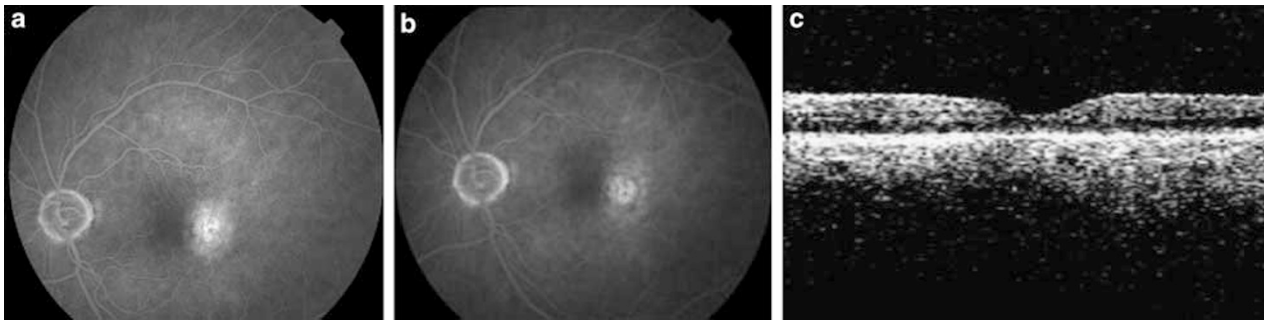

Figure 2 Midphase fluorescein angiography before any treatment, showing hyperfluorescence with leakage temporal to the fovea corresponding to the area of the parafoveal telangiectasis complicated with choroidal neovascular membrane (CNV). (b) Midphase fluorescein angiography after three intravitreal sessions of 1.25 mg of bevacizumab, showing hyperfluorescence but no leakage anymore of the former CNV. (c) Corresponding optical coherence tomography of the foveal area after treatment, showing a foveal depression without macular oedema.

Figure 1 (a) Fundus examination showing microaneurysmal and saccular dilation of the parafoveal capillaries, temporal to the fovea, and involving an area of less than one disc diameter, typical for bilateral acquired parafoveal retinal telangiectasis. Retinal oedema with two small areas of subretinal haemorrhages is associated in the area superiorly to the telangiectasis. (b) Midphase fluorescein angiography showing dilated ectatic perifoveal capillaries, and a distinct early hyperfluorescence with diffusion above this, designating the association of a CNV. Blocked fluorescence superiorly corresponds to the areas of subretinal haemorrhages owing to the CNV. (c) After one session of photodynamic therapy with Visudyne and two intravitreal injections with bevacizumab, fundus examination shows regression of the retinal oedema and of the subretinal haemorrhages superiorly. (d) Midphase fluorescein angiography still showing the dilated perifoveal capillaries, but regression of leakage of the CNV superiorly to it. For figure refer page 1433.

eye (20/30). FA showed a CNV with a pigment epithelium detachment temporal of the fovea. After 3 monthly intravitreal injections with 1.25 mg of bevacizumab, the CNV had regressed on FA, oedema had regressed on optical coherence tomography (Figure 2), and VA improved to 20/25.

Comment

Bilateral acquired juxtafoveal retinal telangiectasis is occasionally complicated with CNV, mostly localized subfoveally.[2] VEGF is a major key player for angiogenesis in the eye[3] in several physiologic and pathologic conditions. Concerning anti-VEGF therapy in ocular disease, clinical trials with pegaptanib sodium and ranibizumab show a beneficial effect for all subtypes of CNV secondary to age-related macular degeneration and in pathological myopia. Preliminary results suggest that intravitreal delivery of anti-VEGF therapy combined with or without photodynamic therapy[4] should be considered as one of the treatment options for CNV secondary to bilateral idiopathic acquired juxtafoveal telangiectasis, but long-term follow-up and larger series are needed.

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Sir,

Successful biologic treatment of ocular mucous membrane pemphigoid with anti-TNF-α

Ocular mucous membrane pemphigoid (MMP) is a rare and challenging condition due to lack of a comprehensive therapeutic evidence base. We describe successful treatment of severe ocular MMP with anti-tumour necrosis factor-α (TNF-α).

Case report

A 55-year-old gentleman, with transitional-cell bladder cancer 8 years previously, presented with bilateral

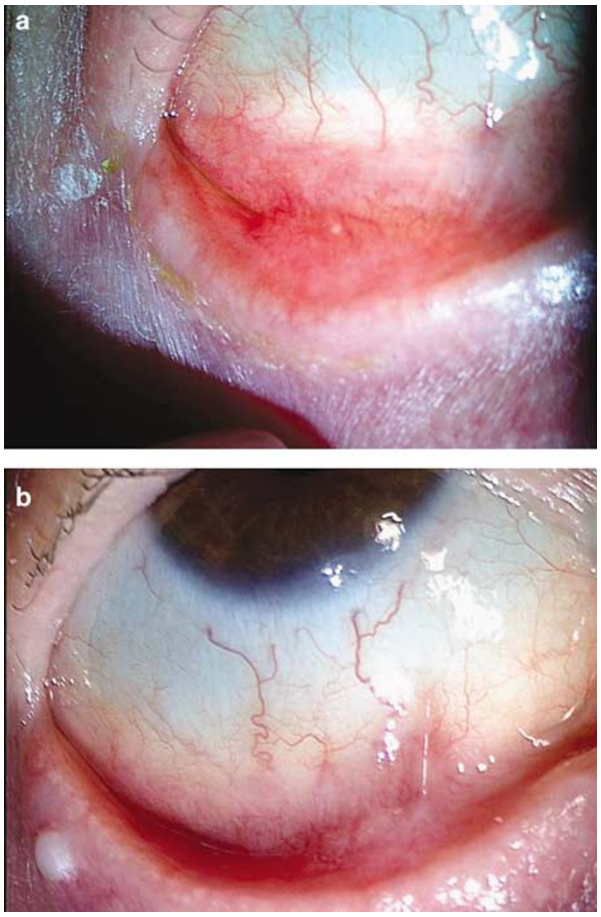

Figure 1 (a) Active ocular MMP. (b) Resolving disease activity 8 weeks after commencing anti-TNF-α treatment.

painful, red, and watery eyes. Examination revealed bilateral inferior fornix symblepharon with active subconjunctival vascular leading edge and progressive loss of inferior fornices. Conjunctival biopsy showed lymphocytes, plasma cells, and leukocytes infiltration with goblet cell depletion consistent with MMP. Immunofluorescence studies were not performed. Cyclosporine eye drops were given for lid margin inflammation. Systemic treatment commenced with dapsone, at which point there was 25% loss of fornix depth. Despite courses of 20–40 mg oral prednisolone with azathioprine and then mycophenalate mofetil, the acute flare continued. Loss of fornices progressed to 50% (Figure 1a). Cyclophosphamide was contraindicated (its metabolite acrolein causes haemorrhagic cystitis and increased risk of bladder cancer). Etanercept anti-TNF-α was commenced (50 mg weekly self-administered subcutaneously). Both eyes responded allowing immediate decrease of the 20 mg prednisolone (Figure 1b). He remains on long-term etanercept; the prednisolone dose decreasing by 1 mg/month. Eighteen months later there have been no flares.

Comment

MMP is a heterogeneous group of autoimmune, inflammatory, subepithelial blistering diseases affecting mucous membranes that heal with scarring.[1] Ocular MMP presents with conjunctival inflammation, symblepharon, and (in one-third of patients) visual impairment. The visual prognosis is poor; corneal scarring and keratinisation may cause blindness.[1,2]

Mild disease is observed. Moderate disease may be suppressed by dapsone.[2] More extensive and progressive scarring disease requires systemic therapy (prednisolone and immunosuppressants). There is evidence for azathioprine, methotrexate, leflunomide, mycophenolate mofetil, and intravenous immunoglobulin.[1,2] However, for severe/rapidly progressive disease (particularly eye or larynx where inadequately treated disease may be devastating), cyclophosphamide with corticosteroids is recommended as first-line treatment.[1,2]

Advances in other autoimmune inflammatory conditions have shown biologic treatments to be effective. A systematic review of biologic therapies for inflammatory eye disease highlighted dramatic remissions in ocular inflammation.[3] Most literature relates to uveitis; inflammatory eye diseases are heterogeneous and research is required to establish which subtypes are responsive.[3] Research in MMP characterising its autoantibody profile supports the potential use of monoclonal antibodies. There are reports describing the successful use of anti-TNF-α therapies[4,5] and daclizumab (reviewed in Lim *et al*[3]).

With rare conditions where treatment is challenging, collating reports of success with novel biological agents is important. We support the development of a patient database[1] for longitudinal follow-up to monitor efficacy and side effects of such treatments.

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