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 significant weight loss, malaise, headache, transient diplopia, myalgia and paraesthesia 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 12.1 g/dl, a white cell count of 20 × 10^9/l with eosinophilia of 60 × 10^9/l (30.5%), a platelet count of 957 × 10^9/l, an erythrocyte sedimentation rate of 78 mm at 1 h, and a C-reactive protein of 161 mg/l (normal < 5 mg/l). Serum albumin was 26.4 g/l (normal 35–50). The p-antineutrophil cytoplasmic antibody (ANCA) was positive. The antيمyeloperoxidase antibodies (MPO-ANCA) was 26 RU/ml (normal < 20). The rheumatoid factor was more than 33 IU/ml (normal less than 7 IU/ml). A temporal artery biopsy showed necrotising vasculitis with erosinophilic infiltrates. 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 12.1 g/dl, a white cell count of 20 × 10^9/l with eosinophilia of 60 × 10^9/l (30.5%), a platelet count of 957 × 10^9/l, an erythrocyte sedimentation rate of 78 mm at 1 h, and a C-reactive protein of 161 mg/l (normal < 5 mg/l). Serum albumin was 26.4 g/l (normal 35–50). The p-antineutrophil cytoplasmic antibody (ANCA) was positive. The antимyeloperoxidase antibodies (MPO-ANCA) was 26 RU/ml (normal < 20). The rheumatoid factor was more than 33 IU/ml (normal less than 7 IU/ml). Blood coagulation tests were within normal limits. Tests for antinuclear antibodies, lupus anticoagulant, antiphospholipid 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 craniofacial 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 eosinophilic 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). He was given intravenous methylprednisolone 1 gram daily for 3 days immediately after the fundoscopic findings, followed by high dose oral prednisolone (1 mg/kg/day). Anterior chamber paracentesis was performed immediately. Oral cyclophosphamide (50 mg/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 hypercoagulable 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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References