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. 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.

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 structure was 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 contents, 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 a thalamic or basal ganglia lesion.

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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. 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”. 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- thesias of the extremities and episodes of blurred vision. In 1988, during a further bout of visual disturbances and bilateral headache, MRI of the brain showed swelling and contrast enhancement of the optic chiasm and of the prechiasmatian portion of the optic nerves. An exploratory frontal craniotomy six months later showed small necrotic optic nerves that were encased by abnormally thick arachnoid but no tumour.

One year later the patient was first seen at our department for rapid onset of left leg weakness and bilateral sensory disturbance. 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 continued 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/mm³ 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 regimens including high dose steroids, immunoglobulins, immune adsorption, and antibody treatments 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 deterioration of visual acuity associated with increasing bilateral leg weakness and urinary 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 sparing of the fovea. Her right arm distal to the elbow was mildly hypaesthetic and sensation to pinprick and pain was reduced distally from T7 bilaterally. The plantar response was upgoing on both sides. A spinal tap gave a colourless CSF with 4 lymphocytes/mm³ and a protein concentration of 50 mg/100 ml. There was no evidence of oligoclonal bands. MRI showed right-sided enlargement of the chiasm and of the right optic nerve with uptake of contrast material. The remainder of the brain was normal. There was also minimal swelling of the cervical medullas and patchy, intramedullary enhancement from C1 to
C4. Laboratory tests for specific causes of myelitis were negative. The patient was given steroids but improvement was minimal.

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.4 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 cervical 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.4

Extensive spinal cord damage seen on MRI as in our patients has already been described in four other patients with suspected neuromyelitis optica.5 7 The MRI follow up of optic neuritis attributed to neuromyelitis optica has also been reported.1 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,5 both our patients exhibited a clinical course of the disease rather different from earlier descriptions,4 and oligodendroglial 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.1 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 nerve (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.
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 aneurysmocoele, Raynaud's phenomenon, and photosensitive skin rashes. At the age of 22 she was admitted with diminished power in her right leg, bilateral extensor plants, and a sensory loss at T2. She improved spontaneously but relapsed a month later, with a positive papilledema, 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 raised erythrocyte sedimentation rate (55 mm/hour), antinuclear antibody titre borderline at 1/40, DNA binding significantly raised at 61%, and a positive lupus erythematosus (LE) cell test. A possible diagnosis of probable systemic lupus erythematosus with transverse myelitis was made. Her neurological deficits improved spontaneously with residual weakness on the left. She had recurrent episodes of myelopathy, similar to those described, in the subsequent years, which resolved either spontaneously or with short courses of methylprednisolone. Repeat DNA binding assays confirmed high titres. Examination of the CSF during acute episodes of her neurological illness revealed no lymphocytes or red cells (normal and Aplysia for multiple sclerosis). In 1987 she was found to have moderately raised anti-cardiolipin antibodies and low levels of C3 and C4. In 1991 another positive antikeratin (IgG) band was detected (normal < 5). Urinary incontinence developed, necessitating insertion of a thoracic epidural stimulator in 1989. At 27 years she had her first ophthalmological illness. Over a period of one year she developed painless loss of vision in her left eye, which she described as a gradual “keyhole” constriction of her visual field. There was no associated headache or diplopia but slight pain on left ocular movement. She was diagnosed as having an acute optic neuritis, but despite treatment with oral and pulsed dexamethasone her visual acuity decreased to no perception of light over a four day period and failed to improve.

Examination in May 1992 revealed visual acuities of 6/24 corrected and N14 in the right eye and N24/200 with 8 diopters of hypermetropia in the left. Her colour vision was reduced to 10/17 correct Ishihara plates. She had a dense left afferent pupillary defect with a pale atrophic left optic disc and attenuated retinal vessels. The right optic disc was normal. Goldman perimetry showed a right inferior altitudinal field defect. Neurological examination showed bilateral extensor plants, residual weakness, and astasia of both lower limbs and reduced vibration sense below the level of the anterior iliac spine. She had two perisplenic splinter haemorrhages and was normotensive. Investigations included anti-nuclear antibody (ANA) 1/10, negative DNA antibody tests, normal immune complex levels, reduced C4 concentrations to 0-07/g/l (normal 0-2-0-5), absence of CSF lymphocytes, a platelet count of 167, a negative venereal disease research laboratory test (VDRL), and normal CT brain and orbit scans. MRI was contraindicated due to the presence of the thoracic epidural implant. Treatment with three daily intravenous pulses of methylprednisolone (500 mg) serially reduced to 10 mg 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 antikeratin (IgG) level of 33-66p 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 aspirin 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 improved on 0-5 mg prednisolone once daily and with an INR reduced to 1-9, she had a recurrence of vision loss in her right eye, which once again responded to a pulse of 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 associated with 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 clinical 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 possibility of our patient's syndrome being related to her period of her patient's eyes was atypical for primary demyelinating optic neuritis; in her left eye, she had the uncharacteristic features of severe vision 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 the aetiology of a vascular pathalogy, 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.14 Ophthalmological oculo-vascular phenomena associated with high levels of phospholipid antibodies include anterior ischaemic optic neuropathy, branch and central retinal artery occlusions, amaurosis fugax, and venous occlusions.15 Splinter haemorrhages have previously been reported to occur in posterior ocular vasculocclusive disease in the antiphospholipid syndrome.16 Oppenheimer and Hoffbrand in 1986, were the first to describe the association of spinal myelopathy and optic neuritis in a case similar to ours.17 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 spinal myelopathy and they suggested that disease in the optic nerve and spinal cord might have a common underlying aetiology. Recruitment of the possible specific association of antiphospholipid antibodies in vasculocerebrovascular diseases 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. This has important therapeutic implications in that long term anticoagulation, reducing the incidence of vascular thromboses, possibly with immunosuppression, may prevent neurological and ophthalmic relapses in...