Branch retinal artery occlusion: a case complicating acute rheumatic fever and dental abscess

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Obstruction of retinal blood flow may cause transient retinal ischaemia or amaurosis fugax, or if obstruction is prolonged, retinal infarction with a corresponding visual defect. In contrast to older patients where retinal occlusion is commonly associated with hypertension or carotid artery disease, retinal artery occlusion in the young should trigger an aggressive search for systemic disease including cardiac sources of emboli (Greven et al, 1995).

Acute rheumatic fever may cause embolic phenomena and although it is an uncommon diagnosis in this country, accurate recognition is essential to reduce long-term morbidity. We describe a case of branch retinal artery occlusion complicating acute rheumatic fever and dental sepsis and briefly discuss the investigation and management.

CASE REPORT

A 15-year-old boy presented with a 2-month history of lethargy, weight loss and migratory arthralgia following an episode of pharyngitis while on holiday in the Mediterranean. Examination revealed the murmurs of mitral and aortic regurgitation which were confirmed on echocardiogram. The serum C-reactive protein (CRP) was high at 124 mg/litre and the streptococcal antibody titre markedly elevated at >1440 units. A diagnosis of acute rheumatic fever was made and penicillin commenced in addition to aspirin for joint pain. Three months after diagnosis he was asymptomatic, the CRP was normal and aspirin was discontinued.

Six weeks later he presented with sudden onset of blurred vision in the right eye with a visual field defect in the nasal upper quadrant. There was no history of migraine and fundoscopy revealed an embolus at the disc with severe oedema below the macula consistent with occlusion of the inferotemporal branch of the retinal artery (*Figure 1*). Warfarin was commenced pending further investigation. Subsequent transoesophageal echo showed mild mitral regurgitation and minimal aortic regurgitation with no vegetations or thrombus. There was no evidence of arrhythmia and brain magnetic resonance imaging was normal. His protein C activity was transiently low at 64% (normal range 70–130%) and his CRP rose to 36 mg/litre. Aspirin was therefore substituted for warfarin. Three days later he presented with two dental abscesses requiring extraction. Fourteen months after presentation the superior altitudinal scotoma has improved and the retinal oedema resolved. Mild mitral valve regurgitation persists but aortic valve function appears normal. He continues on low dose aspirin and penicillin.

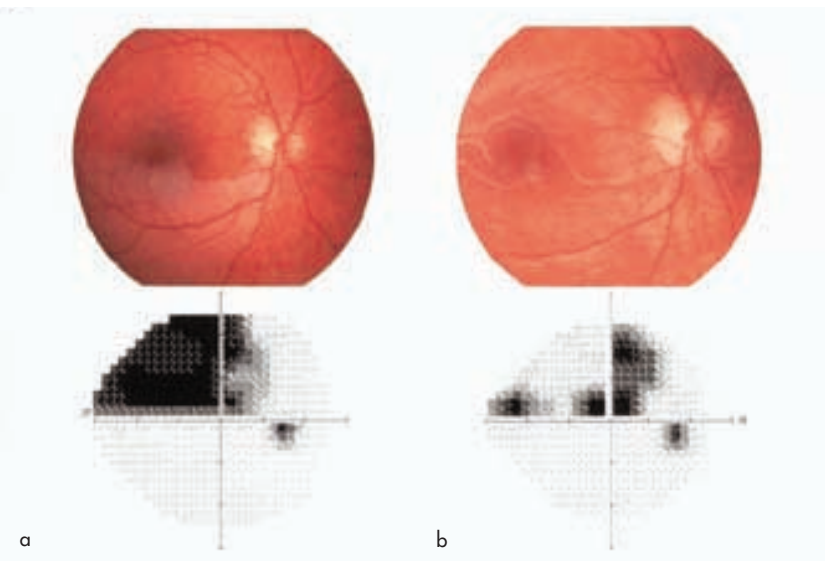

Figure 1. Retinal photograph showing segmental retinal pallor representing oedema and corresponding visual field defect (a) at diagnosis and (b) 14 months later when retina appears normal but scotoma persists.

DISCUSSION

Arteriolar occlusion may occur either in the central retinal artery or one of its branches. It is rare in children and young adults, with only 8% of cases occurring in the 9–29-year-old age group (Brown et al, 1981). In this group it tends to occur in the branch retinal arteries and the aetiology is diverse and includes vessel compression, embolism, thrombosis or reduction in blood flow because of intimal thickening or vasospasm. Congenital anomalies such as preretinal arterial loops predispose to retinal ischaemia by kinking and compression of the loops that extend into the vitreous cavity (Reichel et al, 1994).

Emboli causing arteriolar occlusion may arise from either cardiac or cerebrovascular sources and although cardiac valvular disease accounted for 56% cases of branch artery occlusion in one study, the minority were related to rheumatic valve disease (Wilson et al, 1979). Hypercoagulable states

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including protein S deficiency have been reported as a cause of branch artery occlusion, as have haemoglobinopathies and vasculitic conditions such as polyarteritis nodosa and systemic lupus. Branch retinal occlusion may also complicate head and neck surgery following a metastatic endophthalmitis and has occurred secondary to dental sepsis (Kilmartin and Barry, 1996). A high incidence of migraine has been noted in young adults with retinal artery occlusion and idiopathic recurrent retinal arterial occlusion as part of a microangiopathic syndrome has also been recognized (Johnson et al, 1994). In the above case the likely mechanism was a hypercoagulable state caused by sepsis and recent withdrawal of aspirin.

CONCLUSIONS

Any child or young adult with retinal artery occlusion warrants a vigorous search for an embolic source or thrombophilic state and investigation should include echocardiogram, haemoglobin electrophoresis, a full coagulation/ thrombotic screen and measurement of C-reactive protein or erythrocyte sedimentation rate. Identification of any underlying cause is essential if further episodes of occlusion are to be prevented. **HM**

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Greven C, Slusher M, Weaver R (1995) Retinal arterial occlusions in young adults. *Am J Opthalmol* **120:** 776–83

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Kilmartin D, Barry P (1996) Recurrent septic retinal emboli following dental surgery. *Br J Opthalmology* **80**(12): 1111–2

Reichel E, Duker J, Puliafito C, Hedges III, Caplan L (1994) Branch retinal arterial occlusion caused by a preretinal arterial loop. *Neurology* **44:** 1181–2

Wilson L, Warlow C, Ross Russell R (1979) Cardiovascular disease in patients with retinal arterial occlusion. *Lancet* **i:** 292–4

IN THE PUBLIC'S VIEW...

NICE work?

It was bound to happen. Taxanes had been used so often as prima facie evidence of the unfairness of the NHS, some women with cancer receiving the drugs and others just down the road being denied them, that it was inevitable NICE's verdict would be leaked. And so it was. In a blaze of publicity, the BBC jumped the gun on 11 April. Who did the leak come from? It certainly did not come from Andrew Dillon, Chief Executive of NICE. He released an urgent cascade memorandum to NHS staff the following day, disowning the information. On their news website (http://www.nice.org.uk/updates/upd_ind.htm), NICE declared that it was 'aware of the speculation about its appraisal of taxanes for breast and ovarian cancer', adding that it is not their policy 'to comment on an appraisal until the process is complete and we have issued our guidance to the NHS and the public'.

Now that we're beginning to realize that the NHS cannot provide all things for all people, it's about time that some realism appeared in news reports on health matters. The BBC triumphantly regarded the acceptance of taxanes as only right and proper, which it probably is, but the public need to be told some facts. Prescribing taxanes for women with breast cancer will increase the average survival of those women but, contrary to impressions given by the BBC, it will not guarantee survival for all women. There were interviews with a few women for whom it was implied or even stated explicitly that taxanes would enable them to live until their grandchild's birthday or similar life event. For the interviewed women, that cannot be known. They may survive, disease-free, even if not given taxanes. They may succumb, having had a rotten time with side-effects, despite being given taxanes. They may even die earlier because of those side-effects, but the BBC did not mention side-effects at all.

The BBC also managed to paint the health authorities as the villains of the piece: mean old bodies who won't give women a chance. But not once did the BBC ask where the money for these expensive drugs was going to come from. Or, more correctly, from which groups of patients with what other less fashionable and less telegenic diseases the money was going to be taken. It will help if central funding is made specifically available when NICE recommends expensive drugs (which is what a committee at the Royal College of Physicians has warned will be needed), but that is fudging the issue. Unless the provision of money and control of treatments is to be entirely from the centre, there will still be rationing by postcode. The government are effusive about wanting local health providers to be responsive to local issues, but highly critical of local variation. They can't have it both ways.

The BBC ended its news report on taxanes by interviewing a man with multiple sclerosis. No doubt they will interview him again — ecstatic about his chance of remission or despondent about lost chances — when NICE's verdict on β-interferon is leaked sometime next year. Compared with β-interferon, taxanes are cheap. **HM**

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