

Correspondence: R Ritch, New York Eye and Ear Infirmary, 310 East 14th Street, New York, NY 10003, USA

Tel: +1 212 673 5140;

Fax: +1 212 420 8743.

E-mail: ritchmd@earthlink.net

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Sir,
Scleral granuloma associated with presumed diffuse immune lymphocytosis syndrome

Diffuse infiltrative lymphocytosis syndrome (DILS) is a rare manifestation of human immunodeficiency virus (HIV) infection. It is characterised by HIV seropositivity, a CD8⁺ lymphocytosis, visceral gland infiltration with CD8⁺ lymphocytes, bilateral parotid gland swelling, xerostomia, and xerophthalmia.¹ Peripheral neuropathy, interstitial pneumonitis and a granulomatous panuveitis have also been described in association with this condition.² We present a case of scleral granuloma occurring in the presence of presumed DILS.

Case report

A 10-year-old girl of Congolese origin, who was HIV positive, gave a 1-month history of a red painful right eye. Examination revealed a 15 mm × 15 mm dark scleral nodule above the cornea which was associated with locally injected scleral feeding vessels, as shown in Figure 1. Anterior segments were otherwise unremarkable but dilated funduscopy revealed a bilateral periphlebitis, with no haemorrhage or neovascularisation.

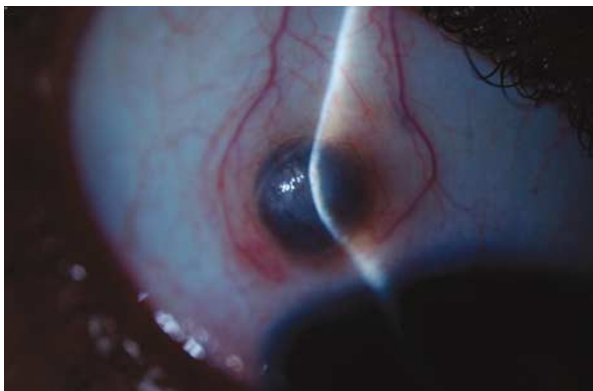


Figure 1 Scleral nodule with surrounding vascularisation.

Systemic examination showed her to be of short stature and to have tinea capitis and tinea unguis, cervical and axillary lymphadenopathy, and bilateral parotid gland enlargement. Serologic analysis revealed absolute CD4⁺ cell count of $0.38 \times 10^9/l$ (normal range, $0.30\text{--}2.0 \times 10^9/l$), absolute CD8⁺ cell count of $1.04 \times 10^9/l$ (normal range, $0.30\text{--}1.80 \times 10^9/l$), and peripheral blood film CD8⁺ positive percentage raised at 47% (normal range, 19–34%). Full blood count was otherwise normal but C-reactive protein was raised at 26 mg/l. Chest radiography findings were consistent with lymphocytic interstitial pneumonitis. Serology for syphilis, cytomegalovirus, and hepatitis B and C showed neither active disease nor previous infection. Measles serology indicated that she had had previous exposure only. Serum angiotensin-converting enzyme (ACE) level was normal.

She was commenced on a course of topical dexamethasone and highly active antiretroviral therapy (HAART) at the following doses: abacavir 450 mg o.d., lamivudine 200 mg o.d., and efavirenz 300 mg o.d. Over 4 months, the scleral granuloma became smaller, less locally injected and the eye comfortable. Subsequent CD8⁺ positive percentage improved sequentially to 37%.

Comment

A presumptive diagnosis of DILS is likely given this patient's HIV seropositivity, relative CD8⁺ lymphocytosis (although this could be due to a relatively lower CD4⁺), bilateral parotid gland enlargement, and normal ACE. Bilateral panuveitis, retinal periphlebitis, and lacrimal gland involvement are recognised ocular associations in DILS,^{2–4} however, a literature search shows no previous report of scleral granuloma occurring in the presence of this syndrome. We appreciate that there was no absolute lymphocytosis, only a reversal of the CD4⁺:CD8⁺ ratio. All cases reviewed in the existing literature have shown absolute lymphocytosis, with numbers much higher than those seen in our patient.

It may be argued that positive tissue diagnosis was required to confirm DILS to show local CD8⁺ infiltration and to exclude granulomatous pathology, especially given the presence of retinal periphlebitis. However, a negative Mantoux test and a normal chest radiograph and serum ACE excluded both tuberculosis and sarcoidosis. Furthermore, it was deemed inappropriate to subject a 10-year-old girl with resolving parotid swelling to a tissue biopsy. Although typing was not performed in our patient, HLA DR11 has been shown to have a higher association with this condition.⁵

Although a diagnosis of scleral staphyloma with uveal prolapse was considered, in which case ultrasonic biomicroscopy would have been useful, we thought it

was more likely that our patient had developed a scleral granuloma as a result of local CD8⁺ infiltration and that this regressed with commencement of HAART and topical steroids.

Scleral granulomas have been found in association with different systemic conditions.^{6–10} To our knowledge this is the first report of scleral granuloma occurring in the presence of presumed DILS. During HIV infection the immune system becomes dysfunctional because of the coexistence of immunodeficiency and immune hyperactivity, and a dysregulated production or activity of cytokines, thereby explaining the development of the DILS. Clinicians need to be aware of this entity, which is gaining in significance.

References

- 1 Nopachai A, Garwacki CP, Moll S. Diffuse infiltrative lymphocytosis syndrome. *Am J Hematol* 2004; **75**(3): 173–175.
- 2 Henderson HW, Davidson F, Mitchell SM. Intraocular inflammation and the diffuse infiltrative lymphocytosis syndrome. *Am J Ophthalmol* 1998; **126**(3): 462–464.
- 3 De Silva DJ, Obi AA, Mitchell SM. Bilateral panuveitis in HIV-1-infected patients with CD8 lymphocytosis. *Ocul Immunol Inflamm* 2005; **13**(4): 311–316.
- 4 Kestelyn P, Lepage P, Karita E, Van de Perre P. Ocular manifestations of infection with the human immunodeficiency virus in an African pediatric population. *Ocul Immunol Inflamm* 2000; **8**(4): 263–273.
- 5 Schuval SJ, O'Reilly ME, Bonagura VR. Increased frequency of HLA-DR11 in pediatric human immunodeficiency virus-associated parotid gland enlargement. *Clin Diagn Lab Immunol* 1997; **4**(3): 258–260.
- 6 Saito W, Kotake S, Ohno S. A patient with sarcoidosis diagnosed by a biopsy of scleral nodules. *Graefes Arch Clin Exp Ophthalmol* 2005; **243**(4): 374–376.
- 7 Gopal L, Rao SK, Biswas J, Madhavan HN, Agarwal S. Tuberculous granuloma managed by full thickness eye wall resection. *Am J Ophthalmol* 2003; **135**(1): 93–94.
- 8 Janknecht P, Mittelviehhaus H, Löffler KU. Sclerochoroidal granuloma in Wegener's granulomatosis simulating a uveal melanoma. *Retina* 1995; **15**(2): 150–153.
- 9 Kielar RA. Exudative retinal detachment and scleritis in polyarteritis. *Am J Ophthalmol* 1976; **82**(5): 694–698.
- 10 Hatsuda TA, Tanaka J. Bilateral necrotizing scleritis. *Am J Ophthalmol* 1978; **86**(5): 710–714.

N Malik¹, SA Aslam¹, S Mitchell¹ and S Walters²

¹Department of Ophthalmology, Chelsea and Westminster Hospital, Imperial College, London, UK

²Department of Pediatrics, St Mary's Hospital Medical School, Imperial College, London, UK

Correspondence: SA Aslam, Eye Department, Chelsea and Westminster Hospital, 369 Fulham Road, London, SW10 9NH, UK

Tel: + 44 20 8746 8000;
Fax: + 44 (0) 2087468846.
E-mail: sheraslam2@yahoo.com

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Sir, Dengue retinopathy manifesting with bilateral vasculitis and macular oedema

Ocular involvement is uncommon in dengue fever, most commonly presenting as blot haemorrhages.^{1–4} We report an unusual manifestation of dengue fever presenting bilaterally with extensive panretinal vasculitis and severe macular oedema, which resolved spontaneously without anti-inflammatory treatment.

Case report

A 37-year-old female with dengue fever diagnosed on dengue immunoglobulin (Ig)M and IgG serology and polymerase chain reaction experienced sudden, bilateral blurring of vision with metamorphopsia 6 days after the onset of fever. Her platelet count was at its nadir ($26 \times 10^9/l$). Visual acuity was 20/400 OD and counting fingers OS. There was bilateral extensive retinal vasculitis at the maculae extending outwards to the peripheral retinae, complicated by macular branch vein occlusion OD. Numerous yellowish retinal infiltrates and blot haemorrhages were scattered throughout both maculae, and there was bilateral macular oedema (Figure 1). Fundus fluorescein angiogram (FFA) demonstrated leakage indicative of extensive panretinal vasculitis, more severe at, but not limited to the maculae (Figure 1). Optical coherence tomography (OCT) showed central foveal thickening of 503 μ m OD and 843 μ m OS.

After platelet transfusion, her platelet count improved to $67 \times 10^9/l$ and gradually returned to normal levels over the next few days. Treatment with anti-inflammatory agents for her posterior retinitis and vasculitis was considered but withheld in consultation with her physician in view of her pyrexia state.

By 2 weeks, the retinal perivascular sheathing had spontaneously resolved, macular oedema had decreased, and visual acuity improved to 20/200 bilaterally. By 10 weeks, without any specific treatment, her vision had recovered to 20/20 bilaterally with no residual inflammation or macular oedema. However, there was