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Visual recovery in orbital vasculitis

A BAGEGNI,[1] R W LYNESS,[2] P B JOHNSTON,[1] AND J F DOUGLAS[3]

[1]*From the [1]Department of Ophthalmology, Royal Victoria Hospital, Belfast BT12 6BA, [2]Department of Histopathology, and [3]Renal Unit, Belfast City Hospital, Belfast BT9 7AB, Northern Ireland*

SUMMARY A 46-year-old woman with polyarteritis nodosa and chronic renal failure developed sudden loss of vision which was associated with orbital vasculitis. Treatment with cyclophosphamide produced rapid improvement in vision, which has been preserved with maintenance doses of cyclophosphamide and prednisolone.

Polyarteritis nodosa is a well recognised cause of orbital vasculitis which may be associated with visual loss due to either optic nerve compression or ischaemic optic neuropathy.[1-3] Corticosteroids, orbital irradiation,[4] and cyclophosphamide have been given as treatment, but, while the disease process and further visual deterioration may be arrested, visual recovery is rare. We report the case of a patient suffering from sudden loss of vision caused by orbital vasculitis due to polyarteritis nodosa, which responded successfully to treatment with cyclophosphamide.

Case report

A 46-year-old woman was referred to the eye clinic at the Royal Victoria Hospital, Belfast, in January 1983 for investigation of bilateral painless proptosis. The visual acuity was 6/6 in both eyes. The proptosis measured 17 mm right and 18 mm left. Both optic discs were swollen, with increased tortuosity of the retinal veins.

The patient did not return to the eye clinic for review, but in June 1983 she attended another hospital, where she was found to have chronic renal failure (serum urea 21 mmol/l and a creatine clearance of 15 ml per minute). A renal biopsy showed fibrous crescents in Bowman's capsule, with compression and hyalinisation of glomerular tufts, some of which were hypercellular. One glomerulus showed segmental necrosis. Immunofluorescence testing was positive for CIq, IgM, and fibrin. A diagnosis of polyarteritis nodosa was made following serological and clinical exclusion of systemic lupus erythematosis and subacute bacterial endocarditis. The patient was treated initially with prednisolone 40 mg daily, which was reduced later to a maintenance dose of 12·5 mg daily.

The patient returned to the eye clinic in October 1985 complaining of sudden loss of vision. The visual acuity was 6/9 in both eyes. The intraocular pressures were 22 mmHg right and 24 mmHg left. Both optic discs were swollen, and fluorescein angiography showed distinct central retinal stasis, with venous return still not complete 10 seconds after filling of the retinal arterioles. The proptosis measured 20 mm right and 21 mm left. Both visual fields showed inferior altitudinal defects (Fig. 1). Both eyes showed a mixed red-green defect with the Farnsworth-Munsell 100-hue test. The pattern visually evoked response (VER) latency was delayed from both eyes, which measured 150 ms right and 130 ms left (normal range 90-120 ms). Computer assisted tomography of the orbits showed abnormal soft tissue masses behind both globes (Fig. 2). A biopsy specimen was obtained from the left orbit through a transconjunctival incision over the left medial rectus muscle. Firm rubbery tissue was felt medial to the lower border of the medial rectus, and a small wedge of tissue was excised. The biopsy showed an acute vasculitis and fibrosis of orbital fat (Fig. 3). The inflammation and

Correspondence to A Bagegni, FRCS.

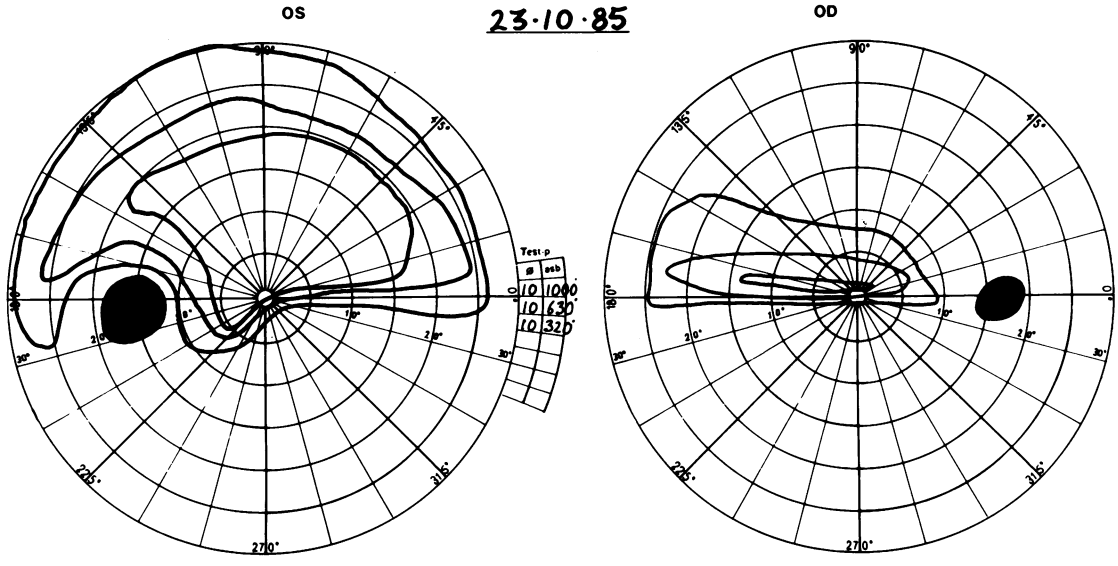

Fig. 1 *Both visual fields showed inferior altitudinal defects in October 1985.*

destruction of the blood vessel walls were consistent with polyarteritis nodosa, though in the small biopsy specimen available the vasculitis had destroyed the features which allow identification of artery from vein.

The patient was initially treated with prednisolone 40 mg daily and subcutaneous heparin injections of 5000 units twice daily. Vision deteriorated rapidly in December 1985, when visual acuity was 1/60 right eye and 6/60 left eye. In view of the rapidly failing vision the patient was treated with cyclophosphamide 200 mg daily and continued on prednisolone 40 mg daily. The visual acuity rapidly improved and in January 1986 measured 6/9 right and 6/12 left. Fluorescein angiography showed a normal circulation time at the optic discs, and late phase angiograms showed widespread choroidal folds involving the entire macular region. Both optic discs were slightly swollen. The patient was maintained on cyclophosphamide 200 mg three days a week and prednisolone 20 mg daily. In March 1987 visual acuity was 6/6 in both eyes. The visual fields showed recovery of the inferior altitudinal defects (Fig. 4), and both optic discs were pale. The patient remains on a maintenance therapy, consisting of cyclophosphamide 75 mg three days a week and prednisolone 20 mg daily.

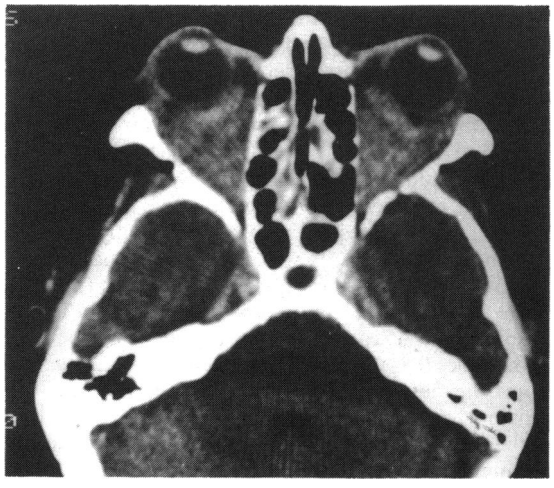

Fig. 2 *Computer tomographic scan of orbits showing abnormal soft tissue behind both globes.*

Discussion

Orbital vasculitis may be the presenting feature of polyarteritis nodosa.[5,6] We describe a patient who presented with bilateral proptosis and within a few months developed renal failure due to polyarteritis nodosa which was confirmed by renal biopsy. She retained normal vision for two years before vision deteriorated rapidly following an increase in the proptosis and consequent retinal vascular stasis. Orbital biopsy revealed an acute vasculitis and diffuse orbital fibrosis which was compatible with type 1 orbital pseudotumour described by Henderson and Farrow.[7] The visual loss was thought to be due to activation of the vasculitis with resulting optic

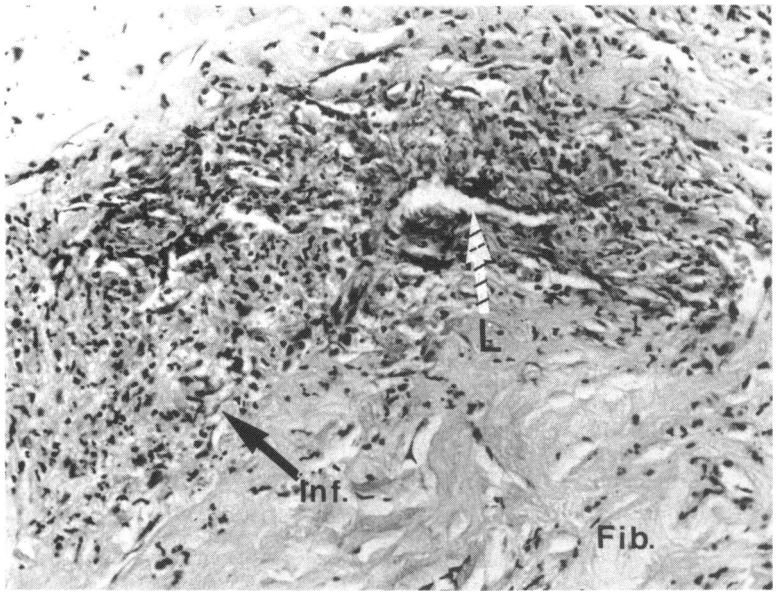

Fig. 3 *Histological section of orbital biopsy showing the lumen* (L) *of a blood vessel involved in an acute vasculitis process* (Inf). *Note the fibrosis* (Fig) *of the surrounding tissue. Haemoloxylin and eosin* ×195.

nerve compression[8] and ischaemic optic neuropathy.[3] Garrity[9] *et al.* reported on three patients with visual loss due to orbital vasculitis resistant to high-dose corticosteroids. They found cyclophosphamide to be effective in controlling the orbital pain and swelling, but visual loss was permanent. The rationale for using cyclophosphamide is its immunosuppressive effect, which is directed mainly against B cells.[10] Orbital vasculitis and polyarteritis nodosa are mediated by deposition of immune complexes in the vessel wall and are considered to be B-cell-mediated diseases.[11] One patient showed a rapid improvement of vision once cyclophosphamide was introduced, and recovery has been maintained for 18 months with a maintenance dose of cyclophosphamide and corticosteroids.

Clinicians faced with the problem of orbital pseudotumour associated with visual loss should first

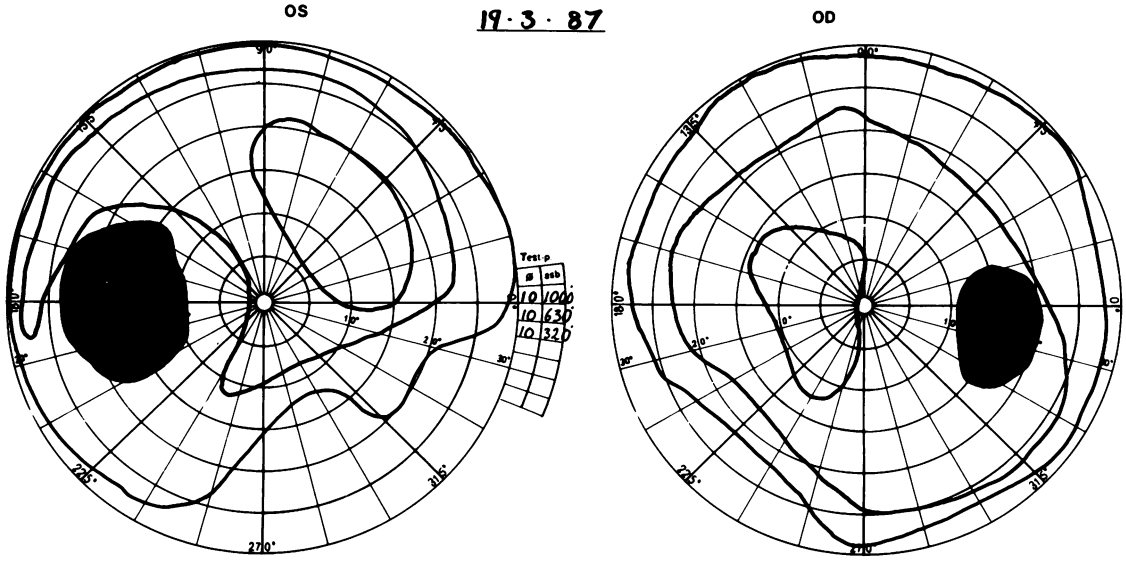

Fig. 4 *Both visual fields show recovery, March 1987.*

obtain an orbital biopsy for diagnosis and classification into either a type I vasculitis or type II lymphocytic inflammatory pseudotumour response as defined by Henderson and Farrow.[7] In cases with type I vasculitis and visual loss prompt immunosuppression of the B lymphocyte response with cyclophosphamide should should be considered as a potentially valuable method of treatment.

However, steroids and cyclophosphamide, both as short and long term drugs, have many side effects. Those of cyclophosphamide include opportunist infection, alopecia, gonadal atrophy, and haemorrhagic cystitis. These are in general reversible, but a more serious long term threat is malignancy, notably leukaemia, lymphoma, and bladder cancer.[12] Clearly long term cyclophosphamide therapy requires frequent re-evaluation. One should always be looking for an opportunity to withdraw the drug permanently or temporarily.

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References

1 Van Wien S, Merz EH. Exophthalmos secondary to periarteritis nodosa. *Am J Ophthalmol* 1963; **56:** 204–8.
2 Harcourt RB. Orbital granuloma associated with widespread angiitis. *Br J Ophthalmol* 1964; **48:** 673–7.
3 Hutchinson CH. Polyarteritis nodosa presenting as posterior ischaemic optic neuropathy. *J R Soc Med* 1984; **77:** 1043–6.
4 Kim RY, Roth RE. Radiotherapy of orbital pseudotumour. *Radiology* 1978; **127:** 507–9.
5 Jones IS, Jakobiec FA. Diseases of the orbit. Hagerstown: Harper and Row, 1979: 214–6.
6 Blodi FC, Gass JDM. Inflammatory pseudotumour of the orbit. *Br J Ophthalmol* 1968; **52:** 79–93.
7 Henderson JW, Farrow GM. *Orbital tumours.* Philadelphia: Saunders, 1973: 512–39.
8 Miller NR. *Walsh and Hoyt's clinical neurophthalmology.* 4th ed. Baltimore: Williams and Wilkins, 1982; **1:** 249–50.
9 Garrity JA, Kennerdall JS, Johnson BL, Ellis DE. Cyclophosphamide in the treatment of orbital vasculitis. *Am J Ophthalmol* 1986; **102:** 97–103.
10 Stockman GD, Heim LR, South MA, Trantin JJ. Differential effects of cyclophosphamide on the B- and T-cell compartments of adult mice. *J Immunol* 1973; **110:** 277–82.
11 Jakobiec FA, Jay L, Knowles DM. B- and T-lymphocytes in ocular disease. *Ophthalmology* 1984; **91:** 635–53.
12 Puri HC, Campbell RA. Cyclophosphamide and malignancy. *Lancet* 1977; i: 1306.

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