LETTERS TO THE EDITOR

Harlequin syndrome due to superior mediastinal neurinoma

The harlequin syndrome was first described by Lance et al. Five patients are presented with sudden onset of unilateral facial flushing and sweating in hot weather or when exercising vigorously. In one patient the site of the lesion was almost certainly brainstem but for the other four cases evidence supports a contralateral peripheral lesion, affecting the sympathetic outflow through the third thoracic root. CT and MRI of the thoracic area failed to disclose a structural lesion in the four patients. The report of this syndrome is still rare. We describe a patient with a left superior mediastinal neurinoma who developed facial flushing and sweating on the right side.

A girl aged 17 years came to our clinic in 1976, because of unilateral facial flushing after exertion. She first noticed asymmetrical flushing at the age of 15 when jogging or walking in hot weather. The right half of her face was flushed and sweat after 10 minutes of jogging while the left half remained pale and dry. Sweating over the trunk and limbs was symmetrical. The same phenomenon also developed on taking a hot bath. On examination, the left pupil was slightly smaller than the right but there was no ptosis and the pupils reacted normally. The nervous system and other systems were otherwise normal. Plain radiographs confirmed a mass in the left superior mediastinum at the level of the third thoracic vertebra.

The tumour was a neurinoma which was totally resected. It was approximately 4 x 4 x 3 cm. The tumour was located at the level of the third thoracic vertebra in the left superior mediastinum. After operation left ptosis developed. Her facial flushing and sweating when exercising have persisted on a 12 year follow up.

This patient showed harlequin syndrome associated with superior mediastinal neurinoma. Although her signs and symptoms did not improve after operation, we could not attribute the syndrome to any other cause except for the tumour. Our case supports the hypothesis by Lance et al. that this syndrome results from a deficit of the contralateral third thoracic nerve. No structural lesion was disclosed in their four patients but this report illustrates the need for thorough investigation of harlequin syndrome.

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Migraine comala

It is rare for attacks of migraine to be complicated by generalised, as opposed to focal, cerebral dysfunction, but when an acute self-limiting encephalopathy recurs, a diagnosis of migraine coma should be considered especially if there is a history of classic or familial hemiplegic migraine.

Our patient, a black Barbadian, was born in 1976. In 1980, he began to suffer from intermittent headaches with vomiting, sometimes associated with drowsiness. During the following two years, he was admitted to hospital on two occasions both following a prolonged episode of clouding of consciousness preceded by severe headaches and vomiting. Examination revealed extensor plantar responses and brisk reflexes, and on each occasion a diagnosis of encephalitis was considered. He was pyrexial, maximum 38°C, during the first 48 hours of the second admission and CSF examination was normal. He made a full recovery and was discharged home on the sixth hospital day on both occasions. Neurological examination was normal.

During the next three years, he continued to have intermittent headaches, mainly right-sided, associated with visual disturbance to the right and vomiting. A diagnosis of classic migraine was made.

In 1984, a generalised headache culminated in hospital admission with profound stupor, mild pyrexia and extensor plantar responses. Again, there was full recovery within one week.

In November 1988, he was involved in a brawl at school during which he sustained a blow to the head. Initially, he complained of a headache, but after about two hours his speech became slurred and he was rushed to hospital. He rapidly became confused, with several episodes of screaming and thrashing of all his limbs followed by longer periods of drowsiness. This sort of disturbed behaviour persisted for three days. His temperature rose to 38°C within 36 hours and fell to normal by 72 hours. At the peak of this illness, he was unresponsive to painful stimuli and showed generalised hyperreflexia with extensor plantar responses. CT brain scan was normal. EEG showed diffuse slowing (0.75-1.5 Hz) compatible with a severe encephalopathy but no epileptiform activity was seen. CSF analysis was normal. He improved gradually after 72 hours, initially complaining of a headache, but after seven days neurological examination had returned to normal and his confusion disappeared.

In summary, between the ages of four and 12 years, this patient with a clear history of classic migraine had four hospital admissions for a self-limiting encephalopathy with normal CSF findings and transient pyrexia. There was no evidence of decline in intellectual capacity as a result of the encephalopathic attacks.

A detailed family history could not be obtained. We believe these recurrent encephalopathic episodes were due to migraine as described by Fitzsimmons and Wolfenden.

Interestingly, our patient's most recent episode appeared to be precipitated by a mild head injury, a characteristic clinical feature in previous reports of coma episodes associated with migraine. CSF findings have been variable with many authors reporting pleocytosis; however, this may be a late feature occurring several days after acute admission and may explain the normal CSF findings in this patient.

Reports of migraine comas are rare but it is important to recognise the condition in the differential diagnosis of stupor with a normal CT scan, since the prognosis is excellent and full supportive measures, including mechanical ventilation may be required.

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Lamellated sensory corpuscles within the endoneurium

A wide diversity of morphological types of corpuscular nerve ending has been reported among different animal species. The unusual finding of an aggregate of lamellated sensory corpuscles within the endoneurium of a sural nerve taken for biopsy (at the level of the lateral malleolus) from a woman aged 77 years.

Transverse ultrathin sections revealed an area within one fascicle which contained a group of densely staining axons lying beneath the perineurium (fig 1). Each axon was surrounded by up to six flattened cytoplasmic lamellae, some of which could be traced into the perineural region of specialised Schwann cells (also called lamellar cells) from which they were derived. The cytoplasm of the lamellar cells was pale and contained few organelles other than intermediate filaments and occasional mitochondria; it was, however, rich in caveolae and pinocytic vesicles. Each layer was invested by basal lamina, and separated from its neighbour by bundles of collagen fibrils.

The axons were ellipsoidal in outline, and some exhibited stubby sidearms which extended between the lamellar cells (fig 2). There were differences in the arrangement of axo-

Figure 1 Transverse section through aggregate of corpuscles. p: perineurium; a: axon; l: lamellar cell. Scale bar 20 μm.

plasmic organelles between the various axons. In some axons the mitochondria were arranged peripherally around a core of filaments and microtubules, whereas in others the entire axonal profile was studded with mitochondria, dense bodies and filaments. These differences have been reported in lamellated sensory corpecules in the oral mucosa of the adult cat and miniature pig, and have been correlated with terminal and ultraterminal segments of the central axons respectively.

The aggregate was loosely encapsulated by fibroblast-like cells, similar to those that have been described forming a "pseudocapsule" around coiled simple corpecules in primate skin. This finding appeared to be incidental to the clinical presentation which was that of a multiple mononeuropathy associated with sarcoidosis. We have not encountered similar structures in any of over 100 other sural nerve biopsies. However, Pacinian corpecules have been described within the connective tissue associated with human peripheral nerve fibres (Hall, Hughes and Atkinson, unpublished observations). Timofeev's corpecules, encapsulated sensory receptors similar to, but smaller than, Pacinian corpecules, have been described as transient structures which occur in close relation to pelvic autonomic nerves and ganglia in late fetal and early post-natal life; their function is unknown. Presumably the corpecules that we have described are mechanosensitive: their situation within a relatively mobile section of peripheral nerve may therefore be significant.

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Permanent oculomotor palsy with occlusion of the internal carotid artery

Transient palsy of the third, fourth, and sixth cranial nerves, as well as retinal ischaemia, have recently been described ipsilateral to occlusion of the internal carotid artery. We report a case of permanent oculomotor palsy occurring in this situation. A 77 year old right handed white woman was admitted having developed sudden left sided weakness two weeks previously. Two days before admission she experienced supraorbital pain with visual loss in the right eye and abnormal eye movements.

Four months earlier she had developed left hemianaesthesia, weakness and then dysarthria in a stepwise manner over two weeks. There was occasional jerking of the left hand, she had a homonymous left hemianopia and left sided pyramidal signs. The fundi were normal. Carotid doppler ultrasound was normal. The CT brain scan showed patchy gyral enhancement in the right frontal and parietal lobes, and a diagnosis of ischaemia in the right internal carotid artery territory was made. Over the next three months her neurological disability improved.

There was also a past history of ischaemic heart disease (treated with verapamil, aspirin and dipryidamole) and a five year history of chronic myelomonocytic leukaemia. Occasional myelosuppression was treated with etoposide the last treatment being given two months previously. The antinuclear factor was known to be weakly positive (speckled) with negative DNA and ENA antibodies, rheumatoid factor, cryoglobulins and circulating immune complexes. Serum complement levels were normal.

On examination the blood pressure was 150/90, and both carotid arteries were palpable with no bruits. The mental state was normal. Vision was NPL on the right and 6/6 on the left with a left hemianopia. The right globe was mildly injected, there was a complete ptosis on the right, and adduction, up and downgaze were absent, with intortion on attempted downgaze. Abduction, and movements of the left eye, were normal. The right pupil was dilated and fixed, the left pupil reacting briskly to direct light and to accommodation. The retina was normal on the left but on the right it had the appearance of a central retinal artery occlusion with diffuse retinal oedema sparing the macula, and attenuated vessels. No haemorrhages or emboli were visible and the optic disc was normal. There was also weakness of the lower face on the left, and in the limbs tone was normal but there was no movement of the left arm or leg, together with left hyperreflexia and an exterior plantar response. Finally, there was sensory inattention on the left.

As before, the blood count and film, erythrocyte sedimentation rate and routine biochemistry were normal. The CT scan (fig A) now showed a large infarct involving the whole of the right middle cerebral and part of the anterior cerebral territory, with swelling of the hemisphere but no abnormal enhancement. In a selective right carotid angiogram (fig B), the right internal carotid artery was occluded 2 cm from its origin. There was no filling of the ophthalmic artery, nor any intracranial filling from the external carotid circulation. Flow through the ophthalmic veins, and the cavernous sinus itself, were normal.

There was no recovery in the ocular findings during a further month in hospital, although motor function improved slowly.

Thus the patient presented with occlusion of the right internal carotid artery, ischaemic visual loss in the right eye, and contralateral hemiparesis. The close temporal association of a complete right oculomotor palsy with these latter deficits suggests that it too was caused by ischaemia following the carotid occlusion. Indeed, there were no signs of brainstem ischaemia, nor indications of active vasculitis clinically or in laboratory tests. Her chronic leukaemia remained in remission, and the CT and angiographic studies showed no cavernous sinus lesion. Finally, the conscious level was not depressed at any stage, there were no ipsilateral motor deficits, and the vital signs remained stable, arguing
Lamellated sensory corpuscles within the endoneurium.

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