Neurogenic tumour of the orbit in a patient with idiopathic chronic inflammatory demyelinating polyneuropathy

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DESCRIPTION

A 49-year-old man had progressive blurring of vision in the left eye for 1 year. He was seen by ophthalmologist and MRI orbit showed left intra-conal orbital lesion extending around the opening of superior orbital fissure and optic canal which is suspicious of neurogenic tumour (figure 1A–C). He was scheduled for a follow-up MRI scan before operation. He was then admitted for numbness and weakness of limbs for a few months. Physical examination showed visual acuity was finger counting at 1 feet on his left eye and left relative afferent pupil defect. There was wasting of muscles of his hands and legs, the upper limb power was Medical Research Council (MRC) grading 5/5 and the lower limb power was MRC grading 5/5 proximally and 3/5 distally. There was reduced temperature and pain sensation in his feet up to the knee level and the jerks were hyporeflexic. Cerebrospinal fluid examinations showed white cell count $<1/\text{mm}^3$ (<5), protein 1.01 g/L (0.15–0.4), glucose 4 mmol/L (2.2–3.9). A nerve conduction study showed prolonged distal latencies and reduction in conduction velocities of the median, ulnar, tibial and peroneal nerves. The amplitude of the compound motor action potential (CMAP) was reduced in the right tibial, left median and both peroneal nerves. There were possible conduction blocks, defined by significant reduction in the CMAP area according to the American Association of Neuromuscular & Electrodiagnostic Medicine criteria,¹ in the right median nerve below the elbow, both ulnar nerves below the elbow and left tibial nerve below the popliteal fossa. The sensory nerve action potentials were not recordable

(table 1). MRI of the cervical spine and plexus showed a diffuse thickening of the nerve roots (figure 2A) and bilateral brachial plexuses (figure 2B). Sural nerve biopsy revealed myelin sheath damage affecting both the large and small fibres. There was inflammatory infiltration of the perivascular connective tissue of the perineurium. Extensive workup for secondary causes of chronic inflammatory demyelinating polyneuropathy (CIDP) including blood glucose, autoimmune markers, immunoglobulin pattern, serum protein electrophoresis and paraneoplastic panels was unremarkable. He was diagnosed with idiopathic CIDP and was given high-dose oral prednisolone (1 mg/kg/day). His limb power improved with treatment.

Common causes of neurogenic orbital tumours include pilocytic astrocytoma, meningioma, schwannoma and neurofibroma.² Association of CIDP with neurogenic tumour of the orbit has not been previously reported. As these two conditions are two different entities, the association is most likely incidental.

Learning points

- ▶ Common causes of neurogenic orbital tumours include pilocytic astrocytoma, meningioma, schwannoma and neurofibroma.
- ▶ Association of chronic inflammatory demyelinating polyneuropathy with neurogenic tumour of the orbit has not been previously reported.
- ▶ As these two conditions are two different entities, the association is most likely incidental.

Figure 1 (A) MRI T1-weighted (T1-W) image without contrast, axial view, shows a 0.6 cm×0.6 cm×1.2 cm hypointense lesion (arrow) extending around the opening of the left superior orbital fissure and optic canal, lying in close proximity to the optic nerve, which is suspicious of neurogenic tumour. The optic nerve is medially displaced with buckling noted. (B) MRI T1-W fat-saturation image with contrast, axial view, shows the lesion has significant contrast enhancement (arrow). (C) MRI T1-W fat-saturation image with contrast, coronal view, shows the lesion has significant contrast enhancement (arrow).

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Table 1 Nerve conduction study

Nerve and site	Latency	Amplitude	Segment	CMAP duration	CMAP area	Conduction velocity	
Motor nerve conduction							
Median R							
Wrist	5.7 ms (N<4)	5.6 mV (N>5.6)	Abductor pollicis brevis-wrist	10.5 ms	20.3 mVms	m/s (N>47)	
Elbow	29.2 ms	2.0 mV	Wrist-elbow	8.9 ms (–15%)	3.7 mVms (–82%)*	10 m/s	
Ulnar R							
Wrist	5.1 ms(N<3.9)	5.7 mV (N>4.5)	Abductor digiti minimi (manus)-wrist	7.0 ms	15.2 mVms	m/s (N>47)	
Below the elbow	15.7 ms	1.8 mV	Wrist-below the elbow	7.0 ms (+0%)	4.0 mVms (–74%)*	21 m/s	
Above the elbow	17.9 ms	1.4 mV	Below the elbow-above the elbow	9.0 ms (+28%)	2.3 mVms (–42.5%)*	36 m/s	
Tibial R							
Ankle	9.1 ms (N<6.3)	3.7 mV (N>5.9)	Abductor hallucis-ankle	8.2 ms	10.5 mVms	m/s (N>44)	
Popliteal fossa	30.1 ms	1.6 mV	Ankle-popliteal fossa	10.8 ms (+32%)	7.6 mVms (–31%)	19 m/s	
Peroneal R							
Ankle	5.0 ms (N<5.9)	0.9 mV (N>2.3)	Extensor digitorum brevis-ankle	8.9 ms	2.4 mVms	m/s (N>43)	
Fibula (head)	13.9 ms	1.1 mV	Ankle-fibula (head)	12.6 ms (+42%)	4.0 mVms (+40%)	34 m/s	
Popliteal fossa	19.7 ms	0.8 mV	Fibula (head)-popliteal fossa	15.1 ms (+20%)	3.2 mVms (–20%)	10 m/s	
Tibial L							
Ankle	5.3 ms (N<6.3)	10.5 mV (N>5.9)	Abductor hallucis-ankle	8.2 ms	23.9 mVms	m/s (N>44)	
Popliteal fossa	20.2 ms	2.7 mV	Ankle-popliteal fossa	9.8 ms (+20%)	7.1 mVms (–70%)*	27 m/s	
Peroneal L							
Ankle	5.6 ms (N<5.9)	1.6 mV (N>2.3)	Extensor digitorum brevis-ankle	7.2 ms	4.3 mVms	m/s (N>43)	
Fibula (head)	15.0 ms	1.7 mV	Ankle-fibula (head)	7.5 ms (+4%)	4.3 mVms (–0%)	32 m/s	
Popliteal fossa	15.9 ms	1.7 mV	Fibula (head)-popliteal fossa	7.7 ms (+3%)	4.1 mVms (–5%)	44 m/s	
Median L							
Wrist	8.1 ms (N<4)	3.2 mV (N>5.6)	Abductor pollicis brevis-wrist	5.9 ms	5.7 mVms	m/s (N>47)	
Elbow	20.8 ms	2.1 mV	Wrist-elbow	6.3 ms (+7%)*	4.2 mVms (–26%)	16 m/s	
Ulnar L							
Wrist	4.4 ms (N<3.9)	5.0 mV (N>4.5)	Abductor digiti minimi (manus)-wrist	6.2 ms	10.7 mVms	m/s (N>47)	
Below the elbow	12.7 ms	1.2 mV	Wrist-below the elbow	12.9 ms (+108%)	6.6 mVms (–38%)*	22 m/s	
Above the elbow	18.3 ms	2.6 mV	Below the elbow-above the elbow	10.1 ms (–22%)	7.3 mVms (+11%)	38 m/s	
Nerve and site	Onset latency	Peak latency	Amplitude	Segment	Latency difference	Distance	Conduction velocity
Sensory nerve conduction							
Ulnar L							
Wrist	NR ms	NR ms	NR μV	Digit V (little finger)-Wrist	ms	140 mm	m/s
Median L							
Wrist	NR ms	NR ms	NR μV	Digit II (index finger)-Wrist	ms	140 mm	m/s
Sural L							
Lower leg	NR ms	NR ms	NR μV	Ankle-Lower leg	ms	140 mm	m/s
Sural R							
Lower leg	NR ms	NR ms	NR μV	Ankle-Lower leg	ms	140 mm	m/s
Ulnar R							
Wrist	NR ms	NR ms	NR μV	Digit V (little finger)-Wrist	ms	140 mm	m/s
Median R							
Wrist	NR ms	NR ms	NR μV	Digit II (index finger)-wrist	ms	140 mm	m/s
Nerve	M latency			F latency		F wave %	
F wave studies							
Median L	6.5					0.0	
Ulnar L	4.6			24.4 (N<28)		90.0	
Tibial L	5.4			99.9 (N<55)		10.0	
Peroneal L	5.1					0.0	
Tibial R	8.9			56.5 (N<55)		10.0	
Peroneal R	5.7					0.0	
Median R	7.3					0.0	
Ulnar R	5.0					0.0	

Nerve conduction study (conducted at room temperature 25°C) shows prolonged distal latencies and reduction in conduction velocities in the median, ulnar, tibial and peroneal nerves. The compound motor action potentials were reduced in the right tibial, left median and both peroneal nerves. There were possible conduction blocks in the right median nerve below the elbow, both ulnar nerves below the elbow and left tibial nerve below the popliteal fossa. The sensory nerve action potentials were not recordable.

*Significant conduction block.

CMAP, compound motor action potential; L, left; N, lower limit of normal ranges; NR, not recordable; R, right.

Figure 2 (A) MRI T2-weighted (T2-W) image short T1 inversion recovery (STIR) sequence, coronal view, shows diffuse thickening of cervical nerve roots (arrows). (B) MRI T2-W image STIR sequence, coronal view, shows diffuse thickening of the brachial plexuses (arrows).

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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