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, as part of investigating a failure to thrive and normal growth, a female child underwent cerebral CT. This showed an unexpected arteriovenous malformation 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 until 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 communicant arteries. These 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 review was undertaken before discharge showing normal fundi and fields.

Ten days after the embolisation the patient presented with a generalised, pounding headache, present since discharge. Examination showed mild left papilloedema, with no focal neurological signs. Brain CT showed a dense discharge showing normal fundi and fields. It was made to attempt embolisation. Complete occlusion of the fistulae was achieved by transarterial platinum coil embolisation.

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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 enables results to be available 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 tuberculous 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 Staphylococcus aureus abscesses. The second patient was an elderly male from Bangladesh who had been in Australia for 5 years, with symptoms for 3 weeks. He was reported to have left sided weakness and left sided headache. The polymerase chain reaction was performed at our laboratory on cerebrospinal fluid samples taken on the day of admission, and a month later. The result was reported as negative. Both patients were treated with antituberculous medications, and the first patient was discharged after 12 months. The second patient continued on treatment for 9 months. Treatment was stopped after 18 months when the polymerase chain reaction was negative in a further CSF sample.

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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 characterized by a slow progression of motor and sensory symptoms starting in childhood or adolescence. CMT type 2 is more severe and usually becomes apparent in adulthood. In CMT type 1, the inheritance pattern is usually autosomal dominant, with mutant alleles in a single gene. Of the CMT type 1 genes, CMT1A is the most common, accounting for about 30% of all CMT1 cases. Other important genes include CMT2A, CMT2B, and CMT2D. The genetic diversity in CMT1A is due to mutations in the myelin P gene, which encodes the protein P0, a major myelin component.

In this study, researchers identified a novel mutation in the myelin P gene from a family with Charcot-Marie-Tooth disease type 1B. The mutation was found to manifest as trigeminal nerve thickening, a symptom not commonly associated with CMT1B. The identification of this mutation highlights the genetic and clinical heterogeneity of CMT type 1.

The mutation identified in this study is a missense mutation that results in a substitution of a single nucleotide. This substitution changes the amino acid sequence of the P0 protein, which is critical for myelin formation and function. The mutation is predicted to alter the protein's structure, potentially disrupting its function in myelin biogenesis and maintenance. The identification of this mutation provides new insights into the molecular basis of CMT type 1B and opens up possibilities for targeted therapies aimed at restoring normal myelin function in affected individuals.

In conclusion, the identification of this novel mutation in the myelin P gene expands our understanding of the genetic diversity underlying CMT type 1B and suggests that trigeminal nerve thickening may be an additional clinical feature associated with this disorder. Further research is needed to elucidate the biological implications of this mutation and its potential impact on the clinical presentation and treatment of CMT type 1B.
been recognised in Dejerine-Sottas disease, peripheral neuropathy with an early onset in childhood, and a more severe phenotype than CMT1. CMT1 and Dejerine-Sottas disease are characterised by thickening of peripheral nerves, and thickening of the cauda equina, nerve roots, and ganglia have often been found. Although cranial nerves are generally spared in CMT, thickening of the acousticoptic 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 P0. The cranial nerve involvements in this patient may be associated with the novel missense mutation of P0 (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, absence of deep tendon reflexes, 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 skin fold 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 showed normal findings on neurological examination and peripheral nerve conduction study. Blood samples were obtained from the patient and her mother with informed consent. DNA was extracted from the blood by a standard phenol/chloroform protocol.

The six exons of the P0 gene were amplified by the polymerase chain reaction using primers, and analysed by single strand conforma
tional polymorphism (SSCP) and sequencing analyses. DNA sequencing of exon 3 showed a novel point mutation (A to C at codon 81) resulting in amino acid substitutions of arginine for histidine only in the patient. A DNA duplication in chromosome 17p11.2-p12, including the peripheral myelin protein-22 (PMP-22) gene, was not present. The patient’s mother did not show any mutations in the P0 gene.

CMT type 1 is caused by abnormalities in myelin protein of Schwann cells. Repeated demyelinating and remyelinating responses in the peripheral nerves result in diffusely enlarged diameters of nerves in CMT type 1, and thickening of the cauda equina, nerve roots, and ganglia has also been found. Although blapharoptosis, ophthalmonplegia, facial weakness, deafness, dysphagia, and dysphonia in CMT have been previously reported, clinical involvement of the cranial nerves is rare and thickening of cranial nerves has not been reported except for the acousticoptic nerve in some cases. In the present study, we observed 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 interval.

Trigeminal neuralgia with CMT has been reported. In these rare cases, trigeminal neuralgia was inherited, suggesting a partial symptomatic 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, Kimura reported that the sensory component of the trigeminal nerve was relatively spared, despite extremely delayed conduction of the motor fibers. 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 Arg81His substitution in the P0 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. Arg81His was located in exon 3, which codes for the extracellular domain of P0. 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 P0 mutation, patients with cranial nerve involvement are rare in CMT with a P0 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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References

Intracranial extracerebral follicular lymphoma mimicking a sphenoid wing meningoia

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 but have been described; 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 meningoia, both radiologically and intraop
eratively.

A 47 year old Ghanaian woman was referred with a 3 year history of worsening bitemporal headache, followed by a 6 month history of daily right frontal headache lasting for 2–3 hours associated with mild photopho
ea. There were no reports of seizures, nausea, or other visual disturbances. Her medical history was 3 years of treated hyper
tension, 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...
A lymphoma, was found to be positive. The kappa light chain restriction. Staining for phocytic phenotype (CD20 positive) with frequent mitotic figures and apoptotic bodies dura. Follicular centres were composed of a size and shape and infiltrated the overlying phoid tissue with an ill defined follicular completely removed.

Primary intracerebral lymphomas repre- sent about 2% of intracranial neoplasms and 2% of all lymphomas. They occur most com- monly in the 6th decade of life with a female to male ratio of roughly 2:1.1

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 peritu- morous oedema and 92% enhancement after administration of contrast medium.7

Leptomeningeal lymphoma is usually en- countered as a late complication of systemic non-Hodgkin’s lymphoma, although primary leptomeningeal lymphoma is occasionally seen. The prognosis for these tumours is poor.8 Diffuse intracerebral lymphomas have been mistaken for more common lesions: solitary primary B cell lymphoma of the cere- bellopontine angle mimicking acoustic neu- rilemoma or meningioma has been reported9; Vighushin et al10 reported a patient with a calci- fied temporaloparietal lymphoplasmacytic lymphoma which resembled a meningioma; however, this tumour was entirely extradural.

There is only one previous report of a follicu- lar rather than diffuse primary intracranial lymphoma. Rubinstein9 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 sphenoid wing meningioma suggest that this entity should be considered as a rare differential diagnosis.

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Determinants of the copper concentration in cerebrospinal fluid

The measurement of CSF copper concentra- tion can serve as an indicator of brain copper concentration.1,8 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 dif- fusion of the free or the bound fraction (bound to albumin or coeruleoplasmin) through the blood-CSF barrier. We studied the factors influencing CSF copper concentra- tion using a stepwise multiple linear regression model. The independent variables were age, plasma coeruleoplasmin, CSF/ serum albumin ratio, total serum copper concentration, and calcified serum free copper concentration (based on serum coeruleoplasmin and total serum copper concen- tration). The CSF copper concentration was measured as a dependent continuous type. We investigated lumbar CSF samples from 113 patients. These patients had dementia, extrapyramidal, or tremor symptoms; lumbar puncture was performed to exclude Wilson’s disease, and none of the patients had the disease. Copper was measured by flameless atomic absorption (Perkin Elmer, HGA 500, Ueberlingen, Germany). Coeruleoplasmin was determined nephelometrically (Beck- man Instruments, Brea, CA, USA). The age of the patients was 50.0 (SD15.5) years; 50 were women and 65 men were mean. Serum coeruleoplasmin concentrations were 394.3 (SD17.7) µg/l. Mean serum copper concentrations were 1194 (SD 335) ng/l. Mean calculated free copper concentrations in serum were 78.5 (SD 1285) µg/l. Mean CSF copper concentrations were 14.6 (SD 6.0) µg/l. The mean albumin ratio (AR) was 6.63×10−13. The mean ratio of calculated 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 predictive value of the CSF copper concentration were found to be AR (p=0.0001) and serum coeruleoplasmin (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 CSF (µg/l)=5.32 × (AR)−3 × serum coeruleoplasmin (mg/l). According to this analysis, CSF/serum albumin ratio and serum coeruleoplasmin 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 coeruleoplasmin concentration. It can be assumed from this that in the case of normal blood-CSF barrier function and a normal serum coeruleoplasmin concentration, 25.3% of the measured CSF copper concentration originates from the brain; the CSF by passive diffusion bound to coeruleoplasmin, and only around 0.09% by passive diffusion bound to albumin. In the case of a markedly raised CSF/serum albumin ratio of AR=10×, this would mean that 60.6% of the measured CSF copper originated from the blood (bound to coeruleoplasmin). 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 an intact blood-CSF barrier and a normal serum coeruleoplasmin concentration, 25.3% of the measured CSF copper concentration originates from the brain; the fraction entering the CSF by passive diffusion (bound to coeruleoplasmin) 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 brain. The investigation here also shows that, after determining the CSF copper concentration, the coeruleoplasmin-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 coeruleoplasmin concentration. A statistical relation with a low correlation (p<0.05) between CSF protein content and CSF copper was already shown in 1988 in various neurological diseases; our study shows a much higher significance and, in addition, the effect of serum coeruleoplasmin (therefore of bound serum copper). Furthermore, we have been able to determine quantitatively the fraction 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 Wilson and Schrum 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 tumour, it was agreed that 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 fibroblastic 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. Involvement of the CNS is exceedingly rare and only one other case of a solitary mass is reported along with few cases of CNS involvement in the generalised form of infantile myofibromatosis. The prognosis 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 microscopic variant of meningioma. Meningiomas are extremely rare in this age group, this lesion was not meningeal based and such lesions are usually reactive for epithelial membrane antigen unlike this tumour. This lesion, unlike schwannomas, did not show immunoreactivity for S-100 protein. Haemangiopericytoma is a diagnosis of exclusion and shows no reactivity for actin, unlike this tumour.

Intracranial 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 reported in which the patient died within 24 hours of surgery, secondary to cardiorespiratory 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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Axonal and polynuropathy and encephalopathy in a patient with verotoxin producing Escherichia coli (VTEC) infection

Escherichia coli serotype O157:H7 causes serious food poisoning worldwide, especially in children and elderly people. It is also called verotoxin producing E. coli (VTEC), which produces a cytotoxic Shiga-like toxin. Gastrointestinal, haemorrhagic, and urogenital effects are well known in VTEC infection, and neurological problems are likely to be more frequent than is generally recognised. Here we describe axonal polyneuropathy and encephalopathy in a young female patient associated with haemolytic-uraemic syndrome caused by VTEC infection.

A 26 year old woman began to have abdominal pain and diarrhoea. She was admitted to hospital and diagnosed as having haemorrhagic colitis due to probable food poisoning. Then her urine volume was increased, and serum creatinine increased, and she was transferred to our hospital. On the 9th day she had a high fever of 39.7°C with increased C reactive protein of 7.6 mg/l and a leukocytosis of 17 800/mm³. She was in a state of anuria and her blood analysis showed severe kidney dysfunction (increased serum creatinine of 6.76 mg/l). She had severe anaemia (haemoglobin 6.0 g/dl), fragmentation, and tear drop deformation of red blood cells in the blood smear and increased lactate dehydrogenase concentration of 4095 IU (normal range 230–460 IU), suggestive of haemolytic anaemia. Her platelet count was also decreased to 21 000/mm³. The culture of her stool showed the growth of E. coli O157:H7 and analysis of the bacterial toxins showed the presence of verotoxin, which confirmed the diagnosis of VTEC infection. She received oral sulbactam (an anti-inflammatory agent) and sildac (an anti-inflammatory agent) and showed normal findings in the distal legs, but preserved for pin prick, light touch, and joint sensation. Routine laboratory data including haematological studies, serum chemistry, urinalysis, and CSF analysis were normal. Serum concentrations of vitamin B1, B6, and B12 were normal. Nerve conduction studies were carried out on her right limbs, and showed normal findings in the distal latencies, motor conduction velocities, and F wave latencies of the median, ulnar, and tibial nerves, and no evidence of conduction block. However, there were distal and muscle action potentials (1.18 mV) and mild slowing of motor conduction velocity (41.0 m/s) in the peroneal nerve. There were also markedly decreased amplitudes of the sensory nerve action potentials (0.47 mV) and sural (0.98aV) nerves. These findings and the clinical features confirmed the diagnosis of sensory dominant, axonal polyneuropathy. She was given 300 mg/day sulbactam (an anti-inflammatory agent) and 1500 µg/day methabolamin (vitamin B12) without effect. Two weeks after administration of 300 mg/day oral mexiletin, her numbness and pain gradually disappeared.

The patient was diagnosed as having VTEC infection, because of a typical history of an acute haemorrhagic colitis, the cultured growth of enterohaemorrhagic E. coli O157:H7, and the detection of verotoxin in her stool. She had haemolytic-uraemic syndrome (haemolytic anaemia, thrombocytopenia, and uraemia, following diarrhoea), which is the main complication of VTEC infection. Experimentally, vero cells, an immortalised primate kidney cell line, can be killed by low doses of verotoxin through the process of apoptosis. Verotoxin shows similar cytotoxicity on human glomerular microvascular endothelial cells via inflammatory stimuli such as tumour necrosis factor α, which induced an increase in the numbers of verotoxin receptors, leading to a microvascular thrombosis. Our patient was treated with antibiotics, plasma exchange, and continuous haemodialysis, with benefit.

During the course of the disease, our patient was in a delirious state with visual hallucinations and tonic convolution. She showed mild brain swelling on CT and diffuse slow waves in the frontal area on EEG, evidence of encephalopathy. Previous reports have shown that the incidence of encephalopathy in haemolytic-uraemic syndrome (most of VTEC infection) is 5%, including seizures in 17%–44%, altered consciousness in 7%–40%, and paralysis in 1%–16%. Many of the patients, including ours, seemed to have metabolic encephalopathy, but some developed encephalopathy without metabolic abnormalities. There is experimental evidence that verotoxin has direct virulence to both endothelial cells and neurons in the nervous system, and its initial lesion is in the hypothalamic areas, then
Crying spells as symptoms of a transient ischaemic attack

In the absence of depression, crying spells associated with neurological 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 transient ischaemic attack with multiple crying spells, the localisation of his attack is unclear; involvement of the right thalamus or neighbouring internal capsule is a possibility. Similar to spells of laughter, spells of crying may occur in relation to unilateral cerebrovascular events. Although most reports of crying after unilateral strokes have reported left hemispheric lesions, crying also may result from right hemispheric strokes. Even more similar to our patient, sudden laughing spells, “le fou rire prodromique,” rarely precede strokes involving the left capsular-thalamic, lenticulocaudate, or pontine regions. Our patient may have had a comparable phenomenon from the right hemisphere. The pathophysiology for this phenomenon may have been transient activation or stimulation of ischaemic motor pathways.

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Continuous drop type of orthostatic hypotension

Orthostatic hypotension has usually been evaluated for 2–10 minutes after standing.1 2 Multiple system atrophy (MSA: Shy-Drager syndrome) is one of the neurodegenerative diseases which show marked orthostatic hypotension. We studied changes of blood pressure for more than 20 minutes after standing in 30 patients with MSA. The patients lay down on a tilting table, and an intravenous cannula was introduced into the cubital vein more than 30 minutes before the 25 minute test of 60° head up tilt. Blood pressure and heart rate were recorded every minute with an automatic sphygmomanometer. Patients could clearly be classified into two groups in terms of the time taken to reach the minimum blood pressure. In 12 patients systolic blood pressure fell rapidly, reached a minimum within 5 minutes, and then remained stable or partially recovered (early drop type); whereas, in 13 patients blood pressure fell immediately after tilting but kept decreasing by more than 8 mm Hg from that at 5 minutes (mean 12.9 mm Hg;
with orthostatic hypotension of the continu-
gate the haemodynamics in three patients
syncope more than 20 minutes after standing.
endurance for more than 10 minutes of exer-

A slight increase in packed cell volume
gng/ml) during the decrease in blood pressure.

between 5 and 20 minutes was noted

noradrenaline (norepinephrine) level (+0.05

type, there were no decreases between 5 and

drop in blood pressure and heart rate seen in

with MSA the blood pressure drops continu-
ously on standing. The continuous blood
decease in blood pressure, cardiac output
proportionally decreased but systemic vascu-
lar resistance did not change (figure).

Most patients with continuous drop type
orthostatic hypotension reported reduced
endurance for more than 10 minutes of exer-
cise (easy fatigability). Two experienced
syncpe more than 20 minutes after standing.

We used a Swan-Ganz catheter to investi-
gate the haemodynamics in three patients
with orthostatic hypotension of the continu-
ous drop type. To prevent the concentration of plasma, saline of calculated volume was
infused during tilting. During the continuous
decease in blood pressure, cardiac output
proportionally decreased but systemic vascu-
lar resistance did not change (figure).

Our results suggest that in many patients
with MSA the blood pressure drops continu-
ously on standing. The continuous blood
pressure drop is caused by continuous reduc-
tion of cardiac output. A part of the
mechanism for continuous reduction of cardiac output should be lack of reflex tachy-

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ously on standing. The continuous blood
pressure drop is caused by continuous reduc-
tion of cardiac output. A part of the
mechanism for continuous reduction of cardiac output should be lack of reflex tachy-
cardia and no significant release of noradren-
aline which are caused by interruption of the
baroreflex arc, as is known in MSA.1

However, further explanation, such as con-
tinuous vasodilatation of the volume vessels,
is necessary for the difference in mechanisms
between the early drop type and the continu-
ous drop type. As we did not record heart rate
and blood pressure continuously and did not
evaluate ventricular function by echocardio-
graphy, the final conclusion and its inter-
pretation require further study.

We think that more than a 20 minute tilt up
study is needed to evaluate orthostatic hypo-
tension and that reduced endurance of
exercise and the syncope that occurs some
time after standing should be considered
symptoms of a continuous drop in blood
pressure.

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CORRESPONDENCE

Respiratory aspects of neurological disease

An account of respiratory aspects of neuro-
logical disease, such as the highly informative
one presented,1 would be incomplete without
mention of breathlessness resulting from
neurogenic pulmonary oedema, character-
ised by an “increase in extravascular lung
water in patients who have sustained a change
in neurological condition”. Neurological
disorders associated with this syndrome
include subarachnoid haemorrhage, middle
cerebral artery stroke, and cerebellar
haemorrhage.2 Brain stem stroke, acute
hydrocephalus due to colloid cyst of the third
ventricle, closed head injury, and status
epilepticus, were also documented as risk
factors in a literature review by Smith and
Matthey,3 who proposed, on the basis of their
own study, that increased pulmonary vascular
hydrostatic pressure might be a more signifi-
cant aetiopathogenic mechanism than in-
creased pulmonary capillary permeability.2 A
more direct link between neurogenic myocar-
dial damage and pulmonary oedema can be
postulated when subarachnoid haemorrhage
is complicated by reversible severe left
ventricular dysfunction, as documented in
two cases reported by Wells et al.4

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Idiopathic cerebellar ataxia associated with celiac disease: lack of distinctive neurological features

Although applauding the contribution of Pellecchia et al., we do not cover neurogenic pulmonary oedema. We agree, however, that it can be a difficult clinical problem and therefore appreciate his contribution.

M P POLKEY


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, 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 their 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 finding could explain the relatively 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 antigiadin by Hadjivassiliou et al.

Our series is too small to estimate the sensitivity of both antigliadin and antiendomysium antibodies in glutten ataxia; unfortunately Hadjivassiliou et al did not report any data on antiendomysium antibodies 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 in contrast with the 2% of antigliadin antibody 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.

M T PELLECCHIA

the EMG pattern in patients with myotonic dystrophy show a multitude of defects including expression of myotonia, myopathies, muscular atrophy, and neural abnormalities.  

The possible management of myotonia and some of its clinical manifestations, such as dysphonia,  

by antimyotonic drugs (disopyramide 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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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 presented signs of lower motor neuron disease in the upper limbs without significant functional involvement of other regions upon clinical presentation. This subgroup of patients is clinically characterised by the display of progressive atrophy and weakness in the arms with little effect on the bulbar muscles or legs. Atrophy and loss of strength affect the upper limb muscles in a more or less symmetric manner, prevalent in the proximal muscles. The comparative study with the rest of the amyotrophic lateral sclerosis group supplies very interesting details for the physician, such as a clear predominance among men, and a longer median survival. They conclude by suggesting that this syndrome could be a new variant of amyotrophic lateral sclerosis.  

Finally, the authors carry out a historical review and refer to the fact that this distinctive amyotrophic 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 Vulpian in 1886, known in Franco-German literature as Vulpian-Bernhart’s form.  

In his book Maladies du Systeme 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, we have come to use the eponym of Vulpian-Bernhart’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 prevalence in the proximal muscles of the upper limbs is unknown. We can furnish little more information in this respect. However, in the 1960s, in the differential diagnosis of this syndrome, it was proposed that the muscles predominantly affected in Vulpian-Bernhart’s form were the deltoideus, the infraspinatus, the supraespinaus, the sternocleidomastoideus, and the teres minor. The predominant involvement in these muscles permitted its distinction from that previously called Erb’s dystrophy.  

As a consequence of the atrophy of these muscles, the upper limbs adopt a characteristic position, with the shoulders slumped, and the arms, forearms, and hands in pronation. As the illness progresses, the hand muscles are affected, with atrophy of the following muscles: opponens pollicis, flexor brevis, adductor pollicis brevis, adductor pollicis, interossei, and lumbricales, which leads to the formation of the characteristic Archen-Duchenne hand.  

Obviously, signs of corticospinal involvement with hypereflexia 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-Bernhart’s, rebaptised as flail arm syndrome from other motor neuron syndromes such as of the spinal muscular atrophies, Kennedy’s disease, multifocal motor neuropathy, and monomelic amyotrophy.  

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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 are.  

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 true inception cohort study where we actually do not have any 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 disorder 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 because it too has only 117 subjects, yet is much quoted. Setting aside for the moment that the Swiss study is hampered by the selection atrocity of advertising for subjects, and has a host of other reportedly fatal faults, and giving some benefit of the doubt, the study is said to be an accurate representation of the state of affairs in Switzerland at that time. Yet, in Switzerland, not even 60% manage to recover fully by 3 months and many of these were reporting total disability during that time, whereas the Lithuanians fully recover in 4 weeks or less, with little or no therapy, and no disability. Studies in other western countries disclose an even greater contrast, with 50%–70% of patients reporting pain even after 3–6 months, despite the fact that all these studies are examining the same grades (1 and 2) of whiplash associated disorders.  

Thus, while the sample size is small in this Lithuanian study, it is comparable with others reporting the prognosis of whiplash, and yet gives a different picture of outcome.
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 with absolutely no vehicle damage, in very low velocity collisions, are just as likely to report chronic pain as those in more severe collisions.6 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 culture may be 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 determine whether 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 cultures 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 acute whiplash injury 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 Germany7 and Greece.8 The time has come 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 our responsibility to utilise this time to help prevent the chronic pain and the suffering we otherwise encounter.9

R FERRARI


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 timely topics 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. I was pleased to see in it the outstandingly useful technique in chronic low back pain and cancer pain will also be of interest to those in other fields.

The chapter on the anatomy and physiology of pain is excellent in that it has clear explanations and a number of very helpful diagrams. Unfortunately it fails to mention increasing understanding of the role of GABA in mediating analgesia within the spinal cord and furthermore does not mention some of the other major changes which are well known to occur in chronic pain states such as central sprouting and phenotypic switching.

The chapter on pharmacology of acute and chronic pain is well written, but unfortunately a lot of time is spent on non-steroidal drugs. There is a review of the adjuvant drugs such as antidepressants and anticonvulsants that are used in chronic pain, however one is left at the end with a sense of knowing about the drugs but not quite to use them. There is no mention of the increasing use of gabapentin or other of drugs that are sometimes used in chronic pain states such as clonidine and other sympathetic agents or calcium channel blockers.

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 as 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 denervations, 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 and that 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 cryosurgery. 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 think one would expect to do to a bigger text for. In summary I think that this volume would make an excellent addition to the bookshelf of those involved in the treatment and management of pain.

RAJESH MUNGALI

BOOK REVIEWS


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 neurophysiologist with a longstanding interest in the dementia, 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.

The title of the book is perhaps a little misleading in that the book includes, as well as traditional neuropathology, a very comprehensive overview of the molecular biology and genetics of the dementias. As would be expected, a considerable proportion of the book is dedicated to Alzheimer’s disease with chapters on both the clinical features, genetic, and the neuropathology. The frontotemporal dementias are also well covered and the book includes a chapter on the recent neupathological and genetic advancements related to chromosome 17 linked dementias. There are also sections on progressive supranuclear palsy, Huntington’s disease, corticobasal degeneration, dementia with Lewy bodies, and prion diseases and vascular dementia.

The editor has managed to persuade many of the world’s experts to contribute. For instance, one of the chapter on prion diseases is by D’Almond and the recent Nobel laureate Prusiner, and the frontotemporal dementias are reviewed by Brun and Gustafson. Genetics of Alzheimer’s disease are dealt with by St George-Hyslop and the neuropathology of Alzheimer’s disease by Price and coworkers.
The standard of illustrations is excellent and the style generally very readable. I shall certainly find it extremely useful as a work of reference and for teaching purposes. The editor is to be complimented on producing such a delightful work.

John Hodge


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 reconstructed 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 textbook 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 précis of the changes occurring in medical negligence claims. This text represents good value for money.

Ian Whitworth
Respiratory aspects of neurological disease

O M P JOLOBE

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