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LETTERS TO THE JOURNAL

Sir,

Giant cell arteritis with a normal ESR and CRP

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Giant cell arteritis is diagnosed when clinical suspicion from characteristic clinical symptoms and signs is supported by simple blood tests, including a raised erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP), and confirmed by a positive temporal artery biopsy. In occult giant cell arteritis, where there is ocular involvement by giant cell arteritis without any systemic symptoms and signs of giant cell arteritis,[1] the diagnosis is more difficult, and the above investigations will help direct the ophthalmologist before a temporal artery biopsy is carried out. However, it is known that a normal ESR does not preclude the diagnosis of giant cell arteritis.[2] A raised CRP may be a more sensitive indicator of the condition.[3] We report a unique case of occult giant cell arteritis with both a normal ESR and a normal CRP.

Case report

A 74-year-old man presented to his optician with a 3-week history of diplopia on right gaze. On direct questioning he also complained of a 2-week history of episodes of monocular visual loss in the right eye lasting 15 min, occurring at least twice a day. There were no precipitating factors and no associated symptoms except minor discomfort behind the right eye. He had a coronary artery bypass graft 11 years previously; his general health was good. He had been told as a young man that he was colour blind. He did not complain of headache, scalp tenderness, jaw claudication, weight loss or arthritis.

On examination his corrected Snellen visual acuity was 6/9 in each eye. Right eye abduction was reduced to 60% with slow abducting saccades. Only the test plate of the Ishihara colour plates could be seen with either eye, in keeping with his previous diagnosis of colour blindness. There was a right relative afferent pupil defect. There was mild constriction of the Goldman I4e isopter on the right visual field to 30°. Intraocular pressures were 7 mmHg OS and 5 mmHg OD. On fundoscopy cotton wool spots were noted on the right (Figure 1a) but his optic discs appeared healthy.

Initial investigations revealed a haemoglobin of 12.4 g/dl, a white cell count of 8.3×10^9/l, and platelets of 279×10^9/l. Renal and liver function tests were normal, as were serum triglycerides, cholesterol and angiotensin converting enzyme. The ESR was 14 mm/h and the CRP was less than 7 mg/l. Fibrinogen was slightly raised at 4.72 g/l (reference range 2.02–4.24). Antinuclear antibody, antineutrophil cytoplasmic antibody and thyroid autoantibodies were negative. Orbital computed tomography, chest X-ray and carotid Doppler ultrasound were all normal. Fundus fluorescein angiography showed severe ischaemic changes with delayed retinal filling at 18 s and delayed choroidal perfusion (Figure 1b), complete choroidal filling not occurring until 40 s. The patient underwent a right temporal artery biopsy, which confirmed a diagnosis of giant cell arteritis (Figure 2).

Comment

Occult giant cell arteritis has been variously said to occur in 8–38% of cases of giant cell arteritis.[4] Some confusion has arisen because not all studies have included only biopsy-positive cases. In Hayreh's study of 85 cases of biopsy-confirmed giant cell arteritis with ocular involvement, 18 (21.2%) had occult giant cell arteritis.[1]

A raised ESR has been regarded as the gold standard of useful investigations in giant cell arteritis,[4] but an ESR of less than 40 mm/h has been described in 8–22.5% of patients with giant cell arteritis.[4] A raised serum fibrinogen may also be helpful in making the diagnosis,[4] and in our patient the serum fibrinogen was slightly raised. CRP levels may be more sensitive in making the diagnosis: in Hayreh's series of 363 patients who had temporal artery biopsy for suspected giant cell arteritis, 223 patients had their CRP values estimated. CRP was 100% sensitive for the detection of giant cell arteritis (ESR was 92% sensitive).[3] In patients with occult giant cell arteritis, both ESR and CRP levels may be significantly lower than in patients with the typical systemic features of giant cell arteritis.[1] Whether these patients with occult disease will develop a raised ESR and CRP in time is uncertain. However, 17 of Hayreh's 18 cases of occult giant cell arteritis presented with anterior ischaemic optic neuropathy, and the other with central retinal artery occlusion,[1] indicating that the disease process affecting the eye can be severe even when the ESR and CRP are only slightly raised. In this case, despite none of the usual systemic symptoms associated with giant cell arteritis, the patient had extremely extensive involvement of the ophthalmic artery circulation as evidenced by his ophthalmoplegia as well as his relative afferent pupillary defect, constricted visual field, low intraocular pressure and ischaemic retinopathy.

Our case, in keeping with many cases of occult giant cell arteritis, posed diagnostic difficulties. There had been no history of permanent visual loss, and on examination there was no anterior ischaemic optic neuropathy, the most common ocular manifestations in giant cell arteritis.[5] The combination of ophthalmoplegia, low intraocular pressure and cotton wool spots made vasculitis likely, which prompted fluorescein angiography. The extremely delayed perfusion time of

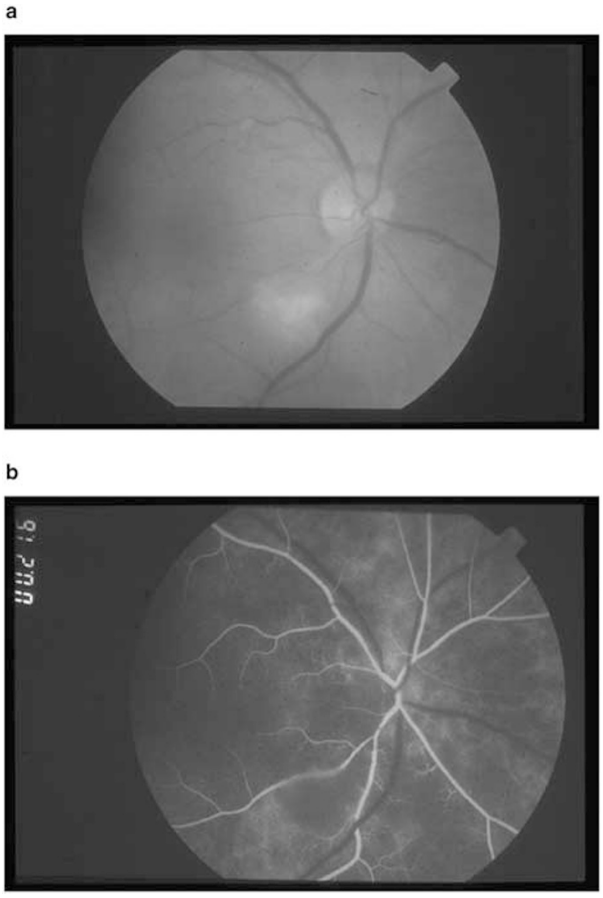

Figure 1 (a) Cotton wool spots of the right fundus. (b) Right fundus fluorescein angiogram at 22 s. The background choroidal fluorescence is still very patchy, indicating marked hypoperfusion.

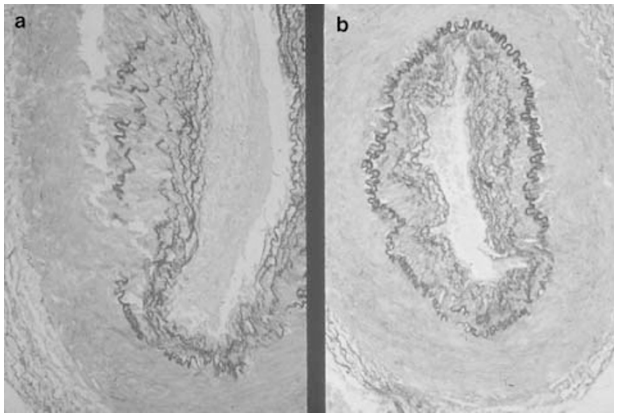

Figure 2 (a) Patient's temporal artery showing disruption of the elastic lamina and of the muscle by giant cell arteritis. (b) Control patient temporal artery biopsy showing arteriosclerosis only. Stain—Elastic van Gieson. Magnification $\times 100$.

both the retina and choroid found on angiography added weight to a diagnosis of vasculitis, particularly giant cell arteritis,[6] and led to a temporal artery biopsy. Clinical suspicion was confirmed with a positive biopsy, despite both normal ESR and CRP.

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Sir,

The role of an ophthalmologist in suspected non-accidental injury

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The diagnosis of paediatric nonaccidental injury (NAI) can have major implications for the future of a child and