LETTERS
TO
THE
EDITOR

Pseudotumour after arteriovenous malformation embolisation

The association between venous outflow obstruction and the development of pseudotumour syndrome is well known, although the mechanism by which the rise in CSF pressure is brought about is less certain. Although there is much evidence that the manifestations are a result of a disturbance of CSF dynamics, previous reports have focused solely on a disturbance to absorption. We present a case in which it is proposed that alterations in CSF formation, and to a lesser extent absorption, are responsible for the development of the syndrome.

At 2 years of age, patient A presented with a generalised pounding headache for 24 hours after a procedure involving the vein of Galen. Although there was no evidence of cardiac failure or hydrocephalus associated with this, assessment by angiography was advised. This, initially declined by the parents, was undertaken at the age of 5 years when vertigo and intermittent numbness of the left arm and leg had been present for about 12 months.

Angiography showed a deep right temporal lobe arteriovenous malformation consisting of three separate fistulae supplied by the right posterior cerebral and posterior communicating arteries and drained into a large venous varix which subsequently drained into the Galenic venous system. A cerebral blood flow study showed a steal syndrome affecting the right frontoparietal area, and a decision was made to attempt embolisation. Complete occlusion of the fistulae was achieved by transarterial platinum coil embolisation.

The patient complained of right sided headache for 24 hours after the procedure, resolving with minor analgesia. Brain CT the next day was reported as normal. A full ophthalmological examination presented with a normal fundus and fields. Examination 2 days later was reported as normal. A full ophthalmological examination presenting with normal fundi and fields.

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False negative polymerase chain reaction on cerebrospinal fluid samples in tuberculous meningitis established by culture

The polymerase chain reaction (PCR) has been reported to be of diagnostic value when performed on CSF samples in tuberculous meningitis.1–4 Rapid amplification of Mycobacterium tuberculosis specific DNA can result in a detectable signal within 48 hours and can influence treatment decisions.

Recently two patients presented to our hospital with symptoms and signs suggestive of tuberculous meningitis. Examination of CSF disclosed a lymphocytic exudate. Repeated samples were sent to a British referral laboratory where CSF PCR for M tuberculosis was reported negative. Despite this, antituberculous treatment was continued for 12 months and both patients responded clinically. Several weeks after the negative PCR result, M tuberculosis was cultured on Lowenstein-Jensen slopes from CSF taken from both patients. False negative CSF PCR in tuberculous meningitis established by culture has rarely been reported. The two patients are described to emphasise the dangers of overreliance on PCR in cases of suspected tuberculosis meningitis. Premature cessation of treatment would have had tragic consequences for the two patients concerned.

The first patient was a 28 year old Asian man, last in India 8 years previously. He was sent from a clinic to hospital for incision and drainage of two deep seated Staphylococci.
M. tuberculosis can vary when different assays and laboratories are used. Claims that PCR can detect 1–10 M. tuberculosis organisms “in vitro” seems not to be the case in clinical samples such as CSF.

In the two patients presented above adequate volumes and repeated samples of CSF were assayed using suitable primers and appropriate controls at a British referral laboratory. Results for these two patients show the dangers of overreliance on PCR CSF when tuberculosis meningitis is clinically suspected.

We are grateful to Dr Deborah Binzi-Gascogne of the Leeds mycobacterium laboratory, where the PCR tests were performed and who provided additional information for the manuscript.


A novel mutation of the myelin P gene segregating Charcot-Marie-Tooth disease type 1B manifesting as trigeminal nerve thickening

Charcot-Marie-Tooth disease (CMT) is the most common type of hereditary peripheral neuropathy. It is classified into two types based on pathological and electrophysiological findings: type 1 and type 2. CMT type 1 is the most common (75%). CMT type 1 has been mapped to chromosome 17 (CMT1A), chromosome 1 (CMT1B), another unknown chromosome, (CMT1C) and the X chromosome (CMTX). CMT1B is a rare form of CMT1 associated with mutations of the myelin protein zero (P) gene. Mutations in the P gene have recently other clinical specimens, particularly respiratory specimens, have reported that PCR may be less sensitive than culture for the detection of M. tuberculosis and that the low sensitivity correlated with low colony counts on culture.
The six exons of the P. gene were amplified by the polymerase chain reaction using primers, and analysed by single strand conformational polymorphism (SSCP) and sequencing analyses. DNA sequencing of exon 3 showed a novel point mutation (His81Arg) of P. The cranial nerve involvements in this patient may be associated with the novel missense mutation of P. (His81Arg).

A 15 year old Japanese girl presented with CMT disease. She showed delayed motor development. Although she became ambulant at 1 year and 8 months of age, she was never able to run. She was referred to our hospital due to progression of her gait abnormality. Her mentality and higher brain function were normal. Neurological examination disclosed weakness in both proximal and distal muscles of the legs, decreased grasping power, sensory disturbance of distal limbs and feet, and ataxia. Facial sensation, mastication power, and hearing acuity were normal. She also had atrophy of the lower limbs, drop foot, a steppage gait, claw hands and club feet deformities. Optic atrophy, incoordination, autonomic dysfunction, and cardiac involvement were not evident.

In laboratory findings, creatinine kinase was 343 IU/l. A peripheral nerve conduction study showed undetectable sensory and motor action potentials in all limbs. Auditory brain stem response disclosed weakness in both proximal and distal muscles of the legs, decreased grasping power, sensory disturbance of distal limbs and feet, and ataxia. Facial sensation, mastication power, and hearing acuity were normal. The thickness of bilateral trigeminal nerves in CMT type 1, and thickening of the cauda equina, nerve roots, and ganglia have often been found.3 Although cranial nerves are generally spared in CMT, thickening of the acoustic or optic nerve has been reported in some cases. We report here on a Japanese patient who exhibited severe polyneuropathy, bilateral trigeminal thickening on MRI, and an abnormality of the auditory brain stem response. Gene analysis disclosed a novel missense mutation (His81Arg) of P. The cranial nerve involvements in this patient may be associated with the novel missense mutation of P. (His81Arg).

In the present study, our patient showed severe clinical manifestations of early onset and undetectable conduction velocities. Therefore, this patient was considered to have a severe form of CMT or Dejerine-Sottas disease. Although her facial sensation, mastication power, and hearing acuity were normal, the thickness of bilateral trigeminal nerves on MRI and prolongation of the I-III interpeak interval in auditory brain stem response were found. The I-III interpeak interval represents the conduction time from the eighth nerve to the pontomedullary portions of the auditory pathway. Prolongation of the auditory brain stem response suggested peripheral conduction delay of the auditory nerve.

Trigeminal neuralgia with CMT has been reported.1 In these rare cases, trigeminal neuralgia was inherited, suggesting a partial symptom of CMT. Although some patients were surgically treated, it was not clear whether a thickened trigeminal nerve was present. Moreover, on electrophysiological studies of facial and trigeminal nerves in CMT, Kimura4 reported that the sensory component of the trigeminal nerve was relatively spared, despite extremely delayed conduction of the facial nerve. However, the MRI study of our patient suggested that the fifth cranial nerves were subjected to the same pathological process that affects other peripheral nerves.

Our patient showed no DNA duplication on chromosome 17p11.2 and we found a novel mutation (A to C) representing an Arg to His substitution in the P. gene. Histidine 81 is conserved among many other species, including cows, rats, chickens, and sharks. This mutant allele was absent in the DNA from 100 controls. Therefore we identified this mutation as pathogenic. Arg^H^is located in exon 3, which codes for the extracellular domain of the P. protein. The extracellular domain plays a part in myelin compaction by homophilic interaction and many mutations in this area have been reported. Although the phenotypic variability is related to the position and nature of the P. mutation, patients with cranial nerve involvement are rare in CMT with a P. mutation. Therefore, the unique thickening of trigeminal nerves and the clinical severity in this patient may be related to this novel missense mutation. A careful comparison of the clinical, electrophysiological, and histopathological data between patients with CMT should be conducted.

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Intracranial extracerebral follicular lymphoma mimicking a sphenoid wing meningioma

Primary lymphoma in the brain is uncommon, accounting for only 2% of primary intracranial neoplasms. Although its incidence seems to be dramatically increasing, leptomeningeal lymphomas are even rarer than expected. Six cases of leptomeningeal lymphoma have been previously reported,1; however, no leptomeningeal lymphoma of the follicular type has previously been reported. We present a case of a primary meningeal follicular lymphoma which mimicked a sphenoid wing meningioma, both radiologically and intraoperatively.

A 37 year old Ghanaian woman was referred with a 3 year history of worsening bilateral headache, followed by a 6 month history of daily right frontal headache lasting for 2–3 hours associated with mild photophobia. There were no reports of seizures, nausea, or other visual disturbances. Her medical history was 3 years of treated hypertension, sickle cell carrier trait, and a cataract extraction. The patient was obese but physical examination was otherwise normal. Neurological examination showed no papilloedema and there were no cranial nerve or long tract signs.

Brain CT showed an enhancing mass consistent with a right sided sphenoid wing meningioma. Axial T1 weighted (TR 600/TE 15) brain MRI at 1.5 Tela of our patient with CMT. Note the thickness of the bilateral trigeminal nerves.

References

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Brain CT showed an enhancing mass consistent with a right sided sphenoid wing meningioma.
chemically confirmed a follicular lymphoma. The patient made an uneventful recovery and was referred for staging investigations and consideration of postoperative therapy. An LDH estimation was within normal limits and HIV serology was negative. Whole body CT including repeat CT of the brain did not show any evidence of lymphadenopathy or lymphomatous deposit. Bone marrow examination was declined. Postoperative adjuvant whole brain or localised radiotherapy was discussed with the patient, however, she declined any further intervention. She has been closely reviewed in the follow up clinic and after 6 months there has been no clinical or radiological change.

Primary intracerebral lymphomas represent about 2% of intracranial neoplasms and 2% of all lymphomas. They occur most commonly in the 6th decade of life with a female to male ratio of approximately 3:1. The association between primary intracranial lymphoma and immunodeficiency has long been established, and it is not surprising, therefore, that the incidence has increased 10-fold over the past 3 decades with the onset of transplant surgery and, particularly, the AIDS epidemic. 2 In postmortem studies, these neoplasms are found, on average, in 5.5% of AIDS cases, and malignant cerebral lymphoma is the most common diagnosis of a focal intracranial lesion in patients with AIDS. 3 Malignant primary lymphoma can occur throughout the CNS and they often have a periventricular distribution. Multifocality seems to be more common in patients with AIDS. The CT scan usually shows hyperdense masses with peritumourous oedema and 92% enhance after administration of contrast medium. 4 Leptomeningeal lymphoma is usually encountered as a late complication of systemic non-Hodgkin’s lymphoma, although primary leptomeningeal lymphoma is occasionally seen. The prognosis for these tumours is poor. 5 Diffuse primary lymphomas have been mistaken for more common lesions: solitary primary B cell lymphoma of the cerebellar pontine angle mimicking acoustic neuroma, or meningeoma has been reported; 8,9 Surgishin et al. 10 reported a patient with a calcified temporal parietal lymphoplasmacytic lymphoma which resembled a meningioma; however, this tumour was entirely extradural. There is only one previous report of a follicular lymphoma with meningioma. 5 The author, Rubinstein described a case of follicular lymphoma metastasis found in the dura of a 61 year old man at necropsy. We found no report of a primary follicular extracerebral lymphoma. Similar radiological and intraoperative appearances of the tumour in our case to meningioma in meningeoma suggest that this entity should be considered as a rare differential diagnosis.

Histologically the lesion consisted of lymphoid tissue with an ill defined follicular architecture (figure B). The follicles varied in size and shape and infiltrated the overlying dura. Follicular centres were composed of a mixture of centrocytes and centroblasts with frequent mitotic figures and apoptotic bodies (figure C). Immunohistochemical staining confirmed that these cells had a B lymphoid phenotype (CD20 positive) with kappa light chain restriction. Staining for Bcl-2 protein, which is an inhibitor of apoptosis and is expressed in 90% of follicular lymphomas, was found to be positive. The histological appearances and immunohistochemical profile confirmed a follicular lymphoma.

The determinants of the copper concentration in cerebrospinal fluid

The measurement of CSF copper concentration can serve as an indicator of brain copper concentrations. However, the complex mechanisms by which copper crosses into the CSF, and the factors determining the CSF copper concentration in humans are largely obscure. Copper can pass into and out of the CSF by various mechanisms. For example, active transport through the blood-brain barrier or the blood-CSF barrier, or passive diffusion of the free or the bound fraction (bound to albumin or coeruleoplasmin) through the blood-CSF barrier. We studied the factors influencing CSF copper concentration using a stepwise multiple linear regression model. The independent variables were age, plasma coeruleoplasmin, CSF albumin ratio, total serum copper concentration, and calculated serum free copper concentration (based on serum coeruleoplasmin and total serum copper concentration). The CSF copper concentration was considered as a dependent variable and a continuous type. We investigated lumbar CSF samples from 113 patients. These patients had dementia, extrapyramidal, or tremor syndromes, or suffered from Wilson’s disease, and none of the patients had the disease. Copper was measured by flameless atomic absorption (Perkin Elmer, HGA 500, Uerberlingen, Germany). Coeruleoplasmin was determined nephelometrically (Beckman Instrument Instruments, Brea, CA, USA). The age of the patients was 50.0 (SD15.5) years; 50 were women and 63 were men. Mean serum coeruleoplasmin concentrations were 394.3 (SE81.7) mg/l. Mean serum copper concentrations were 1194 (SD 335) μg/l. Mean calculated free copper concentrations in serum were 78.5 (SD 1285) μg/l. Mean CSF copper concentration was 14.6 (SD 6.0) μg/l. The mean albumin ratio (AR) was 6.63×10−3. The ratio of calculated mean free copper concentration to total serum copper was 6.6%, the ratio of CSF copper to serum copper was 1.2%, and the ratio of free serum copper to CSF copper was 18%. In the
Correlation of blood-CSF barrier (albumin ratio, (AR)) with total CSF copper concentration (on logarithmic axes). R²=0.46, p=0.0001; 95% confidence bands for the true mean of the total CSF copper concentration are shown.

Stepwise linear regression model (F to enter 4.0, F to remove: 3.996), significant positive predictors of the CSF copper concentration were found to be AR (p=0.0001) and serum coeuloplasmin (p=0.0057). The other independent variables mentioned above showed no statistically significant relation with CSF copper concentration. The figure shows the simple linear regression between CSF/serum albumin ratio and CSF copper concentration (on logarithmic axes; R²=0.46, p=0.0001).

The formula for the CSF copper concentration, derived from the multiple linear regression model, is: copper [µg/l]=5.32 µg/l 0.653 × CSF/serum albumin ratio (×10⁻⁴)+0.012 × serum coeuloplasmin (mg/l).

According to this analysis, CSF/serum albumin ratio and serum coeuloplasmin together determine 25.3% of the variation in CSF copper concentration (adjusted R²=0.253), implying that other (unknown) factors determine the remaining 74.7% of the variation. We have been able to demonstrate here that the CSF copper concentration is determined in a highly significant manner by disturbances in the blood-CSF barrier and by the serum coeuloplasmin concentration. It can be assumed from this that in the case of normal blood-CSF barrier function and a normal serum coeuloplasmin concentration, 20.7% of the measured CSF copper originates from the blood; the CSF by passive diffusion bound to coeuloplasmin, and only around 0.09% by passive diffusion bound to albumin. In the case of a markedly raised CSF/serum albumin ratio of 20×10⁻³, this would mean that 60.6% of the measured CSF copper originated from the blood (bound to coeuloplasmin). A variable fraction of the CSF copper concentration, depending on the degree of damage to the blood-CSF barrier, therefore crosses from the blood into the CSF and can be measured there. Our formula would therefore predict, in patients with Wilson’s disease with intact blood-CSF barrier (assuming a CSF/serum albumin ratio of 6.5×10⁻³), that the CSF copper concentration is actually reduced by 27.4%, when the serum coeuloplasmin concentration falls from its normal value of 394 mg/l to 66 mg/l. In consequence, CSF copper in patients with Wilson’s disease is evidently substantially free, implying that a larger fraction than previously assumed of the raised CSF copper in patients with untreated Wilson’s disease originates from the brain; the fraction entering the CSF by passive diffusion (bound to coeuloplasmin) tends towards zero. It can be concluded from this that, when the aim of therapy is considered in terms of the total CSF copper concentration, a region around 30% lower than the upper limit of the normal range should be aimed for. This is supported by the clinical finding that patients report feeling better when the CSF copper concentration is below this value. This analysis also shows that the raised copper concentration in the CSF can only originate from the brain. In particular, it is not associated with free serum copper, but evidently only via storage in the serum. The investigation here also shows that, after determining the CSF copper concentration, the coeuloplasmin-bound fraction originating from the plasma should be subtracted according to the formula we have given, or better, all measured copper concentrations in the CSF should be adjusted using the CSF/serum albumin ratio and serum coeuloplasmin concentration. A statistical relation with a low correlation (p<0.05) between CSF protein content and CSF copper was already shown in various neurological diseases; our study shows a much higher significance and, in addition, the effect of serum coeuloplasmin (therefore of bound serum copper). Furthermore, we have been able to determine quantitatively the contribution of CSF copper which enters the CSF across the blood-CSF barrier.

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Solitary intracranial myofibroma in a child

A rare case of solitary interhemispheric myofibroma with excellent outcome in a 20 month old boy is described. The clinicopathological features of this unusual condition are reviewed with emphasis on the CNS manifestations.

A case of congenital fibrosarcoma was first diagnosed by William and Schum and was subsequently renamed congenital generalised fibromatosis by Stout in 1954 as a distinct form of juvenile fibromatosis characterised by tumour-like nodules involving the skin, soft tissues, bones, and viscera. Based on the ultrastructural and immunohistochemical features of the cell of origin and the occurrence of this condition in infants, as well as congenitally, it was renamed infantile myofibromatosis by Chung and Enzinger in 1981. This disorder is considered to represent a hamartomatous myofibroblastic prolifer-
myoglobin. Ultrastructural examination showed elongated cells with surrounding collagen fibrils, some showing intracytoplasmic myofilaments.

Solitary lesions of infantile myofibromatosis are more common than multiple lesions, with twice as many males as females being affected, and generally involve the skin and soft tissues, especially of the head and neck. Solitary lesions are less commonly found in viscera or bones.1 Involution of the CNS is exceedingly rare and only one other case of a solitary mass is reported2 along with few cases of CNS involvement in the generalised form of infantile myofibromatosis.3 The prognostic is best for cases with solitary masses and less favourable for multicentric cases, particularly where visceral lesions are present, in which morbidity and mortality derive predominantly from pulmonary involvement or mass effect.

The differential diagnosis for this lesion included meningioma, schwannoma, and haemangiopericytoma. Regionally, the histology was reminiscent of the rare microcystic variant of meningioma. Meningiomas are extremely rare in this age group, this lesion was not meningeval based and such lesions are usually reactive for epithelial membrane antigen unlike this tumour. This lesion, unlike some sarcomas, did not show any immunoreactivity for S-100 protein. Haemangiopericytoma is a diagnosis of exclusion and shows no reactivity for actin, unlike this tumour.

Axial involvement by myofibromatosis includes patients with widespread systemic involvement and multiple leptomeningeal nodules in one patient and extradural masses in another, both of which were fatal at the age of 10 days, a non-fatal extradural mass in one patient, and a patient with systemic involvement, in which there was recurrence of orbital and temporal lesions 2 years after operation. A single previous case of solitary intracranial myofibroma has been reported3 in which the patient died within 24 hours of surgery, secondary to cardiovascular arrest.

We present a patient with a solitary intracranial myofibroma with an excellent postoperative outcome. Although rare, infantile myofibroma should be included in the differential diagnosis of intracranial neoplasms in children.

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spreading into the hippocampus and the brainstem. The convulsions in our patient were successfully treated with 250 mg/day diphenhydantoin, and her encephalopathy gradually improved during plasma exchange and haemodialysis. After recovering consciousness, she began to complain of numbness of her limbs, and a burning pain which exacerbated in the night. Nerve conduction studies and the clinical features confirmed the diagnosis of sensory-dominant, axonal polyneuropathy. At this stage metabolic abnormalities were not detected and serum concentrations of vitamins B1, B6, and B12 were normal. Her numbness and tingling sensation ameliorated after 2 weeks administration of 300 mg/day oral melixin, an agent with a membrane stabilising effect. Up to now, to our knowledge, peripheral neuropathy has not been reported in VTEC infection other than in one patient, by Hamano et al., who showed bilateral phrenic nerve palsy for 2 weeks after recovering consciousness. The above experimental evidence suggests that microcirculatory disturbances and neurotoxicity to the neural cells by verotoxin could cause axonal neuropathy in VTEC infection.

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Crying spells as symptoms of a transient ischaemic attack

In the absence of depression, crying spells associated with neurologic disease usually result from pseudobulbar palsy or, more rarely, from crying seizures. To our knowledge, there are no prior reports of crying spells heralding or signifying a transient ischaemic attack. We report on a patient with crying spells heralding or signifying a transient ischaemic attack. We report on a patient with crying for no apparent reason. There was no history of pseudobulbar palsy or, more rarely, from crying seizures associated with neurological disease usually result from pseudobulbar palsy or, more rarely, from crying seizures. The patient had a 55 year old right handed man who presented with acute, uncontrolled crying spells followed by left sided parasthesias. Around 6 00 am he awoke with a diffuse, pressure headache and suddenly started crying for no apparent reason. There was no accompanying feeling of sadness. This crying was not associated with lacrimation and “sobbing,” abruptly ceased after 5 minutes. Within 30 minutes of his initial crying spell, his headache had resolved but he became aware of numbness over his left face and numbness and pain in his left neck and arm. The numbness was not progressive, and the patient did not complain of paraesthesias in his trunk or left leg. He had photophobia, nausea or vomiting, blurred vision, visual obscurations, difficulty swallowing, dysartria, or focal weakness. Over the next 2 to 3 hours, he had five more crying spells, each lasting 5 to 10 minutes, occurring out of context, without precipitating factors or sadness, with an acute onset and offset, and without alteration of consciousness. The patient’s left face and arm numbness persisted during and between the spells, but abruptly resolved shortly after his last crying spell. This patient had hypertension, diabetes mellitus, coronary artery disease, an old myocardial infarction, raised cholesterol concentrations, and a history of heavy smoking.

On examination between recurrent crying spells, his blood pressure was 143/92 with a regular pulse of 62, and there were no carotid bruits. His mental status was normal. Cranial nerve examination disclosed flattening of the left nasolabial fold and decreased pinprick sensation over his left face, with occasional mild facial twitching. Cranial nerves IX–XII were intact, and gag and reflex and palate elevation movements were normal. He did not have dysarthria or a brisk jaw jerk. The rest of the neurological examination showed mild weakness in his left upper arm, and decreased pinprick and temperature sensation over the left half of his body. Eye movements were +2 symmetric with downgoing toes.

The patient lacked prior depression, new depressive symptoms, or prior crying spells as an adult except for a single episode during dental anaesthesia. At the time of his admission, he had not had any recent adverse events in his life, and was totally surprised by his reaction.

The patient’s crying spells, paraesthesias, and neurological findings entirely resolved within about 3 hours. Routine laboratory tests, ECG, and CT were normal. Two days after admission, MRI disclosed a mild degree of white matter sparing over the right frontal horns and an ECG monitor documented frontal intermittent rhythmic delta activity but no epileptiform changes. Carotid Doppler studies showed atherosclerotic changes without haemodynamically relevant obstruction. He was discharged on antiplatelet therapy with aspirin.

These results suggest that crying spells can be a manifestation of a transient ischaemic attack. He presented with paroxysmal crying spells followed by a left sided hypoaesthesia and a mild left sided weakness, all of which resolved. His crying was non-emotional, inappropriate to the context, and did not correspond to his underlying mood. Moreover, the patient had multiple vascular risk factors supportive of a cerebrovascular aetiology for his episode.

The most common cause of pathological crying is pseudobulbar palsy, a complication of strokes and other diffuse or bihemispheric brain damage.1 Pseudobulbar palsy results from bilateral interruption of upper motor neuron innervation of bulbar motor nuclei and brainstem centres. In addition to crying, pseudobulbar palsy may include dysarthria, dysphagia, bifacial weakness, increased facial and mandibular reflexes, and weak tongue movements. There were no signs or symptoms of pseudobulbar palsy at this inpatient.
maximal 74 mm Hg), taking more than 10 minutes to reach the minimum (continuous drop type) (figure). The other five patients could not remain standing for more than 5 minutes because of symptoms of orthostatic hypotension. No patient showed the sudden drop in blood pressure, cardiac output should be lack of reflex tachycardia and no significant release of noradrenaline (norepinephrine) level (+0.05 ng/ml) during the decrease in blood pressure, cardiac output proportionally decreased but systemic vascular resistance did not change (figure).

Our results suggest that in many patients with MSA the blood pressure drops continuously on standing. The continuous blood pressure drop is caused by continuous reduction of cardiac output. A part of the mechanism for continuous reduction of cardiac output should be lack of reflex tachycardia and no significant release of noradrenaline which are caused by interruption of the baroreflex arc, as is known in MSA.

Continuous drop type of orthostatic hypotension during 25 minute tilt up in a patient with MSA. SBP=systolic blood pressure; HR=heart rate; CO=cardiac output; SVR=systemic vascular resistance; NA=plasma noradrenaline concentration.
Idiopathic cerebellar ataxia associated with celiac disease: lack of distinctive neurological features

Although applauding the contribution of Pellecchia et al. to the more widespread recognition of the association between gluten sensitivity and ataxia, we disagree that ataxia associated with gluten sensitivity lacks “distinctive neurological features”. Both their data and correspondence indicate that this group of patients can be distinguished by the late (non-childhood) onset of gait ataxia with relatively mild upper limb signs, analogous to Harding’s group 1. Again, coexistent neuropathy is common in these patients, found in two out of three of the patients of Pellecchia et al and 21 of our 28. We agree that gastrointestinal symptoms are rare: rather than entitling their paper “lack of distinctive neurological features”, perhaps “lack of distinctive gastroenterological features” might have been more appropriate.

We were surprised at the high specificity and sensitivity of increased antigliadin antibody titres in their hands. Although we found both IgA and IgG antigliadin antibodies to be invaluable screening tools in patients with ataxia, only 11 of our 28 patients with idiopathic cerebellar ataxia had histology of overt coeliac disease on duodenal biopsy, the remainder having normal or non-specific inflammatory changes but with an HLA genotype in keeping with gluten sensitivity. It is interesting to note that despite the often quoted high sensitivity for coeliac disease of increased antiendomysium antibody titres, such was found in only one of three patients of Pellecchia et al with coeliac disease. This concurs with our impression of very modest sensitivity of antiendomysium antibodies in gluten ataxia.

Gluten sensitivity is common in patients with ataxia, and can be identified by increased antigliadin antibody titres in the presence of appropriate histocompatibility antigens. Although the clinical features of gluten ataxia are not entirely specific, they are distinctive.

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Pellecchia et al reply:

We thank Hadjivassiliou et al for their interesting comments on our paper. They suggest that patients with gluten ataxia can be distinguished by the late onset of gait ataxia and the relatively mild upper limb signs. Our results support the finding of a late onset in these patients, but this feature cannot be considered a distinctive one. In fact, it is in our population 11 out of 24 patients with idiopathic cerebellar ataxia had a late onset, but only three of them were affected by celiac disease. Furthermore, we do not think that celiac patients may be distinguished by mild upper limb signs and coexistent neuropathy; in our study 20 out of 24 patients with idiopathic cerebellar ataxia, including the three patients with celiac disease, had ataxic gait as the presenting and prominent clinical feature. Similarly, nerve conduction studies, performed in 17 out of 24 patients, showed a peripheral neuropathy in nine, including two out of the three patients with celiac disease.

We understand that some discrepancies arise comparing our study with that of Hadjivassiliou et al. Firstly, only six out of our 28 patients had evidence of cerebellar atrophy on MRI, whereas all of our patients had cerebellar atrophy. Secondly, many of their patients had a peripheral neuropathy in the absence of cerebellar atrophy. This could explain the mild upper limb signs. Although two of our three celiac patients had a clinically silent peripheral neuropathy, we think that their ataxia was explained by cerebellar atrophy. Thirdly, we found a high prevalence (12.5%) of celiac disease on duodenal biopsy among patients with idiopathic cerebellar ataxia, whereas none of the six patients with cerebellar atrophy described by Hadjivassiliou et al showed histological features of celiac disease.

It would be interesting to know the prevalence of gluten ataxia among all ataxic patients screened for antigliadin by Hadjivassiliou et al. Our series is too small to estimate the sensitivity of both antigliadin and antiendomysium antibodies in gluten ataxia; unfortunately Hadjivassiliou et al did not report any data on antiendomysium antibody screening in their patients. On the other hand, we were surprised at the high prevalence of antigliadin antibody positivity (12%) in the normal population studied by Hadjivassiliou et al in a previous report. This is by contrast with the 2% of antigliadin positivity found in a large population by Catassi et al. Further studies are required to better characterise the syndrome of cerebellar ataxia associated with celiac disease or gluten sensitivity.

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Procanamide for faecal incontinence in myotonic dystrophy

We read with interest the article by Abercrombie et al which describes the pathophysiology and surgical management of faecal incontinence in two siblings with severe myotonic dystrophy.

In the authors’ experience, long term results of both medical and surgical management of the faecal incontinence of pelvic floor dysfunction, as proved by the fact that postanal sphincter repair restored faecal continence only for a brief time.

The authors’ pessimistic conclusions suggest that “faecal incontinence in myotonic dystrophy is difficult to relieve by any currently available treatment other than colostomy”. It should be noted, however, that the medical treatment used is not specified in the text.

Our experience with medical treatment using procarainamide in a patient with severe myotonic dystrophy and faecal incontinence is less disappointing. The patient—a 19 year old man—had his illness diagnosed 4 years earlier on clinical grounds and electrophysiological and genetic tests. Early symptoms of sphincteric impairment developed soon after, including mild stress urinary incontinence and minor episodes of poor control of loose stool.

A complete diagnostic investigation, including physical examination, defecography, and electromyographic tests of pelvic floor musculature, was performed. At physical examination, digital anorectal evaluation showed low squeeze pressures. A reduced rectal diameter (4.5 cm), anal gaping, and barium loss at rest were found in defecography. Motor evoked potentials elicited by cortical and lumbar magnetic stimulation and recorded from the external anal sphincter’ showed normal latency and decreased amplitude. Somatosensory evoked potentials after anal stimulation and sacral reflex latency were normal. EMG recording of the external anal sphincter showed, as in the first patient of Abercrombie et al, a decreased number of motor units and multiple myotonic discharges. Few motor unit potentials presented polyphasic waveforms and decreased duration and amplitude.

A regular treatment with procarainamide (300 mg twice a day) lead to a dramatic improvement of both systemic myotonia and faecal incontinence. A 13 month follow up assessment has shown a stable clinical improvement. Repeated electromyographic investigation showed disappearance of myotonic discharges at the external anal sphincter, whereas defecography disclosed an improved rectal compliance (5.2 cm in diameter) at capacity and no more than a barium leak on straining.

The pathophysiology of motor disorders of the gastrointestinal tract in myotonic dystrophy is still debated and controversial. Histological study of the external anal sphincter and...
the EMG pattern in patients with myotonic dystrophy show a multitude of deficits including expression of myotonia, myopathy, muscular atrophy, and neural abnormalities.1,2

The possible management of myotonia and some of its clinical manifestations, such as dysphonia, by a myotonic dystrophy (dyspnea, dystrophia myotonica and procainamide), justifies the use of the same pharmacological approach in anal sphincter dysfunction manifested in a few cases of myotonic dystrophy.

We conclude that treatment of faecal incontinence with procainamide should always be attempted before any surgical option in patients with myotonic dystrophy.

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Pain after whiplash

This latest study from Lithuania is an answer to many questions—namely, that the previous difficulties that these researchers had with identifying the late whiplash syndrome in Lithuania is that they were not looking “in the right place”. As it turns out, the problem is that Lithuanians simply are not behaving the way many in western countries, and, underlies whiplash associated disorders like. There are some methodological issues which can be considered, as below, but the lesson of discarding “unsightly” data because it is too disturbing to one’s personal view and vested interest in the whiplash controversy has already been taught elsewhere. Suffice it to say that the truth has been laid bare and we (those of us struggling with epidemic proportions of the late whiplash syndrome in our own countries) now need to enlighten ourselves and put this data to practical use in helping whiplash patients rather than resisting the inevitable.

After completion of the first historical cohort study, this more recent study selects an entirely separate, distinct sample of these “misbehaving” Lithuanians, but in a more intriguing fashion. This is the first inception cohort study which people who have not been preselected by their attendance at emergency departments, or contaminated by therapists or lawyers, can be studied to appreciate the natural evolution of the injury which, underlies whiplash associated disorders grades 1 and 2. This is the study’s greatest strength. The study has, however, its limitations.

The first consideration is that there were 98 accident victims who reported acute symptoms, and thus were at risk for the late whiplash syndrome. How does this compare with other studies documenting the natural evolution of the late whiplash syndrome? The Swiss study may be useful for comparison, but in a more intriguing fashion. This is the first inception cohort study which people who have not been selected by their attendance at emergency departments, or contaminated by therapists or lawyers, can be studied to appreciate the natural evolution of the injury which, underlies whiplash associated disorders grades 1 and 2. This is the study’s greatest strength. The study has, however, its limitations.

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Finally, the authors carry out a historical review, and refer to the fact that this distinctive myotrophic lateral sclerosis variant was probably first described by Gowers in 1888, furnished with exquisite graphic illustrations.

To this effect, we draw attention to prior descriptions of the same syndrome, reported by Vulpiani in 1886, known in Franco-German literature as Vulpian-Bernhardt’s form.

In his book Maladies du Système Nerveux Vulpian described a patient who showed signs of weakness and symmetric proximal atrophy of neurogenic origin, and called it chronic anterior poliomyelitis. The patient showed symptoms of proximal amyotrophy, and signs of denervation and upper motor neuron involvement. Since then, in those countries and other countries under their influence,1,4 we have come to use the eponym of Vulpian-Bernhardt’s syndrome to describe those forms of amyotrophic lateral sclerosis with more or less symmetric involvement of the proximal muscles of the upper limbs at the clinical onset.

A certain enigma exists surrounding the characteristic distribution of weakness and muscle atrophy. The reason for the preva-


Flail arm syndrome or Vulpian-Bernhart's form of amyotrophic lateral sclerosis

We read with interest the article by Hu et al concerning flail arm syndrome, a distinctive variant of amyotrophic lateral sclerosis. The authors presented a subgroup of patients affected by amyotrophic lateral sclerosis that predominately showed signs of lower motor neuron involvement in the proximal muscles: opponens pollicis, flexor brevis, abductor pollicis brevis, abductor pollicis, interossei, and lumbricales, which leads to the formation of the characteristic Aran-Duchenne hand.

Obviously, signs of corticospinal involvement with hyperreflexia in the lower limbs and Babinski’s sign both appear. In the initial stages of the illness, there is no effect on the diaphragm. The presence of signs of involvement of the upper motor neuron, its different clinical evolution, and the data supplied by genetic molecular investigation allow us to distinguish the syndrome previously known as Vulpian-Bernhardt’s form from other upper motor neuron syndromes such as the spinal muscular atrophies, Kennedy’s disease, multifocal motor neuropathy, and monomelic amyotrophy.

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A second consideration is that perhaps these Lithuanians are in very minor collisions. True, some of their vehicles were completely wrecked, but perhaps the vehicles were not very good quality and so were easily damaged. Perhaps that is why this cohort had such a good outcome and only minor injuries. This is an unhelpful consideration however, as studies in Canada have shown that those in low velocity collisions, are just as likely to experience of late whiplash syndrome in Lithuania. Perhaps that is why this cohort had some experience of late whiplash syndrome. True, some of their vehicles were completely wrecked, but perhaps the vehicles were not very good quality and so were easily damaged. Perhaps that is why this cohort had such a good outcome and only minor injuries. This is an unhelpful consideration however, as studies in Canada have shown that those in low velocity collisions, are just as likely to experience of late whiplash syndrome as those in more severe collisions. Lithuanians seem to behave appropriately then for minor collisions (if that is what they indeed had), but Canadians seem unable to behave appropriately. Again, another cultural difference in the rate of recovery from whiplash injury is demonstrated.

Thirdly, there are sex differences and even differences in seat belt usage between this population and some others, but even then, it does not seem to matter what sex, age, and use of seat belts there is in other western countries, none of these preclude chronic pain. In Lithuania, those who were female, and who did not wear seat belts, still insisted on behaving as the rest of the cohort.

Finally, perhaps the Lithuanians simply refuse to report their chronic pain, and chronic pain cannot be studied in other countries in this way. The Lithuanians have no reluctance to report acute pain, but perhaps for some reason wish to “suffer in silence” in spite of chronic pain and disability. This would be a potential flaw if it was not simultaneously shown in this study that the general Lithuanian population reports the same prevalence, frequency, and character of neck pain and headache as does the general population in western countries. If there were study design barriers to identifying symptoms, the control population would have grossly underreported their symptoms. Indeed, chronic pain can and is reported by studies in many different cultures and languages, including Japan, France, Italy, and others. If researchers in these non-English speaking populations can use simple questionnaires to document the late whiplash syndrome so effectively there, then the same should be possible in Lithuania.

And so, despite the potential limitations of this study as outlined, there is no way to get around the stark realisation that the natural history of chronic pain in Lithuania is a benign syndrome with 4 weeks or less of pain. Equally compelling is the fact that Lithuania is not the only place where researchers are having difficulty identifying epidemics of chronic pain. Recovery from acute whiplash injury without neurological injury or fracture routinely occurs within 4–6 weeks in Germany and Greece. The time has not yet been set for a reconciliation of these epidemiological observations with our own experience of late whiplash syndrome in western countries. The truth has been laid bare and it is time for us to utilise this truth to help prevent the chronic pain and the suffering we otherwise encounter.

R FERRARI

BOOK REVIEWS


This book purports itself to be a comprehensive reference. Certainly the title would suggest so. However, it is clear that this is not a comprehensive text, but a book that is an update on particular topical issues in the field of pain medicine. There are sections on pain mechanisms, a chapter on the pharmacology of acute and chronic pain, and other chapters on postoperative pain, obstetric pain, and acute paediatric pain. There are three further chapters specifically on the management of chronic low back pain, cancer pain, and an overview of interventional pain techniques.

Many of the authors are internationally known and this is perhaps the book’s strongest point—one does get a state of the art review and to this end I warmly welcome this book as an addition to the bookshelf to update a busy anaesthetist or pain specialist. The chapters are written by different authors and is an excellent summary of the subject. In the section on interventional pain techniques the emphasis was on spinal cord stimulation, radiofrequency, and cryoneurolysis. Again this chapter has been written by an internationally well known author who concentrated on general overview of the techniques rather than a how to do it approach, which I found a little disappointing. This book would be a potential flaw if it was not simultaneously shown in this study that the general Lithuanian population reports the same prevalence, frequency, and character of neck pain and headache as does the general population in western countries. If there were study design barriers to identifying symptoms, the control population would have grossly underreported their symptoms. Equally compelling is the fact that Lithuania is not the only place where researchers are having difficulty identifying epidemics of chronic pain. Recovery from acute whiplash injury without neurological injury or fracture routinely occurs within 4–6 weeks in Germany and Greece. The time has not yet been set for a reconciliation of these epidemiological observations with our own experience of late whiplash syndrome in western countries. The truth has been laid bare and it is time for us to utilise this truth to help prevent the chronic pain and the suffering we otherwise encounter.

R FERRARI

Letters, Correspondence, Book reviews

The chapter on acute postoperative pain management is well written and informative as are the chapters on obstetric and paediatric pain. The chapter on chronic low back pain by Rauck is one of the best I have seen for some time. It is a comprehensive review of both acute and chronic low back pain. It is excellent as it also mentions treatments that are often performed outside the medical specialist arena. I was pleased to see in it the mention of some of the newly evolving techniques such as facet denervation, spinal cord stimulation, and disc denervation. It was a pity that the randomised control trials which have shown facet denervation to be an outstandingly useful technique for acute lower back pain were not mentioned. It was also a pity that the reference to the disc denervation procedure was to another text book rather than any original papers.

The chapter on cancer pain management has been written by internationally known authors and is an excellent summary of the subject. In the section on interventional pain techniques the emphasis was on spinal cord stimulation, radiofrequency, and cryoneurolysis. Again this chapter has been written by an internationally well known author who concentrated on general overview of the techniques rather than a how to do it approach, which I found a little disappointing. This book would be a potential flaw if it was not simultaneously shown in this study that the general Lithuanian population reports the same prevalence, frequency, and character of neck pain and headache as does the general population in western countries. If there were study design barriers to identifying symptoms, the control population would have grossly underreported their symptoms. Equally compelling is the fact that Lithuania is not the only place where researchers are having difficulty identifying epidemics of chronic pain. Recovery from acute whiplash injury without neurological injury or fracture routinely occurs within 4–6 weeks in Germany and Greece. The time has not yet been set for a reconciliation of these epidemiological observations with our own experience of late whiplash syndrome in western countries. The truth has been laid bare and it is time for us to utilise this truth to help prevent the chronic pain and the suffering we otherwise encounter.


The chapter on chronic pain again is well written, but unfortunately the chapter on chronic back pain were not mentioned. It was also a pity that the reference to the disc denervation procedure was to another text book rather than any original papers.

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Rajesh Munglani


This is a really excellent book which is both comprehensive and amazingly up to date, with the inclusion of many references from as late as 1997. As a clinical neurologist and neuropathologist with a longstanding interest in the dementias, I found it extremely valuable. The editor has done a very good job in posing a coherence, format, and style, which is often lacking from multicontributor textbooks.

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I very much enjoyed reviewing this textbook of instrumented spinal surgery written by Giuseppe Tabasso under the auspices of Jürgen Harms. Dr Harms is well known to all spinal surgeons and has made a very important contribution to the development of spinal surgery over the past 20 years, based on strong personal convictions. Many surgeons who manage spinal disorders would not choose to implement all of Professor Harms’ solutions but all who have a serious interest in the surgical treatment of the spine admire and are grateful for his contribution. Within this book spinal surgeons will find a rational and practical approach which will allow them to treat a wide range of spinal disorders according to well thought-out principles.

The opening chapter describes spinal biomechanics under normal and pathological circumstances mainly by using easily understood drawings and diagrams. Some of these drawings reminded me of images that I have recently seen on an interactive CD ROM that I bought for my 4 year old son. This is not a criticism and I fully support any attempt to simplify the science of biomechanics which is often cloaked in seemingly contradictory jargon. Most spinal surgeons will be able to assimilate the two basic principles which underpin much of instrumented spinal surgery—namely, that the anterior column resists load compression forces and that the posterior column acts as a tension band which when disrupted should be reconstituted in compression. The remaining chapters cover fracture management, late kyphosis, metastatic tumours, spondylolisthesis, degenerative spinal disease, and infection. Each chapter sets out the principles of management which are illustrated schematically. There then follow case studies illustrated by radiological images including CT and MRI. These have reproduced well and surgeons will admire the technical precision and excellent anatomical reductions illustrated by these clinical cases. It is, however, a source of constant annoyance to spinal surgeons that perfect postoperative films do not always correlate with good clinical results and this discrepancy remains a source of fascination and mystery.

It is in the degenerative spine that this discrepancy between radiological and clinical findings is most apparent and it is partly for this reason that the management of these conditions is often controversial. It is difficult to disagree with much of the logic presented by the authors in planning their interventions but there is a danger that inexperienced surgeons may be misled into adopting complex solutions when often more simple operations will suffice. The authors’ description of their approach to failed back surgery syndrome illustrates this problem and the inadequacies of attempting to treat a complex clinical problem by focusing on one aspect of it.

This book will be a useful addition to the shelves of spinal surgery textbooks and many orthopaedic and neurosurgical departmental libraries will wish to buy a copy.

RODNEY LAING


I wondered, when I received this book, how I could possibly say anything adverse about a book written by three such world-renowned experts. I have heard them all lecture often and have seen them all at work. They have a vast knowledge and experience of treating disorders of peripheral nerves. In clinic and the operating theatre, they have shown myself and many trainees a clarity in their planning of management of complex problems that humbles one’s own thoughts. That clarity has continued in this text book of over 500 pages. The field of peripheral nerve surgery is covered comprehensively, commencing with descriptions of anatomy, physiology, and pathological reaction to injury. This is followed in subsequent chapters with descriptions of approaches to virtually all the main peripheral nerves, and the operative management of brachial plexus injury and outcomes is covered in three detailed chapters. These are followed by chapters on nerve entrapment, neuropathy, iatropathic injury, and neoplasm within the peripheral nerve. The final section covers electrodiagnosis, pain, nerve recovery, reconstruction techniques, and rehabilitation.

The text is well written, easy to read, and supplemented by some excellent line drawings similar to those used in Lundborg’s text. There are detailed plates showing histology and various imaging techniques. Each chapter is comprehensive, containing important historical aspects as well as up to date techniques, and there is an extensive reference section. I would recommend that trainees of all specialties dealing with peripheral nerve injuries should read much of this text and it would be extremely useful as a regular reference. It would also make an important and necessary addition to most medical libraries. All clinicians would be well advised to read the chapters on iatropathic injuries, not only for the extensive causes of such injuries encompassing all medical and surgical departments, but also for the precis of the changes occurring in medical negligence claims. This text represents good value for money.

IAN WHITWORTH