LETTERS TO THE EDITOR

Creutzfeldt-Jakob disease presenting as complex partial status epilepticus: a report of two cases

Creutzfeldt-Jakob disease is a transmissible human spongiform encephalopathy which may be familial, iatrogenic, or sporadic. The classical clinical features include a rapidly progressive dementia with the patient retaining clear consciousness until the terminal stages of the disease. We report on two patients presenting with a rapidly declining level of consciousness, in whom the clinical picture and EEG were suggestive of complex partial status epilepticus.

The first patient was a 58 year old woman who was admitted to a psychiatric unit with a short history of mood disturbance, confusion, and unsteadiness. A provisional diagnosis of agitation depression was made and she was started on lorazepam. She then became unsteady on her feet and required support when walking. She had had occasional complex partial seizures for 30 years but at presentation was not taking any anticonvulsant drugs.

On examination, she appeared perplexed, tearful, and agitated, and was unable to give a coherent history. She was intermittently confused and her gait was ataxic. There were no other cerebellar signs. The rest of the neurological examination was unremarkable although limited by poor cooperation. She became more withdrawn and uncommunicative with incontinence of urine. She would occasionally jump when sitting in a chair.

Brain CT and MRI were normal, as was her CSF. An EEG showed frequent, almost continuous variable amplitude sharp waves in all areas, although with a right sided emphasis, with a repetitive appearance up to 2 per second (figure). The record was thought to be in keeping with partial status epilepticus.

Her level of consciousness deteriorated despite intravenous valproate and phenytoin and she was transferred to the intensive care unit for continuous EEG monitoring. On arrival, she was deeply unconscious and despite aggressive management of her presumed complex partial status she died 3 weeks later. Histology of the brain was diagnostic of the sporadic form of Creutzfeldt-Jakob disease.

The second patient was a 68 year old man who was admitted to a psychiatric unit with a short history of mood disturbance, confusion, and inappropriate behaviour. He appeared not to recognise his family. Initially he was dysphasic and obtunded. His conscious level then deteriorated and he became mute with evidence of right sided weakness. All investigations including contrast enhanced brain CT and CSF examination were normal. An EEG was reported as showing frequent bilateral epileptiform activity, but there was no improvement in his clinical state after a loading dose of intravenous phenytoin. A repeat EEG was dominated by periodic lateralised epileptiform discharges (PLEDs), more marked on the left side. A third EEG 3 weeks later again showed frequent predominantly left sided epileptiform discharges, which were attenuated by a bolus of intravenous diazepam but without any improvement in his clinical condition.

He was transferred to this hospital for artificial ventilation because of the concern that he was in complex partial status. On admission he was mute, his eyes were closed, and he flexed to pain on the left side only. Intermittent twitching of both sides at a rate of between 1 Hz–2 Hz was seen. Reflexes were brisk and symmetric. His right plantar response was extensor, his left flexor.

A further EEG 5 days later showed generalised synchronous continuous periodic sharp waves occurring at a frequency of 1.3 Hz, at times in the form of biphasic or triphasic complexes. Myoclonic jerks occurred during the recording.

It was considered that overall these features were consistent with a diagnosis of Creutzfeldt-Jakob disease. His condition continued to deteriorate and he died 2 weeks later. A request for a postmortem examination was refused.

These two cases illustrate a previously unrecognised presentation of Creutzfeldt-Jakob disease, namely presumed complex partial status.

In the first case, the interpretation of the EEG findings was made more difficult by the patient’s depressed conscious level and the previous history of complex partial seizures, albeit mild. The initial psychiatric presentation, with mood and behaviour disturbance, as well as fluctuating confusion, was compatible with complex partial status. The initial EEG report, suggestive of partial status epilepticus, prompted treatment, unsuccessfully, with anticonvulsant drugs and subsequent transfer for continuous EEG monitoring. This disclosed marked fluctuations, including discrete runs of rhythmic sharp waves that were considered to be electrographic seizures. Even after sustained burst suppression, the recording fluctuated between generalised periodic discharges and periods of relative inactivity within a matter of seconds.

In the second case, the patient developed focal seizures and PLEDs on the EEG. The initial recordings were suggestive of complex partial status, with asymmetric discharges abolished by diazepam but without any observable clinical change. Subsequent recordings were more characteristic of Creutzfeldt-Jakob disease, particularly as the patient had developed myoclonus. Although the electrographic changes were abolished by diazepam, suggesting seizure activity, the modification of both clinical and EEG activity in Creutzfeldt-Jakob disease by benzodiazepines has been reported giving rise to further confusion with epileptiform sharp wave activity. The focal nature of the patient’s signs and the laterisation on the EEG is well recognised in Creutzfeldt-Jakob disease as are

Initial EEG at presentation in an acute confusional state, showing virtually continuous semirepetitive sharp waves with some right sided predominance. Although seizure-like evolution of discharges was not seen, the electrographic picture was considered to be in keeping with complex partial status.

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[Image: EEG recordings showing periodic lateralised epileptiform discharges (PLEDs)]
periodic PLEDs, which are often associated with contralateral myoclonic jerks. The two cases described here illustrate that a diagnosis of Creutzfeldt-Jakob disease should be considered where a rapid decrease in consciousness is accompanied by EEG changes apparently compatible with complex partial status. When there is a clinical suspicion of Creutzfeldt-Jakob disease, the ideal method of monitoring such patients is with continuous EEG recording, allowing documentation of rapid fluctuations. The present cases are atypical in that the progression from presentation to death was rapid, but they underline the fact that minute to minute changes in EEG rhythm, asymmetry, and electrographic responsiveness to benzodiazepines can all be seen in Creutzfeldt-Jakob disease.

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Childhood demyelinating diseases with a prolonged remitting course and their relation to Schilder’s disease: report of two cases

Schilder’s disease or myelinoclastic diffuse sclerosis is a rare acute or subacute demyelinating disorder which primarily affects children and young adults.1 2 We report the clinical and neuroradiological follow up of two boys affected by a demyelinating disease with a prolonged relapsing-remitting course, response to corticosteroids, and relatively good long term prognosis.

The first patient presented at the age of 12 with a 2 month history of repeated episodes of headache and blurred vision followed by weakness of the left leg, lasting a few hours. Head CT and bilateral carotid angiography were normal. Two weeks later the left hemiparesis and headache recurred. T2 weighted images on brain MRI disclosed a hyperintense signal in the right parieto-occipital white matter, involving the right centrum semiovale, with mass effect.

The second patient was admitted at the age of 4 because of the sudden onset of headache and vomiting with ataxia and drowsiness followed by generalised clonic seizures. Clinical examination on admission showed left hemiparesis, anisocoria (left>right), and dysarthria. Ocular fundoscopy was normal. Head CT disclosed a reduced right lateral ventricle and subarachnoid spaces and, 1 week later, a small hypodense area in the right periventricular white matter. A carotid angiogram was normal. At the age of 5 the child had yearly relapses then every 48 hours for 20 days) induced a rapid and fatal, or a relapsing-remitting course. Most patients have neurological sequelae during follow up and few patients die fully recovered.3 Histological studies typically show a demyelinating process similar to that of multiple sclerosis, with an inflammatory perivascular infiltrate, and in severe cases, cystic lesions. Neuroimaging findings tend to parallel the clinical course. Corticosteroids may improve the outcome of the single relapse and possibly of the disease, as they did in our patients. Some patients respond to immunosuppressive therapy.

In both the patients described the association of headache, signs of diffuse and focal brain dysfunction, a relapsing course, and the response to corticosteroids may raise the possibility of an isolated CNS angiitis, a condition primarily affecting middle aged and elderly people. But neither cerebral angiography nor histological examination disclosed a primary vascular disorder. In addition, the early onset and the sporadic occurrence of the disorder rule out another recently described vasculopathy often associated with familial hemiplegic migraine.1

In conclusion, although demyelinating diseases that do not fulfill the classical definition of multiple sclerosis or encephalomyelitis remain difficult to label in children, the two cases we report here seem to fit Schilder’s description of myelinoclastic diffuse sclerosis. Owing to the current lack of knowledge on the causes of this disease strict diagnostic criteria cannot be applied. Some presentations may warrant brain biopsy. The differing clinical and neuroimaging features seen in these patients may help in delineating Schilder’s disease subtypes.

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1 Schilder P. Zur Kenntnis der sogenannten diffusen Sklerose. Zeitschrift für die Gesamte Pathologie und die Klinische Medizin 1912;63:60.


ing Orbis-Sigma Valve (Elekta-Cordis) may also reduce clinical complications related to overdrainage in the upright body position. It prevents excessive CSF drainage by instantaneously increasing its hydrodynamic resistance when the drainage rate rises.

The new Codman SiphonGuard device is intended to reduce the drainage rate when the flow dramatically increases during transition from a horizontal to vertical body position. It consists of two passages for the flow, which constitutes a high hydrodynamic resistance. This action may help to prevent posture related overdrainage.

We tested a sample of three SiphonGuards (kindly provided by Johnson and Johnson) in the United Kingdom Shunt Evaluation Laboratory to characterise the hydrodynamic performance of the device and its ability to reduce posture related overdrainage.

The pressure flow performance curve consisted of two straight lines of different slopes, both crossing the origin. They represent the two possible states of the SiphonGuard—low resistance (mean of 1.5 mm Hg/ml/min) and high resistance (mean of 42 mm Hg/ml/min, figure A). The differential pressures resulting from the above values, providing the CSF flow is on average 0.3 ml/min in the horizontal body position, would be 0.45 mm Hg and 12.6 mm Hg respectively.

Switching between low and high resistance was initiated by a flow rate, the threshold of which varied between 0.7 and 1.8 ml/min (figure B).

Switching from the high to low resistance was initiated by the differential pressure decreasing below the threshold from 4 to 6 mm Hg.

Overall, the mechanism of the SiphonGuard seemed to work according to the designers’ intention. It is supported by the concept that, during rapid transition from horizontal to vertical body position, initial flow rate increases above 2–3 ml/min. This is enough to switch the valve to the high resistance state, limiting overdrainage. However, in practice, it may not always be the case. In patients with small or slit ventricles previously having overdrainage, CSF may not be available to produce the flow at such a high rate. Moreover, because reliable switching occurs above 1.8 ml/min, in shorter persons or in patients resting persistently in a semisitting position (for example, elderly patients watching TV or reading books) the drainage rate of 1 ml/1.5 ml may cause clinical deterioration without initiating the antisiphon action of the SiphonGuard. Another possible drawback concerns the reverse change—that is, switching back from high to low resistance, to be expected when a patient moves from a vertical to a horizontal position.

The device may not return to its state of low resistance. If the resistance switching mechanism is indeed triggered by a differential pressure (with a threshold of around 5 mm Hg) the SiphonGuard may stay in the high resistance state permanently. If its high hydrodynamic resistance may force the differential pressure to persist higher than 9–16 mm Hg, under conditions when the CSF drainage rate should equal its formation rate (0.2–0.4 ml/min). Hence, it is possible that the device may remain “locked” in the high resistance state, causing underdrainage in the horizontal body position.

In vivo, the device may contribute to the significant fluctuations of pressure resulting from the difference between the operating pressures for low and high resistance—similar to that described for the Orbis-Sigma Valve. Moreover, it may not prevent the overdrainage related to nocturnal vasomotor pressure waves3, as often reported in paediatric cases.

These reservations, based on our short laboratory study, should be taken into consideration both by neurosurgeons and the manufacturer. Whether they cause system malfunction under specific clinical conditions remains to be shown. We advocate a well controlled multicentre study on this new and interesting device together with in vivo measurements of shunt function using a CSF infusion test during tilting.

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Convulsions induced by donepezil

Donepezil, a centrally acting acetylcholinesterase inhibitor, has been recently introduced for the symptomatic relief of cognitive impairment in patients with mild to moderate Alzheimer’s disease. Several adverse events thought to be related to donepezil have been reported so far, the most common ones being gastrointestinal disturbances due to cholinomimetic effects of donepezil. Convulsions have not been reported for donepezil to date. We report on a patient with mild Alzheimer’s disease who presented with convulsions during treatment with donepezil.

The patient was a highly educated, ApoE4 homozygous, 72 year old man, who was diagnosed with dementia of probable Alzheimer’s type (NINCDS-ADRDA criteria) 14 months previously. His medical history, with the exception of non-familial dementia, was unremarkable and his only medication was 100 mg aspirin daily. His mini mental state examination score was 22 points. He was treated with 5 mg donepezil once daily for 2 weeks, and then 10 mg a day for 23 days when he was admitted due to convulsions. The patient was unconscious for 40 minutes with urinary incontinence and bitten tongue. Blood analyses were normal. A contrast CT showed a mild degree of cortical atrophy with no structural lesions. EEG showed mild and diffuse neuronal dysfunction with the absence of grafoelement indicative of epileptic discharges. Donepezil was discontinued and no other therapy was instituted. Six weeks later 5 mg donepezil once daily was restarted. On day 52 of donepezil treatment the patient’s caregiver had reported loss of consciousness and convulsions in our patient. The donepezil was discontinued and 100 mg indomethacin a day was prescribed. For the subsequent 8 months the patient has been convulsion free and his current mini mental state examination score is 18.

Convulsions in Alzheimer’s disease are rare until late in the illness, when up to 5% of patients reportedly have infrequent seizures.1 We think that convulsions reported in our patient could be due to donepezil. It has already reported that some centrally acting cholinesterase inhibitors—that is, tacrine, varenicline, and physostigmine—might induce convulsions in patients with Alzheimer’s disease. The mechanism of convulsive action of acetylcholinesterase inhibitors is not clear. As donepezil seems a useful drug in some of the patients with Alzheimer’s disease who presented with convulsions during the exemption of non-familiar dementia, was thought to be related to donepezil have been reported so far. We think that this report will extend our knowledge of donepezil’s safety profile.

Severe toxic neuropathy due to fibrates

The main adverse effects of lipid lowering agents in the fibrate family involve the gut, the skin, the liver, the blood, and the muscular system. Some of these complications are more frequent when renal failure exists.1 Here we report a case of neuropathy secondary to long term treatment with fenofibrate to a patient without renal failure taking recommended doses.

A 60 year old man was seen in September 1996 complaining of leg pain for 6 months. His relevant medical history included coronary artery disease treated for 10 years with 6 mg molsidomine/ day and 60 mg isosorbide dinitrate/ day, high blood pressure and hyperlipidaemia treated respectively with 100 mg atenolol/ day and 0.5 mg fenofibrate/day for the past 5 years. He complained of paresthesias along the posterior aspect of both thighs, later complicated by progressive muscle weakness.

The physical examination disclosed a patient incapable of standing on his toes or heels. No proximal muscle weakness was present. The deep tendon reflexes were reduced in all limbs. There was no sensory loss to light touch, vibratory sense, pain perception, and joint position sense. There was no disturbance of sphincter control or postural fainting and no impairment of potency to suggest dysautonomia. The rest of the physical examination was within normal limits. The EMG suggested an axonal sensorimotor neuropathy with reduced amplitude of nerve action potentials without any significant slowing of conduction velocity. There were spontaneous fibrillations in the right tibial anterior muscle. The complete blood count, erythrocyte sedimentation rate, fibrinogen, C reactive protein, and serum protein electrophoresis were normal. Muscle enzymes were normal. Immunological studies (antinuclear factor, serum and urinary immunoelectrophoresis, circulating immune complexes, serum complement, ANCA) were negative. Antiaripiphenic and antilygloipid antibodies were not detected. Two CSF examinations were performed: the CSF contained 1 white blood cell/mm³, the protein concentration was 2.5 mmol/l. Accessory salivary gland biopsy eliminated amyloidosis, sarcoidosis, and Gougerot-Sjögren’s syndrome. Nerve biopsy confirmed axonal damage and disclosed a focal perivascular inflammatory lymphocytic infiltrate without vasculitis. No ultrasound study was performed. An adverse drug effect was suggested in May 1997 and fenofibrate was discontinued. Three months later the patient informed us of a remission in the distance he could walk. In December 1997, he no longer complained of myalgia. Improvement in motor function was apparent; the patient could now stand on his toes and heels without help.

Axonal sensorimotor neuropathy was confirmed in this case by electrophysiological and histological findings. Other common causes of axonal neuropathy were excluded and a toxic cause was considered.1 Because the patient had been receiving all of his medications well before the beginning of the clinical manifestations, there was no chronological argument targeting any one drug in particular. However, a review of the literature suggested fenofibrate as the causative agent as neuropathies have been described after treatment with clofibrate and bezafibrate.2–4 In addition, none of the other drugs he was taking have been associated with neuropathy.

The role of fenofibrate was confirmed by regression of the symptoms after discontinuation of this drug without the addition of any other treatment. There are no previous reports of histological findings in neuropathy due to fibrates. The delay between initial treatment with fenofibrate and the appearance of the symptoms as well as the time required for them to regress, suggest a cumulative toxic effect but no other predisposing risk factor such as high dosage or renal failure was present.

In conclusion, fibrates can be responsible for neuropathies even when given in approved doses and in the absence of renal failure.

CORRESPONDENCE

Macs with multiple sclerosis

Rothwell and Charlton have suggested that Scottish ancestry is associated with an increased susceptibility to multiple sclerosis. They make the novel observation that a higher than expected proportion of patients with multiple sclerosis had Scottish surnames as defined by the prefix Mc or Mac. They quote that the percentage of the population in the Highlands and Islands with a surname of Mc or Mac is 22.6%. They then suggest that this is the percentage with Mc or Mac in Orkney and Shetland but these islands are not part of the Highlands and Islands. In Orkney and Shetland, in fact, only 3.5% of the population have a surname beginning with Mc or Mac, which is much lower than the percentage in north east Scotland—namely 7.5%.

Rothwell and Charlton do make the point, however, that an increase in the proportion of surnames prefixed with Mc or Mac with latitude within Scotland is not associated with an increase in the prevalence of multiple sclero-
High incidence and prevalence of multiple sclerosis in south east Scotland: evidence of a genetic predisposition

We read with interest the results of Rothwell and Charlton regarding the incidence and prevalence of multiple sclerosis in south east Scotland. They have identified standardised multiple sclerosis prevalence rates for the Lothian and Border Regions of 203 and 219 per 100 000 respectively, the results challenging the theory that the high prevalence rates previously reported in Scotland are peculiar to the north east and its offshore islands. The authors postulate that the apparent step in prevalence rates between England and Scotland may be due to the distinctive Celtic ancestry of the Scottish population as can be crudely measured by surnames prefixed with Mc or Mac.

In Northern Ireland we have also identified a much higher prevalence rate for the disease than exists in England and Wales and have speculated that the similar rate to that in Scotland is at least partly a function of the common ethnic origins of the two populations. The contiguous region of Coleraine, Moyle, Ballymena, and Ballymoney lies less than 20 miles from Scotland and is closest point and has a standardised prevalence rate for multiple sclerosis, based on the 1961 census population for Northern Ireland of 208 per 100 000, which is one of the lowest in the United Kingdom and yet these islands have the highest percentage of Scottish surnames.

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The author’s reply:
The study of the prevalence of multiple sclerosis in Northern Ireland by McDonnell and Hawkins is interesting.1 The findings are similar to those of the recent study in south east Scotland.2 They suggest that there is an increased prevalence of multiple sclerosis in the north of the British Isles compared with the south. It seems likely, as McDonnell and Hawkins suggest, that this at least partly may reflect a genetic susceptibility of the respective populations.

The south east Scotland study did, as Shepherd suggests, attempt to link the high prevalence of multiple sclerosis to Scottish ancestry. However, the study used a standard text of several hundred surnames which are considered to have originated in Scotland, rather than just those prefixed with Mc or Mac. This is obviously still a very crude approach to the problem and any bias is likely to have weakened rather than strengthened the association. The proportion of cases in the telephone book with a surname pre-fixed with Mc or Mac was simply used as a crude illustration of the fact that the differences in apparent ancestry between the Scotland and England are still considerable. This is supported by major differences in the HLA types of the two populations.3 Contrary to Shepherd’s assertion, the Highlands and Islands telephone book does include Orkney. However, he is correct to point out that the prevalence of surnames prefixed with Mc or Mac is indeed lower on Orkney than in the region as a whole.

Further insights into the high prevalence of multiple sclerosis in the north of the British Isles might come from a prevalence study which is currently being planned on the Isle of Skye.

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4 Swingler RJ, Compston DAS. The distribution of subarachnoid cysts; a family name study in Great Britain. Harrison, 1980.
5 Gorgas HI. Multiple surnames in Great Britain. Harrison, 1980.
out the need to conduct a long term, placebo controlled trial with precise end points, proper randomisation, sample size calculations, and predetermined statistical calculations, to evaluate properly the effectiveness and determine the indications of aetiological treatment for neurocysticercosis.

In the era of evidence-based medicine, we neurologists and general practitioners should be demanding regarding use of sound scientific information with methodological care for improving our clinical decision making. Medical information from reports that do not conform to the minimal requirements of a clinical trial should be avoided.

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The author’s reply:

I celebrate the rigid academic standards of Carpio’s medical practice, but wish they were familiar with the disease. The problem with albendazole therapy just for the sake of science. It has already been demonstrated) but to document if albendazole could also be useful in a severe form of neurocysticercosis that has been associated with a grim prognosis.

Under these circumstances, it is not ethical to deprive a group of patients a safe and inexpensive treatment just for the sake of science. In addition, Carpio’s concerns about the criteria we used for the diagnosis of subarachnoid cysticerci—two typical of those who are not familiar with the disease. The problem with CT is that this imaging method may misdiagnose some subarachnoid cysts as parenchymal cysts, but the opposite is not true.

As a physician interested in the advancement of science, I applaud Carpio’s interest in evidence-based medicine but I completely disagree with him in that information from reports other than clinical trials should be avoided. He must remember that outstanding contributions to medical knowledge have been made through single case reports, small clinical series, and open trials. On the contrary, the “double-dummy” has been the shield of major medical frauds. Medicine is art and science, and wise physicians know that information from clinical findings actually have a “significant” impact on everyday clinical practice.

PETER MARTIN


There are two parallel strands to the development of our understanding of immune mediated disorders of peripheral nerve. The first grew from the demonstration in the 1950s, by Waksman and Adams, that rabbits immunised with homologous sciatic nerve and adjuvant developed an inflammatory demyelinating neuropathy. This model, experimental allergic neuritis, the CSF characteristically shows a raised protein concentration and a paucity of cells. These findings repeated those of Guillain-Barré and Strohl on the CSF abnormalities of Landry’s disease and so spawned the notion that Guillain-Barré syndrome was immunologically mediated. Accordingly, over the past 20 years patients with Guillain-Barré syndrome and chronic inflammatory demyelinating polyneuropathy with the CSF abnormalities of Landry’s disease have been exposed to immunosuppressive regimes borrowed from other inflammatory disorders.

The second important development has been the growing understanding of the relation between plasma cell dyscrasias and peripheral neuropathies. The association of peripheral neuropathy and myeloma was noted in the 1930s and that with an IgM monoclonal gammopathy was reported in the 1960s. Twenty years later IgM antibodies were found that were directed against myelin associated glycoprotein. This work has gathered pace and over the past 10 years, peripheral neuropathies have been described in association with specific angiotensin and antithromboid antibodies.

Alongside this expansion of interest in the immunology of peripheral nerve disorders, new infective neuropathies have emerged such as those due to HIV and Lyme disease, first recognised in 1993. It is an appropriate time then for this authoritative text on immune mediated neuropathies. The scope of the book is wide, including scientific overviews of immune interactions in the peripheral nervous system as well as pragmatic accounts of the use of immunosuppressant drugs and the management of neuropathic pain. The inflammatory demyelinating and axonal neuropathies and antibody associated neuropathies are comprehensively surveyed, as well as more difficult entities such as the post-polio syndrome and the rare toxic inflammatory neuropathies. The dry review of silicone neurotoxicity by Rosenberg is a special treat. British readers may be surprised to find only one United Kingdom contributor to this American-Dutch edited text, whereas 18 authors are American, 11 are from The Netherlands, two each from Italy, Japan, and Israel, and one each from Canada, Nepal, and Switzerland. It is not cheap, but it has no equal as a comprehensive, accessible, and useful resource for the practising neurologist.

ALASDAIR COLES

This is a handsome and liberally illustrated guide to the success and frozen section diagnosis in neuropathology. This aspect of practice remains a central part of a clinical neuropathologist’s role and this book can be recommended to trainees and practitioners for its wealth of illus and an excellently oriented text. It is particularly useful to see a wide range of appearances for each tumour illustrated—for example, 20 figures illustrating metastatic tumours, 13 illustrating pituitary adenomas, and 38 illustrating various grades of astrocytic tumours. This enables the less readily diagnosed examples to be considered as well as more typical varieties. Typical varieties tend to be the only ones illustrated in a less specialised text. There are 18 chapters that cover each of the main types of tumour encountered as well as providing advice on making and interpreting smears and dealing with lesions that do not smear well. The emphasis is on using smears as standard preparations with frozen sections as back up when required—a procedure that is probably adopted in most neuropathology departments.

The success of a book like this depends crucially on the quality of the photographs. These are, appropriately, all in colour. Many are of excellent quality. Some are intentionally obscure—for example, to make the point that dermatofibrosarcomas may be too tough to examine in smears (fig 18.1). A few have rather poorly defined features and these tend to be illustrations of frozen sections which inevitably lack the crispness of smears. However, even these illustrate the points intended. The legends to the figures are full enough to avoid the need for arrows that might otherwise have obscured the images. There is a useful index, but the book would have benefited from more references—I found only six.

Smear diagnosis is best learnt by doing it, with a sympathetic, experienced colleague at one’s elbow. Often this condition cannot be fulfilled and I would strongly recommend this book as a very valuable alternative or adjunct. One’s elbow. Often this condition cannot be fulfilled and I would strongly recommend this book as a very valuable alternative or adjunct. It is impossible to avoid the need for arrows that might otherwise have obscured the images. There is a useful index, but the book would have benefited from more references—I found only six.


Tethered spinal cord comprises a group of dysraphic conditions in which the conus medullaris is located in an abnormally low position. Tethered cord syndrome is a stretch induced symptoms manifested by motor and sensory deficits in the lower limbs and incontinence, and is often associated with musculoskeletal deformities. This book draws together all aspects of the embryology, pathophysiology, diagnosis, and treatment of this rare but important condition in a detailed and readable format. Interestingly there is repetition but this is unavoidable in a multiauthor text in which each chapter can be read as a whole. The text is well illustrated with diagrams, clear radiological images, and well-chosen clinical photographs.

The chapter on pathophysiology of the tethered spinal cord is a fascinating summary of the various experimental studies that have been undertaken in this condition. Although the relevance of some of the models to the condition may be questioned one cannot help but admire the ingenuity and inventiveness of the investigators. Acute traction on the spinal cord has been shown to be associated with impairment of evoked potentials, reduction in spinal cord blood flow, and changes in glucose metabolism. Chronic experiments have shown recovery in neurological deficits after 9 months.

There are well written chapters on diagnosis, investigation, and surgical treatment with plenty of intraoperative detail. The final chapter considers controversies associated with the treatment of the tethered cord syndrome. Most neurosurgeons would now agree that surgical treatment is definitely indicated in patients with progressive neurological deficits and most are increasingly prepared to consider prophylactic surgery in patients with tethering but in whom neurological deficits are absent or established. Although urodynamical testing helps to identify patients with neurogenic bladders, urinary dysfunction may be intermittent and symptomatic tethering may not always be disclosed. A tethered spinal cord is prone to produce problems during periods of rapid growth in childhood, but even when growth is complete patients with an undiagnosed tethered cord may undergo serious deterioration if subjected to sudden flexion movements associated with trauma. A selection process is outlined to help decide treatment in four main categories of presentation. Unfortunately the algorithm can be difficult to interpret—for example, “fluctuating signs and symptoms noted in a patient with stable neurological deficits—patient must be followed closely for detection of minor progression”.

A useful book which I would recommend to all doctors who treat patients with spinal dysraphism and I suspect that many will wish to have a copy in their personal collection.

R LAING


Do you find conference dinners a dull ordeal? Fear no longer! You can sparkle with amusing clinical anecdotes taken from this comprehensive collection. Amaze your colleagues with tales of seizure induced religious conversion or of the patient who ate lunch at his work canteen, had an amnestic seizure, and went to eat another one. He obviously didn’t work in the NHS. Tickle them with the patient for whom safety pins triggered a pleasurable aura that he favoured over sexual intercourse. At last I understand the punk movement.

But what if you want a detailed evaluation of the diagnosis and management of partial seizures and the psychiatric disorders associated with epilepsy? You can find brief chapters adequately covering these issues, but you can read about them in any one of several previous volumes on the subject or in the big, bulky text of epilepsy you use on hot summer days to hold the door open. In this book you can learn about the patient who sang rap music during her seizure—no doubt now residing on Sunshine Boulevard. Or the woman who experienced sexual arousal during her seizures and made excessive demands of her husband until a parietal tumour was removed (from her).

The cases in this book are collected from a wide search of the literature and start with the famous Dr Z. They include ictal and interictal manifestations and are totally delightful. They make this book a good read. These bizarre manifestations of epilepsy provide an opportunity to explore mind-brain relations that is sadly missed by the author. Some controversial issues are covered, such as ictal violence, but if you want a detailed clinical manual, look elsewhere. As a source of anecdotal cases for lectures, this book is ideal.

MARK MANFORD

SHORT NOTICES

Readers may also be interested in:


Creutzfeldt-Jakob disease presenting as complex partial status epilepticus: a report of two cases

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