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 mmH_2O ("dry tap"), which rose to 70 mmH_2O 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 mmH_2O, which rose to 80 mmH_2O 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 (CIDP) 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 CIDP.[2] We describe a case of a autoimmune origin coexisting autoimmune disease with CIDP which supports this hypothesis.

A 77 year old woman noted gradual paraesthesia, hyperalgia 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/μl. 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 CIDP.[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 CIDP were associated with factor VIII antibody and renal abnormalities.

The association of CIDP, 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 CIDP could, however, be the expression of a common autoimmune origin. Only one previous report noted this association[4] and steroids failed to improve the haematological disturbance. The renal abnormalities in our patient were consistent with a glomerulopathy. Renal involvement coexisting with CIDP has been described.[5] In one reported case, the renal involvement was due to *in situ* immune complex formation. CIDP, 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 CIDP showed resist-