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. Less than 5% of patients with PICA infarcts, however, develop periorbital, hemicranial headache and although sympathetically disinhibited in the form of Horner’s syndrome, is a well known manifestation of lateral medullary infarction, 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 hemicranial 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 dysphagia. 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 including new cases.

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 hypohaeaesthesia, left velopatine weakness, right hemicorporal hypotonia, and ataxia as well as impaired sensation over the right limbs. 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 lepirin 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 rhinorrhoea (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, metamizol, and NSAIDs). Verapamil, 240 mg daily, plus sodium naproxen, 1100 mg daily, slightly reduced the pain for 2 or 3 weeks. Medica
tions containing ergotamine, methysergide, and agonists of the 5-HT1B/D receptors were not prescribed. The pain remained unchanged for some time after it began to progressively improve and disappeared 6 months after the stroke.

To the best of our knowledge, this is the first reported case of 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. 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 and the cranial sympathetic and cranial parasympathetic nerves, would be the pathophysiological explanation for these headaches.

As commented on, headache is a frequent complaint in lateral medullary infarction. The posterior is the most common location for headache, 65% in the series of Kuwabara and Hirayama and 49% in Fisher’s series, which concurs with Wolff’s finding that on stimulating the vertebral artery the pain is referred to an occipital-suboccipital-nuchal area. This posterior pain was also reported by our patient during the first week, probably being related to the thrombus formation in the vertebral artery. The most important pain experienced by our patient, however, was a moderate to severe, unilateral, retro-ocular headache. Anterior pain affecting the eye region is much less frequent in lateral medullary infarction, but was already considered by Fisher as typical of this syndrome and probably resulting from a lesion in the nucleus of the descending root of the trigeminal nerve, as anterior pain cannot be induced on stimulating the vertebral artery and as this anterior headache is usually succeeded by numbness. The central medullary lesion also accounts for the autonomic symptoms and signs seen in this patient. Sympathetic dysfunction (Horner’s syndrome) is a well known sign of Wallenberg’s syndrome due to damage of the sympathetic tract within the PICA infarction and it seems logical to try to explain the parasympathetic activation as secondary to irritation of the adjacent superior salivatory nucleus by the infarct area or to an imbalance between the two autonomic systems.

In conclusion, this patient shows for the first time that all the semiology typical of cluster and other trigeminal-autonomic headaches can be secondary to a pure central lesion, located in the lateral medulla, supporting the contention that flow changes occasionally found in cluster headache cases in the cavernous sinus do not generate the disorder, but are in fact a consequence to the pain. This agrees with the proposal that the activation of the cavernous sinus region does not relate specifically to cluster headache, but it is a trigeminovascular autonomic reflex to first division pain. In fact, the only difference between our case and typical cluster headache—the periodicity of the pain attacks—is easily explained by the distinct central generators giving rise to trigeminovascular system activation—the hypothalamus for idiopathic cluster headache and the established lesion of the key structure of this case, the lateral medullary caudal, in this case.

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4 Moskowitz MA. Cluster headache: evidence for a pathophysiologic focus in the superior perica
Chronic autonomic neuropathy in a patient with primary Sjögren’s syndrome

Several investigators have described the autonomic neuropathy in Sjögren’s syndrome.1,2 However, only a few have documented the details of dysautonomia, or the pathology of nerves or of other organs such as eccrine sweat glands.3 We report on a patient with Sjögren’s syndrome in whom dysautonomia was a dominant feature, and describe morphological 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 included 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 amenorrhoea 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 isometric 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, chest X-ray, oral glucose tolerance test, and thyroid function tests were normal. Urinary porphobilinogen (0.5 mg/day), δ-aminolevulinic acid (1.8 mg/day), and vitamin B6 (679 µg/ml) were normal. The concentration of uric acid in blood was increased (24.9%). Serum antinuclear antibody (speckled type×100), anti-SS-A, and rheumatoid factor were positive. Her complement (CH50, C3, and C4) were normal and their sensory nerve conduction velocities were 33.1/30.9 m/s, 28.6/27.2 m/s, and 30.0 m/s, 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 45% (3.0 to 1.7 mm); there was also hypersensitivity (3.0 to 9.0 mm) to 1.25% epinephrine (epinephrine) and a sluggish response to tyramine. The density of active eccrine glands on the dorsal surface of the right foot showed 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 1.0. 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 an increased heart rate (79 to 79 beats/min) and plasma noradrenaline (norepinephrine) concentration (58 to 52 pg/ml). In myocardial "1"I-metabolodobenzylguanidine (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) 884/mm² in 35 patients (3517 (889), 7294/mm² (27 866 (5820)), and 5230/mm² (796 (687)), respectively. There were no onion bulbs, infiltrating inflammatory cells, and fat cells (haematoxylin and eosin). Light microscopical examination of the eccrine sweat glands of the lower posterolateral aspect of the left leg showed atrophic changes of the glands, but no infiltrating inflammatory cells (haematoxylin and eosin). Electron microscopical 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 in 35 patients). 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 showed focal lymphocytic 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-threo-3, 4-dihydroxyphenylserine (200 mg/day). The dose of prednisolone was tapered to 20 mg/day. Fainting attacks on standing disappeared after the treatment. We had no patient with bilateral tonic pupils, bilateral Horner’s syndrome, hyporeflexia, orthostatic hypotension, abnormal cardiovascular reflexes, reduced lacrimation, and segmental anhidrosis. Examination of the autonomic nervous system disclosed parasympathetic and postganglionic sympathetic injury including sudomotor involvement. Since Ross reported the presence of tonic pupils and progressive sudomotor hypohidrosis in his patient (Ross’ syndrome),4 several reports have confirmed that autonomic dysfunction in Adie’s syndrome may be more widespread than previously recognised. However, severe orthostatic hypotension as seen in our patient is rare. The clinical condition of our patient seemed different from that of acute autonomic neuropathy, because the dysautonomia started insidiously and progressed slowly over many years. Our patient did not have any systemic disease except Sjögren’s syndrome. Peripheral neuropathy with Sjögren’s syndrome may represent a distinct syndrome, in which sensory polyneuropathy predominates but there is some autonomic dysfunction or Adie’s pupils.5 Griffin et al6 described autonomic deficits in patients with ataxic sensory polyneuropathy with Sjögren’s syndrome. In their patients, histological examination of sural nerves showed a preferential loss of large myelinated fibres, which is different from the findings of our patient. We concluded that the diagnosis of our patient was 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 useful insight for assessment of autonomic function in patients with anhidrosis.

We thank Dr Shinji 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 spasmodic torticollis

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

Electron microscopic morphometric evaluation of the transverse profiles of secretory coils in the skin and nerve terminals and unmyelinated axons around secretory coils

<table>
<thead>
<tr>
<th>Control†</th>
<th>Patient‡</th>
</tr>
</thead>
<tbody>
<tr>
<td>Perimeter of transverse profile (µm²/profile)</td>
<td>173 (55)</td>
</tr>
<tr>
<td>Area of transverse profile (µm⁴/profile)</td>
<td>207.7 (1100)</td>
</tr>
<tr>
<td>Nerve terminals (n)</td>
<td>9.1 (4.8)</td>
</tr>
<tr>
<td>Unmyelinated axons (n)</td>
<td>12.7 (6.2)</td>
</tr>
</tbody>
</table>

†Mean (SD) of six control subjects in whom 14–38 transverse profiles of secretory coils in each individual were morphometrically evaluated.

‡Mean of 38 transverse profiles of secretory coils. Minimum–maximum values are shown in parentheses. *Number/100 µm² of perimeter of transverse profiles.
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 severe torticollis 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 classical “cock robin” deformity with his head tilted to the left and turning to the right. This was associated with spasm 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 obtained under a general anaesthetic as the patient was extremely anxious. After the induction of the general anaesthetic, the element of spasm and tilting of his neck which was thought to be due to pain was relieved.

The examination and a 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 history 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 with transarticular screw fixation. 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 several weeks during which his pain and spasm had completely resolved. However, shortly after removing the halo, he had a recurrence of the pain and spasm in the left sternocleidomastoid al though the severe occult spinal pain was still completely relieved. His sternocleidomastoid although the severe deformity present for 10 weeks during which his pain and muscle spasm—a type of “post-traumatic” dystonia. 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 alignment of the neck and thus CT was obtained under a general anaesthetic, the element of spasm and tilting of the neck being thought to be voluntary as it subsided when he was relaxed.

A halo-vest was applied. Check radiography showed no loss of the preoperative deformity and thus CT was obtained under a general anaesthetic. The halo-vest was maintained in the position of maximum reduction.

Beyond this the lateral inferior facet of the atlas rocks over the lateral superior articular facet of the axis. When ataxic form of Guillain-Barré syndrome associated with anti-GD1b IgG antibody Richter et al proposed an ataxic variant of Guillain-Barré syndrome, in which patients have severe ataxia of the cerebellar type at the onset of Guillain-Barré syndrome but no ophthalmoplegia or severe loss of proprioceptive sense. Patients with ataxic Guillain-Barré syndrome have distal paresthesias contralateral to the side of the lesion 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 nosology of these patients, however, was not discussed. Of our 340 consecutive patients who had anti-GQ1b IgG antibodies, six had no external ophthalmoplegia and one had minimal extranausal ophthalmoplegia. The serological findings of these seven anti-GQ1b-positive patients were consistent with an “ataxic form of Guillain-Barré syndrome” (Yuki et al, unpublished observations). Tentative diagnoses made by the primary physicians were Guillain-Barré syndrome.
syndrome (n=3), atypical Miller Fisher syndrome (n=3), and acute cerebellar ataxia (n=1). Araki et al, however, reported on a patient with Guillain-Barré syndrome who had prominent cerebellar signs. That patient had high monospecific anti-GD1b IgG antibody titre and an acute phase of the illness but did not have anti-GQ1b IgG. These findings led us to examine whether some patients in whom acute cerebellar ataxia has been diagnosed have anti-GD1b IgG antibodies.

Serum samples were obtained from 39 patients for whom acute cerebellar ataxia or acute cerebellitis had tentatively been diagnosed. One patient who had associated anti-GQ1b IgG was excluded because the tentative diagnosis of acute cerebellar ataxia was the final one ataxic Guillain-Barré syndrome. Serum IgM or IgG antibodies to GM1, GM2, GD1a, GD1b, GT1b, GT1a, or GT1b were measured by an enzyme linked immunosorbent assay as described elsewhere. GT1b is a possible target molecule for serum antibodies from patients with sensory ataxic neuropathy. Serum was considered positive when the antibody titre was 500 or more. One of the 39 patients had a high anti-GD1b IgG titre of 16 000 but carried no antibodies to the other six gangliosides. His clinical presentation is described later. The other 38 patients had no antibodies to those gangliosides.

Finally, it was important to show that serum from the patient who had anti-GD1b IgG did not react with asialo-GM1, fucosyl-GM1, GM1b, GalNAc-GM1b, GalNAc-GD1a, GD3, GT1a, GT1b, or sulfated glucuronol paragloboside.

A 55 year old man had a cough and nasal discharge that disappeared after a few days. After resolution of this illness, he noted paraesthesias 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 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.

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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% ADA 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 inexpensive (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 ADA in the diagnosis of tuberculous meningitis should be further evaluated in larger series, including patients with other lymphocytic meningitides and different settings.

Data were analysed using EpiInfo 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 26 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 77.5% (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. Similar studies have been performed 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 ADA, in the diagnosis of tuberculous meningitis should be further evaluated in larger series, including patients with other lymphocytic meningitides and different settings.

Unilateral focal lesions in the rostralateral medulla influence chemosensitivities and breathing measured during wakefulness, sleep, and exercise

We have read with interest the article entitled Unilateral focal lesions in the rostralateral medulla influence chemosensitivities and breathing measured during wakefulness, sleep and exercise by Morrell et al, which shows that unilateral ischaemic lesions of the rostralateral medulla may lead to an abnormal ventilatory CO₂ response and sleep apnoea. We have recently conducted a similar study on five patients with syringobulbia. Syringobulbia has a predilection for autonomic nuclei of the cardiorespiratory network localised in the caudal medulla and this may cause severe respiratory and cardiovascular abnormalities. On occasion, a syrinx may extend to the rostral and ventral medulla. Four out of five patients with syringobulbia studied with MRI, ventilatory CO₂ response and polysomnography, one patient with bilateral syringomyelic cavities in the caudal dorsal medulla with unilateral extension to the rostralateral 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 dysfunctions 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 any respiratory support, and to date has not developed any cardiorespiratory 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
caudal and dorsolateral medulla with damage to the nucleus tractus solitarius, vagal motor nucleus, and nucleus ambiguus which may impair cardiovascular reflexes; (2) ventral extension with involvement of the intermediary reticular formation; and (3) further ventral and rostral extension to the anterolateral surface of the rostral medulla, involving vasomotor neurons and central chemoreceptors. These last two stages may be accompanied by severe respiratory abnormalities and arterial hypotension.

We have encountered similar difficulties to those described by Morrell et al1 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,1 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 relatively small focal lesions in the rostralateral 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.1

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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:

BOOK REVIEWS


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 may cater to paramedics; 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 transatlantic and this should in no way deter the English reader.

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 fully acquainted with the up to date and appropriate management of a neurological emergency!

GILLIAN HALL


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,

This is the best handbook on sleep disorders in neurology that I know. It gives as 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 and sleep disorders in conditions such as multiple sclerosis and migraine. The forward spells it out – “the British seem never to get bored of discussions about the weather, whilst Americans are equally prepared to talk about 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 in this book.

It may reflect the parsimonious British sleep medicine to complain that there is too much of the neurosorologist who will do a “sophisticated neurological assessment of the type best coordinated by a clinical neuroscientist” in this book. Surely there is still the foremost need to make a definite diagnosis by clinical history rather than sleep laboratory. I still think that a good psychiatrist is of greater value in sleep medicine than an EEG machine. It is surprising to discuss psychological factors is sadly missed. Still, the science presented is state of the art, although surprisingly there is little emphasis on molecular biology, which is an expanding field of sleep research in 2000. HLA systems get attention, but not clock genes or the many gene determined neurological disorders such as the Prader-Willi syndrome, which have important implications for sleep medicine. The book unfortunately was written before the important discovery of the hypothalamic orexin system involvement in sleep and atonia.

There are a few minor criticisms. The index is useful (no Epsom salt, no modafinil). The references are very comprehensive but not selective (a near universal book fault); the illustrations, which are mainly EEGs and actimetry recordings, are adequate but boring. Meaningless sentences are no more common than in most medical writing, but we could do without “the diagnostic criteria for multiple sclerosis served to identify patients with this condition”, as well as “sleep is an active and integral component of the central nervous system (CNS) and may be one of its most important functions”. Treatment information can be inadequate. There are four lines on modafinil in a book of 422 pages. Physicians who want to know how antiepileptic drugs impinge on sleep are referred to “the many excellent texts available”. What about sleep problems with new dopamine agonists, or after stereotactic surgery? Does melatonin deficiency insomnia in elderly people really exist? How does the idea “chronopharmacological considerations will prove to be of importance in TBI” turn into an exact timetable for giving drugs after head injury?

Despite these problems it is most useful to have authoritative writing about conditions as diverse as fatal familial insomnia and narcolepsy collected into one volume. Special mention must be made of an excellent chapter on insomnia by Lugaresi and his colleagues, discussion about dissociated states of brain and mind by Mahowald and Schenck, as well as Zhadanova’s critical review of the role of melatonin in sleep disorders. There is a diverse outstanding historical summary by Thorpy, and the editor, Antonio Culebras, gives a good description of the biology and neuroanatomy of sleep as well as outstanding chapters on neuromotor disorders, head-aches and multiple sclerosis.

Sleep disorders are important in all areas of neurology. Unless you already have one of the many comprehensive texts on sleep medicine, money spent on this book is well worth it. Your practice will improve.

ALASDAIR COLES


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 all very 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 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 simultaneous MEG, and the many known 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, of course 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 competetors for this book on the market in the very near future, but I think that its tidy and concise presentation will make this a popular volume an essential brief reference book for most departments using TMS.

SIMON BONIFACE


An up to date atlas of surface anatomy for electromyography 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 is not 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 concerning 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, intriguingly, 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 a single 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.

LINDA LUXON


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 an emphasis on common and treatable evident throughout with appropriate weighting on risk factors and diagnostic criteria where relevant. Tables and colour illustrations are skilfully deployed around the page, 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
Retro-ocular headache with autonomic features resembling "continuous" cluster headache in lateral medullary infarction
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