CASE REPORT

Recurrent bilateral eyelid and conjunctival granulomatosis in Churg-Strauss syndrome

Nurul Faaiqah Jainuddin, MBBS1, Aliff Irwan Cheong, MOpthal1, Chiew Seow Fan, MPpath1, Norlina Ramli, MOpthal1

1Department of Ophthalmology, Faculty of Medicine, University Malaya, Kuala Lumpur, Malaysia, 2Department of Pathology, Faculty of Medicine, University Malaya, Kuala Lumpur, Malaysia

SUMMARY
A 47-year-old woman with poorly controlled asthma and allergic rhinitis since 2004, first presented to our eye clinic in October 2017 with right eye painless upper lid swelling. She had four previous episodes of bilateral eyelid swelling associated with fornical conjunctival mass over the past 10 years, which resolved after oral high dose steroid treatment. In 2011 and 2017, biopsies were taken from her left eye conjunctival mass and histopathology was consistent with Churg-Strauss syndrome. At presentation, both eyes Snellen visual acuity were 6/9 with no relative afferent pupillary defect (RAPD). On examination, the right upper eyelid appeared swollen with a diffused, mildly injected bulbar conjunctival mass extending from the superonasal to the superotemporal area measuring 3.5mm x 2mm (Figure 1). The left eye also has a similar conjunctival mass at the superotemporal area. Extraocular muscle movements in both eyes were full, and no proptosis was present.Fundus examination of both eyes were unremarkable. The symptoms responded well to oral steroid treatment but recurred following cessation of the therapy. The patient was co-managed with a rheumatologist and the patient currently remains stable and is on oral Methotrexate and low dose oral steroids. The diagnosis of Churg-Strauss syndrome (CSS) was made following biopsy of the conjunctival mass. The symptoms responded well to oral steroid treatment but recurred following cessation of the therapy. The patient was co-managed with a rheumatologist and the patient currently remains stable and is on oral Methotrexate and low dose oral steroids. The diagnosis of CSS was made following histopathological examination of the conjunctival biopsy.

INTRODUCTION
Churg-Strauss syndrome (CSS) is also known as Eosinophilic granulomatosis with polyangiitis (EGPA). It is a rare systemic vasculitis which affects the small and medium-sized-vessels and cause fibrinoid necrosis. CSS usually involves the heart, skin, lung and gastrointestinal tract. It very rarely involves the orbital tissue. Here we describe a patient who presented with bilateral recurrent upper eyelid swelling and conjunctival mass for the past 10 years which responded well to steroid treatment. The diagnosis of CSS was made following histopathological examination of the conjunctival biopsy.

CASE REPORT
A 47-year-old woman with poorly controlled bronchial asthma and allergic rhinitis since 2004, first presented to our eye clinic in October 2017 with right eye painless upper lid swelling. She had four previous episodes of bilateral eyelid swelling associated with conjunctival mass over the past 10 years, which resolved after oral high dose steroid treatment. In 2011 and 2017, biopsies were taken from her left eye conjunctival mass and histopathology was consistent with Churg-Strauss syndrome. At presentation, both eyes Snellen visual acuity were 6/9 with no relative afferent pupillary defect (RAPD). On examination, the right upper eyelid appeared swollen with a diffused, mildly injected bulbar conjunctival mass extending from the superonasal to the superotemporal area measuring 3.5mm x 2mm (Figure 1). The left eye also has a similar conjunctival mass at the superotemporal area. Extraocular muscle movements in both eyes were full, and no proptosis was present.Fundus examination of both eyes were unremarkable. The symptoms responded well to oral steroid treatment but recurred following cessation of the therapy. The patient was co-managed with a rheumatologist and the patient currently remains stable and is on oral Methotrexate and low dose oral steroids. The diagnosis of CSS was made following histopathological examination of the conjunctival biopsy.

DISCUSSION
Churg-Strauss syndrome, eosinophilic granulomatosis with polyangiitis, is a rare systemic autoimmune vasculitis which...
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affects the small and medium-sized blood vessels. It was first described by Churg and Strauss in 1951. The criteria to diagnose CSS according to the American College of Rheumatology include asthma, eosinophilia of more than 10% in peripheral blood, paranasal sinusitis, pulmonary infiltrates, histological proof of vasculitis with extravascular eosinophils and mononeuritis multiplex or polyneuropathy. The presence of four or more criteria yields a sensitivity of 85% and a specificity of 99.7%. Our patient had four of these features.

CSS classically evolves in three stages which starts with a prodromal asthma and allergy phase, followed by peripheral blood eosinophilia and infiltration phase and finally with a vasculitis phase. This phase can occur up to 8-10 years after the asthma phase and is often associated with extravascular granulomas.

CSS usually affects the skin, heart, lung and gastrointestinal tract. Ocular involvement in CSS is unusual. It can manifest as conjunctival nodules, orbital myositis, orbital inflammatory syndrome, dacryoadenitis and cranial nerve palsy. Management of CSS requires a team effort, usually with a rheumatologist and a general physician. Primary therapy for CSS is systemic prednisolone for 6 to 12 weeks until remission and then the dose is tapered gradually. The majority of cases respond well to systemic prednisolone alone, but isolated cases may need immunosuppression to induce remission. In our case, systemic methotrexate was required to maintain remission as her symptoms would recur once the oral corticosteroids were stopped. Although ocular involvement is rare, the ophthalmologist must have high index of suspicion to diagnose CSS from its ophthalmic manifestations. The clue to this in our patient was her history of rapid response to oral steroids and then recurrence once oral steroid is stopped. If in doubt, a biopsy of the lesion is essential for diagnosis. This is crucial as untreated CSS has 50% risk of death within three months of vasculitis onset. With treatment, survival rate improves to 70% to 90% at 5 years.

In conclusion, although CSS is rarely encountered in our clinical practice, it is important to have a high index of suspicion especially in patients with systemic illness and recurrent disease. Prompt medical work up and biopsy of the lesion will help to diagnose CSS which is a potentially life-threatening illness.

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CONFLICT OF INTEREST
All authors declare no conflicts of interest.

REFERENCES