Optic neuritis as an initial presentation of Wegener’s granulomatosis

A 59-year-old male presented to our ophthalmologic emergency room with a 2-day history of blurry vision in the left eye accompanied by generalized scalp tenderness. The patient had had redness and watery discharge for the past 2 months in both eyes, as well as generalized painful joints and muscles for the past 7 months, with a clinical diagnosis of polymyalgia rheumatica. He had lost 15 pounds in this timeframe. The patient did not have any bothersome urinary or respiratory symptoms.

On examination, corrected visual acuity was 6/12 OD and finger counting OS. Confrontational visual field testing showed diffuse field constriction OS. A relative afferent pupillary defect was present OS. Fundus examination was normal OU. The patient had no other cranial nerve impairment.

Erythrocyte sedimentation rate and C-reactive protein were elevated, at 60 mm/h and 96 mg/L, respectively. The patient had general pain in the temporal region. A CT scan of the chest and abdomen then revealed multiple ill-defined pulmonary nodules in all lobes, bilateral renal cysts, and a cyst in the sixth hepatic segment. Bronchial aspirate and renal biopsy were obtained and were compatible with a diagnosis of Wegener’s granulomatosis (WG).

The patient was treated with intravenous methylprednisolone (250 mg) 4 times a day for 3 days, followed by oral methylprednisolone (80 mg) and oral acetylsalicylic acid (81 mg) daily for 7 days. One week after treatment initiation, corrected visual acuity had dramatically improved to 6/7.5 OD and 6/21 OS.

The patient continued treatment with oral methylprednisolone (50 mg) daily.

Our patient probably suffered from optic neuritis secondary to an inflammatory process, which is supported by the quick response to oral methylprednisolone therapy and the absence of compressive signs on imagery. Only 3 other authors1–3 in the English literature have reported visual loss due to optic neuritis as a feature of WG without compressive lesions on imagery. All patients recovered visual acuity following high-dose oral corticosteroid therapy. In all other cases there was upper airway involvement at initial presentation, rendering our patient the first case of isolated optic neuritis as the initial manifestation of WG.

The only ophthalmic sign in our patient was visual loss from optic nerve involvement. Initial ophthalmic involvement occurs in 17% of patients,4 and most frequently manifests as scleritis, conjunctivitis, and eye pain. Peripheral and central neuropathies are present in fewer than 2% of patients at initial presentation,4 and often manifest as mononeuropathy multiplex or cranial neuropathies of the optic, abducens, and facial nerves.5 In WG, initial manifestations include upper airway symptoms in 73% of patients and arthralgias in 32% of patients.6 Other authors2 have raised the possibility that some cases reported as ischemic optic neuropathies in WG might have actually been undertreated optic nerve inflammation complicated by vasculitic infarction.

The optimal treatment for acute-phase WG is a short course of high-dose corticosteroids and cyclophosphamide, which is continued for 9 to 12 months following remission.7 Patients with non-major organ disease may be effectively treated without cytotoxic therapy, with close monitoring for lung and kidney disease.8

REFERENCES


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doi:10.3129/i09-145