6 Deluise VP, Cobo M, Chandler D. Persistent corneal edema in the congenital rubella syndrome. *Ophthalmology* 1983; **90**: 835–839.
7 Sever JL, South MA, Shaver KA. Delayed manifestations of congenital rubella. *Rev Infect Dis* 1985; **7**(Suppl 1): S164–S169.
8 De Rojas Silva MV, Diez-Feijoo E, Rodriguez-Ares MT, Sanchez-Salorio M. Confocal microscopy of stage 4 diffuse lamellar keratitis with spontaneous resolution. *J Refract Surg* 2004; **20**: 391–396.
9 Grupcheva CN, Craig JP, Sherwin T, McGhee CN. Differential diagnosis of corneal oedema assisted by *in vivo* confocal microscopy. *Clin Exp Ophthalmol* 2001; **29**: 133–137.
10 Chiou AG, Kaufman SC, Beuerman RW, Ohta T, Yaylali V, Kaufman HE *et al*. Confocal microscopy in the iridocorneal endothelial syndrome. *Br J Ophthalmol* 1999; **83**: 697–702.

DTL Liu, RWK Law, HHW Lau, KSC Yuen, DSC Lam and W-M Chan

Department of Ophthalmology and Visual Sciences, Hong Kong Eye Hospital, The Chinese University of Hong Kong, 147K Argyle Street, Kowloon, Hong Kong

Correspondence: W-M Chan,
Tel: +852 2632 2879;
Fax: +852 2648 2943.
E-mail: cwm6373@netvigator.com

Eye (2006) **20,** 392–394. doi:10.1038/sj.eye.6701870; published online 1 April 2005

Sir,

Churg–Strauss syndrome in association with proliferative retinopathy

Churg–Strauss syndrome, also known as allergic granulomatosis, was first described by Churg and Strauss in 1951.[1] It is a systemic allergic disease characterised by eosinophil-rich and granulomatous inflammation involving the respiratory tract, and necrotising vasculitis affecting small to medium-sized vessels. The skin, heart, and gastrointestinal tract are occasionally involved, but ocular involvement is unusual. We present the case of a 54-year-old woman who presented with an aggressive unilateral proliferative retinopathy and was subsequently diagnosed as having Churg–Strauss syndrome.

Case report

A 54-year-old female presented to the Ophthalmology department with a 2-week history of hazy vision in her right eye. There was no past ocular history of note. She had a past medical history of hypothyroidism and asthma. Regular medications included thyroxine and salbutamol and becotide inhalers. She smoked 20 cigarettes per day.

On examination visual acuities were 6/24 in the right eye and 6/5 in the left eye. There was a moderate vitreous haemorrhage in the right eye. The left vitreous was clear. There were poor views of the right fundus, but no tears, holes or detachments were seen. The left fundus was healthy. Blood pressure and blood sugar were normal.

The patient was followed up weekly at the retinal out patient clinic. At 3 weeks following her initial attendance the view of the fundus in the right eye had resolved sufficiently to reveal superotemporal new vessels. Fluorescein angiography confirmed the presence of these new vessels (Figure 1) and demonstrated widespread capillary closure (Figure 2).

The patient subsequently underwent right panretinal photocoagulation. Initially, the new vessels appeared to regress. However, 4 months following panretinal photocoagulation she experienced a further vitreous haemorrhage in the right eye. On this occasion the vitreous haemorrhage failed to clear, and therefore 2 months later right vitrectomy combined with endolaser was performed.

The patient was followed up every 2 months in the retinal out patient clinic. At 1 year following her initial presentation, the patient attended clinic complaining of significant systemic symptoms. For the preceding 2 months she had been experiencing dizzy spells and headaches behind her right eye. She complained of lethargy, poor appetite, and had lost a stone in weight. She had noticed progressive numbness in her hands and feet and had developed a nasal discharge. She also

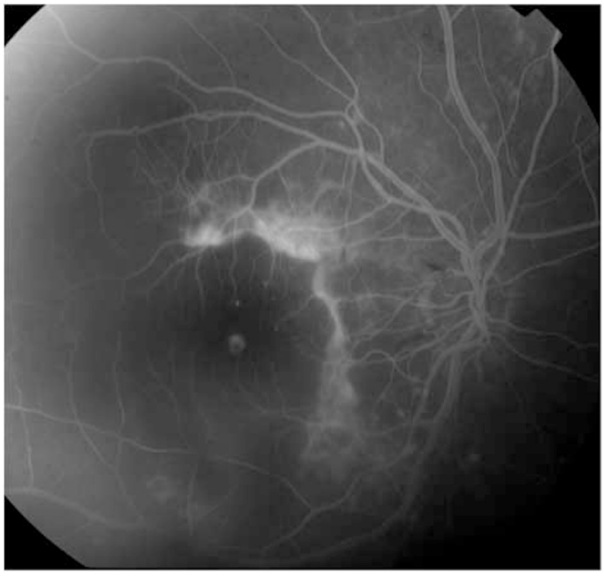

Figure 1 Fluorescein angiography showing superotemporal neovascularisation.

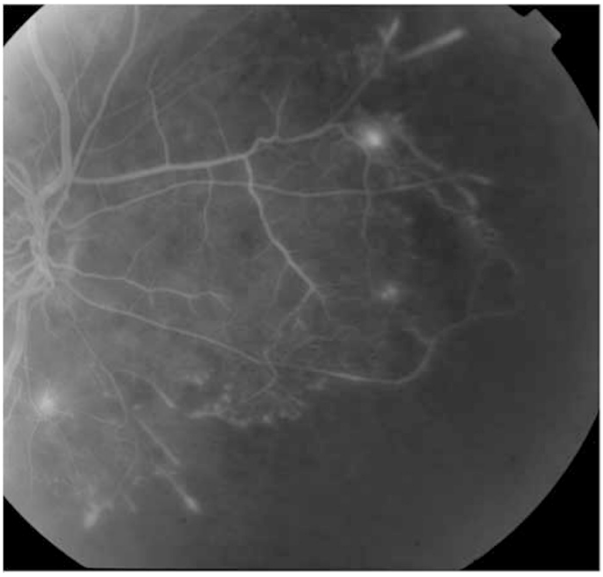

Figure 2 Fluorescein angiography showing inferonasal capillary closure.

described symptoms consistent with episodes of right amaurosis fugax.

On examination she had a mild vitreous haemorrhage in the right eye and had developed further active neovascularisation superotemporally in the right fundus. Left fundus remained healthy. She was noted to have a livedo reticularis rash on her knees and upper thighs. Neurological examination revealed a bilateral glove and stocking peripheral sensory neuropathy.

In view of a suspected vasculitic process, the patient was referred to the rheumatology department. Full blood count revealed a haemoglobin of 104 g/l (normal range 115–165 g/l). Mean cell volume was 72.6 fl (normal range 78–98 fl). There was a hypochromic microcytic blood picture. Eosinophils were elevated at 1.0×10^9/l (normal range 0.0–0.4 $\times 10^9$/l). ESR and CRP were both normal. Serum ACE was normal. ANCA and ANA were both negative. Chest X-ray did not reveal any focal abnormality. On the basis of a history of asthma, hypereosinophilia, peripheral neuropathy, and paranasal sinusitis, a diagnosis of Churg–Strauss syndrome was made.

The patient was initially treated by the rheumatologists with six cycles of chemotherapy comprising intravenous cyclophosphamide (15 mg/kg) and methylprednisolone (10 mg/kg). She was then commenced on oral methotrexate. Further right vitreous haemorrhage and superotemporal neovascularisation was treated with a repeat vitrectomy and endolaser. At her most recent review 4 years following initial presentation visual acuities were 6/9 right eye and 6/5 left eye and there was no evidence of any active neovascularisation in either eye.

Comment

Churg–Strauss syndrome is a rare disease with an estimated annual frequency of 2.4–6.8 per million patient-years.[2] Churg and Strauss reported the cases of 13 patients with severe bronchial asthma and disseminated necrotising vasculitis.[1] These patients were found to have fever, eosinophilia, and multisystem involvement, which was associated with a histologic pattern of necrotising arteritis, eosinophilic tissue infiltration, and extravascular granulomata. They entitled the disorder allergic granulomatosis (Churg–Strauss syndrome). To establish a diagnosis of Churg–Strauss syndrome, four of six criteria must be met (the presence of asthma, hypereosinophilia (>10%), mononeuropathy, or polyneuropathy, nonfixed pulmonary infiltrates, paranasal sinus abnormality, and extravascular eosinophil infiltration in biopsy specimens).[3]

Ocular involvement in the Churg–Strauss syndrome is rare. The reported ophthalmologic manifestations include corneal ulcer, uveoscleritis, conjunctival granuloma, orbital inflammatory pseudotumour, amaurosis fugax, retinal artery occlusion, ischaemic optic neuropathy, oculomotor nerve palsy, and trochlear nerve palsy.[4] Takanashi *et al*[4] classified these ocular manifestations into two types: orbital inflammatory pseudotumour and ischaemic vasculitis. They hypothesised that these two groups may represent the two essential characteristics of the disease processes: granulomatosis and angiitis.

Serum ANCA is positive in 70% of patients with Churg–Strauss syndrome.[5] In the review of ophthalmic cases by Takanashi *et al* the presence of ANCA was characterisitic of the ischaemic type. They advised that this may be a risk factor for sudden visual loss and that prophylactic steroids may be advisable in these cases.

Our case was of the ischaemic type but was negative for ANCA. The proliferative retinopathy in this patient was particularly aggressive and had the potential for marked visual loss. However, with appropriate surgical and medical management she has had a relatively good outcome. To our knowledge this is the first reported case of Churg–Strauss syndrome presenting with a proliferative retinopathy.

Acknowledgements

Proprietary interest: None.

References

1 Churg J, Strauss L. Allergic granulomatosis, allergic angitis, and periarteritis nodosa. *Am J Pathol* 1951; **27**: 277–301.
2 Noth I, Strek ME, Leff AR. Churg–Strauss syndrome. *Lancet* 2003; **361**(9357): 587–594.

3 Masi AT, Hunder GG, Lie JT, Michel BA, Bloch DA, Arend WP *et al*. The American College of Rheumatology 1990 criteria for the classification of Churg–Strauss syndrome (allergic granulomatosis and angiitis). *Arthritis Rheum* 1990; **33**(8): 1094–1100.
4 Takanashi T, Uchida S, Arita M, Okada M, Kashii S. Orbital inflammatory pseudotumour and ischaemic vasculitis in Churg–Strauss syndrome. Report of two cases and review of the literature. *Ophthalmology* 2001; **108**(6): 1129–1133.
5 Lhote F, Guillevin L. Polyarteritis nodosa, microscopic polyangiitis, and Churg–Strauss syndrome. Clinical aspects and treatment. *Rheum Dis Clin North Am* 1995; **21**(4): 911–947.

P Cackett and J Singh

Department of Ophthalmology,
Princess Alexandra Eye Pavilion,
Chalmers Street, Edinburgh EH3 9HA,
Scotland, UK

Correspondence: P Cackett
Tel: +44 131 536 1674;
Fax: +44 131 536 1574.
E-mail: pete@pdcackett.demon.co.uk

Eye (2006) **20,** 394–396. doi:10.1038/sj.eye.6701871;
published online 8 April 2005

Sir,

Tailoring pan-retinal photocoagulation for the haemianopic patient

Numerous studies have showed the effect of panretinal photocoagulation (PRP) on the patients' field of vision.[1–3] Cerebro-vascular accidents (CVA) are often associated with diabetes,[4] and they commonly cause visual field defects.[5] We report the case of a diabetic patient with proliferative diabetic retinopathy (PDR), who had had a stroke causing a homonymous haemianopia — and the manner in which he was treated, to conserve as much of his visual field as possible.

Case report

A 26-year-old male type I diabetic was referred to the eye clinic. He had a stroke causing a right homonymous haemianopia when he was 15 years old (Figure 1). On examination he had disc neovascularisation in the left eye and had scattered PRP to the temporal retina respecting the vertical midline. Heavier more confluent burns than usual were placed to this area of 'nonseeing' retina to attempt to spare the preserved visual field. The new vessels regressed following treatment. After 10 months, he developed further neovascularisation and had more PRP to the temporal retina of the left eye. He then had PRP to the nasal retina of the right eye (respecting the vertical midline) for new vessels elsewhere. Additional laser was applied to the temporal right retina, and nasal left retina, but was placed very sparsely to preserve as much as possible of his residual visual fields. The new vessels have regressed in both eyes following treatment, and we have managed to conserve the visual field unaffected by the stroke.

Comment

The Diabetic Retinopathy Study[1] and the Early Photocoagulation for Diabetic Retinopathy study[2] have clearly demonstrated the benefit of treating proliferative diabetic retinopathy to reduce the risk of severe visual loss. Visual field constriction is a well-recognized side

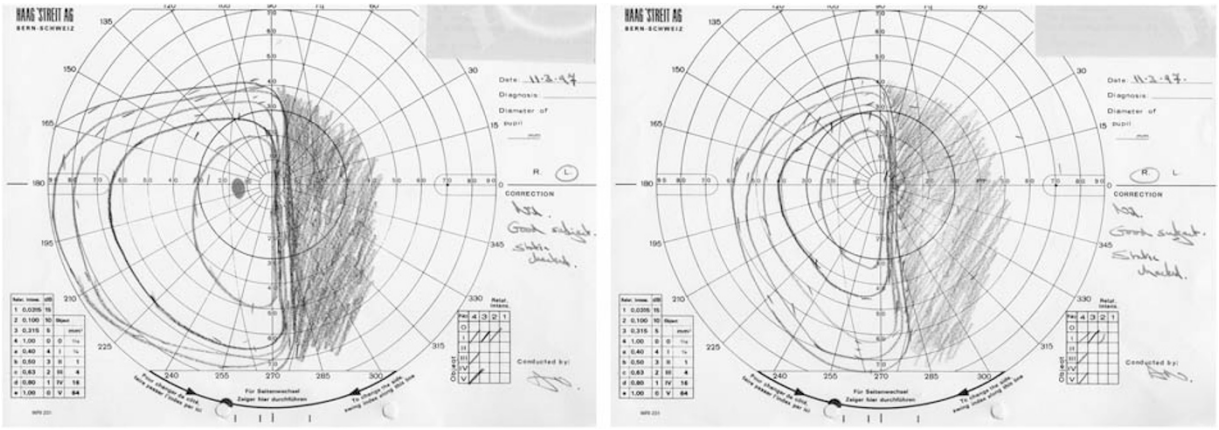

Figure 1 Goldmann's visual field showing a right homonymous haemianopia.