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Paroxysmal torticollis and blepharospasm following bilateral cerebellar Infarction

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Sirs: Paroxysmal secondary dystonia is most frequently due to multiple sclerosis. We here present a case of paroxysmal cervical dystonia following bilateral cerebellar infarction; no similar syndrome has previously been reported.

A 35 year-old woman developed sudden weakness of her legs associated with vertigo, vomiting and blindness. These symptoms resolved within 24 hours of hospital admission; examination revealed a left superior homonymous paracentral scotoma with mild symmetric upper limb and moderate truncal ataxia. Three days later she developed frequent paroxysmal episodes of rotational torticollis to the right side with simultaneous bilateral blepharospasm. These episodes occurred without warning, were stereotyped in nature, and lasted 20-30 seconds without alteration of consciousness; they stopped when she rested in bed but immediately began on sitting and were further increased in frequency by standing and by mental arithmetic. In between attacks at rest there was no dystonic posturing. Over 2 weeks the truncal ataxia and the movement disorder resolved. MRI (T2-weighted axial FLAIR-sequence) demonstrated areas of high signal in the cerebellar hemispheres in the territories of the right superior cerebellar artery (Fig. 1a) and the medial branch of the left posterior inferior cerebellar artery (Fig. 1b), and in the territory of the right posterior cerebral artery in the right occipital lobe and calcarine cortex (Fig. 1c); no abnormalities were evident on sagittal or axial MRI through the brainstem. A diagnosis of multiple posterior circulation cerebral infarcts was made. Trans-oesophageal echocardiography with bubble contrast showed no patent foramen ovale. MR angiography (not formal angiography) of the aortic arch, anterior cerebral circulation, and vertebrobasilar circulation showed no significant extracranial stenosis or dissection. EEG during the abnormal movement, CSF analysis and a thrombophilia screen were normal. There was no family history of dystonia, scoliosis, or tremor; there was no personal history of trauma, migraine with aura, perinatal distress or exposure to any potentially dystonia-inciting drug. Risk factors for stroke included the oral contraceptive pill (OCP) and cigarette smoking; in view of these risk factors and the possibility of an occult source of thromboembolism the patient was treated with anticoagulation for 6 months and advised both to discontinue the OCP and not to smoke. One year later the only abnormality remaining was the visual field defect.

Dystonia secondary to infarction in the cerebellum is rare. The anatomical site of infarction causing dystonia is usually in the lentiform nucleus [1] or ventromedial mesencephalon and red nucleus area affecting the substantia nigra, nigrostriatal and cerebellothalamic fibres;[2] however, a recent review of 25 cases of secondary cervical dystonia [3] included two patients in whom structural brain lesions (hemangioblastoma and cavernous angioma) were localized exclusively to the left and right cerebellar hemisphere respectively, resulting in left-sided torticollis in both cases. On the basis of this series the authors noted that co-lateralization of the anatomical site of the lesion with the dystonic syndrome was inconsistent, especially for lesions in the posterior cranial fossa, and hypothesized that the pathophysiology involved disruption of excitatory projections to the inferior olive. Our case is anatomically consistent with this theory; furthermore, blepharospasm has been reported in the context of bilateral cerebellar lesions,[4] and microinjection of kainic acid into the cerebellar vermis elicited reliable and reproducible dose-dependent dystonic posturing in mice.[5] Both primary dystonia and post-stroke dystonia associated with lesions in the basal ganglia and thalamus have been associated with altered functioning of the cerebellum on fMRI.[6]

Experimental evidence raises the possibility that the postural nature of the movement disorder may be due to vestibular dysfunction. In cats, bilateral vestibular neurectomy resulted in modulation of the firing rate of vestibular nucleus neurons by tilt

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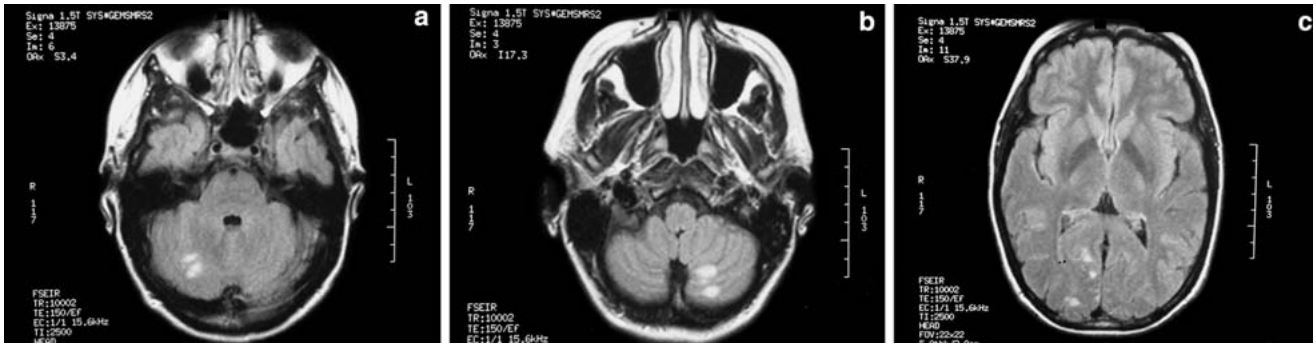

Figure 1

[7] and unilateral vestibular neurectomy resulted in increased muscle tone in the ipsilateral splenius capitis muscle with hypoactivity in the contralateral muscle which resolved after 5 weeks. [8] A haemodynamic explanation seems less likely as no postural disturbance in blood pressure was noted.

Paroxysmal dystonic episodes are most typically seen in the tonic spasms of multiple sclerosis; previous reports have implicated contralateral demyelinating lesions in the posterior limb of the internal capsule or cerebral peduncle but not in the cerebellar hemispheres.[9]

In summary, the present case emphasises the potential role of cerebellar pathology in the pathogenesis of secondary cervical dystonia. Systematic clinico-pathologic analysis of larger numbers of cases is needed in order to discern the relative contributions of different neuronal networks.

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