroleptic treatment at onset of symptoms</td>
</tr>
<tr>
<td>• &gt;1 affected relatives</td>
</tr>
<tr>
<td>• Sustained remission</td>
</tr>
<tr>
<td>• Strictly unilateral features after 3 years</td>
</tr>
<tr>
<td>• Supranuclear gaze palsy</td>
</tr>
<tr>
<td>• Cerebellar signs</td>
</tr>
<tr>
<td>• Early severe autonomic involvement</td>
</tr>
<tr>
<td>• Early severe dementia with disturbances of memory, language and praxis</td>
</tr>
<tr>
<td>• Babinski’s sign</td>
</tr>
<tr>
<td>• Presence of a cerebral tumour or communicating hydrocephalus on computed tomography scan</td>
</tr>
</tbody>
</table>

Patients included in the study were interviewed to obtain demographic data, duration of disease, presence of non-motor symptoms and to assess severity of PD. NMS were assessed using the NMSQuest and the severity of PD was assessed using the Hoehn and Yahr staging. 28 questions were used to identify non motor symptoms and the presence or absence was documented as a “Yes” or “No” response. Further clarification was sought on whether symptoms were “declared” or “undeclared”. Questions on sexual problems were not assessed due to poor response from patients and cultural reasons.

Statistical analysis

Data (NMSQ-T) was scattered in a normal distribution when tested with Shapiro-Wilk test (p 0.087). Descriptive statistics such as percentage was used to determine the frequency of each NMS. NMSQ total score (NMSQ-T) was calculated by computing the number of positive responses in NMSQuest. Frequency of undeclared NMS was computed and expressed as a percentage of patients with each non motor symptom. Association between NMSQ-T and sex, HY stage was assessed using ANOVA test. Comparison of means was also used to assess the association between NMSQ-T and HY stage categories. Correlation was used to identify the association between NMSQ-T and age, duration of illness. Test results were interpreted as significant when p<0.05 with a 95% confidence interval. Analysis was performed using SPSS 19.0 version.

Ethical approval

Informed consent was obtained from all participants. Ethical approval was obtained from the Ethics Review Committee of Faculty of Medical Sciences, University of Sri Jayewardenepura.

Results

Hundred and three patients, age 64.55 (SD 9.62, range 43-86) years, 58 (56.3%) males and 45 (43.7%) females completed the study. Mean duration of illness was 3.16 years and ranged from 1 month to 15 years. Duration of illness was further categorized into four groups. (≤59, 60-119, 120-179 and 180 or more months)

One patient had a family history of Parkinson’s disease. Co-morbidities such as diabetes mellitus, hypertension, ischemic heart disease, and stroke were seen in 57 (55.3%) patients. Resting tremor (102, 99%) and muscular rigidity (96, 93.2%) were the commonest motor symptoms, while bradykinesia and postural instability were reported in 53 (51.5%) and 41 (39.8%) patients respectively. Patient characteristics are summarized in table 2.
Non motor symptoms in patients with Parkinson disease

Presence of non-motor symptoms

Majority of patients (n= 101, 98.1%) had at least one or more non-motor symptoms. Two patients without any NMS were below the age of 50yrs (44yrs and 47yrs respectively). The mean total NMSQ-T was 9.08 (SD 5.09) ranging from 0 to 24 of a maximum of 28. Memory loss was the commonest non-motor symptom (n= 64, 62.1%) while vomiting was the least observed symptom (n=4, 3.9%). Difficulty in concentrating, daytime sleepiness, restless legs, constipation, weight loss and memory loss were seen in more than 50% of patients.

ANOVA test revealed a significant association between the HY stage and NMSQ-T (p<0.001). NMSQ-T gradually decreased in stages 4 and 5 compared to stage 3 but remained higher than mean NMSQ-T of stages 1 and 2. (Figure 1) We further categorized HY stages 1 and 2 as “Mild” and stages 3, 4, 5 as “Moderate-severe”. Comparison of mean NMSQ-T by t-test between these 2 groups revealed a significant difference (7.08 and 12.14, p<0.01) suggesting that NMSQ-T was commoner among moderate-severe disease than mild disease.

No association was found between the NMSQ-T and age (r= 0.127, p 0.149) or duration of disease (r= 0.097, p 0.328). Duration of disease was further divided into four
groups (≤59, 60-119, 120-179 and 180 or more months) and the mean NMSQ-T of each group was compared, which did not reveal a significant difference between the groups. T-test did not reveal a significant association between NMSQ-T and sex. (P 0.095) Chi-square test was used to compare the presence of individual NMS in males and females. Loss of weight, difficulty in swallowing and falls were significantly commoner among males than females with p values of 0.037, 0.03 and 0.038 respectively.

**Undeclared symptoms**

A significant proportion of non-motor symptoms were undeclared to the health care personnel. Proportion of undeclared symptoms varied with the individual symptom ranging from 36.7% to 100%. Mean was 76.1 +/- 17.7 (SD) (Figure 2). Incomplete bowel evacuation, sweating, diplopia and anxiety were the least declared symptoms (Table 4).

### Table 3. Individual NMS assessed by NMS Questionnaire

<table>
<thead>
<tr>
<th>NMS</th>
<th>Male, N (%)</th>
<th>Female, N (%)</th>
<th>Total, N (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nausea/ Vomiting</td>
<td>3 (2.9)</td>
<td>1 (1.0)</td>
<td>4 (3.9)</td>
</tr>
<tr>
<td>Incomplete bowel emptying</td>
<td>4 (3.9)</td>
<td>2 (1.9)</td>
<td>6 (5.8)</td>
</tr>
<tr>
<td>Faecal incontinence</td>
<td>6 (5.8)</td>
<td>3 (2.9)</td>
<td>9 (8.8)</td>
</tr>
<tr>
<td>Loss of taste/smell</td>
<td>6 (5.8)</td>
<td>3 (2.9)</td>
<td>9 (8.8)</td>
</tr>
<tr>
<td>Acting out during dreams</td>
<td>7 (6.8)</td>
<td>5 (4.8)</td>
<td>12 (11.6)</td>
</tr>
<tr>
<td>Leg swelling</td>
<td>6 (5.8)</td>
<td>9 (8.8)</td>
<td>15 (14.6)</td>
</tr>
<tr>
<td>Diplopia</td>
<td>11 (10.7)</td>
<td>9 (8.8)</td>
<td>20 (19.4)</td>
</tr>
<tr>
<td>Nocturia</td>
<td>13 (12.6)</td>
<td>7 (6.8)</td>
<td>20 (19.4)</td>
</tr>
<tr>
<td>Hallucinations</td>
<td>16 (15.5)</td>
<td>9 (8.8)</td>
<td>25 (24.2)</td>
</tr>
<tr>
<td>Delusions</td>
<td>15 (14.6)</td>
<td>10 (9.7)</td>
<td>25 (24.2)</td>
</tr>
<tr>
<td>Dribbling of saliva</td>
<td>20 (19.4)</td>
<td>9 (8.8)</td>
<td>29 (28.1)</td>
</tr>
<tr>
<td>Falls*</td>
<td>17 (16.5)</td>
<td>12 (11.6)</td>
<td>29 (28.1)</td>
</tr>
<tr>
<td>Insomnia</td>
<td>19 (18.4)</td>
<td>11 (10.7)</td>
<td>30 (29.1)</td>
</tr>
<tr>
<td>Fearful dreams</td>
<td>21 (20.4)</td>
<td>10 (9.7)</td>
<td>31 (30.1)</td>
</tr>
<tr>
<td>Anxiety</td>
<td>22 (21.4)</td>
<td>9 (8.8)</td>
<td>31 (30.1)</td>
</tr>
<tr>
<td>Swallowing difficulty*</td>
<td>37 (35.9)</td>
<td>13 (12.6)</td>
<td>40 (38.9)</td>
</tr>
<tr>
<td>Sweating</td>
<td>25 (24.3)</td>
<td>16 (15.5)</td>
<td>41 (39.8)</td>
</tr>
<tr>
<td>Urgency</td>
<td>24 (23.3)</td>
<td>18 (17.5)</td>
<td>42 (40.8)</td>
</tr>
<tr>
<td>Body pains</td>
<td>20 (19.4)</td>
<td>23 (22.3)</td>
<td>43 (41.8)</td>
</tr>
<tr>
<td>Postural dizziness</td>
<td>26 (25.2)</td>
<td>19 (18.4)</td>
<td>45 (43.7)</td>
</tr>
<tr>
<td>Sad/low mood</td>
<td>27 (26.2)</td>
<td>21 (20.4)</td>
<td>48 (46.6)</td>
</tr>
<tr>
<td>Loss of interest</td>
<td>32 (31.1)</td>
<td>19 (18.4)</td>
<td>51 (49.5)</td>
</tr>
<tr>
<td>Difficulty concentrating</td>
<td>32 (31.1)</td>
<td>21 (20.4)</td>
<td>53 (51.4)</td>
</tr>
<tr>
<td>Daytime sleepiness</td>
<td>35 (34.0)</td>
<td>19 (18.4)</td>
<td>54 (52.4)</td>
</tr>
<tr>
<td>Restless legs</td>
<td>35 (34.0)</td>
<td>20 (19.4)</td>
<td>55 (53.4)</td>
</tr>
<tr>
<td>Constipation</td>
<td>31 (30.1)</td>
<td>25 (24.3)</td>
<td>56 (54.3)</td>
</tr>
<tr>
<td>Weight loss*</td>
<td>36 (35.0)</td>
<td>21 (20.4)</td>
<td>57 (55.4)</td>
</tr>
<tr>
<td>Memory loss</td>
<td>37 (35.9)</td>
<td>27 (26.2)</td>
<td>64 (62.1)</td>
</tr>
</tbody>
</table>

* These symptoms were common among males than females with a p<0.05 when analyzed by chi-square test
Table 4. Percentage of undeclared non motor symptoms among the study population

<table>
<thead>
<tr>
<th>Non motor symptom</th>
<th>Total no of patients (n)</th>
<th>No with undeclared symptoms (n)</th>
<th>Percentage undeclared (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Insomnia</td>
<td>30</td>
<td>11</td>
<td>36.7</td>
</tr>
<tr>
<td>Faecal Incontinence</td>
<td>09</td>
<td>04</td>
<td>44.4</td>
</tr>
<tr>
<td>Falls</td>
<td>29</td>
<td>13</td>
<td>44.8</td>
</tr>
<tr>
<td>Body aches</td>
<td>43</td>
<td>21</td>
<td>48.8</td>
</tr>
<tr>
<td>Leg swelling</td>
<td>15</td>
<td>08</td>
<td>53.3</td>
</tr>
<tr>
<td>Restless legs</td>
<td>55</td>
<td>33</td>
<td>60.0</td>
</tr>
<tr>
<td>Postural hypotension</td>
<td>45</td>
<td>28</td>
<td>62.2</td>
</tr>
<tr>
<td>Constipation</td>
<td>56</td>
<td>37</td>
<td>66.1</td>
</tr>
<tr>
<td>Drooling saliva</td>
<td>29</td>
<td>20</td>
<td>68.9</td>
</tr>
<tr>
<td>Vomiting</td>
<td>04</td>
<td>03</td>
<td>75.0</td>
</tr>
<tr>
<td>Acting out dreams</td>
<td>12</td>
<td>09</td>
<td>75.0</td>
</tr>
<tr>
<td>Hallucinations</td>
<td>25</td>
<td>19</td>
<td>76.0</td>
</tr>
<tr>
<td>Dementia</td>
<td>64</td>
<td>51</td>
<td>79.69</td>
</tr>
<tr>
<td>Difficulty in swallowing</td>
<td>40</td>
<td>32</td>
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Discussion

Our study included 103 diagnosed PD patients in all stages of disease, representing a disease duration of 1 month to 15 years. Non motor symptoms were present in the majority of these patients with 98.1% reporting at least one NMS. Patients had an average of 9 non motor symptoms which is comparable to the published literature. Study conducted by Chaudri et al found that patients with PD had 10-12 NMS. A multicenter study on 545 patients by Pablo Martinez-Martin et al revealed a prevalence of 9-12 NMS in patients with PD.

Commonest non motor symptoms were memory loss, weight loss and constipation. Memory loss was the commonest symptom in our group (62.1%) but ranged from 25.1% - 51.2% in other studies. This wide range could be due to the different tools adopted to identify memory loss and cognitive function. However, studies using specific cognitive assessment methods have also found a similar prevalence of cognitive impairment to our study among patients with PD. Studies further reveal that patients with memory impairment frequently complain of other NMS than patients without memory problems. Therefore, memory loss should be identified in these patients for a better overall outcome. Gastrointestinal symptoms such as vomiting, incomplete bowel emptying and faecal incontinence were the least reported NMS in our study population.

We found that non motor symptoms increased significantly with the severity of PD as judged by Hoehn and Yahr stage. A positive association was seen up to stage 3. This finding was consistent with the other international studies demonstrating an increase in NMS with the increasing severity of motor symptoms of PD. Patients with severe motor symptoms (HY stage 4 and 5) had less NMS than moderate disease. It is possible that patients do not declare NMS in the presence of severe motor disability. However, patients with moderate-severe PD had significantly higher NMS scores than patients...
with mild disease. It is well known that NMS contribute to morbidity and impaired quality of life. Therefore, NMS should be identified in patients with moderate-severe disease to minimize disability. Our study also highlights that NMS occur in mild PD patients as well (HY stage 1 and 2) and screening for NMS should not be limited to severe PD.

We did not find an association between NMSQ-T and sex or age but few individual symptoms (loss of weight, falls and difficulty in swallowing) were more common among males. There was no association between NMSQ-T and disease duration in our population which was in contrast to the available data demonstrating an increase in NMS with duration of disease. Severity of disease (Mean and Median HY stage) in our study population was similar to published literature but with a lower duration of disease. Mean duration of illness was 3 yrs as compared to 5-9 yrs in other studies. It is possible that our patients had a delayed presentation to healthcare providers resulting in failure to demonstrate an association between NMS and the disease duration on final analysis.

Most important finding in our study was the significant proportion of undeclared symptoms. 76% of symptoms were undeclared to the healthcare provider on average with symptoms such as incomplete bowel emptying, sweating, diplopia, anxiety and delusions being least declared despite their presence. This highlights the importance of inquiring about these symptoms and the use of screening tools during health visits for early identification of NMS in patients with PD.

Study limitations

Main limitation was the absence of an age matched control group to compare the NMS, as some of these
Non motor symptoms are common among the elderly. NMS are also subjective and could’ve influenced the results.

Conclusion

Non motor symptoms are present in the majority of patients with PD increasing with the severity of disease. Memory impairment, weight loss and constipation are frequently observed. A significant proportion of NMS are undeclared to the healthcare personnel highlighting the importance of using screening tools to identify these symptoms during health visits.

References

Introduction

Dementia is a clinical syndrome characterised by global cognitive impairment, with a decline from previous level of functioning. It is associated with impairment in functional abilities and in many cases with behavioural and psychiatric disturbances. 1-2% of 65 year olds, 10% to 15% of 80 year olds and 40% of 90 year olds have dementia. Pharmacotherapy is often the main intervention used to improve the symptoms as well as delay the progression of dementia syndromes.

Many medications have been tried in dementia patients and they can be classified into 3 broad categories: Cholinergic neurotransmitter modifying agents; non-cholinergic neurotransmitters/neuropeptide modifying agents and other pharmacological agents. Tacrine (1993), donepezil (1996), rivastigmine (2000), galantamine (2001 and memantine (2003) are the only five agents approved by the Food and Drug Administration (FDA) for the treatment of dementia. Other agents have been evaluated in trials and prescribed off-label. The various agents have different levels of evidence for efficacy.

This review focuses on the evidence for pharmacological treatment of dementia in the domains of cognition, global function, behaviour/mood and quality of life/activities of daily living. The main focus will be of the 5 drugs approved by the FDA.

Pharmacological management of Alzheimer disease

Of the five drugs approved for treatment of dementia tacrine, donepezil, rivastigmine and galantamine are cholinergic neurotransmitter modifying agents. Tacrine has been discontinued due to the side effects.

Cholinesterase inhibitors

Numerous trials have been carried out to evaluate the efficacy of the different drugs. Cholinesterase inhibitors are used based on the cholinergic hypothesis of memory impairment where cholinergic deficits were considered to be responsible for the cognitive and behavioural changes and augmentation of central cholinergic function was considered to improve cognitive function.

Donepezil

Donepezil is a long acting reversible acetylcholinesterase inhibitor approved by the FDA based on evidence of efficacy in 2 phase three clinical trials; additional randomized clinical trials of 6 to 12 months duration and trials carried out in severely impaired and nursing home patients. Of the trials that evaluated the efficacy of donepezil compared to placebo most evaluated Alzheimer disease of mild to moderate severity. Doses of 5 or 10 mg were used and the study duration varied from 12 to 56 weeks. These trials were reviewed in a Cochrane review. A single placebo controlled trial followed patients over several years and showed modest cognitive effects over two years but no significant effects on loss of function, nursing home placement or health economics.

Rivastigmine

Rivastigmine is a pseudo-irreversible cholinesterase inhibitor selectively for acetylcholinesterase and butyrylcholinesterase. Two trials evaluated rivastigmine in doses from 1 to 4 mg or 6 to 12 mg and the duration of therapy varied from 14 to 26 weeks. These studies showed that rivastigmine was efficacious. Due to dose decreases not being permitted there was less tolerability and more side effects. A trial evaluated transdermal patches (17.4 mg and 9.5 mg patch vs 6 mg oral rivastigmine twice daily) in patients with moderately severe Alzheimer disease for 6 months and the patch was efficacious with fewer adverse reactions.

Galantamine

Galantamine is a reversible competitive acetylcholinesterase inhibitor with less butyrylcholinesterase inhibition compared to rivastigmine. A Cochrane review showed that 4 trials of 3 to 6 months duration demonstrated the efficacy of galantamine at doses of 8 to 16 mg twice daily. Galantamine showed positive effects with no additional effects at doses over 16 mg/d. The frequency of gastrointestinal adverse events was similar to cholinesterase inhibitors.

Adverse effects of cholinesterase inhibitors

Nausea, diarrhoea, vomiting, anorexia and weight loss are the most common adverse effects of the cholinesterase inhibitors.

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terase inhibitors. Muscle cramps are common with donepezil. The early cholinergic effects are dose related and temporary dose reduction and retritrating reduces re-emergence of adverse effects. Few trials compared adverse effect of cholinesterase inhibitors. Donepezil in doses of 23mg/d had more adverse effects than 10mg/d in a direct comparison and fewer adverse events were noted with transdermal rivastigmine. Long term safety of cholinesterase inhibitors have not been systematically studied and warrant consideration in future studies.

Efficacy of cholinesterase inhibitors

Most trials have been carried out for 3 to 6 months in patients with mild to moderate Alzheimer disease. No evidence exists for difference in efficacy between the 3 cholinesterase inhibitors. In a Cochrane review all drugs showed an overall positive effect over placebo on the Alzheimer Disease Assessment Scale – Cognitive Subscale (ADAS-Cog) over 6 months. The trial design and modest therapeutic effect make it difficult to identify individual treatment responders.

Based on three 6-month, randomised placebo controlled clinical trials donepezil is the only cholinesterase inhibitor efficacious for severe Alzheimer disease. However the effects were modest.

Dosage of cholinesterase inhibitors

Donepezil is started at 5mg/d and can be increased up to 10mg/d after 4 weeks. Both doses were effective but the 10mg dose was more effective when the dosing groups were directly compared. Patients with moderate to severe Alzheimer disease can be treated with the sustained release 23mg/d donepezil if they have been treated with 10mg/d for at least 3 months.

Rivastigmine is commenced at 1.5mg twice a day with meals. It can be increased to 3mg twice daily after 2 weeks. Doses can be increased up to 6mg twice per day. Higher doses have better efficacy. The initial dose of the transdermal patch is 4.6mg per day and the maintenance dose vary from 4.6mg/d, 9.5mg/d or 13.3mg/d.

Galantamine is commenced at 4mg twice a day and can be increased up to 12 mg twice a day. The extended release preparation is commenced at 8mg/d and can be increased to 24 mg/d.

Cholinesterase inhibitors for mild cognitive impairment

Cholinesterase inhibitors are not indicated for mild cognitive impairment. Randomised placebo controlled trials of cholinesterase inhibitors for mild cognitive impairment were not positive on their primary outcomes and showed more adverse events. Uncertainty on the definition of mild cognitive impairment limited the inferences obtained from the trials. Further there were broad variations in cognitive impairment and progression to Alzheimer disease.

Memantine

Memantine a moderate affinity, uncompetitive N-methyl D-aspartate (NMDA) receptor antagonist is approved for moderate to severe Alzheimer disease based on two 6 month long placebo controlled clinical trials. Cholinesterase inhibitors were not allowed in one trial and in the other trial the patients had been taking donepezil for at least 6 months and on an average for over 2 years before being randomised to memantine or placebo. Another trial did not show any significant effect of memantine in moderate to severe Alzheimer disease. Memantine is not approved for mild Alzheimer disease.

Adverse effects include headache, dizziness, confusion, somnolence and sometimes hallucinations. There are no adverse drug interactions with cholinesterase inhibitors. Memantine is commenced at 5 mg/d for 1 week and increased in weekly increments of 5mg/d until a dose of 10mg twice daily. Its effectiveness is not known beyond 6 months.

Controversies of pharmacotherapy

The treatment of dementia with cholinesterase inhibitors and memantine result in statistically significant but clinically marginal improvement in measures of cognition and global assessment of dementia. The United Kingdom’s National Institute for Health and Clinical Excellence (NICE) suggests that cholinesterase inhibitors can delay cognitive impairment for 6 months but they are not cost effective.

Efficacy versus effectiveness

Trends show a statistical significance, but there is overlap in outcomes between drug and placebo patients. There is only small or modest cognitive effects of cholinesterase inhibitors thus making an inference about their effects are difficult. It is difficult to identify individual patients who benefit from cholinesterase inhibitors or memantine because the outcome measures and mean changes on the scale scores do not identify responders.

Duration of treatment

Most trials have been for 6 months with a few lasting 12 months or more. Inferences were made that if the drug was effective over this period they will be effective for longer periods. Some observational studies and open label extension of clinical trials suggest that use of cholinesterase inhibitors over at least 1 year result in a delay in nursing home placement and that addition of
memantine could further delay the progression\textsuperscript{20,21,22,23}. These observational studies were not controlled and were subject to bias. Thus optimal period of duration of treatment is uncertain.

**Effectiveness for treatment of disruptive behaviour**

In a post hoc analysis of 14 cholinesterase inhibitor trials only 3 showed significant effect in improving behaviour\textsuperscript{24} and none of these effects were large. A meta-analysis showed significant but minimal effects on behaviour in the mildly cognitively impaired patients but no effect in the severely impaired\textsuperscript{25}. Memantine and donepezil was not efficacious in improving behaviour\textsuperscript{26,27}.

Discontinuation of cholinesterase inhibitors has been associated with worsening of cognition and confusion in some patients\textsuperscript{28}. However worsening of behaviour or confusion do not appear common in clinical practice, where only 19 to 23% of patients continue to take donepezil or rivastigmine for more than a year and about one third discontinue the drug within 2 months\textsuperscript{29}.

**Overview of treatment of other forms of dementia**

**Frontotemporal dementia (FTD)**

There is no approved treatment for FTD. Neuroleptics and antidepressants have some efficacy. Antidepressants are a safer initial choice due to better side effect profile. Selective serotonin reuptake inhibitors\textsuperscript{30,31} and trazodone\textsuperscript{32} and neuroleptics olanzapine\textsuperscript{33}, risperidone, and aripiprazole have shown to improve behaviour. Cholinesterase inhibitors should be avoided due to behavioural worsening in FTD and lack of cognitive improvement\textsuperscript{34}. Early studies showed some improvement with memantine but subsequent large studies did not show any benefit\textsuperscript{35}.

**Dementia with Lewy Bodies**

There are currently no approved treatments for dementia with Lewy Bodies. Treatment and management are symptomatic. Cognitive features may benefit from acetylcholinesterase inhibitors and memantine\textsuperscript{36,37}.

**Other dementias**

There are no FDA approved drugs for the other common types of dementia. However rivastigmine has been approved for treatment of dementia associated with idiopathic Parkinson’s disease\textsuperscript{38}.

**Conclusion**

The currently available FDA approved therapeutic agents for Alzheimer disease and other dementias are the cholinesterase inhibitors and memantine. Many other agents such as vitamins and food supplements are available in the market, however these have not proven to be efficacious. None of the pharmacological agents prevent or delay onset of minimal cognitive impairment or Alzheimer disease. The available FDA approved agents also produce statistically significant but clinically minimal effects. Thus the necessity to treat and the duration of treatment are mostly guided by the clinicians’ decision.

Development of new drugs and molecules are in the process. But as there are no validated drug targets for Alzheimer disease the development of new drugs faces challenges.

References


Disappearance of a disease is a rarity. Arguably, the only disease to have been erased from the face of the earth is smallpox. Poliomyelitis is hopefully the next in line, but in spite of a concerted effort worldwide, civil strife and political interference, are preventing the eradication of poliomyelitis in a few countries. Many neurological diseases are chronic with little hope of recovery, leave alone worldwide eradication. In this context even a reduction of incidence, in a localized region is significant and deserves a closer look.

The diseases to be described are seen mostly in Asian countries and that too in the last 50 years. Some have been documented only in Sri Lanka.

Distal spinal muscular atrophy (DSMA)

My first contact with DSMA was in 1972. The first patient that drew my attention was a 21 year male with insidious onset, non-familial, bilateral, asymmetrical, wasting of his hands and forearms. There was no lower limb, bulbar, sensory or sphincter involvement. There were no long tract signs. Hitherto, similar patients had not been seen in the country. During my 2 years training in London, Edinburgh and Glasgow, I did not see a single patient with similar manifestations. For a diagnosis, rare conditions like distal myopathy, a pure motor neuropathy and a cervical myelopathy were considered, though they were clinically only remote possibilities. In the next 3 years, 33 patients were seen with similar presentations. There was a remote resemblance to Madras Motor neuron disease. This made me consider it a motor neuron disease in the young and presented it as such for the silver jubilee celebrations of the Institute of Neurology, Madras1. As the years passed and more patients were seen with a longer follow up, its resemblance to the Japanese Hirayama disease-atrophy of the distal upper extremity became apparent, though there were a few distinct features2. Over 100 more patients were seen over the next 10 years, when our series had more patients than that seen in Japan3. Affection of the upper arm and shoulder was rare. Most patients had a spontaneous arrest in 5-7 years. New patients with similar symptoms are now rare and the reason is obscure. Over the last 10 years, the numbers have dropped significantly amounting to less than 10 annually.

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No definite cause for this entity was found and absorption of a pesticide through the skin of the hands was postulated, and the spontaneous arrest may have been due to discontinuation of agricultural activity though in some an exposure to pesticides were not present. This was during an era when mechanized agriculture and routine use of gloves were not common.

Retinopathy in megaloblastic anaemia

In the same location, the University Unit of Peradeniya (where I was registrar), around the same time as my encounter with the first patient with DSMA, there were many megaloblastic anaemia patients from the tea plantations developing a retinopathy. The retinal haemorrhages and exudates disappeared in 7-10 days of treatment – with vitamin B12 initially and with folic acid, if there was no response. The anaemia was accompanied by a depressed leucopoiesis with few megakaryocytes and diminished platelet formation in marrow. A comparable group of 20 iron deficiency anaemia patients had no thrombocytopenia and no retinopathy. There has been a paucity of publications on retinopathy in megaloblastic anaemia, but none from Sri Lanka in the last 45 years. The improved nutrition of the tea plantation workers may have been responsible for the virtual disappearance of megaloblastic anaemia and accompanying retinopathy.

Transient embolic aorto-arteritis (T E A)

Strokes in the young without the accepted risk factors of hypertension, hyperglycaemia, hyperlipidaemia, obesity, smoking and alcohol abuse, appear to be a problem common in Asian countries. The advent of echo cardiography, duplex carotids and Holter monitoring has unearthed a few young stroke patients with a treatable cause but there is still a significant number without a cause. By doing immaculate autopsy studies on 10 young stroke patients who died, the neuropathologist in General Hospital, Colombo was able to identify unusual hitherto undocumented lesions in the aorta and cervical vessels which embolised to the cerebral circulation producing infarcts. There were transient inflammatory lesions in the media with an overlying thrombus which embolised. The lesions in the media were transient leaving a scar which could easily be missed, and are unlikely to be detected by sonography or arteriography. Hence, even in this day and age, with all the modern imaging techniques available, the condition is likely to be missed unless focused and detailed autopsies are carried out, with special staining of suspect healed areas.
Juvenile cervical spondylosis

Cervical spondylosis is essentially a disease of ageing and is uncommon under the age of 40 years. In a short space of 9 months, five patients under the age of 40 years were seen with recent onset, insidious, spastic paraparesis of 1-6 months with soft signs in upper and lower limbs – impaired or inverted supinator jerk, slightly exaggerated triceps and knee jerks. Some gave a history mild indirect cervical trauma.

Clinical diagnosis was a non-compressive cervical myelopathy and the MRI showed a mild C3/C4 or C4/C5, disc protrusion. The neurosurgeon was persuaded to do a discectomy even without ‘adequate indication’ before dooming the patient for a lifetime of gloom. At surgery, one or more soft cervical discherniations were found and patients underwent cervical discectomy with or without bone grafts. Patients made a good recovery and returned to normal life.

The neurosurgeon who operated on our patients migrated to Malaysia and continued the study in Malaysia and described 26 patients with disabling symptoms of less than 6 months, 58% presenting with a spastic paraparesis. Few had bladder symptoms and motor symptoms in upper limbs and may have presented later than the SriLankan counterparts.

No patients have been described in Sri Lanka over the last 30 years – it is possible that some are being missed, due to non-consideration of it in the diagnosis, even with greater availability of MRI scanning.

Cerebellar malaria

A 40-year-old female was transferred from a malarial endemic area for exclusion of a posterior fossa tumour by CT scan because of a disabling truncal ataxia. Examination revealed midline cerebellar disease without raised ICP, brain stem, CP angle or long tract signs.
There was no clinical evidence to suggest a space occupying lesion. A focused detail history revealed that there were others in the same locality with almost identical manifestations. Malaria was rampant in the area at that time and many had taken multiple doses of anti-malarial drugs and had multiple episodes of malaria including falciparum malaria. We were uncertain as to whether it was a hitherto undocumented effect of falciparum malaria or was it an unrecognized form of toxicity to multiple doses of anti-malarials – mostly chloroquine. Fortunately we were able to establish a link with Oxford university and we concluded that it was due to immune activation causing cerebellar dysfunction following plasmodium falciparum malaria. Malaria has been at a single digit level since 2004, and is now completely eradicated, and with it cerebellar malaria.

References


Introduction

Sjögren’s syndrome first described by Swedish ophthalmologist Henrik Sjögrens in 1933 is an autoimmune epithelitis with lymphocytic infiltration of the exocrine glands, resulting in dry eyes (keratoconjunctivitis sicca) and dry mouth (xerostomia). It can occur alone or in association with other connective tissue diseases. One third of the patients presents with extra glandular neurological, rheumatological, gastrointestinal or pulmonary manifestations. Acute autonomic sensory neuropathy (AASN) with dorsal root ganglionitis is a rare association of primary Sjögren’s syndrome (PSS). We report a case of a 29-year-old female who presented with sensory ataxia with autonomic involvement who was found to have PSS.

Case report

A 29-year-old female presented with numbness in all four limbs and face with difficulty in walking and unsteadiness for one week. She had a history of upper respiratory tract infection three weeks back. She had dry eyes and mouth with dry cough and early satiety. On examination, she had resting heart rate of 120 bpm and supine blood pressure of 160/100 mmHg with blood pressure dropping to 130/90 mmHg on standing. Neurological examination revealed sensory ataxia, generalized absence of joint position sense, vibration sense and global areflexia with abnormal autonomic function tests. Muscle power, pain and temperature sensation and rest of the system examination were unremarkable.

Her ESR was 50 mm/1st hr. Antinuclear antibody was negative. Nerve conduction studies revealed absent sensory nerve action potentials in both upper and lower limbs resembling generalized axonal degeneration. Her motor conduction studies were normal. MRI scan of the brain and cervical spine and cerebrospinal fluid analysis was normal. Her sural nerve biopsy did not reveal any evidence of significant inflammation or granuloma formation.

Schirmer’s test and anti SSA antibody were positive (24.3 U/ml) with an equivocal result of anti SSB (3.62 U/ml) antibody. Parotid sialogram was negative. But minor salivary gland biopsy revealed chronic sialadenitis with lymphocytic infiltration.

According to the 2002 American-European classification criteria, the diagnosis of primary Sjögren’s syndrome was made with an acute autonomic sensory neuropathy with dorsal root ganglionitis.

She was treated with intravenous immunoglobulin 200 mg/kg/d for five days and oral prednisolone 1 mg/kg/d and thereafter with supportive rehabilitation. Her autonomic functions returned to normal but the dryness and the ataxic sensory neuropathy persisted.

Discussion

AASN is a rare disorder, usually with an antecedent event of either upper respiratory infection or gastrointestinal infection. In the pathogenesis, immune mediated mechanism may be associated with a predisposed risk of lack of blood nerve barrier in autonomic and sensory ganglion. It is associated with connective tissue diseases (Sjögren’s syndrome, systemic sclerosis, rheumatoid arthritis, systemic lupus erythematosus, mixed connective tissue disorders) paraneoplastic conditions, coeliac disease, HIV, vitamin B6 toxicity, use of cisplatin and other drugs.

Primary Sjögren’s syndrome is diagnosed according to the 2002 American-European classification criteria for PSS, which require at least four out of six (two subjective measures of ocular and oral dryness and four objective measures of ocular or salivary gland involvement or elevated anti-Ro or anti-La auto antibodies) or at least three out of the four objective criteria. She had fulfilled the criteria needed for the diagnosis of primary Sjögren’s syndrome without any association with other autoimmune diseases.

PSS is more common in females with a ratio of 2:1. Neurological manifestations of Sjögrens could be
peripheral nervous system or central nervous system involvement or myopathies. The dorsal root ganglionitis and peripheral nerve vasculitis have been observed in histology. Predominantly sensory and pure sensory neuropathies are most common in PSS. Sensory ataxia, and painful trigeminal neuropathy are due to ganglionitis while multiple mononeuropathy and multiple cranial neuropathy are due to vasculitis. Studies have shown that antibodies against dorsal root ganglion play a role in pathogenesis of sensory neuropathy. In our patient the sural nerve biopsy was normal and with the evidence of autonomic and sensory involvement, the possible site of pathology is at the sensory ganglion.

IV Immunoglobulin is used to treat sensory ataxic neuropathy and small fibre involvement. For multiple mononeuropathies and cranial neuropathies, corticosteroids are used as the first line treatment. Plasmapheresis, D-penicillamine, infliximab, cyclophosphamide, rituximab and Interferon α have been used.

Considering the age, gender, dry mucous membranes and finding of sensory and autonomic neuropathy with laboratory finding of generalized sensory axonal neuropathy, positive Schirmers and anti SSA antibody, the most likely diagnosis is PSS with a rare manifestation of ganglionitis.

References
Introduction

Decompression illness (DCI) is caused by intra or extra-vascular air bubbles that are formed as a result of rapid reduction in ambient pressure i.e. decompression. Severe DCI is a known complication among scuba divers that can lead to neurological injury and death. It is suggested that any new symptom arising shortly after an event of decompression should be considered as possible DCI. Since brain and spinal cord are the most commonly reported sites of grievous injury, neurologists’ role is imperative in proper evaluation and management of DCI. We report a professional scuba diver who developed clinical and radiological manifestations of DCI leading to long term disability following acute decompression and critically analyze the available facilities for managing these patients in Sri Lanka.

Case report

A 41 year old male was transferred from Trincomalee with weakness of both lower limbs, transient visual blurring and reduced level of consciousness of three days. He was a healthy, professional scuba diver with more than 20 years of experience in deep sea diving.

On admission GCS was 15 and he had a sensory level at T-4 with grade 1 lower limb power and sphincter involvement. Basic hematology was normal except for grossly elevated D-dimer level (9000ng/ml). The 2D echocardiography and bubble contrast study didn’t show evidence of intra-cardiac shunt. Magnetic resonance imaging (MRI) of brain revealed B/L cerebral and thalamic hyper-intensities in T2W and flare images, which were suggestive of ischemia (Figure 1). The MR angiogram of brain was normal. Cervical and thoracic spine MRI showed hyper-intensity in T2 images indicating ischemic or hypoxic changes and there were no radiological evidence of spinal cord compression.

In hospital he was given oxygen, IV fluids and though evidence is lacking, high dose IV steroids and anti-coagulation. In view of the severe disability, he was transferred to a long term rehabilitation care facility.

Figure 1. Widespread hyper-intensities seen on MRI brain.

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In hospital he was given oxygen, IV fluids and though evidence is lacking, high dose IV steroids and anti-coagulation. In view of the severe disability, he was transferred to a long term rehabilitation care facility.
Discussion

The scuba divers carry their own air cylinders underwater and breathe air at a higher atmospheric pressure for a longer duration. The gas solubility increases at greater pressure according to Henry’s law and inert gases, mainly nitrogen gets absorbed in to the tissues. The gas solubility decreases during ascent and as a result bubbles are formed in tissues and if no sufficient time is given for “off gassing” via lungs, those bubbles can cause mechanical and biochemical disruptions leading to tissue damage. Therefore dives must always be pre-planned with a controlled rate of ascent and compulsory “decompression stops” in order to allow gas to leave the system. This phenomenon can also be seen when an unpressurized air craft is flying up and when a worker is coming out from a pressurized mine or a caisson.

Decompression Illness takes two major forms; type 1 involving muscles, joints and skin causing so called ‘bends’ and type 2 affecting the CNS, respiratory or cardiovascular systems which could be life threatening. Spinal cord injury usually arises from DCI as a result of nitrogen bubbles obstructing spinal venous plexus and cerebral involvement is mostly due to arterial gas embolism.

This patient had clinical and radiological evidence of both cerebral and spinal involvement of DCI. Despite significant ischemic insult to brain, he regained full consciousness within a short period but the spinal cord syndrome poorly recovered regardless of IWR and HBO. The definitive treatment of DCI is HBO therapy which involves placing the patient in a pressurized chamber where he breathes pure HBO i.e. oxygen at pressure excess of 1 atmosphere. In Sri Lanka HBO therapy is available only at Naval Hospital Trincomalee. Water recompression as practiced here involves the incapacitated diver being taken down with a fresh air cylinder to various depths and durations until the diver feels adequate relief of symptoms. It is not a recommended practice but cannot be completely dejected due to the local situation where access and gaining admittance to the only centre with HBO facility in Sri Lanka is difficult. Prompt HBO treatment gives the best chance of complete recovery, but Fonseka et al reported two cases of type 2 DCI with neurological deficits recovering from recompression therapy even after four and five days after injury.

Conclusion

In Sri Lanka scuba diving is becoming an admired recreational activity and also there is a population of professional scuba divers who daily dive the deep sea. There are risks involved with it thus certain governing regulations do exist. It is imperative to follow standard diving protocols, as DCI is always preventable by correct diving practices. Being an island nation, raising awareness among health care workers regarding management of DCI and upgrading country’s HBO facilities with more than one center can help not only to minimize DCI related disability but also Sri Lanka to become a diving hub in South Asia with state of the art treatment centers for managing diving related medical problems.

References

Sjögren’s syndrome presenting with recurrent strokes

A T M Liyanaarachchi1, V Jeevagan1, S Bandusena1, P S Gunaratne1


Introduction

Sjögren’s syndrome (SS) is an autoimmune rheumatic disease characterized by lymphocytic infiltration and destruction of exocrine glands. The spectrum of clinical manifestations of SS is wide, ranging from mucosal dryness to extra glandular systemic manifestations. Involvement of the central nervous system (CNS) in patients with SS is variable and the peripheral nervous system involvement is well recognized. We report a patient with SS presenting with strokes as the first clinical manifestation.

Case report

A 49-year-old female was admitted with acute onset vomiting, vertigo and difficulty in walking for three days. She also had difficulty in swallowing and worsening weakness of her right upper limb and lower limb. One year ago she had presented with sudden onset right hemiparesis and vertigo. MRI brain at that time had revealed multiple lacunar infarctions in left corona radiata, right cerebellar hemisphere and brainstem. She was given physiotherapy and was started on aspirin, dipyridamole and atorvastatin. She made a good recovery with mild residual weakness of right side but had defaulted follow up after three months. Her medical history was otherwise unremarkable.

On examination she had right hemiparesis with bilateral cerebellar signs, left Horner’s syndrome and left seventh (lower motor neurone), ninth and tenth cranial nerve palsies. Cardiovascular system examination was normal with a blood pressure of 120/80 and regular rhythm. There were no neck bruits.

MRI brain revealed acute and old multiple lacunar infarctions in basal ganglia bilaterally, left corona radiata, pons (Figure 1) and cerebellum. MRA of neck and intracranial arteries was normal. MRI further revealed multiple cystic lesions in both parotid glands possibly due to sialectasis (Figure 2). Subsequent inquiry revealed the patient to have dry mouth (xerostomia) and dry eyes for one year.

Routine stroke work up including lipid profile, fasting blood glucose, transthoracic and transoesophageal echocardiogram, carotid and vertebral duplex studies were normal.

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Basic laboratory testing was normal except elevated ESR of 95mm. Vasculitic screen revealed positive rheumatoid factor and anti SS-A antibodies. ANA, anti DS DNA, anti SS-B, anti cardiolipin, VDRL and HIV antibodies and C-ANCA, P-ANCA were negative and C3, C4 and LDH levels were normal.

Lip biopsy showed marked lymphocytic infiltrate, acinar atrophy and lymphoepithelial lesion formation compatible with SS (Figure 3). Cerebrospinal fluid (CSF) had a high protein content of 125mg/dl but was acellular.

Diagnosis of SS was made and she was given intravenous immunoglobulin (IVIG) 0.4 mg/kg for 5 days, intravenous methyl prednisolone 1g daily for 3 days followed by oral prednisolone and azathioprine together with physiotherapy. During the hospital stay of three weeks she made a good recovery and Barthel index improved from 40 to 80.

Discussion

SS is the second commonest autoimmune rheumatic disease after rheumatoid arthritis, affecting 3-4% of the adult population. Spectrum of CNS SS include focal lesions presenting as motor and/or sensory deficit, aphasia, dysarthria, seizures, cerebellar syndromes and non-focal lesions presenting as encephalopathy, seizures, aseptic meningitis, dementia, and psychiatric disorders. Spinal cord lesions can give rise to transverse myelitis, chronic progressive myelopathy, neurogenic bladder and Brown Sequard syndrome. CNS SS may mimic multiple sclerosis and present as a relapsing-remitting or primary progressive syndrome with increased IgG synthesis and oligoclonal bands in the CSF\(^1\). Immunologically mediated mechanisms, mainly vasculitis is thought to play a role in CNS manifestations of SS. It has also been found that anti SS-A positivity to be associated with severe disease with frank necrotizing angiitis\(^2\). Histopathologically there is small vessel mononuclear inflammatory and ischaemic/haemorrhagic vasculopathy.

Recurrence of stroke in a patient at forty nine years of age in the presence of uncontrolled multiple vascular risk factors may not be uncommon. However, in situations where vascular risk factors are absent one need to be vigilant and should search for uncommon causes. While raised ESR directed towards a systemic pathology, positive Rheumatoid factor and anti SS-A antibodies along with oral and ocular symptoms and lip biopsy findings confirmed Sjögren’s syndrome in the reported patient\(^3\). For diagnosis of CNS SS, MRI brain is nonspecific and CSF commonly shows lymphocytosis, a raised IgG index and oligoclonal bands. Treatment of CNS SS remains largely empirical. There are no randomized clinical trials but intravenous pulses of corticosteroids, cyclophosphamide, plasmapheresis and IVIG have been reported to be beneficial\(^4,5\).

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References


Introduction

Narcolepsy is a syndrome, characterized by a pentad; excessive daytime sleepiness (EDS), cataplexy, hypnagogic hallucinations, sleep paralysis and nocturnal sleep disturbance. It is a chronic brain disorder that involves poor control of sleep-wake cycles. Those affected enter Rapid Eye Movement (REM) sleep within a short period compared to normal sleeper with intrusion of features of REM sleep into wakeful state. The pathophysiological basis of the disease is the loss of hypocretin secreting neurons in the hypothalamus which is postulated to be related to a multifactorial inheritance. A strong genetic association with HLA-DQB1*0602 and polymorphism in the T cell receptor alpha locus is described. Recent evidence also suggests an autoimmune basis due to isolation of antibodies against the anti-tribbles homolog.

Narcolepsy is a rare disorder with an estimated prevalence of 0.07% and presenting in childhood is even rarer. We present an 8-year-old boy who presented with classic features of narcolepsy syndrome followed by good response to therapy.

Case report

An 8-year-old, previously healthy boy of non-consanguineous parents presented initially with abnormal orofacial movements which manifested as brief episodes of lower lip biting. This was followed by a march of events appearing one after the other; lateral tongue movement, episodic change in size of palpebral fissure and episodic protrusion of tongue. After about three months he developed features of excessive daytime sleepiness such as sleeping during school, in the middle of conversation and even during favorite television programs. He was also noted to have abnormal limb movements in sleep with frequent a waking but did not experience snoring or hypnagogic hallucinations. Seven months since onset of first symptoms, he developed episodes of falling down whenever he laughed loud. Retrospective analysis revealed an increase in his body weight by nine kilograms over a nine month period. His school performance also gradually deteriorated with worsening aggressive behavior.

He appeared lethargic during examination with BMI between +2 SD and +3 SD. Systemic examination including neurological examination was normal.

MRI brain with pituitary sliced view did not show any significant focal lesions. EEG was normal and polysomnogram did not show features of obstructive sleep apnoea.

He was treated with methylphenidate and imipramine in addition to non-pharmacological measures such as improved sleep hygiene, organized sleep schedules, caffeine containing food and beverages and regular exercise before sleep. These resulted in improvement of the EDS and the cataplexy.

Discussion

Narcolepsy syndrome is a sleep disorder with subtle and gradual onset. The progression varies but may take several years to develop all clinical symptoms. This results in difficulties in arriving at a definite diagnosis early in the illness. The symptoms of narcolepsy syndrome includes EDS; the commonest experienced by all patients. It is the first symptom to be clinically apparent as well. It consists of involuntary sleep episodes of variable lengths ranging from hours to minutes. Cataplexy is the only symptom specific to narcolepsy which occurs at the beginning of narcolepsy or several years later. It is defined as transient episodes of loss of muscle tone of brief duration (<2 minutes), with preserved consciousness. It is often triggered by emotion. Hypnagogic hallucination (HH) occurs at the sleep onset and includes visual, tactile, kinetic and auditory phenomena. Sleep paralysis (SP) is a transient inability to move or speak while falling sleep or awake and last a few seconds or minutes. There is preserved consciousness during these hallucinations.

Disturbed nocturnal sleep includes difficulties in staying asleep due to insomnia, vivid dreaming, sleep talking, acting out while dreaming, or periodic leg movements. Other symptoms that are less frequently experienced in narcolepsy include weight gain, deterioration of school performances, poor concentration, orofacial movements, periodic limb movement during sleep and emotional lability.
Among the above symptoms, cataplexy is the most specific symptom and is rarely present outside of narcolepsy. When cataplexy is not present, diagnosis must be made after excluding other possible causes of daytime sleepiness and fatigue, along with a positive multiple sleep latency test (MSLT). A PSG also helps to find whether REM sleep occurs at abnormal times in the sleep cycle and can rule out the possibility that an individual’s symptoms result from another condition. When no other serious medical condition is present, low cerebrospinal fluid (CSF) hypocretin-1 can establish hypocretin deficiency as the cause of narcolepsy. Human leukocyte antigen (HLA) typing of narcolepsy patients shows certain alleles located on chromosome 6 are strongly associated with narcolepsy-cataplexy and it supports the hypothesis of possible of autoimmune aetiology.

Treatment of narcolepsy requires both non-pharmacologic and pharmacologic measures. Non-pharmacologic measures are avoiding sleep deprivation, scheduled naps, caffeine containing foods such as chocolate, cola drinks, coffee and tea, undisturbed sleep environment, regular exercise in the evening and fixed sleeping schedules.

The pharmacological treatment strategies include use of several classes of drugs. Amphetamines like compounds are mainly useful to improve EDS. These include drugs like methylphenidate, methamphetamine and amphetamine. Although EDS is improved subjectively, sleep variables are never completely normalized with these compounds. Out of these methamphetamine is well tolerated and effective in many and because of its relatively short duration of action (3-4 h) it can be used on an “as needed” basis.

The other class of medications used in narcolepsy is compounds such as modafinil or armodafinil. They are milder stimulants with relatively lesser side effects and limited tolerance and dependence, thus favoring them as the first line used for EDS. However, it has limited efficacy on cataplexy and other sleep variables. Armodafinil has relatively longer wake-promoting effects.

Gamma hydroxybutyrate, a sodium oxybate belong to another group of drugs used in narcolepsy. It is efficacious for EDS, cataplexy, hypnagogic hallucination, sleep paralysis and disturbed nocturnal sleep. A large-scale, double-blinded, placebo-controlled clinical trial in the United States has shown its efficacy on narcolepsy-cataplexy placing it as the first line treatment for the above. Its effect on EDS is relatively quick, but anti-cataplectic effect may take one to twelve weeks to appear. Antipsychotics are the other group of drugs that are used in narcolepsy. Tricyclic antidepressants (TCA), selective serotonin reuptake inhibitors (SSRIs), selective serotonin-norepinephrine reuptake inhibitors (SNRI) are reported to be effective for cataplexy. TCA such as imipramine is the most commonly used anti-cataplectic agent and it also effective in sleep paralysis and hypnagogic hallucinations. Though SSRIs show comparatively lesser side effects, its potency is not impressive since evidence shows predominance of the norepinephrine system for the control of cataplexy rather than the serotonin system. Venlafaxine; a selective norepinephrine reuptake inhibitor (SNRI) is reported to be effective for cataplexy. Hypocretin replacement, gene therapy, cell transplantation and immunomodulation are emerging therapies in narcolepsy.

This case history highlights the importance of careful clinical evaluation. Narcolepsy syndrome can be easily misdiagnosed as either epilepsy or movement disorder during initial part of illness because of the abnormal orofacial movements and cataplexy. Though we did not have facilities to do MSLT and serological tests – clinical evaluation, normal MRI and treatment response point towards the diagnosis.

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Introduction

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An 8-year-old, previously healthy boy of non-consanguineous parents presented initially with abnormal orofacial movements which manifested as brief episodes of lower lip biting. This was followed by a march of events appearing one after the other; lateral tongue movement, episodic change in size of palpebral fissure and episodic protrusion of tongue. After about three months he developed features of excessive day time sleepiness such as sleeping during school, in the middle of conversation and even during favorite television programs. He was also noted to have abnormal limb movements in sleep with frequent a wakening but did not experience snoring or hypnagogic hallucinations. Seven months since onset of first symptoms, he developed episodes of falling down whenever he laughed loud. Retrospective analysis revealed an increase in his body weight by nine kilograms over a nine month period. His school performance also gradually deteriorated with worsening aggressive behavior.

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References

Summary
Behcet’s disease is a multisystem vasculitis of unknown aetiology, in which neurologic involvement has been reported in the range of 5% to 10%. We report a young patient, who presented with orogenital ulcers, brainstem dysfunction and positive pathergy reaction where the diagnosis of Neuro Behcet’s disease (parenchymal type) was made. His magnetic resonance imaging (MRI) showed bilateral lesions, mainly in brainstem, thalami and periventricular regions. He was successfully treated with immunosuppressive agents.

Index words: Neuro Behcet’s disease, vasculitis, brainstem syndrome

Introduction
Behcet’s disease was originally described by a Turkish dermatologist, Hulusi Behcet in 1937 as a triad of oral and genital ulcerations with uveitis. Now we know that it is a multisystem vasculitis of unknown origin, in which neurologic involvement has been reported in the range of 5% to 10%

According to the widely accepted International Study Group (ISG) criteria, for the diagnosis of Behcet’s disease recurrent oral aphthae (at least three times in one year) are a prerequisite. It should be accompanied by any two out of the following: recurrent genital ulcerations; skin lesions such as erythema nodosum, papulopustular lesions, pseudofoliculitis and acneform lesions; eye involvement such as anterior or posterior uveitis; skin pathergy reaction.

Case report
A 28-year-old male presented with a one week history of unsteady gait, slurred speech, vertigo, diplopia in both directions, mouth deviation to right and right hemiplegia. He had developed painful oral and genital ulcers one week prior to the onset of neurological symptoms. Past history was remarkable for recurrent painful oral ulcers (3-4 episodes per year) and one episode of genital ulcers which had developed few years back. There was no fever, headache, rashes, joint pain, abdominal pain or loose stools. On examination, there were multiple oral ulcers, where each had a well defined border, yellowish base and a diameter of less than one centimetre. Moreover, there were multiple scrotal ulcers. There were no other skin lesions. Neurological examination revealed bilateral lateral rectus palsy, bidirectional horizontal nystagmus, left sided lower motor neuron type facial nerve palsy, bilateral cerebellar signs with scanning dysarthria and mild right sided hemiplegia with anupgoing plantar response. He had normal cognitive functions, pupillary and fundoscopic findings.

Initial haematological and biochemical parameters were within normal range except for moderately raised inflammatory markers. Erythrocyte sedimentation rate (ESR) was 40 mm after one hour and C reactive protein (CRP) was 12 mg/l (<6). Magnetic resonance imaging (MRI) of brain demonstrated multiple focal hyper-intensities in T2 weighted and fluid attenuated inversion recovery (FLAIR) images in bilateral pons, midbrain, periventricular regions, cerebral and middle cerebellar peduncles and thalami with restricted diffusion in diffusion weighted images (DWI) (Figure 1). MRA, MRV and digital subtraction angiogram (DSA) were normal.

Cerebrospinal fluid (CSF) analysis showed marginally high protein levels of 48 mg/dl (ref. range: 15 - 45 mg/dl), lymphocytic pleocytosis (30 lymphocytes / mm³) and two polymor phonuclear leukocytes / mm³ and normal glucose (glucose of 5.1 mmol/l in CSF with a plasma value of 5.9 mmol/l). Workup for infective causes including tests for tuberculosis (TB), viral and fungal pathogens, HIV and syphilis were negative. ANA, c and p ANCA were not detected. As the patient had recurrent oral and genital ulcers with neurological symptoms, pathergy test was done, which became positive after 48 hours (Figure 2).

Based on ISG criteria and by excluding other infective and inflammatory causes, diagnosis of Behcet’s disease was made. He fulfilled the required criteria (recurrent oral ulcerations) and two of the four minor criteria (recurrent genital ulcerations and positive pathergy test).
Eye involvement was excluded by ophthalmological examination. He was started on IV methylprednisolone followed by oral prednisolone 1 mg/kg. In the mean time he was started on azathioprine. Oral prednisolone was tailed off and azathioprine is to be continued for 18 to 24 months. With the immunosuppressive therapy, the patient improved clinically, and interval MRI done 3 months later demonstrated near complete resolution of the brain lesions.

**Discussion**

Behcet’s disease is a chronic, relapsing, occlusive vasculitis of unknown etiology, affecting almost every organ system in the body. Theories behind the pathogenesis of Behçet disease currently suggest an autoimmune aetiology. It has been observed that there is an increase in the risk of HLA-B51/BS carriers to develop Behçet’s disease. Among the systemic vasculitides, Behçet’s disease is remarkable for its ability to involve blood vessels of all sizes (small, medium, and large) and in both the arterial and venous sides of the circulation.
In 1999, Akman Demir and colleagues reported the evaluation of 200 patients with Behcet’s disease who had neurological manifestations. They found out two patterns of neurological involvement: parenchymal and nonparenchymal. Parenchymal involvement is due to lesions in the corticospinal tracts, brainstem, cerebellum, periventricular white matter, spinal cord and basal ganglia. Pathology reveals local perivenular lymphocytic cuffing, inflammatory cell infiltration, gliosis, necrosis and neuronal loss. Frank vasculitis is not always observed in parenchymal lesions. Common presentations of parenchymal involvement are brainstem syndrome with cranial nerve palsy, cerebellar dysfunction and corticospinal tract signs, similar to above described patient. Non-parenchymal type, also known as vasculo-Behcet’s disease, mainly results from vascular involvement and commonly presents as venous occlusion such as dural sinus thrombosis, arterial occlusion and arterial vasculitis. Arteritis may lead to ischemic strokes, dissection, aneurysmal dilatation, and subarachnoid hemorrhage. Other manifestations include aseptic meningitis and epileptic seizures.

Neuro Behcet’s disease (NBD) can be mistaken for other diseases with multi focal presentations, such as multiple sclerosis (MS) and systemic lupus erythematosus (SLE), especially in the absence of systemic manifestations. In the acute phase of NBD, large confluent lesions in the brainstem-thalamic-basalganglia region are highly suggestive of the disease. In SLE, the lesions predominantly involve the subcortical white matter, and in MS, the lesions are predominantly periventricular, with less frequent basalganglia or brainstem involvement. Our patient had MRI lesions in brainstem-thalamic region with normal MRA/V, which favors the diagnosis of parenchymal Neuro Behcet’s disease. One notable feature in our patient was the lack of eye involvement, as 66% of the patients in the case series by Akman Demir and colleagues had ophthalmological manifestations.

Parenchymal involvement, especially of the brainstem, and high CSF cellular and/or protein content, is significantly associated with a worse prognosis. Management of neurological manifestations of Behcet’s disease involves high dose steroids combined with a second immunosuppressive agent. Venous sinus thrombosis requires immunosuppressive therapy simultaneous to anticoagulation. Hence identifying the type of neurological involvement is important for the management and prognosis.

Competing interests

The authors declare that they have no competing interests.

References

A deadly strike of a needlefish

B M T P Nawasiwatte¹, S Bandusena¹, S Wadanamby¹, P S Gunaratne¹


Index words: needlefish, ear injury, cerebral infarction

Introduction

Fatal and near fatal injuries inflicted by needlefish (Tylosurus crocodilus) among worldwide marine dwellers are heard infrequently. We report a young diver who developed a massive fatal middle cerebral artery (MCA) territory infarction following a piercing injury to right ear by a wandering needlefish, which is the first reported case of this nature.

Case history

A 41 year old healthy Maldivian professional diver was airlifted to Sri Lanka for acute management of sudden onset left hemiplegia and rapidly deteriorating level of consciousness.

Two days ago while diving in shallow sea he was accidentally hit by a needlefish and his right ear was penetrated deep in, with its long sharp beak. The patient managed to detach the fish and the beak from himself and later, a friend removed further particles from the ear. Apart from profuse ear bleeding, he remained well and had received a tetanus toxoid from his island health centre.

Three hours later, he developed bi-lateral visual blurring and sudden onset left face arm leg weakness necessitating transfer to a bigger hospital. His GCS dropped rapidly to eight and the right pupil was dilating. The NCCT brain showed acute right MCA territory infarction and patient was severely hypotensive on admission. At this point we received him for further management.

Figure 1. Anatomical proximity of internal carotid artery to the auditory canal.

Figures 2 and 3. NCCT brain of the patient 6h and 48h after.

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He was resuscitated and the NCCT brain 48h later confirmed the massive infarction of the right MCA territory with cerebral edema and mid line shift, plunging his GCS to three. Initial biochemistry, coagulation profile and D-dimers were normal but his inflammatory markers were elevated (WBC 22,000 CRP 44mg/l). There was no evidence of hemolysis or thrombocytopenia. He was intubated and ventilated but by day three he developed intractable diabetes insipidus and deteriorating renal functions.

On examination, brain stem reflexes were repeatedly absent and patient was confirmed brain dead after 72 hours of the initial event. A decompressive craniectomy was not done and he was gradually weaned off from inotropes and ventilator. No specific therapy was instituted other than mannitol, dexamethasone and IV broad spectrum antibiotics.

Discussion

The needlefish (*Tylosurus crocodilus*), sometimes referred to as *Garfish* a slender, long-beaked surface fish found in tropical and sub-tropical oceans including Sri Lankan coastal belt. The needlefish can cause serious bites but is non venomous1. These fish are attracted to bright lights and can propel themselves out of the water towards the light source in a surprising speed and are capable of inflicting deep puncture wounds, in those who happen to cross their trajectory path, beak often breaking off inside the victim during the process.

There are three reported cases of fatal orbital and brain penetrations and one case ending up forming a carotico-cavernous fistula2-3. Another handful of cases of stabbing through heart, neck and abdomen causing near fatal injuries are also published. However, this is the first case that we came across of a cerebral infarction following a penetrative ear injury by a needlefish. His clinical presentation of stroke was confirmed radiologically.

Direct trauma to the MCA or internal carotid artery (ICA) primarily by the fish beak and secondarily during the forceful removal of it by the patient could have led to dissection or arterial spasm of either artery, causing the infarction. As the anterior cerebral artery territory was more or less spared, MCA is the likelier site than the ICA. Possibility of cerebral venous thrombosis causing a venous infarction is less likely because the infarction respected the arterial territory. Fish bites are known to cause a variety of exotic secondary infections that can lead to cerebral infarctions but the swiftness of events did not suggest that possibility. However, cerebral vascular studies could not be carried out due to the rapidity of clinical deterioration.

Diving and surfing are emerging pastimes in Sri Lanka among the locals as well as the foreigners. It may be worthwhile raising awareness of perils that may come along with adventures and educate regarding safety measures one should take while exploring the unfathomable oceans.

References

Foster Kennedy syndrome

A T Maththias¹, A L L Roshan¹, S B Gunatilake¹,²


Index words: Foster Kennedy syndrome, meningioma, papilloedema, optic atrophy

Introduction

Foster Kennedy syndrome is a sign in neurology which is seen rarely but spoken about frequently. Since the development and wide spread availability of numerous neuroimaging modalities, this sign is a rarity in the modern world. Foster Kennedy syndrome is ipsilateral optic atrophy and contralateral papilloedema caused by an intracranial mass. Since it was first described by Sir Robert Foster Kennedy back in 1969, many case reports have been published. Ophthalmologic examination and neuroimaging is essential for the diagnosis of Foster Kennedy syndrome. We present a case of unilateral blindness due to a large sphenoidal wing meningioma causing Foster Kennedy syndrome and discuss the history, etiopathogenesis and clinical relevance of this sign in today’s context.

Case report

A 49-year-old woman admitted to hospital with a complaint of visual impairment for about two weeks. She complained of total blindness in the left eye. She had not experienced any significant headache or any eye pain. She did not have emotional lability, depression nor abnormal behaviour. The patient was not on any medications. The past medical, gynaecological, family, surgical histories were not significant.

On examination, the patient was alert and Glasgow coma scale was 15. The pupillary examination revealed a left relative afferent papillary defect. Sense of smell was normal. Fundoscopy revealed left optic atrophy and right papilloedema. Extraocular eye movements were normal. All other cranial nerves were normal. Her visual acuity was 6/9 in the right eye and 1/60 in the left eye. The retinal photography shows left optic atrophy and right papilloedema (Figure 1).

All her basic blood investigations were normal. The contrast computed tomography of the head showed a large mass 5X5 cm in the left frontal region with some calcification in the middle suggestive of a meningioma. A magnetic resonance imaging confirmed the diagnosis of a meningioma arising from the lesser wing of sphenoid encasing the supraclinoid portions of the left internal carotid and compressing the intracranial portions of the left optic nerve causing mass effect (Figure 2 and 3).

The patient was started on dexamethasone for cerebral oedema and was transferred to a neurosurgical unit for surgical management. The patient is presently awaiting surgery.

Figure 1. Papilloedema in left and optic atrophy in right eye.

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Discussion

Foster Kennedy published his article titled, “Retrobulbar Neuritis as an Exact Diagnostic Sign of Certain Tumors and Abscesses in the Frontal Lobes” in 1991. The six cases he discussed were tumours and abscesses of the frontal lobe and the mass can be a meningiomas of the olfactory groove, falx, sphenoidal wing or subfrontal regions, craniopharngiomas, pituitary adenomas plasmacytomas and large aneurysms.

In them he described the occurrence of retrobulbar neuritis and optic atrophy on the side of the lesion together with papilloedema of the contralateral eye.

Mechanism

The Foster Kennedy sign represents optic atrophy and anosmia on one side, with papilloedema caused by raised intracranial pressure on the other. Majority of cases occur without anosmia. The exact pathogenesis of Foster Kennedy syndrome is unclear. It is postulated that an intracranial mass causes direct pressure on the optic nerve, causing impared ocular venous return and compression in the subarachnoid space of the intraorbital part of the optic nerve. This mechanical compression results in ipsilateral optic atrophy. Therefore the development of papilloedema is prevented in that eye. The elevated intracranial pressure causes papilloedema of the contralateral side.

Variations

Pseudo-Foster Kennedy syndrome is described as unilateral optic disc swelling with contralateral optic atrophy in the absence of an intracranial mass causing compression of the optic nerve. This occurs mainly due to bilateral sequential optic neuritis, ischaemic optic neuropathy and unilateral optic nerve hypoplasia.

Conclusion

Foster Kennedy syndrome is rare. Today any patient complaining of persistent headache undergoes neuroimaging and an intracerebral lesion is found before visual loss develops and hence the rare occurrence of this sign. Typical syndrome due to mass lesions are very unlikely to be seen today especially in the developing world, but rare cases still can come up in countries where imaging facilities are not freely available and where access to proper health care is difficult. Foster Kennedy syndrome should be considered in patients who present with unilateral visual impairment, or a junctional scotoma and headache.

References

Varicella-zoster vasculitis presenting with ischemic stroke is a known entity and has been described as one of the rare but important causes of stroke, especially in children. But primary intracranial haemorrhage has been reported only in few cases. The child reported here had basal ganglia haemorrhage causing acute left hemiplegia two weeks post varicella.

**Case report**

A previously healthy 16-year-old female presented to our ward with acute left hemiplegia. Two weeks prior to admission, the patient has developed an erythematous vesicular rash on her chest and abdomen which spread to her face, arms, and legs. The vesicular lesions on the trunk crusted week after appearance, consistent with varicella.

On examination her vital signs were normal, as is her mental status and cranial nerve examination. Motor strength was 0/5 in right upper and lower extremities, and 5/5 in the left with a mild left hemisensory deficit.

The initial computerized tomography (CT) of the patient’s brain, performed few hours after admission, revealed a left basal ganglia haemorrhage without midline shift or mass effect (Fig. 1a). MRI done next day showed the same haemorrhage, but did not show any underlying structural cause for the bleed (Fig. 1b).

Laboratory investigations showed mild anaemia, normal white cell count, normal coagulation profile (prothrombin time, activated partial thromboplastin time and thrombin time), and normal erythrocyte sedimentation rate. But C-reactive protein levels were 14 mg/L (normal range 0-6 mg/L). Laboratory investigations did not show any bleeding diathesis. Her systemic vasculitis workup was negative for anti-nuclear antibodies, anti-double-stranded DNA, and rheumatoid factor. Echocardiography was also normal. Anti-VZV immunoglobulin M (IgM) was detected in blood but VZV immunoglobulin G (IgG) was not present. She did not have a history of any other systemic illness.

The patient was managed conservatively and patient was referred to physiotherapist and occupational therapist. Four weeks after the episode her weakness improved to a level where she was able to perform activities of daily living.
Discussion

There are many possible causes of non-traumatic intracranial haemorrhage, and at least one risk factor usually can be identified if a complete evaluation is done. A study (Table 1) of non-traumatic hemorrhage in children found at least one potential cause for the hemorrhage in children. These authors failed to identify risk factors in only 15% children whose evaluation included standard cerebral angiography3.

The most common risk factors for any type of intracranial haemorrhage were intracranial vascular anomalies, congenital heart disease, and brain tumors. These were excluded in our patient reported here.

But vasculitis of the intracranial vessels can be difficult to diagnose with confidence. Not all individuals have clinical or laboratory signs of inflammation and the classic angiographic findings of arteritis (arterial beading and alternating areas of constriction and dilatation) are nonspecific. Angiography is also not a freely available investigation as in our case.

<table>
<thead>
<tr>
<th>Risk factor</th>
<th>No of Children (N=85)</th>
<th>% of children</th>
</tr>
</thead>
<tbody>
<tr>
<td>Brain tumor</td>
<td>11 1 1 13 15</td>
<td></td>
</tr>
<tr>
<td>Congenital heart disease with or without surgery or anticoagulation</td>
<td>6 8 14 16</td>
<td></td>
</tr>
<tr>
<td>Intra cranial vascular lesion</td>
<td>21 3 24 28</td>
<td></td>
</tr>
<tr>
<td>AVM</td>
<td>11 11 13</td>
<td></td>
</tr>
<tr>
<td>Cavernous malformation of the brain</td>
<td>7 7 8</td>
<td></td>
</tr>
<tr>
<td>Venous angioma</td>
<td>2 2 2</td>
<td></td>
</tr>
<tr>
<td>Aneurysm</td>
<td>2 2 2</td>
<td></td>
</tr>
<tr>
<td>Moyamoya syndrome (with sickle cell disease or Down syndrome)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Infection</td>
<td>3 3 5 6</td>
<td></td>
</tr>
<tr>
<td>HSV encephalitis</td>
<td>1 1 1</td>
<td></td>
</tr>
<tr>
<td>Sepsis (thrombocytopenia)</td>
<td>2 1 3 4</td>
<td></td>
</tr>
<tr>
<td>Endocarditis</td>
<td>1 1 1</td>
<td></td>
</tr>
<tr>
<td>Leukemia or lymphoma (thrombocytopenia)</td>
<td>5 1 6 7</td>
<td></td>
</tr>
<tr>
<td>ARF, aplastic anaemia or LCHAD</td>
<td>4 1 5 6</td>
<td></td>
</tr>
<tr>
<td>Coagulation factor deficiencies</td>
<td>1 3 4 5</td>
<td></td>
</tr>
<tr>
<td>Genetic (e.g. Menkes syndrome)</td>
<td>1 1 1 1</td>
<td></td>
</tr>
<tr>
<td>Unidentified</td>
<td>10 2 1 13 15</td>
<td></td>
</tr>
</tbody>
</table>

Abbreviations: ARF, acute renal failure; AVM, arteriovenous malformation; HSV, Herpes simplex virus; ICH, intracerebral hemorrhage; LCHAD, long-chain 3-hydroxyacyl coenzyme A dehydrogenase deficiency; SAH, subarachnoid haemorrhage; SDH, subdural haemorrhage.
Over the past few decades there have been an increasing number of reports of vascular disease after VZV reactivation. Unlike early cases of acute hemiplegia after contralateral zoster caused by large-artery disease, the recognized clinical range of this disease has expanded to include transient ischaemic attacks and protracted illness involving both small and large arteries. In addition to ischaemic infarction, VZV can cause aneurysm, cerebral and subarachnoid haemorrhage, and arterial ectasia, and might be a co-factor, along with trauma, in the pathogenesis of cerebral arterial dissection. Furthermore, VZV can also cause peripheral arterial disease. In adults, the exact incidence of VZV vasculopathy is difficult to estimate, although it is more common in immunocompromised individuals. In children, VZV vasculopathy has been proposed to account for 31% of all arterial ischaemic strokes; moreover, stroke was preceded by chickenpox in 44% of children with transient cerebral arteriopathy.

Brain imaging reveals abnormalities in most cases. Abnormalities are cortical and deep, and occur in both the grey and white matter and at grey-white matter junctions in particular a clue to the cause of disease. Although lesions of the grey and white matter are commonly seen in patients with other disorders such as metastatic carcinoma and embolic disease, VZV vasculopathy should be included in the differential diagnosis of patients with lesions at grey-white matter junctions. Most lesions are ischaemic, but haemorrhagic lesions also occur; some lesions enhance on MRI with contrast, indicating breakdown of the blood-brain barrier.

When a clinical diagnosis of VZV vasculopathy is suspected and is corroborated by single or multiple characteristic lesions on MRI or CT, virological confirmation is required.

The clinical diagnosis of VZV vasculopathy is strongly suspected when a patient with a recent history of varicella or zoster has a transient ischaemic attack or stroke corroborated by MRI abnormalities, particularly at the grey-white matter junction.

References
Neurotoxicology

(Compiled by Ajini Arasalingam and Saman Gunatilake)

1. A 30-year-old male was brought to the emergency department (ED) 6 hours after dining in a restaurant complaining of perioral numbness and tingling, a strange metallic taste, and cold allodynia. He was also mildly hypotensive and bradycardic. He had consumed this fish grilled.

What is the diagnosis?
Name the likely sources.

2. A 25 year old female was brought to the emergency department 24 hours after eating a Japanese seafood delicacy at a sushi restaurant. Shortly after ingestion, she developed lip and tongue paresthesia. Four hours after ingestion, she became nauseated, vomited, and had abdominal pain. She presented with decreased strength in both of her legs.

The dish contained the fish shown below.

What is the causative agent?
What are the likely sources?

3. What symptoms and signs would a bite from this spider cause?

4. A 55 year old man from Somalia presented with spastic paraparesis. He had been eating the legume shown for many years.

What is the diagnosis?

5. A 25 year old male from Northern Mexico presented to a hospital in Mexico with a rapidly progressive paralysis. He had consumed the fruit shown.

What is the diagnosis?

(Answers on page 42-43)
The Sri Lanka Journal of Neurology is published half yearly (June and December) by the Association of Sri Lankan Neurologists. Material received for publication in the SLJN must not be submitted for publication elsewhere without the editors’ permission (see below under Previous Publication and under Cover Letter).

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The SLJN publishes original papers and commentaries which have relevance to Neurology and allied sciences.

Papers

Original work concerning the causes, mechanisms, diagnosis, management and prevention of disease belong in this category. So do articles on health systems research, health economics and management, and medical ethics. They should have less than 2000 words, 5 tables and illustrations, and 20 references.

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This category includes case reports of drug adverse effects, of a single event that could lead to a new piece of knowledge, preliminary reports of drug trials, new patient management methods, and reports of new techniques and devices. They should not exceed 1000 words, and contain more than 3 tables or illustrations, and more than 10 references.

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The SLJN will also consider for publication letters (less than 400 words of text, 3 authors, and 5 references), obituaries (less than 400 words), and contributions to the picture-story series (not more than 250 words of text, 3 authors, 3 references and 2 clear black and white or colour photographs).

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Cover letter

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The manuscript should be mailed, with adequate protection for figures, to Prof Saman Gunatilake, the Editor, SLJN, Department of Medicine, Faculty of Medical Sciences, University of Sri Jayewardenepura, Nugegoda. SRI LANKA. Email: saman.gunatilake@hotmail.com

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1. Conceived and planned the work that led to the paper, or interpreted the evidence it presents, or both.
2. Wrote the paper or reviewed successive versions, and took part in revising them.
3. Approved the final version.
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In the cover letter give full details on any possible previous publication of any content of the paper. eg.

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2. Patients in a study already described and published.
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The SLJN will consider all manuscripts prepared in accordance with the uniform requirements for manuscripts submitted to biomedical journals developed by the International Committee of Medical Journal Editors [1]. A summary of these and the requirements of the SLJN are given below.

Manuscript typing
All parts of manuscript, including tables and figure legends, must be typed with double-spacing. References must also be double spaced. Manuscripts should be typed in capital and lower case letters, on white paper, 216 × 279 mm (8 × 11 in), or A4 (212 × 297 mm). Arrange components in the following order: title page, abstract, text, references, tables in numerical sequence, and figure legends. Begin each component on a separate page. Number all pages consecutively, starting with the title page.

Title page
The title page should contain the following:
1. Main title, subtitle (if any) and a maximum of 5 index words (or phrases).
2. Authors listed in the form and order in which they are to appear in the published article.
3. Institutional affiliation for each author, in a footnote on the title page of the article. The institutions listed should reflect the affiliations of the authors at the time of the study, not their present affiliations, if they differ.
4. Financial support information. Include the grant number, if any, and the granting agency. Other financial support, such as that for equipment and drugs, should also be listed.
5. Name, address, e-mail and telephone number of author responsible for correspondence.
6. The number of words in the manuscript, exclusive of the abstract, references, tables, figures, and figure legends.

Abstract
Abstracts for articles are limited to 250 words; those for Brief Reports, to 150 words. Authors of original research articles are asked to submit a structured abstract organised into the following categories (where relevant): Objective(s) Design setting Patients Intervention (if any) Measurements Results Interpretation

Authors are asked to see papers in any recent issue of the British Medical Journal or Annals of Internal Medicine for guidance on structuring the abstract.

Headings in text
Use only three levels of headings in the text. Clearly indicate the levels of headings by using different typographic conventions (such as all capital letters or bold type) or by positioning (flush to margin, indented). Keep headings short (three or four words).

Style
The British Medical Journal, Lancet and Annals of Internal Medicine are recommended to authors as guides to style, clarity of presentation and conciseness.

Units
Use SI units throughout [2], except for systemic arterial blood pressure and haemoglobin content. Other units may be given in parentheses. Use only arabic numbers.

Name of drugs and instruments
Generic names must be used for all drugs. Include the proprietary name only if it is needed for a specific purpose. Instruments may be referred to by proprietary
name, giving the name and location of the manufacturer in the text in parentheses.

References

Number references in the order in which they are first cited in the text. Use superscripted arabic numerals in the text. Note that the SLJN requires the COMPLETE name of journal (and not its abbreviation), year, volume and first and last page numbers.

The reference list should not include unpublished material. Symposium papers may be cited from published proceedings; oral presentation of a paper at a meeting does not constitute publication. References to articles or books accepted for publication but not yet published must include the title of the journal (or name of the publisher) and the year of expected publication. Unpublished work (personal communication, papers in preparation) may be cited by inserting a reference within parentheses in the text; authors must submit a letter of permission from the cited persons to cite such communications.

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Journals: List all authors when 4 or fewer; when 5 or more, list only the first 3 and add et al.

1. Standard article.

2. Corporate author.
   The Royal Marsden Hospital Bone Marrow Transplantation Team. Failure of syngeneic bone marrow graft without preconditioning in posthepatitis marrow aplasia. Lancet 1977; 2: 242-4.

3. Special format.

Books: List all authors or editors when 4 or fewer; when 5 or more, list only the first 3 and add et al.

1. Author.

2. Editors.


Other citations in Reference List:

1. In press (must have journal title).

2. Magazine article.

In-text citations of unpublished material (to be placed within parentheses):

1. Personal communication.
   (Strott CA, Nugent CA. Personal communication).

2. Unpublished papers.

Tables

All tables must be typed double-spaced. Tables should be numbered with arabic numerals, in the order in which they are cited in the text. A table title should describe concisely the content of the table.

Figures

Figures should be professionally drawn or prepared using a computer and high-resolution printer. Lettering should be uniform in style. Free hand or typewritten lettering is not acceptable. Number the figures in the order in which they are cited in the text. Photomicrographs should have scale markers that indicate the degree of magnification. Submit three glossy prints of each figure. Indicate on a label the name of the first author of the paper, the figure number, and the top of the figure: then paste the label on the back of the figure. Do not mount figures on backing board.

Colour figures may be submitted and will be published if essential.

Legends for figures

Reduce the length of legends by using partial sentences. Explain all abbreviations and symbols on the figure, even if they are explained in the text. Stain and magnification should be given at the end of the legend for each part of the figure. If there is no scale marker on the figure, the original magnification used during the observation should be given, not that of the photographic print.

Acknowledgements

Acknowledge only persons who have contributed to the scientific content and provided financial or technical support. Authors must submit written permission from persons acknowledged for other than financial or technical support.

References
