

Persistent Unilateral Conjunctival Inflammation as the First Sign of Granulomatosis with Polyangiitis

Dear Editor,

Granulomatosis with polyangiitis (GPA) or, formerly known as Wegener's granulomatosis, is an autoimmune, small-to-medium sized vessel vasculitic disease that can affect multiple organs, including the eyes.¹ We report an interesting case of atypical persistent unilateral conjunctival inflammation in a patient with previously undiagnosed GPA.

An 83-year-old male presented to the eye casualty with a 4-week history of painful and injected left eye. Previous ocular history included bilateral age-related macular degeneration. Best-corrected visual acuity (BCVA) was 6/60 in the right eye and 6/12 in the left eye. Examination revealed bilateral blepharitis with left conjunctivitis associated with peripheral corneal infiltrates.

The patient was treated for left blepharokeratoconjunctivitis with oral doxycycline, topical steroids and antibiotic drops. The eye improved but there remained a localised persistent, area of fleshy, inflamed conjunctiva superonasally with abnormal-looking vasculatures encroaching onto the cornea after 3 months of treatment (Fig. 1).

A conjunctival biopsy was performed, which revealed chronic inflammatory infiltrates comprising plasma cell, lymphocytes and prominent population of eosinophils in the substantia propria. Such inflammation could be consistent with atopic or rosacea keratoconjunctivitis but an autoimmune/vasculitic screen was recommended as part of the investigation for potential underlying systemic vasculitides affecting the conjunctiva. Vasculitic screen showed low haemoglobin 11.2 g/L (normal: 13-18 g/L), raised erythrocyte sedimentation rate 49 mm/hr (normal: 2-10 mm/hr) and raised antineutrophil cytoplasmic antibodies (ANCA)-proteinase 3 antibodies 25 IU/ml (normal: <2 IU/mL), supporting the diagnosis of GPA. Rheumatoid factor, antinuclear antibodies and ANCA-myeloperoxidase antibodies were negative.

Patient was referred to rheumatology for further assessment, which revealed a recent history of epistaxis and nasal crusting, some extent of Raynaud's disease affecting the fingers and toes, and livedo reticularis affecting the legs and forearms. There was no evidence of pulmonary

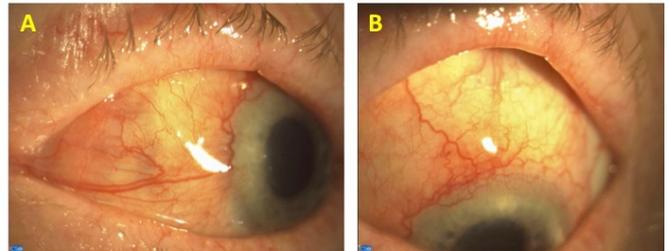


Fig. 1. A and B show patient's persistent conjunctival inflammation superonasally with abnormal-looking vasculatures encroaching onto the cornea of the left eye after 3 months of initial treatment.

or kidney involvement. A diagnosis of GPA was made and the patient started on a tapering regime of oral prednisolone and azathioprine. Patient was started on oral prednisolone 40 mg daily for 1 month, then 30 mg daily for 1 month, followed by 5 mg reduction every month thereafter until 10 mg daily. Oral azathioprine was started at 100 mg daily 1 month later after the initial commencement of oral prednisolone.

The conjunctival inflammation improved significantly within a week following the systemic steroids and completely resolved within 2 months. At 2 years follow-up, the patient had come off from oral prednisolone and remained on azathioprine 100 mg daily under the care of rheumatology with no recurrence of ocular inflammation.

Discussion

GPA is a systemic inflammatory condition that is commonly diagnosed at the age of 40 to 55 years old.² Ocular manifestations of GPA include episcleritis, scleritis, peripheral ulcerative keratitis, uveitis, retinal vasculitis, and orbital inflammation;²⁻⁴ however, non-specific conjunctivitis manifesting as the isolated finding is rare.^{3,4}

Our case highlights the importance of tissue diagnosis and appropriate screening for autoimmune disease in patients with unusual persistent conjunctival inflammation. Collaboration between ophthalmologists and rheumatologists is essential in these cases.

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