Bilateral central retinal artery occlusion in a patient with Churg-Strauss vasculitis

Sirs,

We present the case of a 56-year-old man who presented with acute painless visual loss in his right eye and then left eye 2 days later. A few months before this admission he had had fever, significant weight loss, malaise, headache, transient diplopia, myalgia and parasthesia of legs. He was diagnosed to have had asthma for 1 year and his past medical history was otherwise unremarkable. Ophthalmology examination showed right relative afferent papillary response. As far as visual acuity, he had no light perception in the right eye while in the left eye there was perception of light. Fundoscopic examination showed retinal whitening with a "cherry-red spot" in the fovea in both eyes which was diagnostic of bilateral central retinal artery occlusion. No evidence of retinal vasculitis and no visible emboli were found. The other cranial nerves were intact. He had wasting of muscles in the upper and lower extremities. He had weakness of his distal limb muscles and generalised hyporeflexia. Electrocardiogram showed sinus rhythm. Laboratory investigations on presentation showed a haemoglobin level of 12.1g/dl, a white cell count of 20× 10^9/l with eosinophilia of 6×10^9/l (30.5%), a platelet count of 957×10^9/l, an erythrocyte sedimentation rate of 78mm at 1h, and a C-reactive protein of 161mg/l (normal < 5mg/l). Serum albumin was 26Lg/L (normal 35–50). The p-antineutrophil cytoplasmic antibody (ANCA) was positive. The antimyeloperoxidase antibodies (MPO-ANCA) was 26RU/ml (normal <20). The rheumatoid factor was more than 33IU/ml (normal less than 7IU/ml). Blood coagulation tests were within normal limits. Tests for antinuclear antibodies, lupus anticoagulant, anticardiolipin antibodies, and circulating immune complexes were negative. The CT brain showed sinusitis of ethmoidal sinuses. The MRI brain was normal and MRA did not demonstrate any significant lesions of craniocervical vessels. Nerve conduction studies revealed axonal damage involving multiple individual nerves in an asymmetric fashion. The CT abdomen showed multiple renal and splenic infarcts. Transthoracic and transesophageal echocardiography revealed no cardiac source of emboli. The bone marrow examination showed no haematological malignancy. A temporal artery biopsy showed necrotising vasculitis with esosinophilic infiltrates, consistent with CSS (Fig. 1). Sural nerve biopsy was taken 1 month after initiation of the immunosuppressive therapy. It showed severe demyelination and severe axonal loss due to arterial occlusion. No vasculitis was seen. Our patient fulfilled the American College of Rheumatology criteria for CSS diagnosis (1).

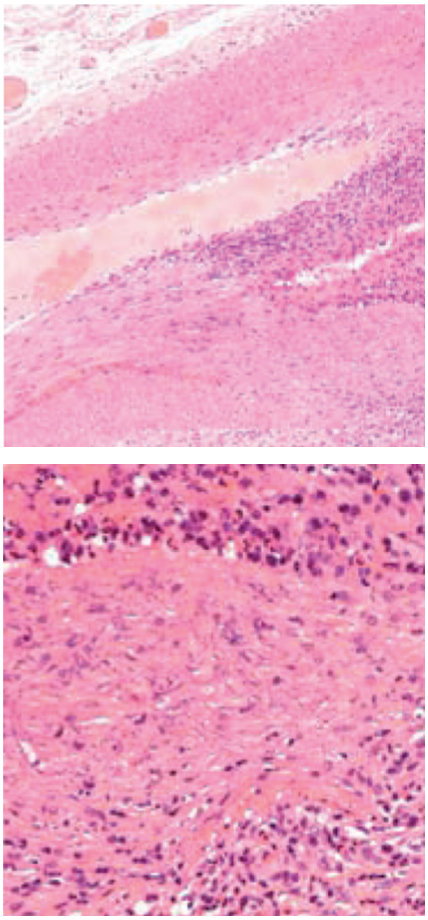

Fig. 1. The temporal artery biopsy showed necrotising vasculitis with erosinophilic infiltrates.

He was given intravenous methylprednisolone 1 gram daily for 3 days immediately after the fundoscopic findings, followed by high dose oral prednisolone (1g/kg/day). Anterior chamber paracentesis was performed immediately. Oral cyclophosphamide (50mg/day) was given after pulse steroid. His visual acuity improved slightly after the treatment. Visual acuity of his both eyes was able to see hand motion. There was a resolution of his constitutional symptoms and the eosinophil count was normalised.

Central retinal artery occlusion is rare in patients with CSS and there are only 5 reported cases (2-6). The mean age of onset is 53 (ranges from 48 to 68). Male and female are equally affected. The combination of vasculitis and hypercoaguable states may be a possible cause. The visual recovery is usually poor, ranging from no light perception to dense central scotoma, despite aggressive treatment. Ophthalmological involvement is uncommon in CSS. Other manifestations include conjunctival granuloma (7), anterior uveitis (8), orbital inflammation (9), retinal vein occlusion (10).

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