LETTERS TO THE EDITOR

Retro-ocular headache with autonomic features resembling “continuous” cluster headache in lateral medullary infarction

Headache is a common, although underemphasised, complaint of lateral medullary infarction. More than half of patients with posterior inferior cerebellar artery (PICA) infarcts develop headache. Fisher found headache in 22 out of 41 (54%) patients with lateral medullary infarction and, more recently, Kuwabara and Hirayama reported headache in 26 out of 34 (76%) patients with Wallenberg’s syndrome.1,2 Less than 5% of patients with PICA infarcts, however, develop periorcular, hemicranial headache and although sympathetic dysfunction in the form of Horner’s syndrome, is a well known manifestation of lateral medullary infarction,1,3 signs of parasympathetic overactivity, such as lacrimation, eye injection, and nasal congestion, have never been described in lateral medullary infarction. Here we report on a patient with a lateral medullary infarction who developed anterior hemiscleral pain accompanied by severe and persistent autonomic parasympathetic activation.

This heavy smoking 37 year old man came to our hospital due to acute vertigo, lack of coordination of his left limbs, numbness in his right limbs, dysarthria, and dysphasia. An angiographic study performed in another hospital 3 months earlier due to intermittent claudication and decrease in left radial pulse had disclosed atherosclerotic changes in the lower limbs and in the left subclavian arteries. General examination showed reduced pulses in the left radial and right dorsalis pedis arteries. Abnormal signs at neurological examination included hiccup, nystagmus, left Horner’s syndrome, and facial hypoaesthesia, left temporal weakness, right hemicorporal hypotonia, and facial hypoesthesia, left velopharyngeal weakness, right hemihypertrophy, and left palatine weakness, right hemihypertrophy, and left palatine weakness, right hemihypertrophy. Cranial MRI showed an acute infarction restricted to the left PICA territory (fig 1), and an occlusion of the left vertebral artery, with no sign of dissection, was seen on angiography. He was treated with heparin and then oral warfarin.

From the beginning of his clinical picture this patient complained of two clearly differentiated head pains. The first one was located in the right occiput and progressively disappeared during the first week after the acute stroke. The second was a very disturbing, continuous headache located anteriorly, mainly in the left retro-orbital and temporal region. This pain was described as moderate to severe, steady, or boring and constantly accompanied by ipsilateral conjunctival injection, lacrimation, and nostril blockage or rhinorrhea (fig 2). The patient also had one or two dramatic daily exacerbations of unbearable pain intensity together with an increase in autonomic symptoms and signs lasting about 2–4 hours. The pain did not significantly respond to either oral or intravenous analgesics (paracetamol, aspirin, metimazol, and NSAID). Verapamil, 240 mg daily, plus sodium naproxen, 1100 mg daily, slightly reduced the pain for 2 or 3 weeks. Medications containing ergotamine, methysergide, and agonists of the 5-HT1B/D receptors were not prescribed. The pain remained unchanged for 4 months and then it began to progressively improve and disappeared 6 months after the stroke.

To the best of our knowledge, this is the first reported patient with a lateral medullary infarction with unilateral anterior headache, sympathetic dysfunction, and parasympathetic autonomic activation, all this resembling a “continuous” cluster-like headache syndrome. The pathophysiology of trigeminal autonomic cephalalgias, including cluster syndrome, is largely unknown.4 It has been proposed, on anatomical grounds, that an inflammatory process in the cavernous sinus, as a point of intersection of the first division of the trigeminal nerve, as anterior pain cannot be explained by trigeminal pericranial pain. This is compatible with the hypothesis that the pain is triggered by the direct central generators giving rise to trigeminalovasovascular system activation—the hypothalamus for idiopathic cluster headache and the established lesion of the key structure of this lesion in the trigeminal nucleus caudalis, in this case.

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Chronic autonomic neuropathy in a patient with primary Sjögren’s syndrome

Several investigators have described the autonomic neuropathy in Sjögren’s syndrome.1 However, only a few have documented the details of dysautonomia, or the pathology of nerves or of other organs such as eccrine sweat glands.2 We report on a patient with Sjögren’s syndrome in whom dysautonomia was the dominant feature, and describe histological findings for the sural nerves and the eccrine glands.

A 39 year old woman visited our hospital because of frequent fainting attacks and diminished sweating. Her history indicated attacks of dizziness on standing since the age of 27. At the age of 30, she experienced a fainting attack on standing. At the age of 36, she noticed dry eyes and focal loss of sweating on the left forehead. She developed amenorrhea at the age of 27. The family history was unremarkable.

She was 164.5 cm tall and weighed 36 kg (body mass index 13.4). Neurological examination showed bilateral ptosis and isocoric pupils (3 mm in diameter; measured using infrared photography) with an irregular margin. The light reflex was absent and the accommodation reflex was tonic. Muscle tone and power were normal. The tendon reflexes were absent and plantar responses were flexor. Sensation and coordination were normal. The skin was generally dry, and spontaneous sweating was present only over the right forehead and T-10 and T-11 dermatomes in hot circumstances. The urinary system was normal. Results of routine laboratory examinations, including urinalysis, and oral glucose tolerance test, and thyroid function tests were normal. Urinary porphobilinogen (0.5 mg/day), δ-aminovaleric acid (1.8 mg/day), and serum vitamin B12 (679 pg/ml) were normal. The mean ocular concentration of γ-2-globulin and α2-microglobulin was increased (24.9%). Serum antinuclear antibody (speckled type160), anti-SS-A, and rheumatoid factor were positive. Her complement and serum reaction potentials in the extensor digitorum brevis were 1.90/0.77 mV (right/left). Sensory nerve action potentials in the bilateral median, ulnar, and radial sural nerves were 3.73/3.54 μV, 3.74/6.52 μV, and 1.47 μV, respectively. The sensory nerve conduction velocities were 33.1/39.0 m/s, 29.8/32.7 m/s, and 14.7 μV, and their sensory nerve conduction velocities were 3.73/8.54 μV, 3.74/6.52 μV, and 1.47 μV, respectively. The sensory nerve conduction velocities were 33.1/39.0 m/s, 29.8/32.7 m/s, and 14.7 μV, respectively. Distal latencies for median and ulnar motor nerves were 3.24/ 3.02 ms and 2.46/2.50 ms. Brain MRI showed no abnormality.

The pupillary response to 2.5% methacholine chloride showed hypersensitivity; the miotic ratio was 43% (3.0 to 1.7 mm); there was also hypersensitivity (3.0 to 9.0 mm) to 1% atropine and 2% pilocarpine (epinephrine) and a slugish response to tyramine. The density of active eccrine glands on the dorsal surface of the right foot shown by iontophoretically applied pilocarpine (1%) was 19 cm² (age matched controls: 141–277 cm²). Staining with rose bengal disclosed erosion. Results of Schirmer’s test and a chewing gum test were 2 mm and 8 ml respectively. Cardiographic R-R interval showed an expiration-inspiration ratio of 1.03. In a cold pressor test, her blood pressure did not change (112/80 to 112/80 mm Hg). In a head up tilting test at an angle of 60 degrees, her blood pressure decreased from 122/74 to 84/44 mm Hg without the development of an increased heart rate (79 to 79 beats/min) and plasma noradrenaline (norepinephrine) concentration (58 to 52 pg/ml). In myocardial 111*I-metabolobenzylguanidine (MIBG) scintigraphy, the heart to mediastinum ratio was 1.52. Histological examination of the left sural nerve showed no degenerating abnormality on teased fibre analysis. In epon embedded sections of the left sural nerve (toluidine blue staining ×1200 for light microscopy and ×7600 for electron microscopy), the densities of small myelinated fibres, large myelinated fibres, unmyelinated fibres, and denervated Schwann cell clustering were 2591/mm² (mean (SD) 2591/mm² (3527 (889)), 7294/mm² (27 866 (5820)), and 5230/mm² (796 (687)), respectively. There were no onion bulbs, infiltrating inflammatory cell cells (haematoxylin and eosin). Light microscopic examination of the eccrine sweat glands of the lower postero lateral aspect of the left leg showed atrophic changes of the glands, but no infiltrating inflammatory cells (haematoxylin and eosin). Electron microscopic examination showed a reduction of the perimeter and area of the transverse profile of secretory coils compared with the controls, indicative of atrophy of the glands (>2591/mm²) and there was a marked reduction in the number of nerve terminals and unmyelinated axons around the secretory coils (<12 500, table). Histological analysis of the minor salivary glands of the lip mucosa showed focal lymphoctic sialoadenitis with degeneration of the ductular epithelium (haematoxylin and eosin), suggesting coexistence of Sjögren’s syndrome.

The patient was treated with oral prednisolone (40 mg/day) and 1-3cho-4, 4-dihydroxyphenylserine (200 mg/day). The dose of prednisolone was tapered to 20 mg/day. Fainting attacks on standing disappeared after the treatment. Our patient had bilateral tonic pupils, bilateral Horner’s syndrome, hyporeflexia, bilateral polyneuropathy associated with primary Sjögren’s syndrome. A case with coexisting Adie’s syndrome. This paper reports a subtype of polyneuropathy with Sjögren’s syndrome. The marked reduction in the density of unmyelinated fibres seen in our patient may reflect damage of the postganglionic sympathetic efferent projection. We showed the severe degeneration of postganglionic sympathetic cholinergic nerves innervating the eccrine glands by histopathological morphometric analysis. Examination of the eccrine glands would provide further insight for assessment of autonomic function in patients with anhidrosis.

We thank Dr Shinti Oono, Department of Ophthalmology, Saga Medical School, Japan, for his help and advice in the examination of pupils.

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C1/C2 rotary subluxation due to spasmatic torticolis

Rotary subluxation of the atlantoaxial complex is encountered more often in children than in adults. It is usually associated with a clear history of cervical trauma, upper respiratory infection, recent head or neck surgery, or rheumatoid arthritis. This paper reports on an adult presenting with C1/C2 rotary subluxation in whom all of these causes were absent. The subsequent clinical course...
showed that the patient in fact had spasmodic torticollis which we think was the cause of the rotary subluxation at C1/C2.

A 37 year old man presented in November 1997 with a history of an insidious onset of progressive deformity of the neck associated with spasms and tenderness of the left sternocleidomastoid and trapezius muscles and occipital pain from April of that year. He had had mild neck pain for several years. The occipital pain was left sided and became increasingly severe. Electromyography was not performed. He had, been off work for 6 months and found it increasingly difficult to sleep in a comfortable position. The only relevant history was one of anxiety attacks. On examination there were no signs of rheumatoid arthritis. The patient had a classically “cock robin” deformity with his head tilting to the left and turning to the right. This was associated with spasms and tenderness, but no obvious hypertrophy, of the left sternocleidomastoid and trapezius which was thought to be voluntary as it subsided when he was relaxed. Plain radiography of the atlantoaxial region was essentially normal and the left sided atlantoaxial subluxation was not apparent. Pelvis and knee examination showed persistence of the “cock robin” deformity. Spinal CT in the neutral position confirmed a C1/C2 rotary subluxation which reduced with the head turned towards the left but was exaggerated when turning to the right. There was no history of recent neck injury, rheumatoid arthritis, or pharyngeal infection and thus a cause for the C1/C2 rotary subluxation was not apparent at that stage. In view of the severity of the pain surgical stabilisation of C1/C2 was suggested.

The patient was placed in halo-traction for a week, and further CT was performed. This showed significant improvement but not total correction of the rotary subluxation. As reduction was not total, it was decided not to perform transarticular screw fixation of C1/C2 but a posterior modified Gallie fusion was performed instead. He had a significant reduction in pain and spasm. A halo-vest was applied. Check radiography was satisfactory and the patient reported a very pleasing relief of the pain and spasm which he had preoperatively. The halo-vest was maintained for 10 weeks during which his pain and spasm reduced, possibly due to the extra somatosensory input from the halo (a mechanical geste antagonique). Removal of the halo was followed by recurrence of the dystonic spasm but the occipital neck spasm was present due to the stabilisation of the atlantoaxial complex. In this case it was not possible to determine at what stage the rotary subluxation occurred. It is possible that the subluxation was the primary event leading to the dystonia which developed over time. Surgery and application of the halo abolished the occipital pain and the spasm was reduced, during the length of time the halo was in place. The evolution of the clinical findings with relief of occipital pain and “cock robin” deformity followed by a more typical appearance of the neck and muscle spasm—a type of “post-traumatic” dystonia. However, in patients with atlantoaxial rotary subluxation, the normal neck deformity is the classic “cock robin” deformity and activation of sternocleidomastoid and trapezius does not occur. The surgical treatment in this case resolved the “cock robin” deformity and occipital pain but the typical clinical findings of spasmodic torticollis reappeared once the halo was removed. It is most likely that the halo provided sufficient somatosensory input to inhibit the sternocleidomastoid spasm during the length of time the halo was in position. The occurrence of clinical findings with relief of occipital pain and “cock robin” deformity followed by a more typical appearance of spasmodic torticollis strongly suggest that it was the dystonia which caused the rotary subluxation.

Spasmodic torticollis is a focal and usually idiopathic dystonia of the cervical muscle spasm causing involuntary neck posturing and movement. It can occur at any age. Chemical denervation of the overactive muscles with botulinum toxin is now the usual treatment and is effective in most patients. Dystonia can cause subluxation or dislocation of the joints. For instance, the temporomandibular joint can undergo recurrent or chronic dislocation in idiopathic or tardive oromandibular dystonia. Angeli et al described subluxation of the subaxial cervical spine resulting in a cervical myelopathy in a child with spastic dystonic cerebral palsy and Tunkel et al reported cervical subluxation causing improvement in the dystonia in a patient with longstanding idiopathic tor- son dystonia. To our knowledge adult onset spasmodic torticollis presenting as a rotary atlantoaxial subluxation has never been reported in the literature. Prolonged rotation beyond the physiological limit is likely to be the cause of this subluxation.

When atlantoaxial subluxation appears after prolonged involuntary neck posturing an underlying diagnosis of dystonia should be considered. Botulinum toxin will not resolve the subluxation, but was necessary in this case to control the underlying dystonia. Externally braces and collars rarely control the forceful movements of cervical dystonia, and the toxin may take some days or even weeks to work, so we recommend treatment as soon as dystonia is diagnosed. In theory botulinum toxin might enhance the effect of or interfere with recovery from acute muscle paralysis due to Guillain–Barré anaesthesia. However, no such reactions have been reported in 12 years of extensive experience worldwide with botulinum toxin, and therefore it is probably safe to give injections even before neck surgery. When a patient with spasmodic (variable posturing) torticollis develops a fixed and sufficiently extreme “cock robin” posture, the clinician should consider investigation by plain radiography and CT to exclude rotary subluxation, even if the muscle spasm is intermittent.

Dystonia can cause subluxation or dislocation at various joints. For instance, the temporomandibular joint can undergo recurrent or chronic dislocation in idiopathic or tardive oromandibular dystonia. Angeli et al described subluxation of the subaxial cervical spine resulting in a cervical myelopathy in a child with spastic dystonic cerebral palsy and Tunkel et al reported cervical subluxation causing improvement in the dystonia in a patient with longstanding idiopathic torsion dystonia. To our knowledge adult onset spasmodic torticollis presenting as a rotary atlantoaxial subluxation has never been reported in the literature. Prolonged rotation beyond the physiological limit is likely to be the cause of this subluxation.

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Ataxic form of Guillian-Barré syndrome associated with anti-GD1b IgG antibody

Richter et al proposed an ataxic variant of Guillian-Barré syndrome, in which patients have severe ataxia of the cerebellar type at the onset of Guillian-Barré syndrome but no ophthalmoplegia or severe loss of proprioceptive sense. Patients with ataxic Guillian-Barré syndrome have distal paresthesias, are ataxic, and raised CSF protein concentrations. Kusunoki et al reported that of 149 patients who had anti-GQ1b IgG antibodies without profound weakness, five had acute self limited ataxia without ophthalmoplegia. The nosol-ogy of these patients, however, was not discussed. Of our 340 consecutive patients who had anti-GQ1b IgG, six had no external ophthalmoplegia and one had minimal exter- nal ophthalmoplegia. Of the patients with seven of these seven anti-GQ1b-positive patients were consistent with an “ataxic form of Guillian- Barré syndrome” (Yuki et al, unpublished observations). Tentative diagnoses made by the primary physicians were Guillian-Barré syndrome associated with anti-GD1b IgG antibody.
The outcome of tuberculous meningitis is influenced by the stage of disease at the start of treatment. Initiation of antituberculous therapy is often delayed because of the inaccuracy of currently available laboratory tests. Management of patients with possible tuberculous meningitis would thus be advanced by the development of an accurate, reliable, and rapid diagnostic test, particularly if it could be applied in settings with poor resources.

Analysis of adenosine deaminase isoenzyme-2 (ADA) in cerebrospinal fluid in the diagnosis of tuberculous meningitis

The presence of adenosine deaminase (ADA), an enzyme involved in purine catabolism, exists in at least three forms. ADA is a monomeric protein with a molecular mass of approximately 35 kDa and two ADA molecules joined via a connecting protein form the dimeric immunoglobulin. Serum samples from patients for whom acute cerebellar ataxia or acute cerebritis is diagnosed should be tested for anti-GD1b and anti-GQ1b IgG antibodies in order not to overlap cases of ataxic Guillain-Barré syndrome.

Acute cerebellar ataxia and polyneuropathy associated with IgG anti-GD1b antibodies have been reported in a patient with Guillain-Barré syndrome who had prominent sensory ataxia and in a patient who had acute distal areflexia and areflexia after upper respiratory tract infection. Whether the presence of monospecific anti-GD1b IgG is correlated with a particular clinical condition, therefore, is uncertain. By contrast, a patient with cerebellar ataxia and polyneuropathy, who has been reported to have IgM M-protein to GD1b, GM1, and asialo-GM1. Both the monoclonal IgM with anti-GD1b activity and murine monospecific anti-GD1b monoclonal antibody bind to the human cerebellar granular layer. The binding of anti-GD1b IgG to the cerebellar granular layer or spinocerebellar 1a fibres in the peripheral nerves may have produced the patient with cerebellar ataxia reported by Araki et al and our present patient.

A 55 year old man had a cough and nasal discharge that disappeared after a few days. After resolution of this illness, he noted paraesthesia in his fingers and toes (day 1) which worsened, and he developed an unsteady gait on day 3. He was apathetic and fully conscious. Blepharoparalysis was absent. Ocular movement was not limited, but smooth pursuit was saccadic. His pupils were normal, and light reflexes present. Neither facial nor oropharyngeal palsy was present. Limb weakness was insignificant. Deep tendon reflexes were absent. Babinski’s sign was absent. Finger to nose and heel to knee tests were normal and uncoordinated. Romberg’s sign was negative, but standing in a tandem position was unsteady. Tandem gait was impossible, and assistance was needed to walk. Posture of the body and standing type were present. There was no impairment of pinprick, touch, position sense, or vibratory sensation. Autonomic nervous function was normal except for hyperhidrosis in the palmar and plantar surfaces. On days 3, 4, 6, 8, and 10, he underwent immunosuppression treatment. The neurological signs rapidly disappeared. Motor and sensory nerve conduction values were normal on days 7 and 21. Protein in CSF was 32 mg/dl on day 3 and 60 mg/dl on day 10 with normal cellularity. On day 24 he was discharged without clinical signs, but still with mild paraesthesias in the right fingers. No external ophthalmoplegia was recorded. However, during the course of the illness the patient developed an unsteady gait on day 3.

Acute cerebellar ataxia had been tentatively considered, but the ataxic form of Guillain-Barré syndrome, as proposed by Richter,1 could be diagnosed. This is important because some patients in whom acute cerebellar ataxia has been diagnosed may have ataxic Guillain-Barré syndrome, and they could benefit from undergoing established treatment for this syndrome—namely, plasmapheresis or aavenous administration of interferons. Serum samples from patients for whom acute cerebellar ataxia or acute cerebritis is diagnosed should be tested for anti-GD1b and anti-GQ1b IgG antibodies in order not to overlap cases of ataxic Guillain-Barré syndrome.
Data were analysed using EpInfo 6.04 (CDC, Atlanta) and PRISM 2.01 (GraphPad Software, USA). Continuous variables were compared using analysis of variance (ANOVA) and a 5% level of significance was used.

Comparison of total CSF and serum ADA and CSF ADA, in the diagnostic categories is shown in the table. Total CSF ADA was highest in patients with tuberculous meningitis. Using a cut-off of ≥6 U/l, the test was 90.9% sensitive in detecting tuberculous meningitis (10 of 11). The specificity was 94% (47 of 50) in all patients and 77.3% (17 of 22) compared with those with cryptococcal meningitis or acute bacterial meningitis. There were no significant differences between those with tuberculous meningitis established by culture and probable disease. Similarly, there were no significant differences in the CSF ADA concentrations in HIV positive and negative patients in the acute bacterial meningitis (mean 4.88 U/l vs 3.71 U/l; p=0.49) and normal lumbar puncture group (mean 0.74 U/l vs 0.12 U/l; p=0.14).

Serum ADA concentrations were highest in patients with tuberculous meningitis or cryptococcal meningitis and significantly lower in those with acute bacterial meningitis.

There were significant differences in the mean proportion of total ADA that comprised ADA, in patients with tuberculous meningitis, cryptococcal meningitis, or acute bacterial meningitis (p<0.001). Using a cut-off of 80% for the proportion of CSF ADA, the test was 100% sensitive and 86.4% specific in detecting tuberculous meningitis (positive predictive value 100%). An ADA of >80% was found in three of nine of those with cryptococcal meningitis and none with acute bacterial meningitis. A cut-off of >90% changed the sensitivity and specificity to 36.4% (4 of 11) and 95.5% (21 of 22) respectively.

The diagnosis of tuberculous meningitis remains a challenge. Acid fast bacilli are typically identified by microscopy in less than a quarter of patients and mycobacterial cultures with limited resources. Measurement of the use of the laboratory technique for measuring ADA2 produces results rapidly, thus potentially differentiating the of 22) compared with those with cryptococcal meningitis or acute bacterial meningitis. The use of CSF ADA2, in the diagnosis of tuberculous meningitis should be further evaluated in larger series, including patients with other lymphocytic meningitides and different settings.

We were able to differentiate patients with tuberculous meningitis from those with aseptic meningitis or a normal lumbar puncture on the basis of the total CSF ADA. However, there was overlap between patients with tuberculous meningitis and those with cryptococcal meningitis or acute bacterial meningitis. A proportion of ADA, isoenzyme of >80% seems to be a reliable marker of tuberculous meningitis, yielding a sensitivity of 100% and specificity of 86.4%. The only other diagnostic category with patients who had >80% ADA2, was cryptococcal meningitis, which is easily diagnosed on Indian ink staining and serology. Serum ADA concentrations were not useful in differentiating the cause of meningitis.

The laboratory technique for measuring ADA, is insensitive (about £1 per test), relatively simple to perform, and can be adapted to an autoanalyser. It may thus be used in laboratories with limited resources. Measurement of ADA, produces results rapidly, thus potentially decreasing delays before therapy for tuberculous meningitis is initiated. These results seem promising and may make a valuable contribution to the early and accurate diagnosis of tuberculous meningitis. The use of CSF ADA2, in the diagnosis of tuberculous meningitis should be further evaluated in larger series, including patients with other lymphocytic meningitides and different settings.

Out of five patients with syringobulbia studied with MRI, ventilatory CO2 response and polysomnography, three had bilateral syringomyelic cavities in the caudal dorsal medulla with unilateral extension to the rostral medulla (figure). This 40 year old patient showed the following respiratory abnormalities: end tidal CO2 47.3 mm Hg; p 0.1, 0.21; ventilatory CO2 response, 1.78 l/mm Hg; apnoea index, 52 events/hour of sleep, with a total number of 212 obstructive sleep apnoeas, four central apnoeas, and 39 hypopnoeas. Maximal duration of obstructive sleep apnoeas was 125 seconds and oxygen saturation values during apnoeic episodes lower than 50%. There was also evidence of severe autonomic dysfunction with orthostatic hypotension, arterial hypotension at rest, and complete loss of sinus arrhythmia. Despite the severity of the respiratory abnormalities recorded, the patient refused to receive respirator support, and to date has not developed any cardiac respiratory complication during a follow up of 9 years.

It seems that extension of the syrinx to ventral and rostral medullary areas may lead to more severe respiratory and cardiovascular abnormalities. Three stages in the progression of syringobulbia may be described in involvement of autonomic and respiratory structures: (1) initial involvement of the
We were interested to read the paper by the authors reply: Morrell et al. We have encountered similar difficulties to those described by Morrell et al in outlining small lesions in the medulla by using MRI. Unless the cervicomedullary junction is studied with thin slices, small cavities may be easily overlooked. In such patients, chest and abdominal movements due to respiratory difficulties contribute to poor MRI definition.

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The authors reply:

We were interested to read the paper by Nogues et al, and we are grateful to the authors for bringing it to our attention. One of the aims of our study was to investigate the effect of unilateral lesions in humans at sites previously defined as being important for chemosensitivity in animals. For this reason we studied patients with relativity small focal lesions in the rostrolateral medulla. Nogues et al have investigated the effect of syringomyelia and syringobulbia on chemosensitivity and breathing during sleep; the MR images of their patients show the lesions to be more extensive than in our study. Nevertheless, both papers highlight the gross sleep apnoea in these patients and the importance of carrying out nocturnal polysomnography to identify any abnormalities in breathing during sleep; this point is emphasised in an excellent editorial by Malow.

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The content covers that dealt with by the neurologist, neurosurgeon, and casualty officer. It should be compulsory reading for those on the front line receiving such emergencies and may then play a part in preventing an emergency becoming a catastrophe. On the other hand, while providing the experienced neurologist with an enjoyable read, hopefully most are already familiar with the up to date and appropriate clinical neurology. For the general practitioner, the evidence, or lack of it, for a management decision is described and referenced. Although American, the principles are transferable and this should in no way deter the English reader.

One is naturally a little weary when asked to review a book entitled Neurologic Catastrophes in the Emergency Department. Why me? springs to mind. This said, swallowing my pride, I thoroughly enjoyed this book. Although the content is predictable enough; ischaemic stroke, haemorrhage, cord compression, infections etc, the presentation is special. It is an extremely accessible book, clearly laid out with key points of management and pharmacology appearing in shaded boxes (called capsules). The book is generously illustrated with informative diagrams (and I refer the reader, in particular, to that explaining assessment of the Glasgow coma scale) depicting procedures and anatomy. There are bountiful radiological images and a few select colour plates of clinical cases.

The text is a monograph but Dr Wijdick’s opinions and practices are clearly stated as such and on the whole the evidence, or lack of it, for a management decision is described and referenced. Although American, the principles are transferable and this should in no way deter the English reader.

For those on the front line receiving such emergencies and may then play a part in preventing an emergency becoming a catastrophe. On the other hand, while providing the experienced neurologist with an enjoyable read, hopefully most are already familiar with the up to date and appropriate clinical neurology. For the general practitioner, the evidence, or lack of it, for a management decision is detailed and referenced. Although American, the principles are transferable and this should in no way deter the English reader.

GILLIAN HALL.

**BOOK REVIEWS**


This is volume 71 in the series of the Handbook of Clinical Neurology. It is part III, the third and last volume dedicated to the neurology of systemic disease and, therefore, updates the previous volumes on this subject,

**CORRECTION**

Davies NP, Eunson LH, Gregory RP, et al. Clinical, electrophysiological, and molecular genetic studies in a new family with paramyotonia congenita. J Neurol Neurosurg Psychiatry 2000;68:504-507. During the editorial process, figure 1 was reproduced incorrectly. The correct figure is shown here:
Their efficacy are that they “reduce attack rates and severity... may be helpful in preventing progression in secondary progressive disease... and have positive effects that are highly significant statistically and clinically”. Surely doctors managing patients with multiple sclerosis will find this 234 page book is apparently addressed, need a more penetrating analysis. Similarly, the authors recommend that all patients with optic neuritis be treated with intravenous steroids to reduce the 2 year risk of multiple sclerosis. They do not say that this advice is based on a controversial interpretation of the Optic Neuritis Treatment Trial and that the 5 year risk of developing multiple sclerosis (in the same study) was unaffected by treatment. Perhaps the next edition will be better.

ALASDAIR COLES


This is the best all-inclusive book on sleep disorders in neurology that I know. It gives clear as well as comprehensive information about the many and common sleep problems that accompany neurological illness. The text is well organised with chapters on sleep symptoms such as insomnia as well as sleep disorders in conditions such as multiple sclerosis and migraine. The forward spells it out - “the British never seem to get bored of discussions about the weather, whilst Americans are equally prepared to discuss their own sleep patterns”. This is fair comment on the cottage industry state of United Kingdom sleep medicine versus the GB (Giga-buck) values of the United States. Indeed, we owe much of what we know about sleep to the American pioneers, who are well represented here. Much of what we know about sleep medicine versus the GB (Giga-buck) money spent on this book is well worth it. Your practice will improve.

DAVID PARKES


The applications of transcranial magnetic stimulation (TMS) have broadened significantly in the past few years and the number of publications involving this technique have increased dramatically. This supplement is very well timed, therefore, and the editors are well known in this field. It is a book of about 360 pages and is published as a supplement to the journal Clinical Neurophysiology.

It is divided into five sections, covering methodology, physiology, clinical neurophysiology, psychiatry and cognition. Most of the authors have written extensively on TMS, so the chapters represent a fairly good distillation of relevant details into brief texts, each accompanied by a set of references, which will lead new readers to pursue their own special interests and allow those who have been involved in TMS for some years to catch up with recent developments. The latter applies in particular to the community of neurophysiologists who have been using TMS in clinical and non-clinical studies of the motor system, who are now interested to see how the technique being applied outside this system.

The various sections include modelling the stimulating field, its haemodynamic effect and role in mapping, the acquisition of single- and multi-site TMS, the relationship between animal and human studies. The paired pulse and rapid rate paradigms are discussed, with clinical, physiological, and pharmacological applications and in the clinical section there is a series of chapters describing several
applications. This section in particular shows how diverse the efforts have been to apply TMS in different ways, but reinforces the point that many of the most prominent advances have been in non-clinical applications. The exception to this is in psychiatry, where a new TMS has become of particular interest for the controversial treatment of selected patients with depression. This and other potential psychiatric applications are described, moving the reader to the final section on TMS and cognition, including its use in the study of language.

I think that the editors have pitched the standard of this book very well, so that it will be of interest to those who have been involved in this field for many years and to all newcomers. There are one or two chapters, unfortunately, which represent work of a slightly lightweight or pilot nature, and there is no group editorial on controversial issues such as the treatment of depression. There will be some hot competitors for this book on the market in the very near future, but I think that its tidy and concise presentation will make it an essential volume an essential brief reference book for most departments using TMS.

SIMON BONIFACE


An up to date atlas of surface anatomy for electrolymography is often useful for someone in training and also remains useful on those occasions when an unusual muscle is to be sampled, and some of the relevant details need to be rekindled. There are one or two old atlases which are no longer in print and this book by Hang and DeLisa provides a useful replacement for these. It is the right size to be handled in a busy clinic and is well laid out, covering the head and upper limb, the lower limb and trunk and pelvis, and head and neck. Information about positioning the patient, inserting the needle, and activating the muscle are provided with notes on innervation and anatomy.

My only reservation is the style of the drawings, which are not always very clear. This is a major failing, unfortunately, particularly when this atlas is compared with one or two of its older rivals. For supervised training or occasional reference, however, I am sure this atlas will remain helpful.

SIMON BONIFACE


Thomas Brandt's masterpiece Vertigo remains mandatory reading for clinicians interested in this symptom. The basic sciences underpinning vertigo, together with the clinical diagnosis and management of all disorders characterised by this symptom, are comprehensively reported, extensively referenced, and beautifully illustrated. The book is detailed and exhaustive in its subject matter. The only area which seems to have been omitted in any depth is that of the cardiovascular causes of dizziness and vertigo.

There is a broad introduction covering the pathophysiology underpinning vestibular disorders, vertigo, dizziness, and falls, the clinical assessment of such a patient; and the management strategies, together with their rationale. Common disorders—namely, acute peripheral vestibular episodes and Menière's disease, are well covered, in addition to the rarer disorders such as the Ramsay-Hunt syndrome, bilateral vestibular failure, and autoimmune disorders associated with vestibular dysfunction. In this section, the author's views on certain disorders are reflected particularly in the chapters on vestibular neuritis and peripheral vestibular paroxysmia, and it is regrettable that a little more of the controversy in the literature is not highlighted. The management section of the Menière's chapter shows little in depth discussion of trials of medication which, with one or two exceptions, have been extremely poorly designed. Moreover, the incidence of hearing loss in association with gentamicin installation has not been emphasised, although surgical treatments in general are well discussed. The chapter on perilymph fistulas is somewhat didactic in the light of many surgeons' views that the validity of this entity must be questioned.

Central vestibular disorders are clearly explained with an excellent and easily understood introduction outlining vestibular disorders in the different planes of action of the vestibulo-ocular reflex. Illustrations and diagrams make some difficult concepts accessible. There are two particularly valuable chapters on the vestibular cortex and its disorders and vestibular epilepsy, both areas that to date have been poorly investigated and understood, but which represent areas ripe for research with new imaging techniques and perceptual tests Brandt then moves on to positional and positioning vertigo which is possibly the “topic of the decade” for those with a vestibular interest, as the introduction of particle repositioning manoeuvres has provided a “cure” for a very common vestibular syndrome although, unfortunately, the underlying science of exactly what we are doing with these procedures remains elusive. The pathophysiology and mechanisms of such disorders are clearly defined and atypical presentations discussed.

Vascular aetiologies are considered in chapters on stroke and the often overlooked association of migraine and vertigo. The overlap between migraine and familial periodic ataxia is also considered, although this latter entity is fully discussed in a separate chapter. For the general clinician interested in vertiginous disorders there are chapters of particular value, including trauma, vertigo in childhood and in elderly people, and vertigo in association with drugs. For the more specialised clinician, the chapter on visual vertigo is of particular value. Importantly, the relation between psychiatric disorders and vertigo is explored in one of the final chapters.

Overall, this is an excellent readable book that can be dipped into for those requiring information about a particular patient, or read cover to cover for those who wish to extend their knowledge in vestibular medicine. Inevitably, in such an author book, idiosyncratic views are revealed, but, that aside, I cannot recommend this book too highly to guide both the experienced and the uninitiated through the minefield of pitfalls that beset the clinician trying to sort out vertigo. The limiting factor will be the price but certainly it is a book, and possibly the book, which should be in every library used by neurologists and otologists.

SIMON BONIFACE

Instant answers—from pocket books to reduced, handy text—this is the theme of our times. Nothing captures this zeitgeist better than the success of the Penguin 60s or in medicine the proliferation of summary or concise texts. These books are exceedingly difficult to do well and require a distinct approach to that of writing a conventional textbook. All too often a summary or concise text is a poorly edited version of a larger tome.

A model example of what a concise specialist book should look like in style and content is provided by David Perkin, in Mosby's Color Atlas and Text of Neurology. This book uses an imaginative combination of illustrations, colour coded boxes which highlight “the take home points”, and bullet pointed narrative to deliver a very readable book. Part of the success of the book is the focus on common conditions with a sufficient nod to minutiae within this. The emphasis on common and treatable is evident throughout with appropriate weighting on risk factors and diagnostic criteria where relevant. Tables and colour illustrations are skilfully deployed around this central point perhaps, but not always done—to complement the text and emphasise the important.

Inevitably a book such as this will not please the purist. This in many ways undoes the very success of the book—notably, to provide a broad overview of neurology suitable for both the undergraduate and MRCP candidate. It is testament to the skill of the author that such a goal is achieved in a deceptively easy to follow and, therefore, easy to remember style. A claim not made by the author—but one that I would venture is that the book would also be valuable to the junior specialist registrar, and to those that teach Neurology.

SIDDHARTAN CHANDRAN