cerebellar connections with the brainstem, especially the superior cerebellar peduncle, should be spared for a postural type of tremor to occur. An intention tremor results if these connections are involved.\textsuperscript{4} The postural tremor caused by cerebellar lesions is a slow tremor of 4 to 5 Hz and is thought to result from hypotonia of the affected limbs.\textsuperscript{4}

The postural tremor in our case was unexpected considering the location of the mass in the thalamus with compression of the adjacent basal ganglia. The tremor is likely to be affected by the cyst include the basal ganglia and their connections, dentatorubrothalamic fibres, and the thalamic nuclei. Lesions of these structures are not known to produce isolated postural tremors. The mass in our patient could be expected to produce a rest tremor. Even an intention tremor is conceivable considering the possible compression of the superior cerebellar peduncle by the cyst. As mentioned, only discrete lesions of the cerebellar hemisphere have been considered to cause postural tremors and our patient had neither hypotonia nor involvement of the cerebellar hemispheres.

The tremor in our case was contralateral to the side of the tumour and responded to aspiration of the cyst, suggesting that the pressure exerted by the cyst on adjacent neural structures was in some way responsible for the production of the tremor. To the best of our knowledge there has been no previous report of unilateral postural tremor caused by athalamic or basal ganglia lesion.

At follow up three months after surgery, the patient was asymptomatic and had no tremors of the left upper or lower limbs. A small residual cyst was seen on CT but it was not producing any mass effect or ventricular dilatation. No further treatment was offered and she was advised clinical and CT monitoring.

Intracranial tumours are on occasions known to produce a parkinsonian syndrome with rest tremors, bradykinesia, and rigidity.\textsuperscript{2} Another type of tremor that has been described in association with intracranial masses is “rubral tremor”. Involvement of the red nucleus and the decussating fibres of the superior cerebellar peduncle by lesions in the midbrain region has been implicated in the production of “rubral tremor”.\textsuperscript{2} In both these instances, the tremor is present at rest, although rubral tremors may be aggravated by maintenance of a posture or goal directed movement.

Postural tremors can theoretically occur in isolated cerebellar hemispheric lesions including mass lesions, but this manifestation is rarely seen in clinical practice. The

tude, as we show by the presentation of two cases.

Starting in 1981, case 1, a 28 year old woman, had repeatedly experienced pares-theses of the extremities and episodes of blurred vision. In 1988, during a further bout of visual deterioration MRI of the brain showed swelling and contrast enhancement of the optic chiasm and of the prechiasmatic portion of the optic nerves. An exploratory frontal craniotomy six months later showed granulomatous optic nerves that were encaused by abnormally thick arachnoidica but no tumour.

One year later the patient was first seen at our department for rapid onset of left leg weakness and bilaterally ascending pares-
theses. At that time visual acuity was reduced to finger counting. MRI showed swelling of the upper thoracic cord with irregular intramedullary enhancement after giving gadolinium-DTPA. The brain seemed normal, including the chiasm. Lumbar puncture and a spinal angiogram were uninformative. Laboratory tests were negative for involvement of any other organ, systemic infection, or collagen vascular disease. No specific antibodies except for an intermittently raised titre against Toxo-
plasma gondii was found.

Over the next four years the patient con-
tinued to have bouts of myelitis at times involving almost the entire spinal cord, as shown by MRI. Cell counts in the CSF ranged from 0 to 10/mm\(^3\) and consisted mainly of lymphocytes and activated monocytes. Sometimes fat containing macrophages indicating tissue necrosis were also seen. The protein content of CSF was mostly increased and ranged from 71 to 153 mg/100 ml but there was no evidence of intrathecal immunoglobulin production. Repeated search for oligoclonal bands was negative.

Various therapeutic regimes including high dose steroids, immunoglobulins, immune adsorption, and antibiotic treat-
ments failed to stabilise the patient’s clinical condition. When she was last seen in mid-
1992 she was bedridden because of a spastic tetraparesis with a bilateral sensory level at C2 and had unchanged severe visual impairment. MRI of the brain continued to be normal whereas the spinal cord at the cervical and upper thoracic region was very atrophic. The patient refused the administration of contrast material at that time.

For five months case 2, a 22 year old woman, had experienced continuous deteri-
oration of visual acuity associated with increasing bilateral leg weakness and uri-
inary incontinence before she was seen at our department. Visual field testing showed concentric narrowing on the right and a temporal hemianopia on the left with spar-
ing of the fovea. Her right arm distal to the elbow was mildly hypaesesthesia and sensa-
tion to pinprick and pain was reduced dis-
tally from T7 bilaterally. The plantar response was upgoing on both sides. A spinal tap gave a colourless CSF with 4 lymphocytes/mm\(^3\) and a protein concentra-
tion of 50 mg/100 ml. There was no evi-
dence of oligoclonal bands. MRI showed right sided enlargement of the chiasm and of the right optic nerve with uptake of con-
trast material. The remainder of the brain was normal. There was also minimal swelling of the cervical medullos and patchy, intramedullary enhancement from C1 to


MRI of nevromyelitis optica: evidence for a distinct entity

Neuromyelitis optica was originally described as a distinct demyleinating disorder characterised by visual disturbances and spinal cord signs occurring closely in time.\textsuperscript{1} Later studies on patients presenting with this syndrome often reported the subsequent evolution of additional neurological deficits,\textsuperscript{2} found pathological evidence for a more widespread demyelination, or disclosed other specific aetiologies such as acute disseminated encephalomyelitis, Behét’s disease, or systemic lupus erythe-
matosus.\textsuperscript{3} From these findings it was con-
cluded by many that neuromyelitis optica was inseparable from multiple sclerosis and its existence as a specific entity was ques-
tioned.\textsuperscript{4} MRI promises to alter this atti-
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A repeat MRI two months later showed a further increase in chiasmal pathology with extension into both optic nerves (fig 1). Spinal cord involvement seemed almost unchanged (fig 2). After a waxing and waning of symptoms for the next 18 months her last MRI confirmed still active optochiasmatic and spinal cord disease which spared the remainder of the CNS.

The MRI findings in both our patients exactly paralleled the pattern of damage described as characteristic for neuromyelitis optica in pathological studies. They consisted of swelling and congestion of the optochiasmatic region and the spinal cord due to severe demyelination which often progresses to whole tissue necrosis. Medullary inflammation extended over many segments, tended to involve both grey and white matter, and had a predilection for the cerebral and upper thoracic cord. The brain itself remained free from demyelination. This pattern was very different from the distribution of lesions found in multiple sclerosis. There MRI typically shows numerous round or oval signal hyper-intensities, which are scattered throughout the brain with a predilection for the periventricular region.

Extensive spinal cord damage seen on MRI as in our patients has already been described in four other patients with suspected neuromyelitis optica. The MRI follow up of optic neuritis attributed to neuromyelitis optica has also been reported. Yet our study for the first time presents the full diagnostic potential of MRI for this disorder—that is, to show disease activity in both optic chiasm and spinal cord of the same patient and to rule out other abnormalities in the brain. It is also important to stress that the disease of our patients remained limited strictly to the optic nerves, chiasm, and spinal cord during a relapsing-remitting course of at least seven and two years, respectively. We also found no evidence for any other disorder previously associated with neuromyelitic optica such as acute disseminated encephalomyelitis and vasculitic or granulomatous diseases. In accord with more recent findings, both our patients exhibited a clinical course of the disease rather different from earlier descriptions, and oligodendral bands were not found in their CSF.

Our findings provide further evidence for the existence of Devic’s Disease as a nosological entity and document the important contribution of MRI in separating it from a “syndrome” of associated symptoms due to other pathology. This notion will enable new research into the aetiology and treatment of this often devastating demyelinating disorder.

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**Figure 1** Case 2: Magnification of T1 weighted coronal cut through the chiasm (left) and optic nerves (right) before (upper panel) and after (lower panel) application of gadolinium-DTPA. There is pronounced asymmetric swelling of the chiasm with contrast enhancement extending into both optic nerves (arrows).

**Figure 2** Case 2: Sagittal (upper panel) and axial (lower panel) contrast enhanced T1 weighted MRI of the spine shows multiple irregular bright areas within the entire cervical cord.
Ischaemic optic neuropathy, transverse myelitis, and epilepsy in an anti-phospholipid positive patient with systemic lupus erythematosus

We report a 37 year old woman who had severe, bilateral visual loss in association with transverse myelitis, epilepsy, serological evidence of systemic lupus erythematosus, and the presence of anti-phospholipid antibodies. Recognition of a possible underlying thrombotic aetiology suggests that long term anticoagulation and immunosuppressive treatment could prevent progression and relapses of neurological and ophthalmic complications in patients with antiphospholipid related disease.

The 37 year old housewife was admitted in May 1992 with a three day history of visual loss in her right eye, preceded by a severe frontal headache lasting 24 hours. There were no associated symptoms of retro-orbital tenderness or pain on eye movement. She had a complex history. Aged 13 months she had had febrile convulsions and at 11 years of age developed epilepsy. At 16 years of age she was mildly hypertensive and developed recurrent attacks of angioneurotic oedema, Raynaud’s phenomenon, and photosensitive skin rashes. At the age of 22 she was admitted with diminished power in her right leg, bilateral extensor plantar responses, and a sensory loss at T2. She improved spontaneously but relapsed a month later with a paresis of the left arm and leg, bilateral extensor plantar responses, and a sensory level below T9 on the right and between C4 and T2 on the left. All investigations including a myelogram were normal apart from an erythrocyte sedimentation rate (55 mm/hour), antinuclear antibody titre borderline at 1:40, DNA binding significantly raised at 61%, and a positive lupus anticoagulant on a venereal disease research laboratory test (VDRL), and normal CT brain and orbit scans. MRI was contraindicated due to the presence of the thoracic epidual implant. Treatment with three daily intravenous pulses of methylprednisolone (500 mg) with oral aspirin (75 mg daily), was started on admission with no initial visual improvement. On day five she was given an intravenous pulse of cyclophosphamide (250 mg) but by day 11 the visual acuity was reduced to hand movements. Further results showed a strongly positive anti-cardiolipin IgG level of 33 6Gpi units, negative IgM antibodies, and a positive lupus anticoagulant. A diagnosis of an ischaemic optic neuropathy related to systemic lupus erythematosus was made. She was given a further pulse of cyclophosphamide (500 mg) and anticoagulated with heparin and warfarin. On day 18 her vision was reduced bilaterally to no perception of light and she was registered blind. She was given two daily pulse doses of methylprednisolone (2 g) with no immediate effect. On day 25 she showed an improved visual acuity on the right of hand movements and was discharged fully anticoagulated and on oral prednisolone (10 mg once daily) and aspirin (75 mg once daily). Three weeks later the right visual acuity had improved to 6/9, N5 and 16/17 Ishihara colour plates unaided. Goldman perimetry showed complete recovery in the right visual field. The left eye remained unchanged. There was widespread retinal haemorrhage and a possible underlying aetiology. On May 11, 1992 the prednisolone was stopped once daily and with an INR reduced to 1-9, she had a recurrence of vision loss in her right eye, which on admission was visual field loss and increased methylprednisolone and increased anticoagulation (INR 3>0). On recovery, her visual acuity was 6/9 but there was a persistent right altitudinal field defect. She has subsequently found her vision to be subject to occasional anticoagulation and steroid treatment, stabilising at 10 mg prednisolone once daily and an INR of 3-0, with preservation of visual acuity and field.

The differential diagnosis of this patient’s illness includes multiple sclerosis and the syndrome of lupus anticoagulant. Lupus anticoagulant can give rise to neurological disease and in such cases may be associated with systemic lupus erythematosus, as in our patient.

Systemic lupus erythematosus only very rarely produces identical symptoms to typical demyelinating optic neuritis.* The patient’s illness revealed a right optic disc which was initially pale and then yellow on the first day of her eye’s atrophy for primary demyelinating optic neuritis; in her left eye, she had the uncharacteristic features of severe visual loss with failure to improve and later pronounced retinal vascular attenuation, and in the right eye no associated pain on ocular movement, no CSF lymphocytes, and progressive visual loss to no perception of light over 18 days despite substantial treatment with immunosuppressants, more suggestive of an ischaemic optic neuropathy. Her subsequent response and sensitivity to anticoagulation and steroids, we believe favours a thrombotic aetiology. Pathologically, most likely ischaemia of the microvasculature of the optic nerve in relation to antiphospholipid antibodies, and not primary demyelination.

Transverse myelitis and epileptiform seizures are known to occur in the presence of antiphospholipid antibodies.8 Ophthalmic vaso-occlusive phenomena associated with high levels of phospholipid antibodies include anterior ischaemic optic neuropathy, branch and central retinal artery occlusions, amaurosis fugax, and venous occlusions.9 Splinter haemorrhages have recently been reported to occur with antiphospholipid vasoocclusive disease in the antiphospholipid syndrome.4 Oppenheimer and Hoffbrand in 1986, were the first to describe the association of spinal myelopathy and optic neuritis in a case similar to ours.4 In the same paper, they reviewed 13 previously reported cases of patients with known systemic lupus erythematosus and optic neuritis: six had at some point had a visual ischaemia and they suggested that disease in the optic nerve and spinal cord might have a common underlying aetiology. Describe the association of spinal myelopathy in patients with stroke and antiphospholipid antibodies was reported with affected patients having human brain microvascular endothelial reactive antibodies present.18

The association of antiphospholipid antibodies with transverse myelopathy, epilepsy, and optic neuritis may be part of the spectrum of antiphospholipid syndrome.18 This has important therapeutic implications in that long term anticoagulation, reducing the incidence of vascular thromboses, possibly with immunosuppression, may prevent neurological and ophthalmological relapses in


