

Letters

Benign intracranial hypertension and essential thrombocythaemia

Sir: A number of neurological syndromes are associated with essential thrombocythaemia.^{1,2} 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 $15.9 \times 10^9/l$ with 68% neutrophils and a platelet count of $1162 \times 10^6/l$. Routine biochemistry was normal and the antinuclear factor negative. A diagnosis of benign intracranial hypertension was made and the patient started on acetazolamide 500 mgs bd. After lumbar puncture the patient's headache was no better and she remained symptomatic on the acetazolamide. A digital intravenous 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 leucocytosis and the abnormal morphology of the megakaryocytes in the marrow suggested that the thrombocytosis 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 $1000 \times 10^6/l$ 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 $339 \times 10^6/l$ 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 recurred 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 $845 \times 10^9/l$. She was restarted on hydroxyurea 1.5 g daily. After one week the platelet count had fallen to $644 \times 10^9/l$ and she became asymptomatic. A repeat bone marrow aspirate and trephine biopsy showed a normal cellular marrow with numerous platelet clumps and increased reticulin. 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 thrombocythaemia³ 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.⁴

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

Sir: Epidural haematoma 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,¹ and accounts for only 3-4.7% of all intracranial epidural haematomas.¹⁻⁷ Mortality rates have varied from 33.3 to 100%.^{4,7} 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 100 patients with a posttraumatic epidural haematoma were treated in the Neurosurgical Department of the University Hospital of Rotterdam. The haematoma 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

showed nuchal rigidity; three patients had uni- or bilateral abducens paresis. Three patients had an occipital fracture on skull radiograph. Besides the haematoma, CT showed mild hydrocephalus in three and slight frontotemporal contusion in two patients. Five patients were treated operatively with suboccipital craniotomy, immediately after the diagnosis was made. The patient who did not present until the second week was treated conservatively. All patients had good recovery: only one patient had a mild abducens paresis at discharge from the hospital.

Clinical diagnosis of an epidural haematoma in the posterior fossa is difficult because the symptoms and signs are usually not specific, especially in acute cases. The major sign is deterioration of consciousness: this was present in five of our patients but occurs with all types of traumatic haematoma. Although the classical lucid interval is reported to occur in only a minority of patients,^{1-3,5,6} four of our patients had a lucid interval varying from one to 24 hours. Diagnosis is more easy when patients present more than a day after injury as in three of our cases: the signs may then be either of raised intracranial pressure or of a posterior fossa lesion (lower cranial nerve dysfunction, cerebellar signs, nuchal rigidity), or both. A haematoma in the posterior fossa was suspected in five of the reported patients because of deterioration of consciousness after occipital injury and in three patients because of either an abducens paresis or nuchal rigidity, or both. In all our patients who presented in the first week the haematoma was diagnosed by CT immediately after admission. Early recognition and diagnosis of a posterior fossa clot is possible if the possibility is considered in every patient with occipital injury. If such a patient develops neurological symptoms, especially deterioration of consciousness or deterioration of vital signs, CT should be undertaken, even if there is not a skull fracture. Before CT was available mortality was high because diagnosis was difficult; often, the extradural clot in the posterior fossa was not found until necropsy and many series included post-mortem cases,^{7,8} CT enables prompt diagnosis and treatment and improved results: it may show the associated intracranial lesions which are important for prognosis. In the report of Brambilla *et al* two of the three patients who had died, had bilateral frontal lacerated-contusive areas and haemorrhages of the brainstem and basal ganglia.¹ Three of our patients had only mild hydrocephalus and two had slight frontotemporal contusion. The lack of mor-

tality and minimal morbidity in our patients reflects prompt diagnosis with CT, followed without delay by surgical evacuation of the haematoma and to the lack of serious associated intracranial lesions.

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Disappearing CT lesions in epilepsy: is tuberculosis or cysticercosis the cause?

Sir: Goulatia *et al*¹ reported a series of patients, with seizures, who showed CT lesions which resolved over a 6-12 week period, when seizures were controlled with anticonvulsant medication. These were labelled as "Disappearing CT lesions". Similar CT findings in epilepsy have been reported by others.²⁻⁴ There has been considerable speculation regarding the nature of these lesions. The lesions cannot be solely attributed to the postictal state as they are

focal and take several weeks to resolve. Furthermore, such lesions are not common in patients with seizures. Since almost all cases are reported from India, an inflammatory aetiology is considered likely. Tandon and Bhargava⁵ considered these to be tuberculomas. They treated their patients with antitubercular drugs and advanced the argument that since the lesions then resolved, their contention was confirmed. Other possibilities to be considered are cysticercosis,¹ focal encephalitis² and microabscess.⁴ In the present study we investigated such patients for evidence of tuberculosis and cysticercosis by testing serum for antibodies against *M tuberculosis* and cysticercus using ELISA.

Thirty eight patients with seizures who on contrast enhanced CT of head done within 2 weeks of a seizure showed a single enhancing ring lesion or a hyperdense lesion (with or without surrounding hypodensity) and a complete or substantial resolution on CT scan 8-12 weeks later were studied. There were 16 male and 22 female patients. The age of the patients was from 7 to 65 years. Eighteen had generalised convulsions and 20 had partial motor seizures. None of the patients had neurological deficit on examination. Chest radiographs were obtained in all. Patients who had overt evidence of tuberculosis or subcutaneous nodules suggestive of cysticercosis were excluded. Also excluded were patients in whom the CT lesion remained unchanged after 8-12 weeks. Serum was tested for tubercular and cysticercus antibodies using ELISA. Antibodies to cysticercus and tubercle bacilli were measured by ELISA as described previously.^{6,7} Serum ELISA was also done in healthy controls, patients with systemic tuberculosis, CNS tuberculosis, documented cerebral cysticercosis and epilepsy.

The results of the ELISA tests are shown in the table. Twelve out of 38 patients with "disappearing lesions" had serology positive for cysticercosis while two were positive for tuberculosis. None of the healthy controls showed a positive reaction for cysticercosis. Nineteen out of 22 proven patients with neurocysticercosis were positive for cysticercus antibodies while none showed evidence of tubercular antibodies.

The prevalence of epilepsy is not significantly different in different areas of the world. CT scan is frequently done to detect underlying structural lesion in the brain. For reasons which are not understood, the "disappearing lesions" have almost exclusively been reported from India. Because of this geographic feature, an infection or infestation has been the prime suspect. Tandon and Bhargava considered these to be tuber-