otherwise normal. Motor nerve conduction velocity (MNCV) in the right ulnar nerve was slowed at 47 m/s and 48 m/s in the forearm and around the elbow respectively, but severely reduced to 10 m/s in the upper arm where there was conduction block. The evoked compound muscle action potential (CMAP) at the wrist, below elbow, above elbow, and in the upper arm was 4.3, 3.3, 2.4, and 1.4 mV respectively. MNCV was moderately reduced, to 37 m/s in the left ulnar nerve in the forearm, but to 16 m/s around the elbow where there was conduction block. The amplitude of the CMAP fell from 3.6 mV at the wrist to 2.5 mV below and 1.2 mV above the elbow. The distal motor latency (DML) was normal in both nerves; F waves were absent. MNCV in the right peroneal and tibial nerves was borderline at 40 and 42 m/s without block and with normal DML values. F wave latency in the tibial nerve was increased at 60–64 ms; no peroneal nerve F responses were obtained. The right median sensory nerve action potential (SAP) was of normal amplitude (22 μ V) and velocity (63 m/s). The left sural SAP was 5.5 μ V with a velocity of 46 m/s; it was absent on the right.

The P100 visual evoked potential latencies were increased bilaterally (117 ms on right, 112 ms on left). Cranial MRI showed lesions of high T2 signal in cerebral white matter (figure), many abutting the walls of

the lateral ventricles, especially their posterior portions. Lesions were also present in the corpus striatum, pons, and middle cerebellar peduncles.

The electrophysiological results indicated a patchy demyelinating disorder with conduction block affecting the peripheral nerves consistent with CIDP with concomitant CNS involvement. Clinical examination also showed CNS abnormalities. The cranial MRI showed clear evidence of multifocal demyelinating pathology. The patient was treated with prednisolone with resolution of her symptoms apart from persistent paraesthesiae distally in the limbs and objective evidence of improvement on examination.

This patient is of interest in relation to the onset of relapsing symptoms in early childhood. It is uncertain whether the two previous episodes represented peripheral or central nerve disease. Cranial nerve lesions are known to occur with either in CIDP with central involvement.⁴ The rarity of multiple sclerosis in Afriid populations suggests that the central demyelinating lesions in this type of case may reflect a different form of demyelination than occurs in multiple sclerosis. Experimental allergic neuritis has been considered to provide a model for the Guillain-Barré syndrome and its chronic variant for CIDP.⁵ Possibly the chronic variant of experimental allergic encephalomyelitis⁶ provides an equivalent for the

central demyelination seen in the present case and other instances of CIDP with central involvement. The question as to whether chronic relapsing experimental allergic encephalomyelitis is a valid model for multiple sclerosis continues to be controversial.

P K THOMAS
A VALENTINE
B D YOUL

Royal Free Hospital and School of Medicine,
London, UK

Correspondence to: Professor P K Thomas, Department of Clinical Neurosciences, Royal Free Hospital School of Medicine, Rowland Hill Street, London NW3 2PF, UK.

- 1 Thomas PK, Walker RWH, Rudge P, *et al.* Chronic demyelinating peripheral neuropathy associated with multifocal central nervous system demyelination. *Brain* 1987;110:53–76.
- 2 Mendell JR, Kolkin S, Kissel JT, *et al.* Evidence for central nervous system demyelination in chronic inflammatory demyelinating polyradiculoneuropathy. *Neurology* 1987;37:1291–9.
- 3 Rubin M, Karpatis G, Carpenter S. Combined central and peripheral myelinopathy. *Neurology* 1987;37:1287–90.
- 4 Waddy HM, Misra VP, King RHM, *et al.* Focal cranial nerve involvement in chronic inflammatory demyelinating polyneuropathy: clinical and MRI evidence of peripheral and central lesions. *J Neurol* 1989;236:400–10.
- 5 Pollard JD, King RHM, Thomas PK. Recurrent experimental allergic neuritis. An electron microscope study. *J Neurol Sci* 1975;24:365–83.
- 6 Raine CS, Snyder DH, Valsamis MP, Stone SH. Chronic experimental allergic encephalomyelitis in inbred guinea-pigs—an ultrastructural study. *Lab Invest* 1974;31:369–76.

Neurothekeoma of the cauda equina

Neurothekeoma or nerve sheath myxoma is a rare benign tumour of peripheral nervous tissue. Histogenesis from Schwann cells¹ or undifferentiated nerve sheath precursor cells² has been suggested. Intrathecal location is rare.

An 84 year old woman presented in May 1994 with severe low back pain and pain radiating into both legs. She had fallen on to the floor at home three weeks previously. The pain was exacerbated by coughing, sneezing, and straining. She had increasing difficulty walking, requiring the aid of one person. She had difficulty initiating micturition and there was occasional faecal soiling. Clinical examination disclosed a flexed posture. She was unable to stand erect due to pain. There was pain on femoral stretching and on straight leg raising bilaterally. Tendon reflexes were absent in the lower limbs. There was sensory loss below the right knee and in the left foot. Saddle sensation and anal tone were normal. Power testing was not possible due to pain.

Blood biochemistry and haematology were normal. Magnetic resonance imaging disclosed a 3 × 1.7 × 1.2 cm intrathecal mass displacing the lower conus anteriorly and to the left. The tumour had high signal intensity on T2 weighted images (figure, a). After gadodiamide there was pronounced enhancement of the tumour mass with a small central non-enhancing component (figure, b). No adjacent bone or extradural abnormality was identified.

At operation a thoracolumbar laminectomy was made and the thecal sac was opened longitudinally. The tumour seemed exophytic from the right side of the caudal conus and displaced the roots of the cauda equina. The major exophytic part of the

Cranial MRI showing lesions of high T2 signal in cerebral white matter as detailed in the text.

tumour was removed with an ultrasonic aspirator. A thin layer of tumour remained attached to the conus. A good plane of dissection between the tumour and the surface of the conus could not be safely achieved.

Postoperatively, she made a slow but steady recovery. She was relieved of low back and radicular pain. At follow up four months after surgery, there was an area of numbness in the L4 dermatome on the right. She had returned home, where she looked after herself.

The tumour specimen was an irregular, soft, greyish portion of tissue $2.2 \times 1.5 \times 0.7$ cm in size. Microscopically it displayed a multinodular structure, in which pale, sparsely cellular islands were separated by connective tissue septa or bands of more compactly cellular neoplastic tissue (figure, c). The pale nodules had a myxomatous appearance composed of stellate or elongated cells in a strongly alcianophilic, mucoid matrix. The tumour cells in the

intervening areas were mostly spindle shaped and had a more bulky eosinophilic cytoplasm. Both components stained positively for S-100 protein (figure, d).

Electron microscopy showed that the tumour cells in the myxomatous nodules and those in the internodular areas had elongated cytoplasmic processes, mostly covered by a continuous external lamina. Wide spaced collagen fibres (Luse bodies) were also identified.

Neurothekeomas most often occur in the skin of the face and arms of young adults. They arise from small cutaneous nerve branches and not from the major peripheral nerves. They are benign lesions which are cured by excision. Rare recurrences have been reported after incomplete resection. Cutaneous neurothekeomas have been classified into cellular and myxomatous subtypes, the second arising at an older age.³

Neurothekeomas have rarely been found intrathecally. There is one report of the

tumour arising from a single nerve root in the lower cauda equina.⁴ In that case the tumour was completely excised. In the present case complete excision was not possible due to adherence to the conus and multiple nerve roots. The early postoperative course has, however, been satisfactory with relief of pain and restoration of ambulation.

The MRI appearance is similar to that of other intrathecal extramedullary tumours. Ependymoma, neurofibroma, and meningioma were the major differential diagnoses.

Connective tissue myxomas have occasionally been described as arising from paraspinal muscle⁵ and from the vicinity of a facet joint.⁶ An intramedullary location has also been described.⁷ The compact cellular schwannomatous component, the positive staining for S-100 protein, and the ultrastructural appearances clearly distinguish the present tumour from connective tissue myxoma.

GEORGE F KAAR
SAAD H BASHIR
JAMES M N'DOW
Department of Neurosurgery
PHILIP V BEST
Department of Pathology
LESLIE N GOMERSALL
Department of Radiology,
Aberdeen Royal Hospitals NHS Trust,
Aberdeen AB9 2ZB, UK

Correspondence to: Mr G F Kaar, Ward 40, Aberdeen Royal Infirmary, Foresterhill, Aberdeen AB9 2ZB, UK.

- 1 Webb JN. The histogenesis of nerve sheath myxoma: report of a case with electron microscopy. *J Pathol* 1979;127:35-7.
- 2 Angervall L, Kindblom LG, Haglid K. Dermal nerve sheath myxoma. A light and electron microscopic, histochemical, and immunohistochemical study. *Cancer* 1984;53:1752-9.
- 3 Barnhill RL, Mihm MC Jr. Cellular neurothekeoma. A distinctive variant of neurothekeoma mimicking melanocytic tumours. *Am J Surg Pathol* 1990;14:113-5.
- 4 Paulus W, Jellinger K, Perneczky G. Intraspinal neurothekeoma (nerve sheath myxoma). A report of two cases. *Am J Clin Pathol* 1991;95:511-6.
- 5 Tahmouresie A, Farmer PM, Stokes N. Paraspinal myxoma with spinal cord compression. Case report. *J Neurosurg* 1981;54:542-4.
- 6 Bell WO, Gill A, Babiak T, et al. Epidural myxoma causing compression of the cauda equina. Case report. *Neurosurgery* 1983;12:325-6.
- 7 Pasaoglu A, Patisroglu TE, Orhon C, Yildizhan A. Cervical spinal intramedullary myxoma in childhood. Case report. *J Neurosurg* 1988;69:772-4.

Voluntary facial palsy with a pontine lesion

Emotional voluntary dissociation is not uncommon in patients with central facial palsies.¹⁻³ The neuroanatomical basis of this dissociation is not well understood. A recent case report in this *Journal* suggested that a pontine stroke could result in unilateral voluntary facial palsy.⁴ We have recently had the opportunity to study a very similar syndrome.

A teacher aged 57 was referred three days after the onset of progressive neurological symptoms and signs that had included, in order, unsteadiness, dysarthria, dysphagia, and weakness of the right arm, leg, and face. On admission, the patient was awake, in order, unsteadiness, dysarthria, dysphagia, and weakness of the right arm, leg, and face. On admission, the patient was awake, dysarthric but not aphasic, and had normal vision and oculomotor function. He complained of impaired swallowing and right facial weakness. Neurological examination disclosed a right central facial palsy for vol-

(a) Sagittal T2 weighted MR image (TR = 4500 ms, TE = 130 ms) showing tumour distorting the region of the conus (arrow). (b) Postgadodiamide injection T1 weighted MR image (TR = 450 ms, TE = 15 ms), showing tumour (black arrow) displacing conus (white arrow) anteriorly and laterally. (c) Photomicrograph (haematoxylin and eosin × 10) showing pale myxomatous nodules separated by narrow septa. (d) Photomicrograph (× 40) of the tumour stained with the immunoperoxidase method to demonstrate S-100 protein. Note the dark (positive) staining in nuclei and cytoplasmic strands.