Letters

Benign intracranial hypertension and essential thrombocythaemia

Sir: A number of neurological syndromes are associated with essential thrombocythaemia. We report a case of essential thrombocythaemia and benign intracranial hypertension. A 21 year old girl presented with a 2 month history of constant occipital headache associated with nausea and vomiting but no visual disturbance. She had been on an oral contraceptive pill 14 months prior to the onset of her symptoms. She took no other medication. On examination, the patient was overweight. Neurological examination revealed bilateral papilloedema, enlarged blind spots and normal peripheral visual fields (bedside testing). Visual acuity was normal being 6/6 L and 6/5 R. There were no other abnormal signs. Her blood pressure was 140/80 mmHg. Investigations on admission included a normal CT brain scan, a CSF opening pressure 24-5 cmH2O, an acellular fluid with a protein of 0.5 g/l and sugar 3-5 mmol/l (blood sugar 5-5 mmol/l). Full blood count showed a haemoglobin of 13.5 g/dl, a white cell count of 15.9 × 10^9/l with 68% neutrophils and a platelet count of 1162 × 10^9/l. Routine biochemistry was normal and the antinuclear factor negative. A diagnosis of benign intracranial hypertension was made and the patient started on acetazolamide 500 mg s bd. After lumbar puncture the patient's headache was no better and she remained symptomatic on the acetazolamide. A digital intraventricular angiogram (DIVA) looking for dural sinus blockage was performed. This was normal. The haematological problem was further investigated with bone marrow aspirate which showed a cellular marrow with increased numbers of megakaryocytes. Morphologically the megakaryocytes appeared abnormal. Platelet function tests were normal as was the leucocyte alkaline phosphatase score. The height of the platelet count together with the neutrophil leucocytes and the abnormal morphology of the megakaryocytes in the marrow suggested that the thrombocytopoiesis was primary rather than secondary. Two weeks after admission to hospital the patient complained of a painful left calf. A left popliteal vein thrombosis was confirmed on venography. The patient was anticoagulated with heparin and then warfarin. Antithrombin III levels were normal. She remained on warfarin for three months. During this time the headaches increased in severity. The papilloedema and blind spots appeared unchanged and the visual acuity remained normal. Because the patient was anticoagulated repeat lumbar puncture was not performed. She was given steroids for two weeks but continued to have headaches. The platelet count at this stage was still above 1000 × 10^9/l and a decision was taken to reduce the platelet level in the hope that this would relieve symptoms. She was commenced on hydroxyurea 1.5 g orally daily. One week after starting treatment the patient noticed a strikingly significant improvement in severity and frequency of the headache. The optic discs appeared less swollen and the blind spots returned to normal. Visual acuity was unchanged. The platelet count fell to 339 × 10^9/l and the hydroxyurea was stopped after three weeks.

On review in out-patients the platelet count showed a consistent tendency to increase and with this the headaches occurred although not as bad as previously. There was no evidence of papilloedema at this stage and repeat lumbar puncture was not carried out. She was commenced on aspirin which tended to reduce the platelet count somewhat but six months after the initial presentation the platelet count was again elevated at 845 × 10^9/l. She was restarted on hydroxyurea 1.5 g daily. After one week the platelet count had fallen to 644 × 10^9/l and she became asymptomatic. A repeat bone marrow aspirate and trephine biopsy showed a normal cellular marrow with numerous platelet clumps and increased reticulocytes. These findings were consistent with essential thrombocythaemia.

We postulate that the elevated platelet count may have given rise to intermittent dural sinus blockage plus calf vein thrombosis. There has been one previously reported case of lateral sinus thrombosis and essential thrombocythaemia treated with busulphan. We chose hydroxyurea in preference to busulphan because of the former's lesser side effect on ovarian function. There has also been one previously reported case of benign intracranial hypertension associated with thrombocythaemia post splenectomy.

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References

Epidural haematoa of the posterior fossa: good results after prompt diagnosis with CT

Sir: Epidural haematoa of the posterior fossa may cause rapid and fatal deterioration if not promptly treated. Early recognition is therefore extremely important but the lesion is rare and easily missed. An extradural clot in the posterior fossa is present in 0.3% of all head injuries, and accounts for only 3–4% of 7% of all intracranial epidural haematoa.1–7 Mortality rates have varied from 33.3 to 100%.4 We report six cases in order to emphasise the value of CT scanning in early diagnosis and in improving outcome.

Between January 1978 and June 1987 131 patients with a posttraumatic epidural haematoa were treated in the Neurosurgical Department of the University Hospital of Rotterdam. The haematoa was shown by CT in the posterior fossa in six patients (5%; 95% CI: 1–9%). Patients' records were studied for age, sex, type of injury, duration of lucid interval, neurological signs and symptoms, delay between signs and symptoms and CT scanning, the presence of occipital fracture on skull radiographs, CT findings, treatment, delay between diagnosis and treatment, and neurological signs and symptoms at discharge.

There were four males and two females, aged 4 to 38 years (median 25 years). Two of our patients were in the first decade. Five patients had sustained a fall on the back of the head during a mild injury; one patient had had a car accident. Symptoms developed within 24 hours in three, after 2–7 days in two and after 1 week in one patient. A typical lucid interval was present in four patients. The lucid interval varied from 1 to 24 hours. Five patients had impaired consciousness on admission; one of these was in coma. The Glasgow Coma Scale score was between 7 and 15 on the 3–15 point scale. Two patients...