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 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 erythrocyte sedimentation rate and cerebral angiography were all normal. His cranial nerve palsy 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 Menière'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 erythrocyte sedimentation rate, 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 cervical spine. 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, hypoaesthesia, 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, miosis and ptosis of the left eye, bulbar speech, paresis of the left velum palatinum, and perioral hypoaesthesia on the left side. The gait was ataxic with propulsion to the left. Left-sided 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.

The MR of the brainstem showed a small ischaemic lesion in the left dorsolateral medulla without any further pathology.

Until confirmation of the total occlusion of the left vertebral artery the patient was treated with cumarine to prevent arterial embolism. The clinical symptoms progressively disappeared over the next 30 days and the patient left hospital with only a slight ataxic gait and mild dysaesthesia within the left C6 segment.

Discussion

The patient presented with an anterior displacement and fracture of the left facet joint between level C5 and C6 and radicular pain in the left C6 distribution, and subsequently developed a left vertebral artery dissection with left brainstem ischaemia. It is suspected that the dissection occurred during the 19 day delay with subsequent occlusion including the posterior infracerebellar artery. By contrast with our case, most reports during the past decade concerning posterior circulation ischaemia connected with vertebral artery occlusion occurred spontaneously or were loosely related to minor trauma during sport or neck manipulation during chiropraxy. These typically occurred in young or middle aged adults, equally between the sexes.¹ Although traumatic fractures of the cervical spine occur often, injuries of the vertebral arteries with or without clinical symptoms are rarely evident. The incidence is postulated to be between 3% and 10%.²

In a retrospective study Parent and coworkers reviewed some 640 patients sustaining fractures of the cervical spine. Of these, 96 had facet involvement and in only five was injury of the vertebral artery diagnosed by initial major neurological deficits such as cerebellar infarction, cortical blindness, or pontine infarction, which have been documented by postmortem examination in two cases.³ All these patients had cervical fractures located at C5-C6 and in one case in combination with a fracture at C6-C7. Radiographs showed anterior dislocation at C5-C6 in four cases and at C6-C7 in one case. Bilateral facet fractures were evident in four cases.

Only one prospective study exists that considers the combination of facet joint dislocation of the cervical spine, the incidence of vertebral artery injury, and the neurological deficit.⁴ From 12 consecutively examined patients with facet joint dislocation (C5/C6 in seven, C6/C7 in three, and C4/C5 in two) nine showed an occlusion of at least one vertebral artery. Of these nine patients only two with bilateral facet joint dislocations had a transient neurological deficit. Further indications of a traumatic or spontaneous dissection of the vertebral artery are neck pain and symptoms of a C6 root compression.⁵

Thus a combination of neurological and radiological findings could lead to an early diagnosis and may indicate development of a dissection of the vertebral artery. We suggest that patients with the clinical symptoms and type of injury described here are prone to development of a dissection or occlusion of the vertebral artery. Early diagnostic procedures by non-invasive diagnostic techniques such as MR angiography and ultrasound techniques coupled with treatment at the onset of a possible dissection may help to prevent the formation of a microembolism or arterial occlusion.

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Herpes simplex virus type 2 ascending myeloradiculitis: MRI findings and rapid diagnosis by the polymerase chain method

Although neurotropic viruses are often suspected of causing spinal cord injuries, confirmation by early diagnosis is difficult. Ascending myelitis related to herpes simplex virus type 2 (HSV-2) infection has seldom been reported and the diagnosis could be established only at postmortem examination.¹⁻⁵ We report the case of an elderly woman with a subacute ascending myeloradiculitis. MRI showed spinal cord and sacral root involvement and the polymerase chain reaction allowed the rapid identification of HSV-2 DNA in the CSF.

A 76 year old woman was referred because of urinary retention and paraparesis. Three weeks previously, she had noted the progressive onset of anorexia, fever (38°C), weight loss (4 kg), and low back pain. Evaluation performed in another hospital showed negative bacterial cultures from blood and urine and CT of the thorax, abdomen, and lumbosacral spine was normal. Three days before admission she complained of right sciatalgia and rapidly developed lower extremity weakness and sphincter disturbances.

Neurological examination showed a flaccid paraplegia, a T10 sensory level, and a distended bladder. Deep tendon reflexes were absent in the lower limbs and plantar responses were both extensor. In the upper extremities strength was normal but reflexes were brisk and a bilateral Hoffman sign was noted. Mental state and cranial nerves were normal.

Non-enhanced T1 weighted images of the spine were normal. T2 weighted sequences showed a linear hyperintense signal at the T10 level and within the conus medullaris. T1 weighted images with contrast injection showed an enhancement of both the posterior meninges and the roots of the cauda equina.

Her CSF contained 73 leucocytes/mm³ (97% lymphocytes), 132 mg/dl protein and 68 mg/dl glucose. Electrophoresis of CSF protein showed 26% γ -globulins with an oligoclonal distribution and a raised IgG/albumin ratio (0.56; normal <0.25). A polymerase chain reaction was performed on CSF with a pair of primers that allowed the simultaneous detection of four viruses

of the herpes group. A strong signal was obtained on ethidium bromide stained gel. Characterisation of HSV-2 DNA was achieved by restriction analysis of the amplified product. α -Interferon in CSF was normal. The patient had no history of recurrent herpes genitalis. There was no serological evidence for borreliosis, HIV-1 or HIV-2, HTLV-I, Q-fever, listeriosis, cytomegalovirus, measles, varicella zoster, or Epstein-Barr virus infection. CD4 counts were normal and no cause for immunodepression could be identified.

Parenteral acyclovir (30 mg/kg daily) was given for 10 days and the patient's neurological state remained unchanged. Six days after admission, sparse vesicular lesions appeared on the patient's buttocks, internal aspects of the thighs, and lower part of the abdomen. Ten days later, numbness in both hands appeared. Examination showed bilateral arm and shoulder weakness and the disappearance of upper limb reflexes. The patient then became confused and drowsy, developed hyponatraemia and hyperazotaemia and died on day 21 after admission. Necropsy examination was not performed. Subsequently, CSF cultures were reported as positive for HSV-2. Analysis of serial serum samples showed a sevenfold rise in anti-HSV antibodies between admission and death. IgM antibodies were detected in one early serum sample. Analysis of the CSF and serum anti-HSV antibodies ratio showed the existence of specific intrathecal synthesis.

Various clinical syndromes have been linked to HSV-2 involvement in the nervous system. HSV-2 encephalitis typically occurs in the newborn, but accounts for less than 5% of herpetic encephalitis in children and adults. Acute, self limited meningitis is found in young adults with primary genital HSV infection. Sacral radiculitis with perineal dysaesthesias, autonomic dysfunction, and sometimes mild lower limb weakness may also be associated with herpes genitalis in young adults. In most cases, neurological symptoms occur two to seven days after the genital eruption and patients recover within three weeks.

By contrast, HSV-2 involvement in the spinal cord is rare and severe. Two cases of HSV-2 extensive myeloradiculitis have been reported in patients with AIDS simultaneously infected with cytomegalovirus,^{1,2} and two others in diabetic patients.^{3,4} HSV-2 necrotising myelopathy has also been found in association with malignancy.⁵ In all cases the outcome was fatal within four to seven weeks, and the diagnosis could only be made at necropsy, when HSV-2 was recovered from the spinal cord. There has been a single report of HSV-2 myelitis with a favourable outcome in which the virus was isolated from the CSF.⁶

In our patient, MRI clearly showed myeloradiculitis, on T2 weighted and gadolinium enhanced T1 weighted sagittal sequences. Although well correlated with the clinical features, however, MRI findings lacked specificity.

The direct diagnosis of nervous system infection by HSV is difficult as isolation of the virus from CSF is most often unsuccessful and serological confirmation is too late. Recently, the polymerase chain reaction has proved to be a powerful tool in the rapid diagnosis of meningoencephalitis due to herpes viruses. In this case, it allowed us to identify HSV-2 in the CSF immediately