Comment

Central retinal vein occlusion is extremely rare in children. In a review of 17 patients with CRVO under the age of 40,[5] only one was in the paediatric age group. We speculate that there was an initial optic disc swelling in association with neuroretinitis which precipitated a CRVO. Solley *et al*[6] reviewed 24 patients with cat scratch disease, of whom 13 had unilateral disease and three were under 16 years of age. They reported one case of branch retinal vein occlusion. Epidemiological data suggest that higher cat infection rates are associated with a warm climate, fleas and kittens. Most authors advocate treatment of *B. henselae* with antibiotics, and Rifampicin and Doxycycline are often used. Because of the age of this patient we used Clamythromycin. There is some debate as to whether antibiotics alter long-term outcome in *B. henselae* associated neuroretinitis which is good in most cases.[3]

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Sir,

Spontaneous orbital haemorrhage in purpura fulminans secondary to meningococcal septicaemia

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Spontaneous orbital haemorrhage is a very rare event.[1–3] Law in 1971 defined this condition to be the occurrence of haemorrhage within the orbit not caused by local trauma and not referrable, as far as can be ascertained, to any constitutional causative condition.

Meningococcal infection is associated with the development of an imbalance of haemostatic pathways.[4,5] A general shift towards activation of the pro-coagulant pathways leads to disseminated intravascular coagulopathy (DIC).

DIC is followed by a secondary bleeding diathesis, as pro-coagulant factors are rapidly consumed.[4–6] This bleeding tendency is thought to be responsible for the characteristic purpuric rash in meningococcaemia.[5]

In this clinical scenario, thrombosis and haemorrhage are known to occur in several organs and tissues,[5,6] but haemorrhage into the orbit from this cause has not been described before. We present a case of spontaneous orbital haemorrhage in a patient with purpura fulminans secondary to meningococcal septicaemia.

Case report

A 14-year-old female presented to the paediatric unit with a 12-h history of fever, irritability and diarrhoea. A few hours later this was followed by swelling around the left eye. The patient was previously fit and well with no significant past medical history. She had previous vaccination for group C meningococcal disease. On the morning of admission the patient developed widespread petechial and purpuric haemorrhages. Based on the clinical findings, a diagnosis of meningococcal septicaemia was made and intravenous benzyl penicillin started. The patient was then transferred to the ITU.

Four hours after admission the patient developed a productive cough, respiratory distress and peripheral cyanosis. Clinical examination confirmed the presence of pulmonary oedema. Endotracheal intubation and mechanical ventilation was instituted along with inotropic support using dobutamine and adrenaline infusions.

When the patient was stabilised, it was then noted that the swelling around the left eye appeared to have increased along with increased haemorrhages around the globe. An ophthalmic opinion was then sought.

During this time, gram staining confirmed the causative organism to be meningococcus. Subsequent cultures isolated the microbial pathogen to be Group B meningococcus.

When seen by the ophthalmic team, the patient was sedated and on ventilatory support. The left eye was surrounded by marked periorbital oedema and subconjunctival haemorrhage temporally (Figure 1). There was no conjunctival discharge.

The left eye was proptosed. With the Keeler exophthalmometer the left eye measured 24 mm and the right 19 mm respectively. There was no afferent pupillary defect. Resistance to retropulsion was noted on the left. The IOP by Perkins tonometry was left 28 mmHg and right 19 mmHg respectively. Fundoscopy showed normal disc and macula bilaterally. The vitreous was quiet and clear. There was no optic disc oedema and no choroidal folds. No retinal haemorrhages were seen.

A CT scan of the orbits demonstrated proptosis of the left eye. There was periorbital high intensity signal, which extended around the globe on the temporal side (Figure 2). The recti muscles were enlarged. This appearance was consistent with peribulbar and orbital haemorrhage.

Haematological investigations revealed leucocytosis and marked thrombocytopaenia: haemoglobin, 8.8 g/dl; white cell count, 27.58×10^9/l; platelets, 35×10^9/l. There were also abnormalities of the bleeding indices with prothrombin time (PT) 22 s (normal 9–15); and activated partial thromboplastin time (APTT) 99 s (normal 27–41). Fibrinogen levels were at the lower limit of normal 2.8 g/l (normal 2–40). There was no clinical evidence to suggest that there was the threat of optic nerve compression and the eye was managed conservatively with daily follow-up.[2,7] The IOP normalised 48 h later and the proptosis had almost completely disappeared by one week post admission.

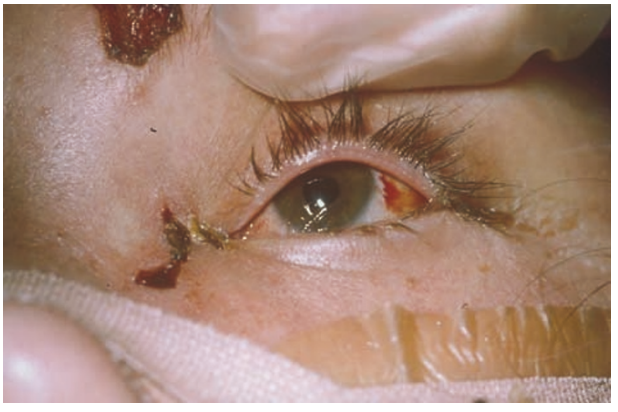

Figure 1 Photograph showing periorbital oedema and temporal subconjunctival haemorrhage.

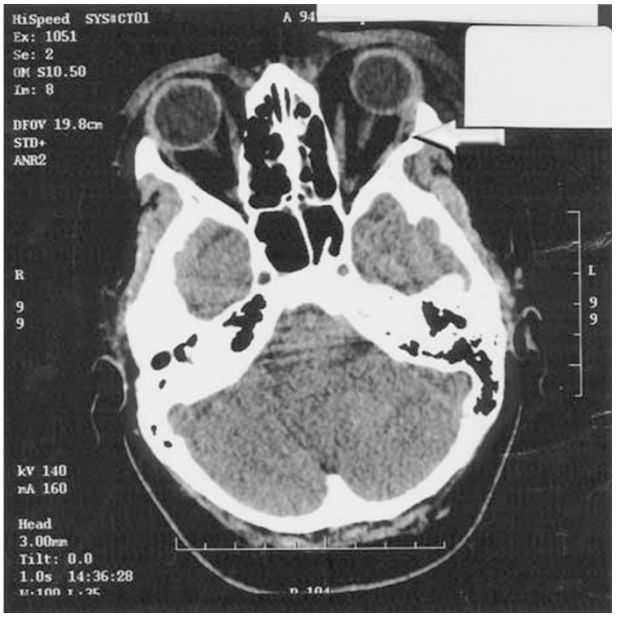

Figure 2 CT scan showing proptosis of the left eye. Arrow denotes area of orbital haemorrhage.

After 7 days of intravenous antibiotics and fresh frozen plasma, the platelet count and bleeding indices returned to normal. Platelets, 219×10^9/l; PT 14.5 s and APTT 44 s. Adequate progress was made and the patient was eventually taken off the ventilator and transferred out of the ITU.

Six weeks post admission, the left eye was quiet and there was no proptosis. The visual acuity in the left eye was 6/6.

Comment

Law described spontaneous orbital haemorrhage in 1971 as the occurrence of haemorrhage within the orbit not caused by local trauma and not referrable, as far as can be ascertained, to any constitutional causative condition.[1] It is well accepted that this is an uncommon event and published literature on this subject reflects the rarity of such cases.[8]

Spontaneous orbital haemorrhages characteristically present acutely with sudden onset of pain, periorbital haemorrhage and ecchymosis, proptosis, and in severe cases; diplopia and decreased vision.[8] Constitutional symptoms may be present and include nausea and vomiting.[9] When severe, orbital haemorrhage can cause optic nerve compression with reduced colour vision, reduced visual acuity, relative afferent pupillary defects and optic nerve swelling.[2] The causes for orbital haemorrhage may be classified into: (i) local and (ii) systemic.

Local causes include: orbital varix, haemangioma,

lymphangioma, ruptured intraorbital aneurysm, orbital myositis, Valsalva manoeuvre, carotico-cavernous fistula and idiopathic inflammatory pseudotumour.[1,2,9–12]

Systemic causes usually result from bleeding disorders and include: haemophilia, von Willebrand's disease, heparin treatment, thrombolytic treatment and liver disease.[13–16] Other systemic causes described are anaemia, leukaemia, sickle cell disease, and hypertension.[2,3]

Meningococcal infection is caused by the organism, *Neisseria meningitidis*, a gram negative diplococcus.[5,6] The clinical features of meningococcal infection are variable and range from asymptomatic carriage to multi organ failure and death.[5]

Ophthalmic complications of meningococcal infection include conjunctivitis, conjunctival petechiae and ecchymosis, cellulitis and endophthalmitis.[6,17–19] As far as we are aware, periorbital and retrobulbar haemorrhage have not been previously described as complications of meningococcaemia.

Meningococcal bacteraemia is associated with endotoxin release, which causes a procoagulant haemostatic environment due to reduction of anticoagulant factors (protein C, protein S, antithrombin III and tissue factor pathway inhibitor).[4–6] Up-regulation and increased expression of procoagulant factors (tissue factor, platelet activator inhibitor and inhibition of fibrinolysis) adds to the imbalance. This results in widespread DIC.

DIC then causes rapid depletion of existing coagulant factors with a subsequent bleeding tendency, the so called 'consumption coagulopathy'. Marked thrombocytopaenia also occurs in this type of infection and as a consequence prothrombin and partial thromboplastin times are increased.

Reduced coagulation factors and thrombocytopaenia are responsible for the characteristic clinical features of purpura fulminans.[4–6]

Purpura fulminans describes the presence of severe purpura plus ecchymoses, with vascular thrombosis and gangrene.[5] Purpura fulminans is not unique to meningococcaemia and is seen in three main clinical settings: (1) in neonates as a result of protein C or protein S deficiency; (2) acute infectious causes, which in the presence of septic shock is nearly always caused by meningococcal infection; (3) rarely, post infectious syndromes from viruses or bacteria.[5]

Our patient had characteristic manifestations of the bleeding diathesis of meningococcal septicaemia and this was confirmed by abnormalities of the relevant haematological indices (platelets, PT, APTT). The likely mechanism for the orbital haemorrhage would be spontaneous rupture of a subconjunctival petechial haemorrhage, with retrobulbar extension as a result of thrombocytopaenia and prolonged bleeding time, secondary to DIC that occurred in the presence of meningococcal infection.

Cases of orbital haemorrhage in young patients carry a good visual prognosis, but in elderly patients visual loss may occur in up to 50%.[2] Acute, sight-threatening orbital haemorrhage should be decompressed by lateral canthotomy and cantholysis.[20] Non sight threatening cases can be safely managed conservatively with good long term visual outcome.[2,7,16]

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Sir,

Premacular gliosis in a patient with tuberose sclerosis
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Ocular involvement occurs in approximately half of those affected by tuberose sclerosis: variably elevated astrocytomas being the most common association. Pseudocolobomas, depigmented fundus lesions and papilloedema/optic atrophy associated with intracranial involvement also occur.[1,2] We report an additional ocular feature.

Case report

A 32-year-old woman with known tuberose sclerosis (TS) (displaying the adenoma sebaceum, depigmented skin macules, periungual fibromas and epilepsy typical of this condition) was referred for ophthalmic review with an ill-defined period of bilateral visual blurring without distortion or other visual symptoms. Her unaided bilateral 6/12 acuities improved to 6/6 bilaterally with a low myopic lens correction. Both fundi showed features consistent with TS: a slightly elevated, whitish astrocytic hamartoma nasal to the disc in the right eye, and bilateral small midperipheral flat translucent lesions probably representing smaller astrocytic hamartomas. Of interest was the striking right premacular epiretinal membrane in the absence of a posterior vitreous detachment (Figure 1). This was separate from the other retinal lesions.

Comment

'Idiopathic' premacular gliosis is rare in patients younger than 50 years. Secondary premacular gliosis in children and young adults may occur with retinal vascular disease, previous intraocular trauma and surgery, Coat's disease and ocular inflammatory disease.[3] There is a recognised association between premacular gliosis and combined hamartoma of the retina and pigment epithelium—a distinct developmental lesion not associated with TS.[2,4] With the demonstrable typical TS-associated lesions in this patient, the premacular gliosis is unlikely to be coincidental, particularly in view of her young age.

In premacular gliosis, retinal pigment epithelium and glial cells proliferate along the surface of the inner limiting membrane. Disruption of the inner retinal surface is believed to be a precipitating event. In the more common idiopathic premacular gliosis seen in

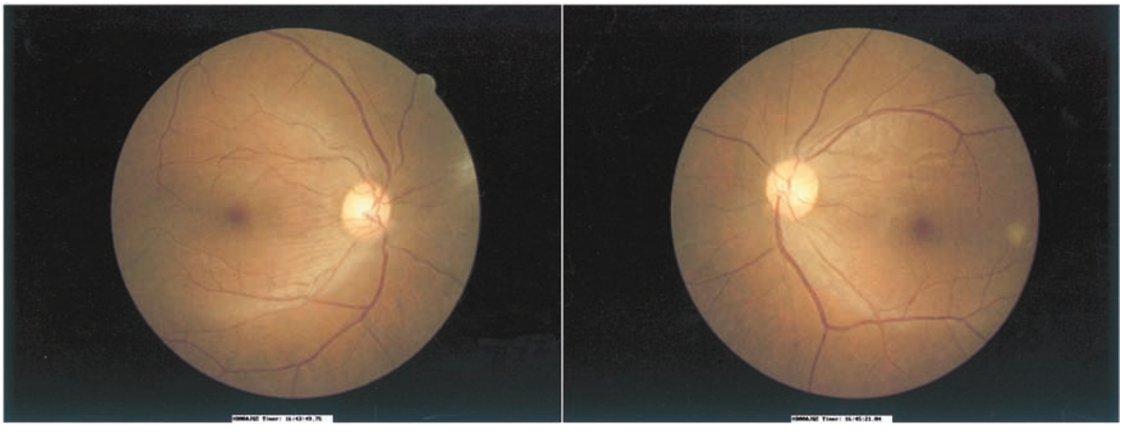

Figure 1 Fundal appearances of both eyes showing the marked right premacular epiretinal membrane and flat astrocytic hamartomas in the right nasal and left temporal areas. Similar small hamartomas are present more peripherally in both eyes as well.