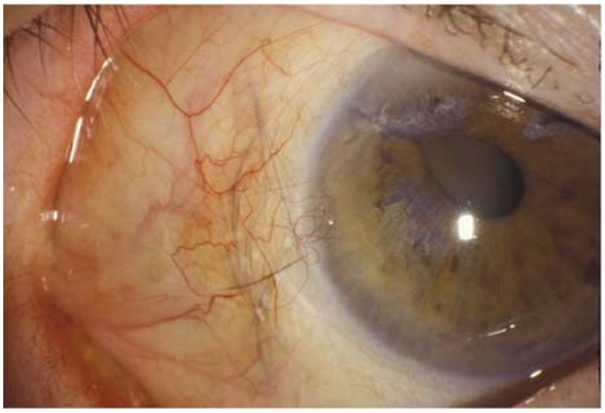

Figure 1 Subconjunctival cilia near temporal limbus of right eye.

right eye. Each time, multiple subconjunctival lashes in the temporal aspect were epilated.

In October 2004, the patient underwent conjunctivoplasty with *en bloc* removal of the lashes and cautery to the underlying sclera. So far, there has been successful resolution of symptoms.

Comment

Ectopic cilia are rarely encountered. Some reports describe congenital lash tufts in the temporal aspects of upper eyelids.[4,5] Eyelashes have also been reported emerging from the iris *de novo* and following trauma.[6] The aetiology of the former remains uncertain, posteriorly located dermoids or teratomas have been postulated. In the latter case, displacement of lash follicles is felt to be causative.[6]

The few reports of subconjunctival cilia mainly concern single lashes[1,7,8] and include granuloma formation secondary to conjunctival embedding of a cilium[7] and dermolipomas allowing lash ingress to the conjunctiva.[1] To the authors' knowledge, there is only one other published report of ectopic cilia in the setting of previous intraocular surgery, that patient having had retinal surgery, pterygium removal, and cataract extraction.[3] In our case, given that the ectopic cilia were first observed 1 year following surgery, it is likely that displacement occurred perioperatively. The mechanism may have been a cumulative effect of the fall suffered by the patient preoperatively, and surgery. Following traumatic displacement of lash tissue to the conjunctiva, the peritomy folds gave recess for any dislodged follicles. The yearly recurrence contrasts with trichitic lash regrowth. This unusual case illustrates the need for such patients to be reviewed for recurrence necessitating surgical intervention.

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Sir,

Occurrence and reactivation of cytomegalovirus retinitis in systemic lupus erythematosus with normal CD4 counts

Cytomegalovirus retinitis is the most common opportunistic ocular infection in patients with acquired immune deficiency syndrome (AIDS), accounting for up to 30–40% of all ocular manifestations.[1] CMV retinitis is also known to occur in patients with rheumatic disease, postorgan transplant and leukaemia on immunosuppressive therapy. A strong risk factor for the development of CMV retinitis is a $CD4^+$ T-lymphocyte count of <50 cells/μl.[2] With counts greater than 100 cells/μl, reactivation or occurrence of this disease is unusual.[2,3] We report two cases of CMV retinitis in patients with systemic lupus erythematosus (SLE)

undergoing immunotherapy despite normalised $CD4^+$ T-lymphocyte counts.

Case report 1

A 47-year-old Chinese lady with a 12-year history of SLE developed active diffuse lupus nephritis and was started on mycophenolate mofetil (CellCept®, Roche, NJ, USA) 500 mg b.i.d. with dose increase to 750 mg b.i.d. after 3 months. She complained of deterioration in vision of her right eye 1 month later. Visual acuity was 20/40 in the affected eye and 20/20 in the fellow eye. Anterior segment was normal. No relative afferent pupillary defect (RAPD) was elicited. Fundal exam of the affected eye showed retinal necrosis with flame haemorrhages over the central macula region consistent with clinical zone 1 CMV retinitis (Figure 1a). Blood investigations revealed a low absolute $CD4^+$ T-lymphocyte count of 53 cells/μl (normal 280–1430 cells/μl) with a CD4 : CD8 ratio of 0.33 (normal 0.50–2.50). HIV status was negative. Mycophenolate was withdrawn and replaced with low-dose prednisolone and hydroxychloroquine. Intravitreal ganciclovir therapy was started: 2 mg/0.04 ml twice weekly for the first month (induction) followed by 2 mg/0.04 ml weekly for the second month (high-dose maintenance) and thereafter 1 mg/0.02 ml weekly intermediate-dose maintenance therapy. CMV retinitis regressed and visual acuity improved to 20/30. Intravitreal treatment was discontinued when her CD4 count increased to 394 cells/μl after 6 months. Despite a normal CD4 cell count, reactivation of CMV retinitis occurred 1 month later at the border of previous CMV retinitis involving the superior edge of the macula (Figure 1b) and reinduction with intravitreal ganciclovir was started. Progression was halted, but her vision dropped to 20/120.

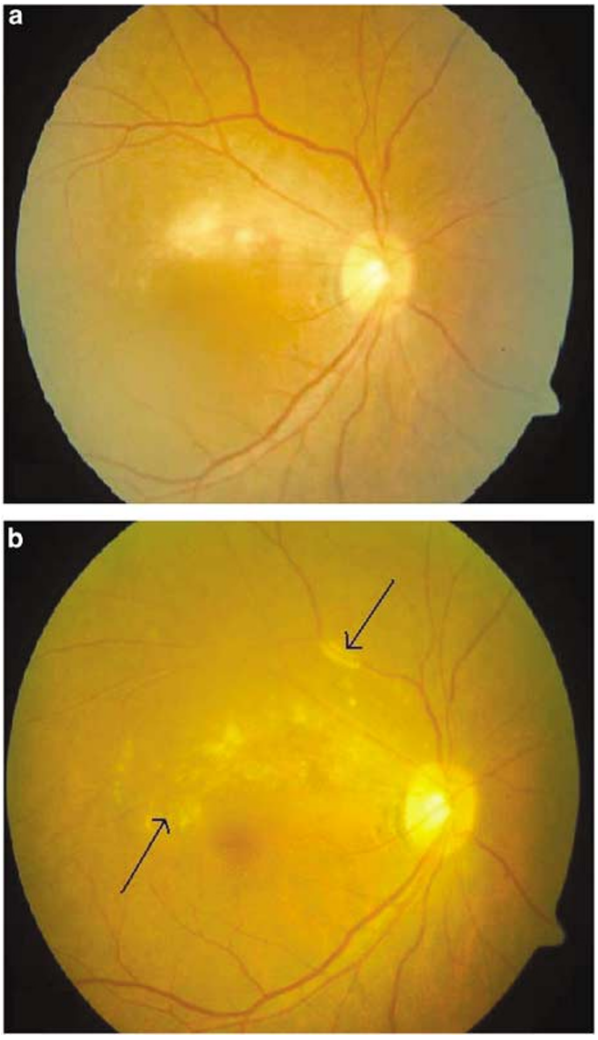

Figure 1 (a) CMV retinitis occurring at the macular area. (b) Reactivation of CMV retinitis along previous borders extending towards the superior edge of the fovea. Arrows point to sites of recurrence.

Case report 2

A 32-year-old Malay woman with a 4-year history of SLE and secondary antiphospholipid syndrome (APS) was treated with oral prednisolone and hydroxychloroquine since 1998. She was however noncompliant to medication and follow-ups, resulting in multiple episodes of relapses. She developed neuropsychiatric lupus in September 2002 that required treatment with intravenous pulse methylprednisolone and cyclophosphamide, followed by long-term high-dose oral prednisolone (60 mg o.d.). After 3 months, she presented with acute blurring of vision in right eye over 3 days with a visual acuity of hand movement with RAPD.

Anterior segment exam and intraocular pressures were normal with no evidence of rubeosis iridis. Fundal exam revealed extensive retinal haemorrhages with thrombosed and attenuated retinal arteries and veins consistent with vaso-oclusive disease. The nasal retina also had granular retinal infiltrates and necrosis with adjacent retinal haemorrhage advancing centrally consistent with the clinical diagnosis of zone 2 CMV retinitis (Figure 2). No vitritis was seen. The left eye was normal with a visual acuity of 20/20.

Investigations showed CD4 count of 586 ells/μl. HIV serology was negative. CMV serology was *positive* for IgG, but nonspecific for IgM. She was restarted on anticoagulation. Oral prednisolone was reduced to 18 mg o.d., and hydroxychloroquine 300 mg o.d. Prior to institution of local anti-CMV treatment, fundal examination showed mild resolution of retinal infiltrates associated with overlying vitritis and mild anterior uveitis. A decision was made to withhold local anti-CMV

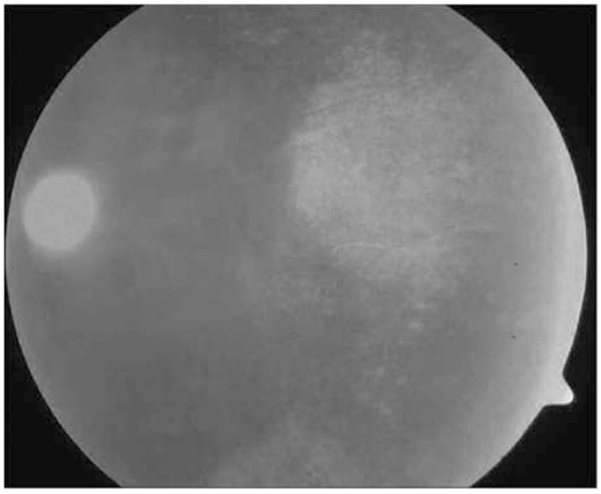

Figure 2 Retinal infiltrates and necrosis consistent with zone 2 CMV retinitis. Extensive haemorrhage with severe vaso-occlusive disease also noted.

treatment, as there was clinical evidence of immune recovery uveitis documented on serial fundal examinations and 9-view digital photography. Over the next 3 months, there was spontaneous resolution of CMV retinitis and uveitis responded to topical prednisolone acetate 1%. However, visual acuity remained at hand movements only.

Discussion

We have illustrated two SLE patients who developed CMV retinitis while on heavy immunosuppressive therapy. There have been few previous cases reported in the literature.[4–6] CMV retinitis has also been described in three cases associated with Wegener's granulomatosis,[7] rheumatoid arthritis[8] and Behcet's disease.[9] All were under chronic immunosuppression with cytotoxic drugs such as azathioprine, high-dose steroids, cyclophosphamide and adenine arabinosine. Unlike AIDS patients, these patients typically do not manifest any clinical symptoms of systemic CMV involvement despite the ocular manifestations.

In our first patient, CMV retinitis was triggered possibly due to an impaired T-cell function with decreased counts caused by mycophenolate mofetil. This drug is well known to have a potent cytostatic effect on lymphocytes, which resulted initially in a significant decrease in her CD4 count.[10] Interestingly, reactivation of CMV retinitis still occurred despite recovery of CD4 counts 6 months *after* withdrawal of mycophenolate. The occurrence of CMV retinitis associated with high CD4 count was also seen in our second SLE patient who was on chronic high-dose steroid therapy for treatment of her complications and multiple flare-ups. These suggest that occurrence and reactivation can still occur despite a high absolute CD4 count possibly due to disruption of functional integrity of the immune system with depression of leucocytic and complement function by chronic cytotoxic drugs and possibly due to an underlying dysfunction in the autoimmune system from the disease itself.[11] This is unlike AIDS where CD4 count is commonly used as an indicator of immune status that predicts the risk of opportunistic infections, for example, CMV retinitis. In SLE patients complicated by CMV retinitis, absolute $CD4^+$ T-lymphocyte count may not be a good marker for cessation of specific anti-CMV therapy and they may need a longer duration of treatment till stabilization of their underlying systemic condition to prevent reactivation. However, with gradual tapering of immune-suppressants and recovery of their immune system, we observe that it is possible to mount an immune response as demonstrated by the immune-recovery uveitis associated with resolution of the CMV retinitis in our second patient.

It was also noted from the literature review that all the SLE patients who developed CMV retinitis, including our first patient, had lupus nephritis requiring chronic immunotherapy. It is likely that these patients may have undergone more intensive or prolonged immunosuppressive therapy that further impaired their immune function, thereby rendering them more susceptible to the condition. Margo and Arango also suggested that SLE patients with secondary APS develop CMV retinitis in ischaemic retina due to widespread microvascular disease aggravated by antiphospholipid antibodies.[4] The presence of APS and severity of lupus nephritis may be indirect risk factors in SLE patients for CMV retinitis. However, larger epidemiological studies will be required to confirm these postulations.

Conclusion

Our two case reports illustrate that CMV retinitis can occur in SLE patients who are severely immunosuppressed due to chronic high-dose immunosuppressants or medications with potent cytostatic effects such as mycophenolate. Treatment options may include immunotherapy dose reduction or substitution, possibly combined with specific anti-CMV treatment. The presence of lupus nephritis and secondary APS may be risk factors for developing CMV retinitis, as it is often associated with intensive immunosuppression. Owing to the impaired immune system, the absolute $CD4^+$ T-lymphocyte count may not be a reliable indicator of the immune status and function in an SLE patient. As such, we recommend that heavily immunosuppressed SLE patients who develop sudden visual symptoms should be evaluated early for CMV

retinitis. For patients who have developed CMV retinitis, monitoring of treatment protocols should be based on the patient's clinical systemic condition with less reliance on blood results and CD4 counts.

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Sir,

Silicone oil migration causing increasing proptosis 13 years after retinal surgery

Silicone oil is necessary for endo-tamponade in selected cases of retinal detachment. Oil granuloma is a recognised condition in which there is a granulomatous response to mineral oils in the body tissues. In the past, this has been a well-documented complication of breast augmentation surgery. We report a case of silicone oil leaking into the periorbital and retro-orbital tissues, causing increasing proptosis and red eye many years after retinal surgery.

Case report

A 54-year-old Caucasian male presented with a 6-month history of a red, 'unsightly', more prominent right eye. He had suffered trauma to this eye 40 years previously and developed cataract and retinal detachment 13 years ago. He underwent a lensectomy and vitrectomy with silicone oil insertion at that time. The eye had been blind since this surgery and he was lost to follow-up until this recent complication. He was a non-smoker and had no other medical history and was on no medication. On examination, the eye was divergent and proptosed with a large subconjunctival gelatinous mass medially and an opaque, vascularised cornea (Figure 1). There was no regional lymphadenopathy. A CT scan

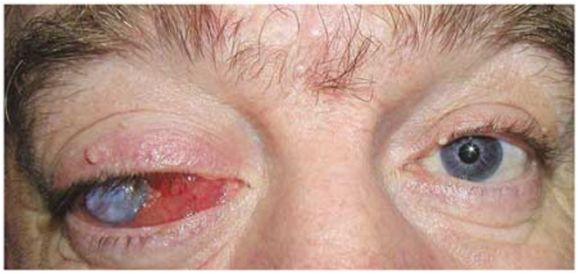

Figure 1 Clinical photograph showing gelatinous subconjunctival mass medially in the right eye.