

LETTERS TO THE EDITOR

Delayed postexertional headache, intracranial hypotension and racket sports

Benign exertional headache (BEH) is a well described condition of controversial pathophysiology. Recently, the International Headache Society separated this from the headache that is associated with sexual activity.¹ Runners' and footballers' migraine or weightlifters' cephalalgia are examples of exertional headaches related to sports. BEH has been described as a brief episode, lasting for a few seconds or minutes, during or immediately after exercise.

We describe two patients, one a tennis and one a squash-player. Both had an intense postural headache associated with intracranial hypotension after vigorous exercise. This suggests that strenuous brachial effort could induce a traumatic rupture of a nerve root sleeve with transitory leakage of CSF.

Case 1, a 32 year old woman without previous history of headache or cranial trauma, developed severe occipital headache and diplopia 12 hours after a hard tennis match. The pain was dull, continuous, and was relieved by lying down. Clinical examination was normal except for a right abducens palsy. A lumbar puncture showed a CSF pressure of 0 mmH₂O ("dry tap"), which rose to 70 mmH₂O on sitting. CSF analysis, cranial CT and MRI were normal. The headache and diplopia gradually resolved with rest, over 6 days.

Case 2, a 36 year old man without previous history of headache or cranial trauma, developed intense generalised headache 6 hours after playing in a squash match. The pain was severe and continuous, forcing him to lie in bed. The next day he complained of horizontal diplopia. Neurological examination showed a right abducens palsy, without any other abnormal signs. A lumbar puncture performed showed a CSF pressure of 25 mmH₂O, which rose to 80 mmH₂O in the sitting position. CSF analysis, cranial CT and MRI were normal. The headache disappeared in 7 days, and the abducens palsy in one month.

The development of delayed benign postural headache in our patients, together with the low CSF pressure and the normal analysis, cranial CT and MRI, are typical features of intracranial hypotension.² This headache usually develops following lumbar punctures, and it is attributed to the leakage of CSF from the spinal tap. This headache can also be a consequence of slight cranial trauma.³ It is surprising that less than 20 apparently "primary" cases of this type of headache have been described, without comment on any relationship with sport or exercise. One of these cases was similar to ours, and began after "4 hours of vigorous tennis",⁴ although the author did not elaborate on this. The onset of the headache in our patients also occurred after prolonged exercises in racket sports. It is possible that the pathogenetic mechanism is repeated traction of the brachial plexus and disruption of a nerve root sleeve. This could not be sufficient to cause a

lesion, but could cause a transitory leakage of CSF, and consequently intracranial hypotension. The repeated increment of CSF pressure, and possibly concomitant dehydration, could also facilitate the development of the BEH.⁵ Our patients refused to be subjected to isotope cysternography which may have shown a spinal fluid leak.⁶

We suggest that the development of a postural headache, with or without nerve palsy, several hours after the practice of a racket sport is a benign syndrome secondary to intracranial hypotension.

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Chronic inflammatory polyneuropathy associated with nephropathy and anti-factor VIII antibody: improvement with intravenous immunoglobulin

Chronic inflammatory polyneuropathy (CIPD) is thought to be a variant of Guillain-Barré syndrome (GBS), distinguished by its chronic and relapsing course.¹ Humoral and cellular immunity may be involved in both GBS and CIPD.² We describe a case of a autoimmune origin coexisting autoimmune disease with CIPD which supports this hypothesis.

A 77 year old woman noted gradual paraesthesia, hyperalgesia and weakness of all four limbs over a period of five months. Examination showed a slight symmetrical weakness of the distal and proximal muscles. There was no wasting or fasciculation. There was a glove and stocking decrease in proprioceptive and nociceptive sensation. She was areflexic but plantar flexor responses were normal. Gait was not possible without aid but physical examination was otherwise normal. Laboratory tests showed a raised partial thromboplastin time (greater than 70 s with control of 12.6 s) and hypoalbuminaemia (30 g/l) with proteinuria (1.5 g/day). Blood tests, including glucose, oral glucose tolerance test and creatinine, were normal. CSF protein was 84 mg/dl with one lymphocyte/ μ l. Electromyography showed fibrillation and positive waves in the distal and proximal muscles. Motor conduction velocities were decreased in the peroneal (31 m/s) and median (33 m/s) nerves. Distal motor latencies were slightly prolonged in the median (6 ms), ulnar (6 ms) and peroneal (5.7 ms) nerves. There were conduction blocks in the median, ulnar and peroneal nerves.

Sural nerve biopsy showed severe segmental demyelination and perivascular mononuclear infiltrates affecting the small vessels. Continuous bleeding from the scar was observed. Further haematological tests showed that IgG factor VIII coagulant activity was less than 8% of normal and that a factor VIII inhibitor was present. Healing of the scar was achieved after repeated intravenous infusion of factor VIII.

A search for occult malignancy and other haematological disorders was negative. HIV serology, rheumatoid factor, complement levels, fluorescent antinuclear antibody, anti DNA antibody, cryoglobulinaemia, circulating immune complexes were all negative or normal. Prednisone, 60 mg daily for two months was given, without clinical improvement. Her hypoalbuminaemia (24 g/l) and proteinuria (up to 4.2 g/d) deteriorated. However, partial thromboplastin time and factor VIII coagulant activity became normal after one month of treatment with prednisone, and factor VIII inhibition disappeared. The patient was then treated with high dose intravenous immunoglobulin (IVIG) (0.4 g/kg/day for 5 consecutive days). Her condition improved dramatically after two infusions. Paresthesiae and sensory loss disappeared and she was able to walk unaided. There was no muscle weakness on neurological examination. Motor and sensory conduction velocities were unchanged in all four limbs but the conduction blocks were not found at the second examination.

Her condition improved after three further courses of IVIG given over a period of three weeks. After the first course of IVIG, albuminaemia was 30 g/l and proteinuria 1.3 g/l. One year later, albuminaemia was 38 g/l, proteinuria 0.05 g/l. After gradual withdrawal of prednisone, coagulation tests remained normal.

This patient fulfilled the diagnostic criteria for CIPD.¹ These include proximal and distal weakness, tendon areflexia, sensory loss, marked slowing of motor nerve conduction with conduction block, albuminocytological dissociation in CSF, and progressive clinical deterioration for more than two months. The diagnosis was supported by sural nerve biopsy findings of demyelination with perivascular inflammation. These clinical and pathological features of CIPD were associated with factor VIII antibody and renal abnormalities.

The association of CIPD, factor VIII antibody and nephropathy had not previously been reported. However, factor VIII antibody has been seen during pregnancy, and in collagen vascular diseases, multiple myeloma, lymphoma, multiple sclerosis and drug reactions.³ These conditions were excluded in our case. The association of factor VIII antibody with CIPD could, however, be the expression of a common autoimmune origin. Only one previous report noted this association⁴ and steroids failed to improve the haematological disturbance. The renal abnormalities in our patient were consistent with a glomerulopathy. Renal involvement coexisting with CIPD has been described.⁵ In one reported case, the renal involvement was due to *in situ* immune complex formation. CIPD, factor VIII antibodies and nephropathy suggest an autoimmune disease. Each of these conditions are thought to be autoimmune, but their mechanism is unclear. Each improved with different types of treatment. Factor VIII antibody entirely disappeared with prednisone, while CIPD showed resist-

ance to this treatment but was improved by IVIG. There was a temporal relationship between neurological and renal improvement and IVIG, and this could reflect a common pathogenesis. Recognition of such coexisting disorders provides further evidence of an immune dysfunction in CIDP.

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Acute stage Bell's palsy and narrowing of the palpebral fissure caused by drooping of the eyebrow and the upper eyelid

Widened palpebral fissure on the affected side has been regarded as one of the hallmarks of peripheral facial palsy.^{1,2} The palpebral fissure of the affected eye may, however, be narrower than that of the healthy eye.³⁻⁵

Twenty five patients with Bell's palsy in the acute stage were examined at our outpatient

clinic from January 1990 to May 1991. No other neurological signs were observed in these patients. Ten patients had a narrowed palpebral fissure rather than widened palpebral fissure on the affected side looking straight ahead. Their ages ranged from 32-59 years with a mean of 46.8 years. Seven patients were men and three women, four being diabetic. All 10 patients complained of a drooping of the upper eyelid on the affected side when opening their eyes. All had a furrowless forehead and the corner of the mouth drooped on the affected side, both characteristics of peripheral facial palsy. They were unable to close the affected eye tightly, and five patients had lagophthalmos (incomplete closure). The position of the eyebrow and the upper eyelid on the affected side was lower and the width between their lower margins was shorter than on the non-affected side (fig). The patients experienced difficulty in elevating the eyebrow and the upper eyelid when gazing upwards. The position of the eyes was intact, however, and the extraocular movements were full. The pupils were round, equal in size and reactive to light.

All the patients had CT or MRI brain scans. All were free from intracranial lesions. Neuroelectrophysiological examinations were performed in five patients and in these patients the distal latency of the facial nerve was prolonged and surface electromyogram of the frontal muscle showed decreased activity on the affected side.

The principal muscle involved in opening the upper eyelid and maintaining normal eyelid posture is the superior palpebral levator, which is innervated by the oculomotor nerve. Two accessory muscles are Müller's muscle via the oculosympathetic pathway and the frontal muscle by the facial nerve. The upper eyelid is indirectly elevated by the attachments of the frontal muscle into the eyebrow with the superior orbital portions of the orbicularis oculi muscle.⁶ Thus it is possible that the weakness of the frontal muscle causes drooping of the eyebrow and the upper eyelid, resulting in narrowing of the palpebral fissure. This phenomenon might be more noticeable in Japanese than in

white patients, because Japanese patients are more "heavy lidded".

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Familial recurrent cranial nerve palsy

Familial recurrent cranial nerve palsy is a syndrome of unknown aetiology in which affected family members experience recurrent paresis of the oculomotor, abducens, and facial nerves in an apparent autosomal dominant inheritance pattern.¹ Typically, these individuals experience acute onset of a cranial nerve palsy followed by gradual resolution of the deficit over several months. This presentation and natural history is similar to that of patients with diabetic vasculopathic cranial nerve palsies and prompted us to search for a vascular aetiology in the patient and family members we describe.

A 36 year old man developed binocular, horizontal diplopia in August 1990. He had experienced an idiopathic, peripheral, right facial nerve palsy (Bell's palsy) in October, 1985 and a left Bell's palsy in May 1986. He was an otherwise healthy nurse anesthetist taking no medications. On examination, best corrected visual acuity was 6/6 in each eye and colour vision was normal. Pupils were briskly reactive without anisocoria or a relative afferent pupillary defect. Visual fields were full. Facial sensation was intact, and there was no ptosis or proptosis. There was mild orbicularis weakness bilaterally and evidence of aberrant regeneration of both facial nerves. The patient had an ocular motility disturbance consistent with a left abducens nerve palsy: abduction of the left eye was decreased and there was a 25 prism diopter (PD) esotropia in primary gaze that increased to 45 PD in attempted left gaze. The remainder of the examination was unremarkable.

A complete blood count, thyroid function tests, rheumatoid factor screen, serum protein electrophoresis, assay for acetylcholine esterase antibodies, assay for angiotensin converting enzyme, Lyme titres, edrophonium test, chest radiograph, and magnetic resonance imaging were all normal with the exception of a mildly elevated cholesterol level in the serum (252 ng/dl) and a mildly elevated CSF protein (55 mg/dl). The left

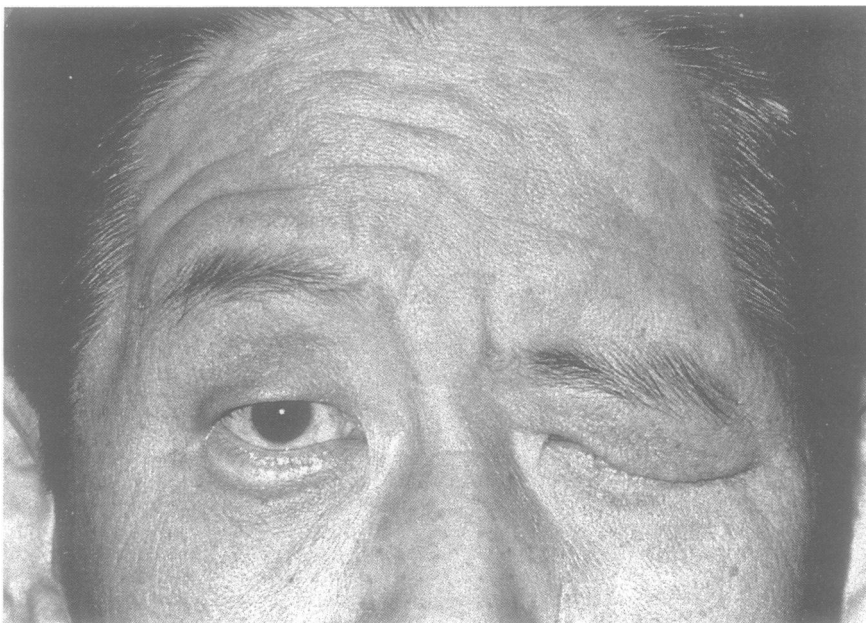


Figure Patient looking straight ahead. Narrowing of the palpebral fissure is seen on the left side.