Figure 2 Gadolinium enhanced coronal MRI scan demonstrating large intracranial mass.

beyond the sensitivity of CT or MRI cannot be eliminated.

Transient visual obscurations, usually lasting seconds, are a well-known accompaniment of raised intracranial pressure. They may occur with or without the presence of optic disc swelling. The pathogenesis remains something of an enigma. Hayreh⁴ explained the phenomenon as being due to ischaemia of the optic nerve head and downward herniation of the parahippocampal gyrus in the tentorial notch. Another theory is of intermittent occipital lobe ischaemia related to compression of the posterior cerebral artery usually from uncus herniation.⁵ Sadun *et al*⁶ suggested that transient ischaemia of the optic nerve head consequent to increased tissue pressure could explain the presence of unilateral visual obscurations occurring with or without raised intracranial pressure. This theory would, however, not explain the presence of bilateral visual obscurations in the presence of unilateral papilloedema.

This case serves to remind us that unilateral papilloedema, though rare, does occur and that transient

visual obscurations can occur without or prior to development of optic disc swelling.

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Sir,

Visual loss precluding acute onset of Budd–Chiari syndrome

Budd–Chiari Syndrome (BCS) occurring as a result of occlusion of the major hepatic veins manifests either acutely or chronically with a triad of abdominal pain, hepatomegaly and ascites. The natural course is progressive with a mortality rate of 75%.¹ Most patients require anticoagulation and urgent decompression or shunting to avoid progressive liver damage. We are unaware of any previous reports of retinal vein occlusions precluding this life threatening complication.

Case report

A 39-year-old lady of Nigerian ancestry presented to the eye casualty department of a London hospital with a 1-week history of painless visual loss in her left eye (LE).

She had been diagnosed with multiple sclerosis (MS) four and a half years previously after presenting with bilateral lower limb weakness, nystagmus, and paraesthesia but was currently without significant morbidity and taking no medication. A complaint of abdominal cramps and distension for the previous 2 months was also elicited. On examination, her Snellen visual acuity (VA) was 6/6 unaided in the right eye (RE) and 3/60 in the LE. Examination revealed mild bilateral posterior uveitis with evidence of active vasculitis associated with haemorrhages in the fundus of the LE (Figure 1a). There was obliteration and sclerosis of venous and arterial vessels with peripheral areas of retinal whitening. The fundus of the RE also showed a sclerosed vessel (Figure 1b). She was under investigation for ischaemic vasculitis, when a week later she was acutely unwell and admitted to the hospital with painful abdominal ascites, swelling of her left leg, diarrhoea, and vomiting. Blood parameters showed a markedly abnormal liver function (AST 745 IU/l (Normal; 10–50 IU/l)). Doppler ultrasound and computerised tomography showed inferior vena cava thrombosis extending to but not involving the right atrium. The hepatic veins were occluded with thrombus extending inferiorly to the bifurcation of the iliac veins and involving the renal veins. A working diagnosis of BCS was made and she was anticoagulated and transferred to the liver unit of our hospital where an emergency thrombectomy with liver transplantation and vena caval grafting was undertaken. She had a slow recovery surviving several episodes of systemic sepsis. A full thrombophilia screen, genotype analysis, and autoimmune profile were normal, as was the bone marrow examination. BCS secondary to BD was confirmed after the patient recalled a long history of painful oro-genital ulcers that the general practitioner had been treating as Herpes Labialis.

She was discharged on immunosuppressants, oral prednisolone, and remains on life-long anticoagulation treatment.

Comment

Doppler ultrasonography of the liver with a sensitivity and specificity of 85% or more² is the technique of choice for initial investigation of BCS but therapy remains controversial, usually requiring transjugular intrahepatic porto-systemic shunting or liver transplantation (LT). LT restores function and outcomes do not differ from that of other aetiologies of liver failure.¹ BCS as a complication of Bechet's disease (BD), was first reported by Maruyama³ in 1967 and is a rare but recognised severe complication of BD. Other predominant causes include; hypercoagulable states,

Figure 1 (a) Left fundus showing inferior vein occlusion and active vasculitis. (b) Right fundus showing sclerosed vessel inferonasally.

either inherited (eg antithrombin III deficiency) or acquired (eg myeloproliferative disease); infiltrative disease (eg hepatocellular carcinoma); trauma; and idiopathic.

BD has a well-described diagnostic criteria⁴ with ocular abnormalities occurring in 70–85% of patients.⁵ Acute anterior uveitis with hypopyon is a classical feature but posterior segment complications can result from vasculitis and ischaemia. This patient is having further neurological investigations to exclude central nervous system involvement, which could have been misdiagnosed as MS.

The pathogenesis of thrombus formation in Behçet's disease is poorly understood but endothelial injury leading to thrombus formation secondary to vasculitis is regarded as the initiating event. The effects of thrombophilic parameters, such as protein C, protein S, antithrombin deficiencies, antiphospholipid antibodies, factor V Leiden, and anticardiolipin antibodies have been studied and published reports have conflicting conclusions⁶ with no specific factor having a high correlation with thrombosis.

Systemic vascular complications occur in approximately 25–48%⁷ of cases and are the predominant cause of mortality. Vascular occlusion with posterior segment inflammation must alert the practitioner to the diagnosis of BD. These patients, frequently presenting to ophthalmologists, should be questioned specifically about abdominal symptoms and further investigation undertaken appropriately. BCS is a life threatening complication and clinical outcome can be improved if diagnosed early.

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Sir, Subinternal limiting membrane and subhyaloid haemorrhage in Terson syndrome: the macular 'double ring' sign

Dome-shaped posterior pole haemorrhages have been described in Terson syndrome (TS).^{1,2} However, the exact location of these posterior pole haemorrhages have been unclear.³ Friedman *et al*² showed that these haemorrhages are located beneath the internal limiting membrane (ILM) of the retina. The purpose of this report is to highlight the concurrent occurrence of a sub-ILM and subposterior hyaloid haemorrhage in TS producing a macular 'double ring' sign. We are unaware of any previous reports describing this sign and could find no reference to it in a computerized search utilizing the PubMed. Nd:YAG laser membranotomy of the ILM was successful in draining the sub-ILM haemorrhage and resorting the vision.

Case report

A 46-year-old female underwent endovascular coiling following a subarachnoid haemorrhage caused by a ruptured anterior communicating artery aneurysm. Following this, she complained of decreased vision in her left eye (LE). On ophthalmic evaluation, visual acuities were 6/6 right eye (RE) and counting fingers LE. Intraocular pressures and anterior segment examination were unremarkable. Fundus examination of the RE showed multiple scattered superficial and deep retinal haemorrhages involving the posterior pole (Figure 1a). The macula was normal. The LE showed a large dome-shaped macular sub-ILM haemorrhage characterized by a glistening light reflex and fine striae on the surface of the haemorrhage and a small dome-shaped subhyaloid haemorrhage located anterior and inferotemporal to the sub-ILM haemorrhage. These two haemorrhages caused a macular 'double ring' with the 'inner ring' presumably formed by the detached ILM and the 'outer ring' by the detached posterior hyaloid (Figure 1b).