The neuropathological abnormalities in the spinal cord in this case have been agreed by all observers, but Salazar et al. considered that non-specific, age-related, spongiform change can be confused with specific Creutzfeldt-Jakob changes and assigned this patient to such a category in their 1983 publication. In our opinion, the cortical spongiform change observed in this case is specific though of mild degree. The pathogenesis of the neuropathology in the amyotrophic form of Creutzfeldt-Jakob disease is probably similar to that observed in scrapie in which the infection can be specifically anatomically targeted, thus determining clinical and pathological abnormalities. This anatomical variation may be the result of differing routes of entry of the agent to the CNS.

Various infectious agents have been postulated in Creutzfeldt-Jakob disease. Intra-cytoplasmic spiroplasma-like bodies have been described in the axoplasm, primarily in presynaptic terminals in this disease. These loosely spiral structures measured up to 1000 nm in length and varied from 40–137 nm in width. Although we described a number of unidentifiable structures including one which was tightly coiled, intranuclear, and which measured 570 nm in length by 45 nm in width, no spiroplasmas-like shapes were seen. Also, Leach et al. were neither able to cultivate spiroplasmas from brains of 18 cases nor to detect antiboody to seven strains of spiroplasma in sera from 15 patients. From all the available evidence it is unlikely that spiroplasmas are involved.

Since this patient's brain contained a transmissible agent which produced Creutzfeldt-Jakob disease in an inoculated squirrel monkey the combination of dementia and motor neuron disease should now be included in the transmissible Creutzfeldt-Jakob group.

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Horner's syndrome due to superior-medial chiasmal schwannoma

Sir: A benign schwannoma of the superior-medial sympathetic chain was discovered in a 21 year old man seven years after the onset of Horner's syndrome and unilateral headaches. The patient was initially misdiagnosed as having Raeder's syndrome.

At the age of 14 he presented with a 2 year history of a left sided ptosis, anhidrosis, miosis (Horner's syndrome) and unilateral headaches. The headaches were localised over the left eye, occurred two to three times a week, were of variable severity, lasted several hours, were occasionally throbbing in nature and associated with blurring of vision. Detailed clinical examination, full blood count, WR and skull radiograph were all normal. In the absence of a demonstrable cause a diagnosis of idiopathic Raeder's syndrome was made (the association of miosis, ptosis and unilateral peri-orbital headache). The fact that anhidrosis is not part of Raeder's syndrome was overlooked at this stage. The patient's headaches and associated symptoms resolved over a one month period on prophylactic migraine therapy (sanomigran 0.5 mg tds) but the Horner's syndrome persisted. After 6 months the medication was stopped without recurrence of headache.

Seven years later he was referred with a 6 month history of dysphagia and a 3 kg loss of weight. On examination he had a mass in the left thoracic inlet with displacement of trachea. The Horner's syndrome was still present on the left although he no longer suffered from headaches. On examination the irises were noted to be markedly different in colour. The miotic pupil failed to dilate with cocaine instillation. Plain radiographs and CT of the thoracic inlet confirmed a mass in the left superior mediastinum displacing the oesophagus, trachea and great vessels to the right.

After an incision biopsy had confirmed the presence of a benign schwannoma, this was excised via a transverse cervical incision dividing the sternocleidomastoid. The tumour was well encapsulated, 5 × 5 × 6 cm in size approximately and was easily enucleated. It appeared to arise from the sympathetic chain at the level of the T2/3 nerve root. After operation the patient's dysphagia resolved over 3 months although the left Horner's syndrome has predictably persisted on 2 year follow-up.

The finding of a schwannoma of the sympathetic chain explained both the original presentation of Horner's syndrome and the subsequent dysphagia. Parapharyngeal and particularly superior mediastinal schwannomas are very unusual. When they do occur in this site they arise from either the vagus or cervical sympathetic nerves. This patient was originally incorrectly diagnosed as Raeder's syndrome. This syndrome differs from Horner's syndrome in that the miosis and ptosis are associated with retro-orbital headache; in addition, there is no anhydrosis. Facial sweating is retained in Raeder's syndrome because the hypothetical interruption to the sympathetic pathways is distal to the bifurcation of the common carotid artery so that the distribution of the external carotid retains its sympathetic innervation. The fact that this patient had anhydrosis and that the pupil failed to dilate with cocaine instillation indicates that the functional sympathetic lesion was between the superior cervical ganglion and the carotid bifurcation.

Raeder recognised the incomplete form of Horner's syndrome in 1924. Two types of Raeder's syndrome are recognised. Type I is associated with para-sellar nerve involvement (that is III, IV, and VI) and is usually due to a demonstrable mass. Type II does not involve cranial nerves, is commonly idiopathic but has once been shown to be caused by an aneurysm of the internal carotid artery.

Heterochromia iridis can be congenital
Thrombosis of cerebral veins following intravenous application of clomipramine

SIR: Thromboembolic complications, mostly within the veins of the legs, following treatment with antidepressants are well known. We report a patient, who developed a lethal thrombosis of the cerebral veins shortly after the beginning of a treatment with intravenous clomipramine.

A 61 year old woman suffering from an agitated depressed state did not respond to oral applications of different antidepressants and electroconvulsive therapy. She had a history of thrombosis of the veins of the right leg and of cardiac failure treated adequately with digitalis. Physical examination revealed a systolic apical murmur and a resonant chest percussion. A chest radiograph showed an enlargement of the left ventricle. Further investigations (ECG, EEG, laboratory reports) were unremarkable. On the 273rd day after admission to the hospital, treatment with clomipramine infusions (50 mg in 500 ml Ringer's solution) was started. The next day the patient vomited, then had a seizure, afterwards she became comatose and died at the 275th day of treatment.

CT on the day she died showed haemorrhage in the left hemisphere. Autopsy revealed complete occlusion of the superficial cerebral veins, the superior longitudinal sinus, the right lateral sinus, the right sigmoid sinus, partial occlusion of the left sigmoid sinus and the left lateral sinus, and intracerebral haemorrhage of the left hemisphere with oedema.

Thrombosis of cerebral veins are rare compared with other diseases of the cerebral vessels. We found only one other case report concerning thrombosis of the Galenic system veins after the start of intramuscular injections with amitriptyline-pamoate.1 Besides the slowdown of the blood flow by prolonged bed rest we assume that changes of the blood coagulation and of the vessel endothelium played a part in the pathogenesis of thrombosis in our patient.2 This case draws attention to the importance of antithrombotic prophylaxes during treatment with infusions of antidepressants, such as low-dose heparin (5,000 units three times a day), bandaging of the legs, and exercise.3

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Myotonic dystrophy with unilateral bulbar involvement

SIR: Myotonic dystrophy is a hereditary autosomal dominant multisystem disease characteristically affecting skeletal muscle, but also involving the heart, gastrointestinal smooth muscle, respiratory system, skull, brain, eyes, and skin.1

Among the bulbar signs, weakness of the pharyngeal muscles is quite common and not as a late manifestation of the disease.2 On the other hand, laryngeal abnormalities have been reported late in the course of the disease but are not common, and are mild and bilateral.3–5

We present here an unusual case of myotonic dystrophy in which the laryngeal symptoms were the presenting signs combined with unilateral atrophy of the neck and shoulder. A 29 year old unmarried man was admitted to the Ear-Nose-Throat department, because of progressive dysphonia in the past year and dysphagia for 4 months. On examination a unilateral left vocal cord paralysis together with severe weakness of the left pharyngeal muscles were observed. The rest of the examination was normal. Thyroid scan, plain radiographs of the foramen jugulare, and computed tomography of the brain stem and the base of the skull were negative.

He was transferred to the neurological department for further investigation. On examination myopathic facial appearance together with unilateral atrophy of the left sternocleidomastoid and trapezius muscles were observed. Myotonic reaction of both hands, decreased deep tendon reflexes in the arms without any decrement of strength or atrophy of the arms and hands were found. Bilateral posterior polar mild cataract, small testicles and first degree A-V block in an ECG were the additional findings. Chest radiographs were normal and lung function tests showed mild proximal airway obstruction. A barium swallow radiograph showed bilateral pooling of barium in the valleculae, more pronounced on the left side, with normal oesophageal peristalsis. Electro-myographic study showed the typical myotonic discharges sounding like a “dive bomber” with normal conduction velocities in peripheral nerves of the limbs.

The diagnosis of jugular foramen syndrome was eliminated by normal CT of the base of the skull, and local disease in the larynx was excluded by 2 years of surveillance without any new finding in an ENT examination.

We have checked nine more siblings and found another brother 40 years old with cardiac pacemaker for 7 years, with advanced myotonic dystrophy. Five more siblings had partial syndrome, with cataract and diabetes mellitus.

The laryngeal involvement in myotonic dystrophy is well accepted even though it is
Horner's syndrome due to superior-mediastinal schwannoma.
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