Posterior ischaemic optic neuropathy after a spontaneous extradural haematoma

Most extradural haematomas (EDH) occur after head injury but cases of “spontaneous” EDH have been reported. These are a result of either local sepsis, a dental arteriovenous malformation or an abnormality of clotting. We report the case of a spontaneous EDH resulting in posterior ischaemic optic neuropathy as a consequence of tentorial herniation.

A 15 year old boy developed a headache which built up over a period of 30 minutes. In the evening he was seen by his general practitioner who diagnosed a viral infection as the headache was associated with some fever. On the second day he vomited in the morning. By the morning of the third day he had fever and was unable to get up because of headache and giddiness. His mother saw him at approximately 1.00 am on the fourth day of his illness when he seemed to be sleeping normally. By 7.00 am the same morning he was found unconscious and was admitted to the District General Hospital on 17 March 1988. There was no other predisposing illness or head injury.

In the accident and emergency department, he was noted to be cyanosed with vomit in his nose and throat. He was in coma with a Glasgow Coma Score of 5. Both pupils did not respond to light and the right was 6 mm, the left 3 mm. His auxiliary temperature was 40°C, a regular pulse of 110, blood pressure 120/55 mmHg and normal heart sounds. Auscultation of his chest revealed added sounds in the left base suggesting an aspiration pneumonia which was confirmed on chest x ray. His blood results were as follows: Hb 1·58 g/dl, white cell count 11·7 x 10⁹/l, platelet count 180 x 10⁹/l, sodium 140 mmol/l, potassium 4·2 mmol/l and urea 4·1 mmol/l. Clotting was within normal limits with an INR 1·0 and KCCCT (control 48).

Before being transferred, the patient was intubated, ventilated, paralysed, sedated and given 100 ml mannitol 20%. On arrival in the neurosurgical unit on 17 March 1988, a CT was performed (figure) which showed a 3 cm maximum depth EDH over the right frontal convexity which was of mixed density. There was no radiological evidence of head injury. In view of his condition and the radiological findings, an immediate right frontal craniotomy was performed. There was a moderately sized EDH was found. The haematoma was considered to be of some days duration. No evidence was found of either a fracture or a dental arteriovenous malformation.

On the evening of arrival he developed the patient a transient diarrhoea which resolved and there were no further episodes. The patient was extubated four days postoperatively and his conscious level continued to improve. Initially his vision was very restricted and he could see only a few objects. Examination showed that the pupils, although unequal, were reacting to light. He appeared to have a left homonymous hemianopia very early after surgery. This was not demonstrable subsequently as he has remained completely blind since then. He was also noted to have a mild left hemiparesis (Medical Research Council grade 4). One month later he was discharged and his conscious level had returned to normal, his hemiparesis had resolved but he was blind.

On review in the outpatient department three months later, his vision was unchanged with no light perception in either eye. He had put on 19 kg in weight. He had polydipsia and, according to his mother, prolonged and excessive periods of sleeping. On examination he was alert and orientated with no limb weakness. His pupils did not react to light and funduscopy revealed bilateral optic atrophy.

Visual evoked potentials showed no response. A CT scan showed moderate atrophy of the right frontal lobe and mild dilatation of the ventricular system compared with the previous scan. The orbits, sella and suprasellar cisterns appeared normal. MRI showed slender optic nerves consistent with atrophy and confirmed the frontal atrophy. There was no evidence of hypothalamic abnormality on the scan. He had an endocrine assessment six months postoperatively which showed an isolated defect of growth hormone production.

One year postoperatively the patient remains blind with fixed pupil, protect substance gain from MPP+ neurotoxicity7 in rats. Injection of the NDMA antagonist MK 801 within the medial pallidum reverses Parkinsonian symptoms in MPTP-treated monkeys. The mechanism of action remains unclear but it has been suggested that blockade of NDMA receptors would result in a facilitation of dopamine action by preventing the glutamate-induced dephosphorylation of DARPP-32, a dopamine- and cAMP-regulated phosphoprotein. However, no clinical data are yet available in Parkinson’s disease (PD).

We investigated in PD patients the effects of an add-on therapy with ifenprodil, a non competitive antagonist of the NDMA receptor at the polypeptide modulatory site, which inhibits [3H] MK 801 binding.8 This drug is, as far as we know, the sole NDMA antagonist currently available on the market. Ifenprodil also possesses alpha-adrenoceptor blocking properties and is already in clinical use as a central antihypertensive agent.

Two groups of patients with idiopathic PD were studied after informed consent: the first one included nine non-fluctuating patients, mean age (SD) 67 (10) years, mean (SD) duration of levodopa therapy 7 (1) years,