given 1 g of intravenous methylprednisolone and then commenced on oral prednisolone 80 mg daily as a reducing course. The patient's anterior segment inflammation responded well to topical steroids. Cyclosporine 125 mg twice daily orally was commenced when the patient's vision showed no significant improvement with oral steroids after 2 weeks. A fluorescein angiogram was performed at this time that showed persistent leakage of dye from the optic discs (Fig. 1).

The exudative retinal detachments gradually resolved. Her oral prednisolone was slowly reduced to 15 mg daily and her cyclosporine discontinued. Marked subretinal fibrosis was noted which probably reflects the severity of the ocular involvement. Her visual acuity improved to 6/18 with pinhole in each eye and this was maintained 1 year later, still on 15 mg of prednisolone daily.

Comment

Classically Vogt–Koyanagi–Harada syndrome follows four stages: an initial short prodromal stage is followed 3–5 days later by an acute uveitic phase; several weeks later there is a convalescent or chronic phase which may be followed months or years later by a chronic recurrent phase.[1,2]

The prodromal illness itself typically consists of headache, nausea, vertigo, slight fever, stiff neck, orbital pain and other non-specific symptoms and usually lasts for only a few days.[1,3] Extraocular signs and symptoms other than headache are very rare, hence the unusual presentation of our patient.[4]

Typical posterior segment findings include bilateral exudative retinal detachments with a granulomatous panuveitis and optic disc swelling. Yellow-white well-defined lesions may also occur in the peripheral retina. These findings are not pathognomonic of Vogt–Koyanagi–Harada syndrome and a similar picture can occasionally be seen with miliary tuberculosis.

Choroidal tubercles present in between 28% and 60% of cases of miliary tuberculosis; 80% are unilateral and overlying serous detachment may sometimes occur. Usually there is no accompanying uveitis but panuveitis may occur with choroidal tubercles, as may optic nerve swelling.[5]

This case demonstrates an unusual presentation of Vogt–Koyanagi–Harada syndrome with severe systemic malaise which required urgent hospital admission to exclude an infectious aetiology.

Ophthalmologists and physicians should be aware that Vogt–Koyanagi–Harada syndrome may present as a surprisingly severe systemic illness, but nevertheless the rapid introduction of high dose systemic steroids and cyclosporine may result in useful recovery of vision and improvement in systemic symptoms.[1]

References

1. Goto H, Rao NA. Sympathetic ophthalmia and Vogt–Koyanagi–Harada syndrome. Int Ophthalmol Clin 1990;30:279–85.
2. Moorthy RS, Inomata H, Rao NA. Vogt–Koyanagi–Harada syndrome. Surv Ophthalmol 1995;39:265–92.
3. Snyder DA, Tessler HA. Vogt–Koyanagi–Harada syndrome. Am J Ophthalmol 1980;90:69–75.
4. Beniz J, Forster DJ, Lean JS, Smith RE. Variations in clinical feature of the Vogt–Koyanagi–Harada syndrome. Retina 1991;11:275–80.
5. Helm CJ, Holland GN. Ocular tuberculosis. Surv Ophthalmol 1993;38:229–56.

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Sir,

Aggressive retinal vasculitis in polyarteritis nodosa

Polyarteritis nodosa (PAN) is an idiopathic systemic vasculitis which affects medium-sized and small arteries. It is more common in males than females (3:1) and usually occurs between 20 and 50 years of age.

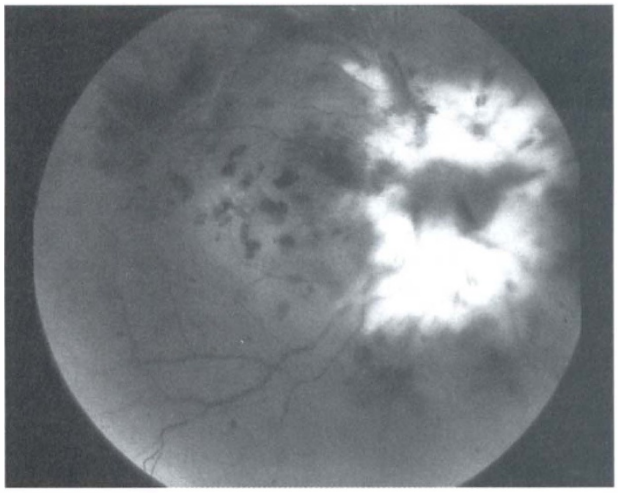
Ocular manifestations occur in 10–20% of patients with PAN. The spectrum of ocular manifestations includes scleritis, peripheral ulcerative keratitis, pseudotumour of the orbit, retinal vasculitis and central retinal artery occlusion.[1]

PAN has a high mortality, with a 5-year survival rate of 13% in untreated patients. Treatment with corticosteroids and cytotoxic immunosuppressive agents in combination has been reported to increase the 5-year survival rate to 80%.[2]

Case report

A 28-year-old man was referred for an ophthalmologic examination with a 2-week history of vision loss in the right eye. No history of other medical problems was elicited at presentation.

At presentation, he had visual acuities of hand movements in the right eye and 6/9 in the left. Anterior segment examination showed fine keratic precipitates with 2+ cells in the anterior chamber of the right eye, and no inflammation in the left anterior chamber. Retinoscopy revealed a large area of retinal opacification in the peripapillary region and fluorescein angiography showed the presence of arterial and venous leakage and disc oedema in the right eye (Fig. 1). In the left eye, there was mild disc swelling and peripapillary venous sheathing. The clinical appearance of the lesion was



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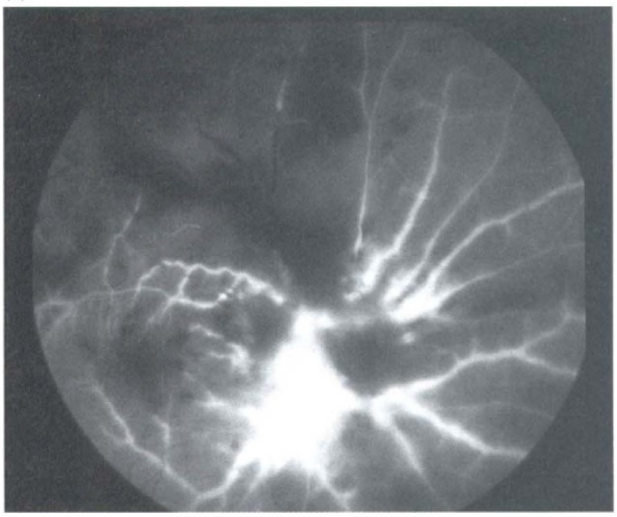

(b)

Fig. 1. *(a) Red-free fundus photograph of the right eye showing a large area of retinal opacification in a peripapillary location. Haemorrhages in the macula and peripapillary regions imply vein occlusion. (b) Late-phase angiogram confirming involvement of veins and also arteries, with closure of the superotemporal main vessels.*

compatible with a viral retinitis. As the visual acuity in the right eye was very poor, we decided to proceed with an intravitreal injection of ganciclovir 2 mg/0.1 ml as this agent has a spectrum that covers for cytomegalovirus and also herpes simplex and varicella zoster. A vitreous biopsy was also obtained and sent for both polymerase chain reaction (PCR) for viral and *Toxoplasma* DNA, and cytology for cell phenotype. Blood tests were ordered including syphilis serology, *Toxoplasma* antibodies, rheumatoid factor, ANA, ANCA, SSA, SSB, HIV, $CD4^+$ and $CD8^+$. The blood tests were positive only for *Toxoplasma* IgG. SInce the patient was HIV negative and his $CD4^+$ count was normal (1660×10^9/l) but an infection by herpes simplex or varicella zoster could not be excluded, he was started on acyclovir 10 mg/kg every 8 h. The vitreous PCR result was negative for herpes simplex, herpes zoster, cytomegalovirus and *Toxoplasma*. The cytology result showed only chronic inflammatory cells without any evidence of lymphoma. At this stage the patient was started on prednisolone (60 mg/day).

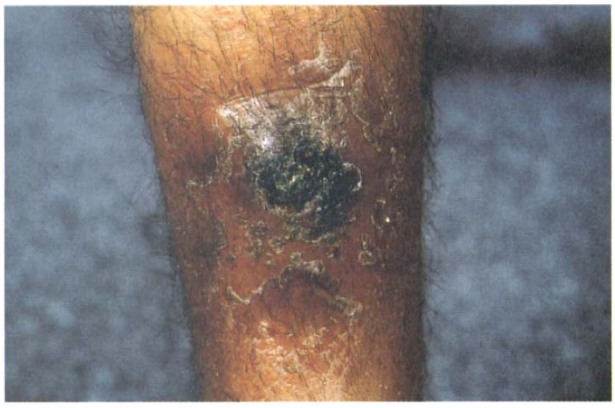

Fig. 2. *Necrotic appearance of the leg ulcer from which the diagnosis of polyarteritis nodosa was confirmed following a biopsy.*

The right eye did not show any obvious response to therapy, with vision further decreasing to light perception. The left eye showed a good response with resolution of the vasculitis and disc oedema. In another attempt to establish the diagnosis, a vitrectomy with retinal biopsy was performed and the sample sent for immunohistochemical analysis. The retinal biopsy was inconclusive. The patient continued to be observed and the systemic steroids were slowly tapered. The visual acuity in the right eye deteriorated to no light perception due to a secondary retinal detachment.

At this point the patient stated that he had an ulcer on his right leg, which had appeared 2 months earlier and for which he had been given systemic antibiotics while on holiday in Bangladesh due to a suspected infectious aetiology. The patient was referred to dermatologists who started him on Dapsone for the treatment of his leg ulcer (Fig. 2), with some improvement. They also performed a skin biopsy, which established the diagnosis of PAN. The patient was, at this point, started on high-dose systemic steroids and azathioprine.

Comment

The real incidence and prevalence of PAN are unknown, due to difficulties in establishing the diagnosis. The diagnosis of PAN is based on clinical findings and histological demonstration of necrotising lesions of the involved vessels.[3] Our patient had the diagnosis established by the histological examination of a skin lesion, in the presence of a severe vasculitis in one eye.

Akova *et al.*[1] reported 5 cases of PAN with ocular involvement with only one case of retinal vasculitis amongst them. The first case of combined arterial and venous vasculitis was described by Morgan *et al.* in 1986.[4] Our patient presented with a severe vasculitis involving both arteries and veins.

Ocular involvement may be the first manifestation of the disease. In our patient, even though the ocular picture was not the first manifestation of the disease, it was this finding that stimulated the dermatologists to proceed with biopsy of the skin ulcer.

PAN is a rare but important condition to recognise due to its potential life-threatening character. It is very important to carry out a proper systemic enquiry, as this

case was diagnosed on the basis of a leg lesion which the patient himself did not mention as he felt it was unrelated to his ocular problem, and the ophthalmologist may be the first to encounter these patients.

References

1. Akova YA, Jabbur NS, Foster CS. Ocular presentation of polyarteritis nodosa: clinical course and management with cytotoxic therapy. Ophthalmology 1993;100:1775–81.
2. Lieb ES, Restivo C, Paulus HE. Immunosuppressive and corticosteroid therapy of polyarteritis nodosa. Am J Med 1979;67:941–7.
3. Lightfoot RW Jr, Michel BA, Bloch DA, *et al.* The American College of Rheumatology 1990 criteria for the classification of polyarteritis nodosa. Arthritis Rheum 1990;33:1088–93.
4. Morgan CM, Foster CS, D'Amico DJ, Gragoudas ES. Retinal vasculitis in polyarteritis nodosa. Retina 1986;6:205–9.

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Sir,

Posterior scleritis mimicking birdshot retinochoroidopathy

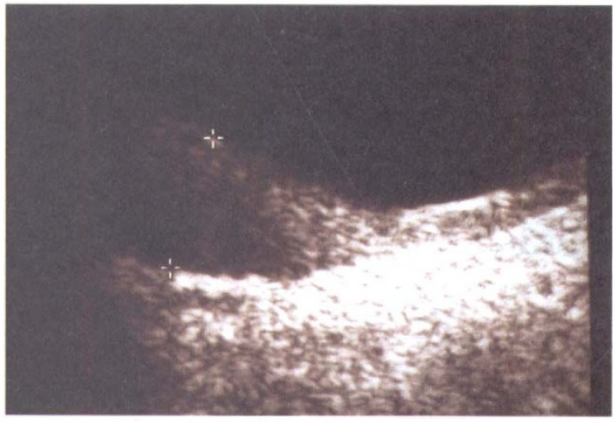
Scleritis is a severe and often painful inflammation of the connective tissue wall of the eye.[1] Scleral inflammation may be diffuse or nodular, necrotising or non-necrotising, and anterior and posterior. Posterior scleritis is less common than anterior scleritis, but since the diagnosis can be difficult to make, its precise prevalence is unknown.[2] Fundus findings that support the diagnosis of posterior scleritis include optic disc oedema, serous retinal detachment, and choroidal thickening, which can produce choroidal folds that are visible on fluorescein angiography. Less commonly, focal mass-like lesions have been described in patients with posterior scleritis, and may resemble a choroidal tumour[3] or, when anterior, a scleral buckle.[4]

We describe two patients who developed posterior scleritis associated with choroidal thickening. On resolution, both patients developed multiple, de-pigmented choroidal spots resembling birdshot chorioretinopathy.

Case reports

Case 1. A 49-year-old Caucasian woman presented with pain and decreased vision affecting her left eye. Prior ocular and medical histories were unremarkable. Best-corrected visual acuity was 6/6 on the right and 6/24 on the left. Examination of the right eye was entirely unremarkable. Anterior segment examination on the left revealed diffuse scleral injection, and mild anterior chamber and anterior vitreous inflammation. Posterior segment examination on the left showed cystoid macular oedema. Serological testing revealed a normal complete blood cell count, a negative FTA-ABS, and negative anti-nuclear antibody, rheumatoid factor and anti-neutrophil cytoplasmic antibody titres. A chest radiograph was also negative. The patient was treated with topical and periocular corticosteroids and showed marked improvement.

Over the following 5 years, the patient had recurrent episodes of anterior and posterior scleritis that became increasingly difficult to control with topical, periocular and oral corticosteroids. A particularly severe episode of posterior scleritis was associated with marked thickening of the superonasal choroid. Ultrasonography measured the posterior fundus wall thickening at 5.0 mm (Fig. 1a). Histological examination of a trans-scleral choroidal biopsy showed numerous macrophages and



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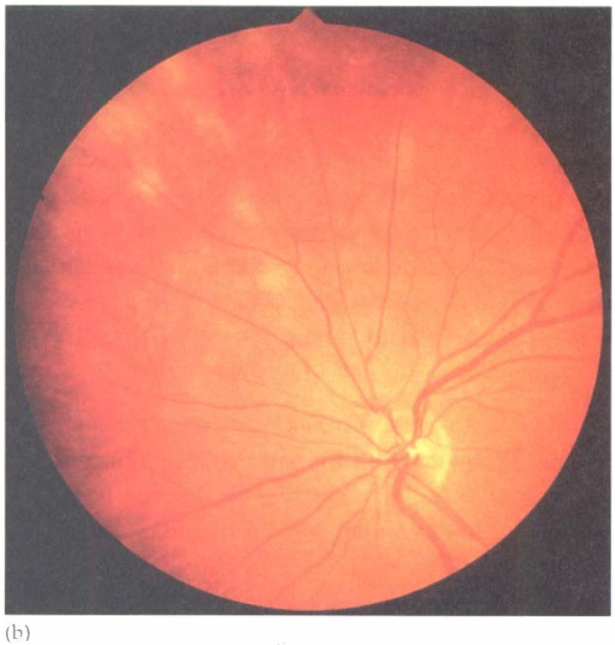

(b)

Fig. 1. *Case 1. (a) B-scan ultrasonography reveals thickening of the superonasal posterior fundus wall measuring 5.0 mm (crossmarks). (b) Colour fundus photograph taken months after the resolution of the patient's posterior scleritis shows multiple, depigmented choroidal lesions mimicking birdshot retinochoroiditis.*