LETTERS TO THE EDITOR

Familial recurrent multiple cranial nerve palsies

Two brothers presented in their mid-40s, each with an isolated cranial nerve palsy. Both had a history of cranial nerve lesions. Of particular interest was the fact that one had an eighth nerve lesion, previously unreported, as part of a familial recurrent multiple cranial nerve syndrome. A 45 year old man presented in 1986 with a 14 day history of right frontal headache and three days of diplopia. He gave a history of a left seventh nerve palsy in 1968 and a right seventh nerve palsy in 1976, each episode lasting about six weeks and resolving spontaneously.

Examination showed a right sided third nerve palsy with slight ptosis. His pupils were normal, as was his general neurological examination. Investigations including evoked response studies were normal. His cranial nerve palsies resolved within six weeks and he has had no further symptoms since. In 1992, the 46 year old brother of the previous patient presented with a 10 day history of frontal headache, diplopia, and pain behind the left eye. He gave a medical history of several attacks of acute vertigo in 1975. These were associated with tinnitus and initially numbness of the right side of his face and right arm. These episodes occurred during a two week period with further attacks three months later. Ear, nose, and throat examination was normal. He was diagnosed as having Meniere’s disease but the sequence of events was not typical: nor was the non-progressive nature of the symptoms, which resolved without recurrence within a few months.

Examination showed a fourth nerve palsy. Investigations including evoked response studies, CT and cerebral angiography were all normal. After three months he was asymptomatic and the palsy had resolved.

Familial recurrent multiple cranial nerve palsies have rarely been reported. Stone1 described recurrent seventh nerve palsies in three brothers, one of whom also had a third nerve palsy. Lisch2 reported two families. In the first, a pair of twins had recurrent seventh nerve palsies and one of them also had an episode of “eye muscle paresis”. In the second family three generations had recurrent facial palsies and one member also had an “eye muscle palsy”. Currie3 described a family with a history of diabetes mellitus in which four siblings had recurrent seventh and fourth nerve palsies. Klee and Moller4 described a family in Denmark in which all known (seven) members of three generations developed pareses of their third, and sixth or seventh nerves.

The brothers described in our report have the following clinical features in common with the patients of Lisch2 and Klee and Moller4: (1) No other neurological signs were present apart from the cranial nerve palsies. (2) There was no associated systemic upset. (3) Investigations including neuroradiology were normal. (4)

Spontaneous resolution of all symptoms occurred within a few months. (5) The episodes of nerve palsy were often separated by many years.

Recurrent non-familial multiple cranial nerve palsies have been widely reported. In almost all such studies, the nerve most commonly affected is the seventh, the next being the third. Fourth and eighth cranial nerve palsies, which occurred in the brothers reported here, are rarely seen as part of a recurrent cranial polyneuropathy. An eighth nerve lesion has not previously been described in familial recurrent cranial nerve palsies.

Clinicians should be aware of the possibility of a familial predisposition to cranial nerve lesions. The connection between our brothers’ histories was only made as one of them, leaving the clinic on his day of discharge made the passing comment that he had just learned that his brother “had the same thing” some years before.

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Wallenberg’s syndrome with delayed onset after cervical spine fracture: a case report

Spontaneous and traumatic dissection or occlusion of the vertebral artery is uncommon and may be asymptomatic, so that the true incidence of this disorder is unknown. Typical symptoms are neck or occipital pain together with other non-specific symptoms such as vertigo, nausea, and vomiting. Although the neurological deficits may be delayed for hours or days, they help to indicate the appropriate clinical diagnostic procedure to be used to distinguish between a non-specific posttraumatic syndrome and brainstem ischaemia. The time lapse between the injury and onset of symptoms is important as appropriate treatment might successfully prevent the development of further unwanted clinical symptoms. Thus it would be helpful if symptoms, other than those already known, could be described to aid in the early diagnosis of this condition.

Case history
A healthy 30 year old engineer had an accident while riding a snowboard resulting in indirect trauma to the left side of the neck. He immediately complained of neck and occipital pains. These were associated with motor and sensory impairments in the left arm. The radiograph of the cervical spine showed an anterior displacement of C5 over C6 and CT demonstrated a facet joint fracture at C6 on the left side (figure A). The neurological examination showed symptoms of a left C6 root lesion with radicular pain, hypoesthesia, and mild paresis. There was no evidence of cerebellar or cortical involvement. Osteosynthetic stabilisation was performed with a hookplate (AO) on the day of the accident and 12 days later the patient left the hospital in a fit condition with signs of improvement of the left C6 deficit.

Nineteen days after the trauma he suddenly complained of vertigo, nausea, and vomiting. On readmission to the hospital, neurological examination showed spontaneous nystagmus with a rotatory component to the right, nistagmus and paresis of the left eye, bulbar speech, paresis of the left palatum velum, and perioral hypoesthesia on the left side. The gait was ataxic with propulsion to the left. Leftsided limb dysmetria was present; C6 symptoms were unchanged. The symptoms suggested ischaemia of the left dorsolateral medulla oblongata, a condition corresponding to Wallenberg’s syndrome.

A control CT of the cervical spine confirmed that the osteosynthetic material had not changed position and there was no encroachment on the vertebral artery canal by a screw. Ultrasound (colour duplex) of the extracranial arteries indicated a proximal occlusion of the left vertebral artery, which was confirmed by MR angiography with signal missing below C2 (figure B). The proximal occlusion was documented by four vessel angiography with no filling of the left posterior infracerebellar artery and without any other vessel injury or abnormality.

(A) Fracture of vertebra C6 with fracture of the facet joint and the foramen transversum at the left side (arrow). (B) MR angiography shows a missing signal of the left vertebral artery under the level C2 (arrow), the occlusion of the artery between C6 and C2 could be verified by angiography with no filling of the left posterior infracerebellar artery.
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