antibodies were, however, present and a labial salivary gland biopsy showed numerous lymphoid aggregates consistent with a diagnosis of the sicca syndrome. The patient subsequently admitted to rather dry eyes, and had moderate caries. A Schirmer’s test confirmed poor tear production.

This patient provides another example of an association between the sicca syndrome and inflammatory polyneuropathy, and lends further support to the hypothesis that, like the sicca syndrome and systemic lupus erythematosus with which they are also associated, the inflammatory polyneuropathies have an auto-immune basis. Our patient developed a clinical neuropathy at a time when symptoms of the sicca syndrome were minimal. This finding illustrates the value of maintaining a high index of clinical suspicion, and of performing a serological auto-antibody screen in patients suspected of having an inflammatory polyneuropathy, but in whom there are no overt features of an associated disease. By this means, associated auto-immune disorders may be uncovered prior to the development of serious manifestations.

Serotal pain with testicular jerking: an unusual manifestation of epilepsy

Sir: I was interested in Dr Bhaskar’s description of a man presenting with focal epilepsy in his scrotum. I am afraid it has been described before, but only to the students and junior staff of Dr Michael Kremer. Dr Kremer is at present unable to relate the story himself, but he described a man whom he saw during the 1939–45 war who had a bullet wound to the brain. Bullets in those days created damage only between entry and exit wounds, and he survived the experience to present with a focal painful epilepsy affecting one half of his scrotal sac.

We told him at the time he should have published it.

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Thymoma without myasthenia gravis: electrophysiological study after thymectomy

Sir: Martinez and Jimenez reported a patient with thymoma without clinical evidence of myasthenia gravis, in whom single-fibre EMG (SF-EMG) studies demonstrated abnormal neuromuscular transmission. We had previously reported increased jitter on SF-EMG studies in three patients with thymoma, none of whom had clinical myasthenia gravis. One patient had a malignant thymoma with red cell aplasia (table, Pt 1) but there was no weakness on examination. Acetylcholine receptor antibody (AChR-Ab) levels were normal and jitter was increased in the extensor digitorum communis (EDC) muscle. After receiving prednisone for treatment of anemia, he developed mild weakness of ocular and shoulder muscles which improved after edrophonium. SF-EMG studies performed on four occasions over a 6 week period demonstrated persistently abnormal neuromuscular transmission (table). He died of infection 2 months after his initial SF-EMG study.

Table Details of patients

<table>
<thead>
<tr>
<th>Patient Diagnosis (Normal value)</th>
<th>Date</th>
<th>Mean MCD (µSec) (&lt;34)</th>
<th>% Fibre pairs with Blocking</th>
<th>Normal jitter (&gt;90%) AChR-Ab (nMoles) (&lt;0-1)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Malignant thymoma, red cell aplasia</td>
<td>10/7/76</td>
<td>43</td>
<td>11%</td>
<td>83%</td>
</tr>
<tr>
<td>2. Benign thymoma</td>
<td>7/13/77</td>
<td>49</td>
<td>12%</td>
<td>76%</td>
</tr>
<tr>
<td>3. Probable thymoma</td>
<td>5/13/78</td>
<td>43</td>
<td>0</td>
<td>91%</td>
</tr>
</tbody>
</table>

In the second patient (table, Pt 2), a mediastinal mass was discovered on routine chest radiography and a benign, encapsulated thymoma was removed. The patient had no weakness on examination before surgery or six months later. He had had no symptoms of myasthenia gravis 10 years after surgery. AChR-Ab was normal. Jitter was increased in the EDC before surgery and miniature end-plate potential amplitude was decreased in an intercostal muscle biopsy obtained at thymectomy. Jitter was still increased in the EDC three weeks after surgery. Six months after surgery there was a decremental response to stapled reflex fatigue testing, and this was partially reversed after the administration of edrophonium.

The clinical manifestations of myasthenia gravis may be subtle and may not be detected unless carefully sought. These patients demonstrate that neuromuscular transmission may be abnormal in patients with thymoma even when clinical examination is normal. We feel that all patients with thymoma should be suspected of having myasthenia gravis and should be evaluated for this possibility before surgery. Since the history, physical examination, AChR-Ab level and repetitive nerve stimulation may all be normal in such patients, increased jitter may be the only evidence of abnormal
neuromuscular transmission. Our experience, like that of Martinez and Jimenez, indicates that these patients may never demonstrate clinical manifestations of myasthenia gravis, thus therapeutic decisions should be made based on clinical findings, rather than EMG abnormalities.

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Matters arising

Stridor during dystonia phases of Parkinson’s disease

Sir: Doctors Corbin and Williams’ report of respiratory stridor co-occurring with limb and orofacial dystonia in two patients with idiopathic Parkinson’s disease is noteworthy. However, their statement that “…spasmodic dysphonia is now recognised as being a form of focal dystonia of the laryngeal muscles” (p. 821) needs clarification.

The cardinal signs of spas tic (spasmodic) dysphonia, principally (1) intermittent or regular adductor voice arrests secondary to vocal fold/laryngeal hyperadduction, (2) moments of strained, effortful vocal quality interspersed within apparent normal phonation, and (3) intermittent or regular breathy moments secondary to adductory glottal arrests may represent psychogenicity, essential tremor and, as implied by Corbin and Williams, other disorders of movement.

Co-occurring impairment of one or more of the other components of motor speech, namely articulation, resonation, respiration, and prosody (speech rhythm) is consistent with dysarthria rather than a focal laryngeal disorder.

The response of spasmodic dysphonia to treatment seems to depend upon aetiology, type of disorder (adductor, abductory, mixed), and mode of therapy.

For those types of spasmodic dysphonia for which a recognisable aetiology cannot be identified, the term “idiopathic spasmodic dysphonia” has been recommended along with regular follow-up, which may eventually reveal an underlying substrate for the disorder.

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References


Corbin and Williams reply:

Sir: Dr Hartman reminds us of the possible mechanisms of vocal cord dysfunction in spasmodic dysphonia; however, in our final paragraph we sought to emphasise the need for clinicians to consider dystonia as a cause of what may otherwise be an enigmatic disorder; we refer not only to some cases of spasmodic dysphonia but also to problems such as piano-player’s dystonia and the occupational dystonias. Many such cases were previously classified as hysterical. Dr Hartman’s policy of accepting that some cases of spasmodic dysphonia lack recognisable aetiology is safer in that it does not deny the possible existence of an organic cause.

References


Book reviews


This is the fifth monograph in a series on management and treatment in specialities in medicine and serves as a perfect riposte to those who would believe that the neurologist has a major role in diagnosis but only a minor role in therapy. It contains a practical approach to major neurological disorders which is spiced by the pharmacological expertise of Dr Jenner in explaining not only when, but why, a particular agent should be prescribed.

The initial chapter is a review of the actions of drugs on the nervous system and briefly considers the role of the blood brain barrier. It contains useful tables of drugs which are agonists and antagonists for the various neuro-transmitter receptors and examples of compounds which act upon specific receptor sub-types. There follow chapters on each of the common neurological disorders with the appropriate therapy and, where known, a summary of their mode of action. The authors’ own interest and expertise in the treatment of movement disorders and sleep disorders is apparent in these, the best, chapters in the book and the whole provides a useful practical guide to logical therapy in neurological disorders. Inevitably the particular biases of the authors are revealed and not everyone will agree with their suggestion that steroids have a role in ischaemic stroke. Indeed the authors seem somewhat uncertain themselves in that on one page they state “it may therefore be beneficial to treat focal oedema around an infarct using osmotic diuretics or steroids” and on the next page “post-infarction oedema does not respond to steroids”.

In the chapter on infections it would have been useful to have a suggestion as to the most reasonable combination therapy in the infant, adult or aged patient presenting with a presumed but unidentified or partially treated bacterial meningitis rather than the bald statement that therapy “depends on isolation of the causative organism”. No
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D B Sanders and J F Howard, Jr

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