Wegener’s granulomatosis (WG) with inflammation involving his kidneys, nose and lungs with positive c-ANCA (EIA) and negative workup for ANCA-associated systemic vasculitis (Wegener’s granulomatosis, microscopic polyangiitis, polyarteritis nodosa). Indirect immunofluorescence for both IgG and IgA were also negative, excluding MMF.

Discussion

Our case demonstrates the heterogeneity in clinical expression of WG. Conjunctival disease has been reported in 4–16% of patients with ocular manifestations [1–4]. This can vary from conjunctival hyperaemia to granulomatous lesions, tarsal conjunctival necrosis, active fibrovascular proliferation or inactive scar tissue. Cicatricial conjunctivitis in WG is extremely rare [4]. An important differential diagnosis is MMP, an autoimmune disease whose target antigen is the β2 peptide of the α6β1-integrin of the basement membrane zone of conjunctiva and epidermis [5]. The conjunctival autoantigen in WG is currently unknown.

Chronic inflammation and fibrosis of the conjunctiva can induce dry eyes via occlusion of the ducts of the lacrimal and accessory glands, eyelid and eye lash abnormalities (entropion, lagophthalmos, trichiasis and dystrichiasis). The aetiology of fibrosis is unknown but these changes can lead to corneal scarring, infection, perforation and loss of vision. The location of conjunctival disease predominantly at the eyelid borders may further the understanding of the pathogenesis of WG. The eyelids are supplied by terminal branches of the marginal and peripheral arcade vessels [6]. An occlusive vasculitis of these peripheral vessels, branches or both may lead to ischaemia or infarction [4].

Physicians should be aware that ‘conjunctivitis’ may represent serious eye involvement from WG and liaison with an ophthalmologist is desirable. Mucous membrane pemphigoid is an important differential diagnosis and should be excluded in all cases. A significant association exists between conjunctival disease and subglottic stenosis [4]. Subglottic stenosis can rapidly progress, leading to laryngeal obstruction and respiratory failure [7]. Our patient continues to be closely monitored and is currently stable.

Conflict of interest statement. None declared.


References

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The significance of cicatricial conjunctivitis in Wegener’s granulomatosis

Sir,

Wegener’s granulomatosis exhibits large heterogeneity in clinical expression. The spectrum of ocular disease can occur at any stage in its natural history. Cicatricial conjunctivitis, a fibrotic conjunctival scarring response is rare and associated with subglottic stenosis.

Case

A 70-year-old man was referred to the ophthalmology department with symptoms of ocular discomfort and abnormal conjunctival appearances. He had no previous ophthalmic history. He was recently clinically diagnosed with Wegener’s granulomatosis (WG) with inflammation involving his kidneys, nose and lungs with positive c-ANCA (EIA) levels of 7.6 units (<2.0). Nasal mucosa biopsy revealed mixed inflammatory infiltrate. He was too ill to undergo a renal biopsy.

Ocular examination revealed bilateral sub-tarsal conjunctival fibrosis with symblephara appearing as vertical folds between bulbar and palpebral conjunctiva (Figure 1) representative of conjunctival cicatrization. Conjunctival biopsy demonstrated minimal inflammation with no specific C3, IgA or IgG distribution as occurs with mucous membrane pemphigoid (MMP), an important differential. Serum indirect immunofluorescence for both IgG and IgA were also negative, excluding MMP.

Discussion

Our case demonstrates the heterogeneity in clinical expression of WG. Conjunctival disease has been reported
and pro-inflammatory molecules [5]. Administration of MR tubular damage and results in an increase in albuminuria. In addition, it causes interstitial leucocyte infiltration, and glomerular sclerosis, fibrinoid necrosis and thrombosis. Damage in experimental models of hypertension. In hyper-mineralocorticoid receptor (MR) also contributes to kidney effects on the heart, it is known that activation of the treatment as well as the heart. Apart from deleterious renal disease, when the kidneys could profit from this treatment in low doses and under tight supervision and potency, but it lacks the anti-androgenic side effects. Higher doses and in the treatment of therapy-resistant hyperaldosteronism decreased from 200–400 mg in the 1970s to 12.5–50 mg daily in recent years. It has been raised as a potential deleterious side effect, particularly in patients with reduced renal function, heart failure or diabetes [13,14], but careful titration [15] and the use of lower doses of MR antagonists minimizes this risk. Therefore, there is now an obvious need for larger trials with antimineralocorticoid treatment in patients with chronic renal disease as well as end-stage renal failure.

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