

of ocular pathologies and following procedures such as laser peripheral iridotomy and macular translocation (MT) surgery.<sup>1,3-5</sup> The prevalence of CBS ranges from 0.4 to 13% in various study populations,<sup>2</sup> but its incidence following ocular surgery is not known.

Two cases of CBS after MT surgery have provided the strongest direct observation supporting the 'sensory deprivation' theory for CBS.<sup>4</sup> The visual hallucinations in these patients started within 24 h after MT and ceased after retinal reattachment and visual improvement 3 and 7 days later, respectively. Our patient developed hallucinations in the early postoperative period when her vision worsened owing to the intravitreal gas. These hallucinations ceased with a decrease in the size of the gas bubble and improvement in vision. The acute reduction in vision rather than the low visual acuity itself may have triggered the visual hallucinations, as suggested by Shiraishi *et al.*<sup>5</sup>

Our patient had good visual acuity in the left eye. Although CBS occurs more commonly in patients with bilateral visual loss, it has been reported in the presence of good visual acuity in the better seeing eye.<sup>1,2,4</sup> The postoperative prone positioning may have contributed to the occurrence of hallucinations owing to occlusion of vision in the better eye in our case. This report extends the spectrum of ocular procedures associated with CBS.

## References

- 1 Menon GJ, Rahman I, Menon SJ, Dutton GN. Complex visual hallucinations in the visually impaired: the Charles Bonnet syndrome. *Surv Ophthalmol* 2003; **48**: 58–72.
- 2 Tan CSH, Lim VSY, Ho DYM, Yeo E, Ng BY, Au Eong KG. Charles Bonnet syndrome in Asian patients in a tertiary ophthalmic centre. *Br J Ophthalmol* 2004; **88**: 1325–1329.
- 3 Tan CSH, Yong VYK, Au Eong KG. Onset of Charles Bonnet syndrome (formed visual hallucinations) following bilateral laser peripheral iridotomies. *Eye* 2004; **18**: 647–649.
- 4 Au Eong KG, Fujii GY, Ng EWM, Humayun MS, Peiramici DJ, de Juan Jr E. Transient formed visual hallucinations following macular translocation for subfoveal choroidal neovascularisation secondary to age-related macular degeneration. *Am J Ophthalmol* 2001; **131**: 664–666.
- 5 Shiraishi Y, Terao T, Ibi K, Nakamura J, Tawara A. Charles Bonnet syndrome and visual acuity: the involvement of dynamic or acute sensory deprivation. *Eur Arch Psychiatry Clin Neurosci* 2004; **254**: 362–364.

AM Wagle<sup>1,2</sup> and K-G Au Eong<sup>1,2,3,4,5</sup>

<sup>1</sup>Department of Ophthalmology and Visual Sciences, Alexandra Hospital, Singapore, Singapore

<sup>2</sup>The Eye Institute, National Healthcare Group, Singapore, Singapore

<sup>3</sup>Department of Ophthalmology, Tan Tock Seng Hospital, Singapore, Singapore

<sup>4</sup>Department of Ophthalmology, Yong Loo Lin School of Medicine, National University of Singapore, Singapore, Singapore

<sup>5</sup>Singapore Eye Research Institute, Singapore, Singapore

Correspondence: AM Wagle,  
Department of Ophthalmology and Visual Sciences, Alexandra Hospital, 378 Alexandra Road, Singapore 159964, Singapore  
Tel: + 65 63793512;  
Fax: + 65 63793540.  
E-mail: Ajeet\_Wagle@alexhosp.com.sg

*Eye* (2007) **21**, 106–107. doi:10.1038/sj.eye.6702411;  
published online 12 May 2006

## Sir, Ocular myopathy of Wegener's granulomatosis

Wegener's granulomatosis (WG) is a systemic disease with obscure aetiology that can frequently affect the eye.<sup>1</sup> An appropriate diagnostic evaluation, follow-up, and treatment can prevent progressive systemic manifestations of this disease, which may rarely result in death.<sup>2</sup> Extraocular muscle involvement presenting as diplopia may be the only presenting symptom of WG before the disease becomes more generalised. We report and discuss here a case of WG who presented with diplopia as a consequence of extraocular muscle involvement. This has not been reported earlier in the literature.

## Case report

A 67-year-old woman presented to the eye clinic with gradual onset of painless intermittent diplopia, particularly on down gaze, over a period of 3 months. Initial ocular examination showed a mild left inferotemporal conjunctival injection with no other signs of inflammation or proptosis. Her vision was 6/9 in both eyes. Orthoptic assessment of the left eye showed restricted ocular movements on elevation as well as depression (Figure 1). Owing to the presence of suppression, there was little evidence of diplopia in any position of gaze. A tentative diagnosis of thyroid eye disease was entertained and the patient was sent for further tests. The left conjunctival redness resolved within a fortnight on topical antibiotic instillation.

Investigations including thyroid function tests, chest X-rays, FBC, ESR, serum electrolytes, complements, and

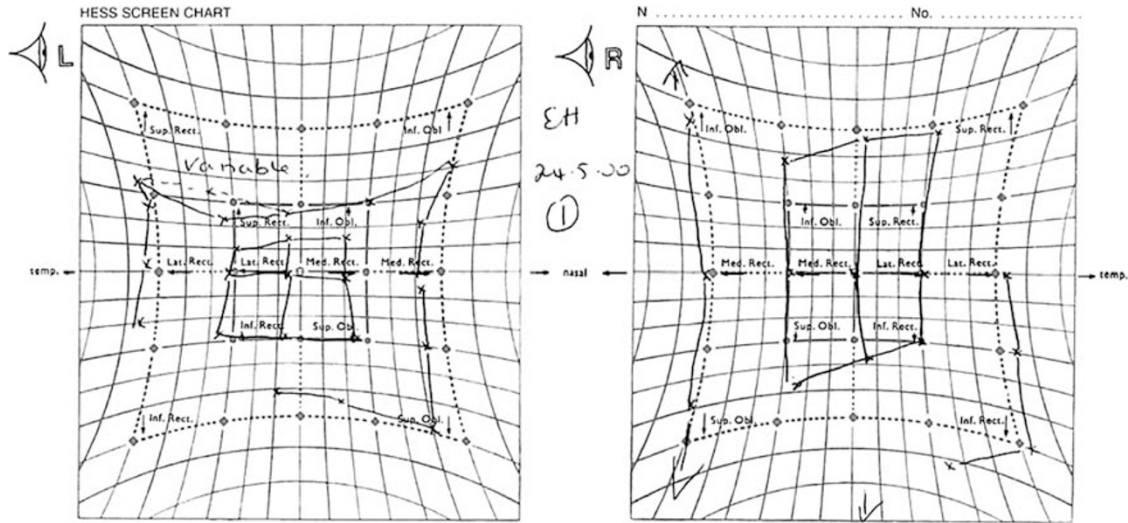


Figure 1 Restricted ocular movements in elevation as well as depression in left eye.

immunoglobulins, as well as electrophoresis were normal. Serum autoantibodies including antineutrophil cytoplasmic antibody (ANCA) screening test was also normal.

CT scan (Figure 2) showed extensive mucosal thickening of the paranasal sinuses involving the antrum, ethmoids, sphenoid, and frontal sinuses. There was a breach of cortex in the medial wall and the floor of the orbit on the left, suggesting a possible malignant sinus disease. The patient was therefore referred to the ENT specialist who performed a sinus biopsy. The biopsy report showed sinus mucosa heavily infiltrated with large number of inflammatory cells including plasma cells and eosinophils. There was no evidence of malignancy. The patient was prescribed steroidal nasal spray, which gave her some relief.

A 6-month follow-up showed little change in her symptom of intermittent painless diplopia. During that time, her redness had returned with mild proptosis in the same left eye. She had fullness of her left lower lid along with marked conjunctival injection. A repeat CT scan showed infiltrative sinus disease with a left inferior orbital mass. The possibility of the right orbital extension as well as the nature of infiltration made the radiologist contemplate a diagnosis of WG. An alternative diagnosis was orbital pseudotumour when there was a dramatic improvement with a trial dose of oral Prednisolone 20 mg o.d.

A low dose of Prednisolone was maintained over the next few months till she presented to the casualty department with sudden onset of haematuria. Till that moment she never had any systemic symptoms. Although a renal biopsy was inconclusive, but a positive ANCA on serology tests and significant MRI scan

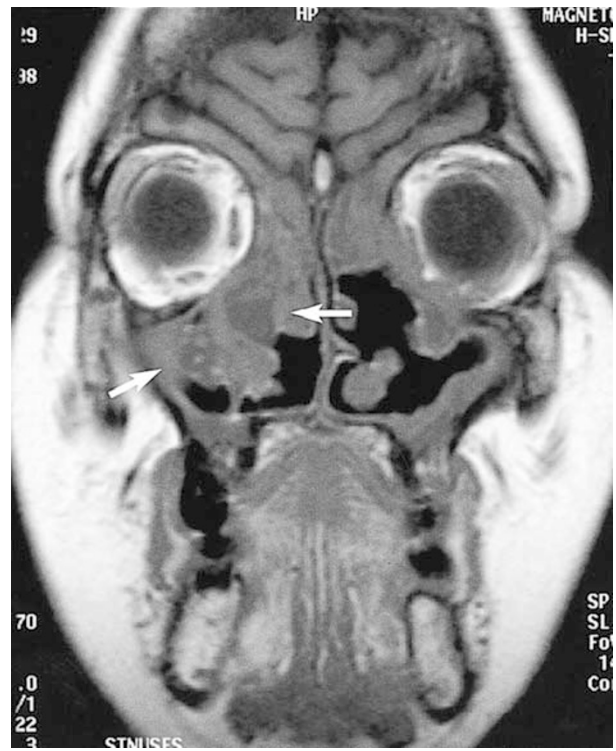
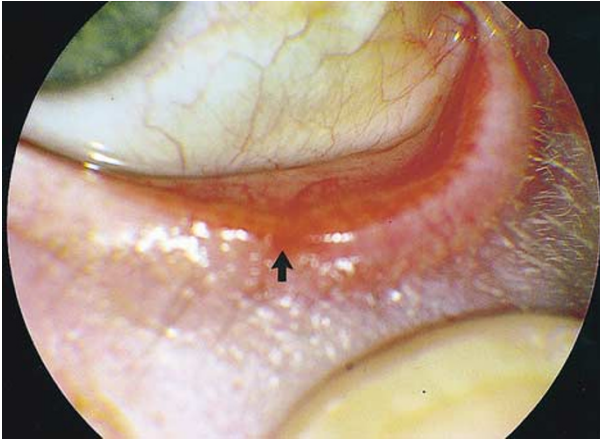


Figure 2 Extensive mucosal thickening of the paranasal sinuses.

changes of the sinuses and the orbits helped the rheumatologist confirm the clinical diagnosis of WG. She was commenced on oral Azathioprin and Prednisolone therapy. Her systemic symptoms improved dramatically with treatment.



**Figure 3** Tarsal conjunctival ulceration of left lower lid.

Since then, her ocular motility problems have remained stable. She subsequently developed a small tarsal conjunctival ulceration on her left lower lid (Figure 3), which resolved spontaneously.

#### Comment

The extraocular muscle involvement in an already confirmed case of WG has been reported earlier.<sup>2,4</sup> In our case, the patient's only symptom, to begin with, was gradual onset of painless diplopia. All the initial appropriate tests including chest X-ray as well as sinus biopsy were inconclusive. The absence of an initial distinctive pathological feature as well as negative ANCA test as reported by previous authors<sup>2,5</sup> made the diagnostic confirmation more difficult and appropriate treatment could not be initiated.

Ocular involvement can occur with or without obvious systemic manifestations of WG. This patient's initial clinical presentation was similar to 'very Limited form of WG' as described by Harper *et al*<sup>2</sup> and she then progressed to a more generalised form. The evolution of the disease process in this case over a period of 18 months will fit in the Harper *et al*'s<sup>2</sup> study Group II.

In their study of 47 patients of WG over a period of 10 years, the Group II patients presented initially with ocular lesions only and subsequently developed systemic manifestations. All patients in this group were in their sixth or seventh decade and so was ours. As in their study, our patient's age of 67 was a risk factor for the development of systemic WG.<sup>2</sup>

The lady subsequently developed a lower lid tarsal conjunctival ulceration (Figure 3), which did not require any intervention. It has been suggested that this may be a sign of renewed disease activity,<sup>6</sup> but fortunately that was not the case in this instance.

To summarise this case, after nearly a year of her initial presentation of diplopia, this 67-year-old lady developed renal symptoms and a positive ANCA. An initial diagnostic evaluation for WG case was difficult and therefore a proper treatment could not be initiated.

We agree with other authors that the probable pathological explanation of diplopia could be either from a contagious granulomatous sinus disease as in our case or a diffuse vasculitis of the extraocular muscles.<sup>1-4</sup>

Following treatment, the lady did not regain full motility. This was perhaps owing to the presence of a prolonged underlying muscular inflammation as confirmed by the development of suppression.

This case is typical of WG, in that the ocular symptoms and signs were subject to relapse depending on the progression of the disease.<sup>2</sup> As seen in this case, the long-term prognosis for recovery from the restrictive myopathy is unpredictable.

From the case, we would like to point out that a restrictive ocular myopathy may be the first sign of WG and this may help the physician to evaluate this lethal disease. We recommend a need for multidisciplinary approach with active communication between specialties to diagnose WG. This case tells us that diplopia may be the only ocular presenting symptom of WG.

#### References

- 1 Haynes BF, Fishman ML, Fauci AS, Wolff SM. The ocular manifestations of Wegener's granulomatosis. Fifteen years experience and review of the literature. *Am J Med* 1977; **63**: 131-141.
- 2 Harper SL, Letko E, Samson CM, Zafirakis P, Sangwan V, Nguyen Q *et al*. Wegener's granulomatosis: the relationship between ocular and systemic disease. *J Rheumatol* 2001; **28**: 1025-1032.
- 3 Spalton DJ, Graham EM, Page NG, Sanders MD. Ocular changes in limited forms of Wegener's granulomatosis. *Br J Ophthalmol* 1981; **65**: 553-563.
- 4 Pinchoff BS, Spahlinger DA, Bergstrom TJ, Sandall GS. Extraocular muscle involvement in Wegener's granulomatosis. *J Clin Neuroophthalmol* 1983; **3**: 163-168.
- 5 Sadiq SA, Jennings CR, Jones NS, Downes RN. Wegener's granulomatosis: the ocular manifestations revisited. *Orbit* 2000; **19**: 253-261.
- 6 Fortney AC, Chodosh J. Conjunctival ulceration in recurrent Wegener's granulomatosis. *Cornea* 2002; **21**: 623-624.

P Chakraborty and R Mukhopadhyay

Department of Ophthalmology,  
Darlington Memorial Hospital,  
HollyHurst Road,  
Darlington DL3 6HX, UK

Correspondence: P Chakraborty,  
Tel: + 44 1325743417;  
Fax: + 44 1325743413.  
E-mail: pceye@yahoo.co.uk

*Eye* (2007) 21, 107–110. doi:10.1038/sj.eye.6702413;  
published online 11 August 2006

Sir,  
**A case of endogenous *Candida albicans*  
endophthalmitis resolving with itraconazole treatment  
without recourse to vitrectomy**

We report the case of a 73-year-old woman who developed endogenous, bilateral, endophthalmitis during a protracted admission to I.T.U. following bowel resection for suspected tumour.

She presented with symptoms of decreased acuity and floaters. Indirect ophthalmoscopy revealed intraretinal and preretinal white lesions as well as large 'snowball' lesions in the vitreous in both eyes. Her best-corrected visual acuity was 6/9 in the right eye and 6/12 in the left. Repeated blood cultures showed no growth.

The severity of her systemic condition did not allow for early vitrectomy, therefore oral fluconazole was given, but despite 400 mg b.d. for 15 days, her vision deteriorated to counting fingers in the right eye and 6/18 in the left.

She subsequently underwent right vitrectomy and intravitreal injection of 10 µg amphotericin B into each eye. A vitreous sample taken from the right eye during the procedure grew *Candida albicans* sensitive to fluconazole; however, despite postoperative treatment with oral fluconazole, 200 mg b.d., the endophthalmitis deteriorated in both eyes, with increased fungal lesions and a further decrease in visual acuity.

Because of her recent bowel surgery and recurrent episodes of bowel obstruction, we were concerned about adequate drug absorption. To ensure that a therapeutic serum level of antifungal agent was achieved, itraconazole was used in preference to fluconazole as serum levels can be monitored and oral dose adjusted in case of any malabsorption.

A radical improvement was seen on examination after 10 days of intravenous (i.v.) itraconazole 200 mg b.d. Serum concentrations were maintained in the therapeutic range. An oral maintenance dose of 200 mg b.d. was then given and tapered down to 100 mg b.d. over 3 months.

Treatment with itraconazole led to complete resolution of *C. albicans* endophthalmitis in both the vitrectomised

and nonvitrectomised eye. Best-corrected visual acuity 4 months after presentation was 6/9-2 in both eyes.

Endogenous endophthalmitis is often associated with high mortality and poor visual acuity outcomes.<sup>1</sup> Bowel surgery for tumour is a common predisposing condition for endogenous endophthalmitis,<sup>2</sup> as is long-term i.v. catheter placement.<sup>3</sup> Compared with postoperative or post-traumatic endophthalmitis, patients with endogenous endophthalmitis are more likely to have fungal isolates with a predominance of *C. albicans*.<sup>3</sup>

**Comment**

The current, established treatment for this condition is pars plana vitrectomy with intravitreal injection of amphotericin B.<sup>1-4</sup> Fluconazole is the antifungal shown to achieve the highest concentration in the vitreous following oral administration to white rabbits.<sup>5</sup> In this case, the right eye failed to respond to conventional treatment including vitrectomy, but both eyes responded to therapeutic serum levels of itraconazole.

This case suggests that a remarkable recovery in visual acuity is possible with antifungal treatment alone, without performing therapeutic vitrectomy.<sup>6</sup> The location of the fungal lesions may be predictive of the success of medical treatment. It has been suggested that only chorioretinal lesions respond to medical treatment, whereas extension into the vitreous requires surgery.<sup>7</sup> In this case, vitreous seeding responded to intravitreal amphotericin and i.v. itraconazole without vitrectomy. Furthermore, although itraconazole is not currently first line in the treatment of *Candida* endophthalmitis, it should be considered when first-line agents fail to control the infection.<sup>8</sup>

**References**

- Schiedler V, Scott IU, Flynn Jr HW, Davis JL, Benz MS, Miller D. Culture-proven endogenous endophthalmitis: clinical features and visual acuity outcomes. *Am J Ophthalmol* 2004; 137(4): 725–731.
- Zhang YQ, Wang WJ. Treatment outcomes after pars plana vitrectomy for endogenous endophthalmitis. *Retina* 2005; 25(6): 746–750.
- Essman TF, Flynn Jr HW, Smiddy WE, Brod RD, Murray TG, Davis JL *et al.* Treatment outcomes in a 10-year study of endogenous fungal endophthalmitis. *Ophthalmic Surg Lasers* 1997; 28(3): 185–194.
- Stoffelns BM. Endogenous *Candida* endophthalmitis combined with severe general diseases. *Klin Monatsbl Augenheilkd* 2005; 222(3): 214–217.
- Savani DV, Perfect JR, Cobo LM, Durack DT. Penetration of new azole compounds into the eye and efficacy in