Sir,

Wegener's granulomatosis presenting with amaurosis fugax

Wegener's granulomatosis (WG) is a rare disorder of unknown aetiology, that is characterised by a necrotising vasculitis and granulomatous infiltration of the respiratory and renal systems.[1] Ocular manifestations are reported to occur in up to 77% of patients[2] and include conjunctivitis, episcleritis, peripheral ulcerative keratitis, scleritis and uveitis.[2] Choroidal and retinal involvement is rare, with only four previous cases of central retinal artery occlusion being reported.[3] We present the case of a 55-year-old male who presented with amaurosis fugax and was subsequently diagnosed as suffering from WG. To our knowledge this is the first reported case of WG presenting in this manner.

Case report

A 55-year-old male presented to the Ophthalmology department complaining of two episodes of visual loss affecting his left eye over the previous 3 days. Each episode lasted several minutes, after which he described his vision rapidly returning 'like a blind going up'. He also had a 2-week history of generalised myalgia, arthralgia and a dull frontal headache. In addition his appetite had been poor and he had experienced night sweats in the days preceding his visual symptoms. He had no significant past ocular or medical history of note.

On examination his visual acuities were 6/6 bilaterally and his ocular examination was normal. The temporal arteries were pulsatile and non-tender. He had a pyrexia of 38.5°C, with a sinus tachycardia of 114 beats/min and blood pressure 150/80 mmHg. His erythrocyte sedimentation rate (ESR) was 134 mm in the first hour and a C-reactive protein (CRP) was 274 mg/l. His full blood count showed a leucocytosis (15.5×10^9/l) and a thrombocytosis (751×10^9/l). His blood glucose was normal (4.5 mmol/l) as was his renal function. He was initially treated with intravenous methylprednisolone on consecutive days, which resulted in a dramatic improvement in his symptoms. He then underwent a left temporal artery biopsy, which proved to be normal. Echocardiography and carotid dopplers were also normal. By this stage the results of his c anti-neutrophil cytoplasmic antibody (cANCA) test were available and found to be positive to a 1:160 dilution. His serum was also found to be strongly positive for anti-proteinase 3 (anti-PR3) antibodies. These blood results were suggestive of a diagnosis of WG. In view of this, a chest CT was performed and revealed nodular pleural thickening in the right lung and multiple nodules bilaterally approximately 1–2 cm in size in the mid and upper zones, some of which showed signs of cavitation. These appearances were consistent with a diagnosis of WG.

The patient was then commenced on a reducing course of oral prednisolone and monthly intravenous cyclophosphamide. Within 4 weeks of treatment his ESR had fallen to 15 mm in the first hour, CRP to <10 mg/l and platelet count to 415×10^9/l. His systemic symptoms have resolved and on regular follow-up he has experienced no further episodes of visual loss.

Comment

The ocular manifestations of WG are seen in 40–77% of patients[2] and are thought to be the result of a necrotising obliterative vasculitis. In addition, thrombocytosis is a recognised finding in WG,[4] and in this particular case may well have contributed to the occlusive pathology causing amaurosis fugax. In summary, this case demonstrates that in patients presenting with amaurosis fugax, WG should be considered in the list of potential differential diagnoses.

References

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P Cackett and C Weir

Tennent Institute of Ophthalmology
Gartnavel General Hospital
1053 Great Western Road
Glasgow G12 OYN, UK

Correspondence: P Cackett
Tel: 0141 423 8958
E-mail: pete@pdcackett.demon.co.uk

Eye (2002) **16,** 676. doi:10.1038/sj.eye.670107