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LETTERS TO THE JOURNAL

Sir,

Optic neuritis as a feature of Wegener's granulomatosis

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In a patient with optic neuritis, sinusitis, orbital pathology and a history of renal failure, Wegener's granulomatosis (WG) has to be included in the differential diagnosis. However, without a high index of suspicion the clinical diagnosis of WG may not be entertained unless the clinician is familiar with isolated and uncommon manifestations of the disease. This may result in delayed or inadequate treatment.

Case report

A 39-year-old Caucasian man, with a history of renal transplant due to Goodpasture's syndrome, was admitted to the ENT ward with sinusitis. The diagnosis of the Goodpasture's syndrome (12 years previously) was based on the clinical findings, the renal biopsy which showed focal glomerulonephritis, and the results of the immunofluorescence test showing the presence of anti-glomerular basement membrane antibodies. Intravenous cefuroxime and flucloxacillin were administered. Ophthalmological opinion was sought for a non-tender soft tissue swelling at the superonasal aspect of the right orbit. Initial examination revealed visual acuities of 6/6 bilaterally, equal and reactive pupils, normal fundi and no signs of orbital pathology. The CT scan had shown extensive sinusitis of ethmoidal and sphenoidal sinuses and abnormal soft tissue swelling anterior to the right globe without any bone destruction (Figure 1). Biopsy of the orbital space-occupying lesion was arranged.

The patient developed visual loss in the left eye 6 days later and was sent back to the eye department for re-evaluation. On examination, visual acuity had dropped to hand movements in the left eye. Relative afferent pupillary defect in full was present; however, both discs appeared normal. Visual field testing showed extensive field loss in the left eye of no typical pattern. Repeat CT scan did not reveal any new abnormalities. Aspiration biopsy of the lesion in the right orbit was performed; FBC, ANCA and ESR were ordered. The results were pending.

A working diagnosis of retrobulbar optic neuritis due to sinusitis (not responding to antibiotic treatment) was made. Intravenous methylprednisolone 1 g was recommended, but due to the patient's prophylactic immunosuppression with cyclosporin 300 mg daily and prednisolone 5 mg, physicians advised oral prednisolone 60 mg daily.

The results of the biopsy taken from the mass and the nasal mucosa showed necrotising vasculitis and thrombosis of small vessels, the inflammation consisting of lymphocytes, histocytes and Langerhans cells. ANCA screening was positive with c-ANCA over 72.3 Elisa units (normal value up to 3) and p-ANCA negative. WBC were 12×10^9/l. ESR was 84 mm/h. Chest radiograph was normal. The above were consistent with a diagnosis of WG. One week after steroid treatment was initiated, the visual acuity returned to normal level.

Comment

Wegener's granulomatosis (first described in the 1930s) is a rare disease, related in some way to a hypersensitivity reaction to an as yet unidentified allergen or autoimmunity.[1] It is characterised by:

(1) necrotising granulomatous vasculitis of the upper and lower respiratory tracts;
(2) focal necrotising glomerulonephritis;
(3) systemic small vessel vasculitis involving virtually any organ in the body, including the eye and orbit.

The mean age at onset is 40 years with a male to female ratio of 3:2. C-ANCA is positive in almost 90% of active generalised WG but is not entirely specific.[2] Remissions and exacerbations of WG may be monitored by changes in these serum titres during the course of the disease.[2] P-ANCA is positive in a few cases of WG, but is more often present in other

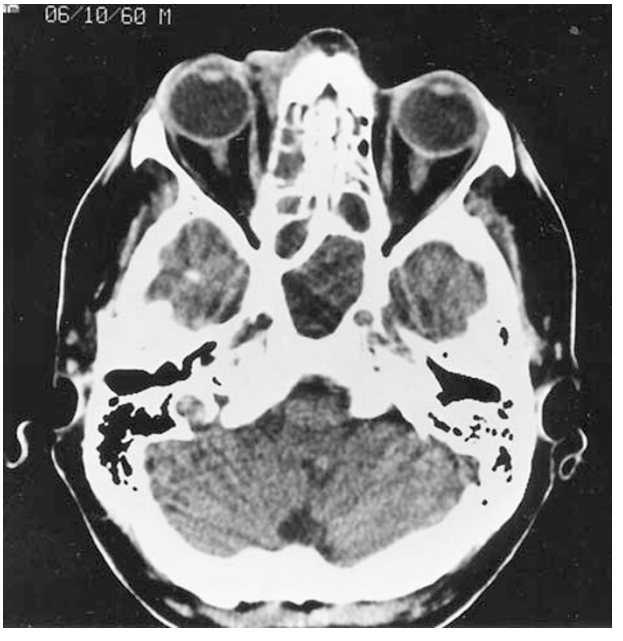

Figure 1 CT scan of the brain and orbits showing extensive sinusitis and soft tissue swelling anterior to the right globe.

vasculitides. Upper respiratory tract and eye involvement occur in 75% and 14% of cases respectively, according to one study.[3] Ocular manifestations include conjunctivitis, episcleritis, scleritis and uveitis. Corneal ulceration has also been reported. The orbit may be involved by granuloma formation causing pseudotumour and proptosis or vasculitis causing internal and external ophthalmoplegia. Optic nerve may be affected by occlusive vasculitis which may lead to optic nerve ischaemia, compressive granulomatous lesion or direct spread of inflammation from the sinuses. Treatment of WG includes corticosteroids and a cytotoxic agent. Cyclophosphamide is the drug of choice.

Although in this case the neuritis showed excellent therapeutic response to oral prednisolone, it must be emphasised that close liaison with physicians is required as these patients need pulsed cyclophosphamide to halt disease activity.

It is important to mention that although the initial results of immunofluorescence testing supported the diagnosis of Goodpasture's syndrome, it is likely that the patient had WG all along, especially since the ANCA test was not available at the time to exclude the diagnosis of WG. There are cases reported in which patients were initially diagnosed as having Goodpasture's syndrome on the basis of renal failure but later proven to have WG.[4,5] However, it should be borne in mind that some overlap does exist between WG and Goodpasture's syndrome.[6]

Thus, although it is known that optic nerve involvement may occur with contiguous granulomatous sinus disease in WG,[7] ophthalmologists must be particularly aware when a patient who has previously had serious renal disease presents with sinusitis and orbital pathology. In this scenario, WG must be excluded.

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Sir,

Visual field defects in adults secondary to pre-term delivery

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It is well recognised that a prematurely born child can present with problems in adult life.[1] Here we present two cases with repeatable visual field defects noticed on routine visual field assessment at the optician. No immediate cause was apparent, but extensive history-taking revealed premature birth in both cases.

Case reports

Case 1 A 35-year-old woman was noted to have a repeatable visual field defect on routine visual field testing at the optician. She was referred to the hospital by her general practitioner for the same problem. Her visual acuity was 6/6 right and 6/5 left with correction. No significant past ocular or general medical history was present and she did not have any history of familial disease. On examination her pupils were normal. Her ocular movements were full. She was orthophoric. Threshold visual field testing on the Humphrey automated visual field analyser showed repeatable field defects suggesting right homonymous hemianopia. Dilated fundus examination did not show any abnormality. A CT scan showed ill-defined areas of low attenuation close to the inferior horn of the left lateral ventricle, indistinguishable from its margin and