CASE REPORT

Recurrent bilateral eyelid and conjunctival granulomatosis in Churg-Strauss syndrome

Nurul Faaiqah Jainuddin, MBBS[1], Aliff Irwan Cheong, MOphthal[1], Chiew Seow Fan, MPatho[2], Norlina Ramli, MOphthal[1]

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

SUMMARY

A 47-year-old woman with poorly controlled asthma and allergic rhinitis presented with recurrent episodes of bilateral upper eyelid swelling associated with forniceal conjunctival mass for the past 10 years. Routine blood investigations showed raised IgE levels and raised eosinophil counts. 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. Ocular involvement in CSS is unusual but this unique presentation of CSS was successfully managed, and the patient remains in remission.

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 a 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. Our differential diagnosis at this point included conjunctival granuloma, conjunctival lymphoma or idiopathic orbital inflammatory disease.

Laboratory investigation of the blood of the patient was negative for perinuclear anti-neutrophil cytoplasmic antibody (p-ANCA), cytoplasmic anti-neutrophil cytoplasmic antibody (c-ANCA) and anti-double stranded DNA antibody (dsDNA). Titres of antinuclear antibody (ANA) and complement C3/4 were within normal range, as were the erythrocyte sedimentation rate (ESR). However, eosinophil count and IgE was elevated at two hundred four. A repeat biopsy was done under local anaesthesia for the right conjunctival mass. The histopathology report diagnosed the condition as granulomatous vasculitis. Microscopic examination showed fibrocollagenous tissue which contained multiple large granulomas with central necrobiotic collagenous core surrounded by palisade of histiocytes and macrophages, lymphocytes, multinucleated giant cells and eosinophils. The interstitium showed infiltrate of eosinophils. There was no fibrinoid necrosis (Figure 2). These histological features were compatible with that seen in CSS as she had four of the six criteria to the diagnosis of CSS.[1]

The patient was treated with oral Prednisolone 20mg OD and topical tobradex QID. The patient was co-managed with the rheumatologist. The symptoms improved, and lesion reduced in size after oral and topical steroid treatment which was slowly tapered over a 1-year period. The lesion recurred however while tapering the oral steroid after one year. Thus, oral Methrotrexate was commenced to control the inflammation and as a steroid sparing agent. The patient was last seen in February 2019 and the conjunctival mass had resolved completely. She remains stable. Her asthma was also well controlled after starting on oral steroid and methotrexate.

DISCUSSION

Churg-Strauss syndrome, eosinophilic granulomatosis with polyangiitis, is a rare systemic autoimmune vasculitis which

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Corresponding Author: Dr. Nurul Faaiqah Jainuddin
Email: nurulfaaiqah239@gmail.com

Figure 1: A, Appearance of patient at presentation with almost full mechanical ptosis on right eye. B, Right bulbar conjunctiva lesion seen on downgaze with lid held up. C, Same lesion on side view with extension to lateral forniceal conjunctiva.

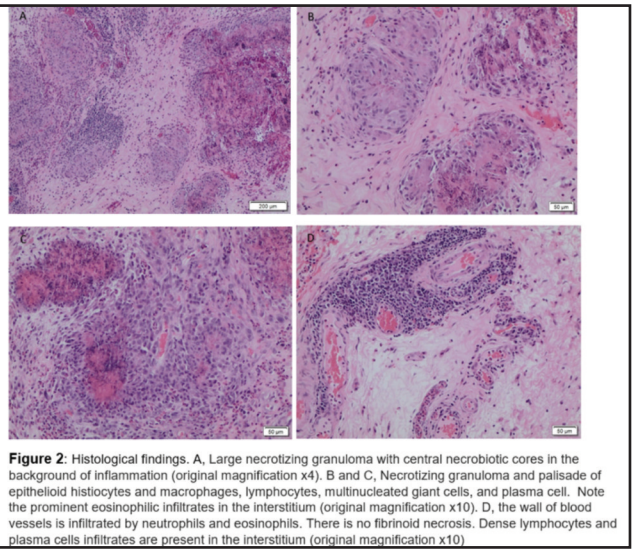

Figure 2: Histological findings. A, Large necrotizing granuloma with central necrobiotic cores in the background of inflammation (original magnification x4). B and C, Necrotizing granuloma and palisade of epithelioid histiocytes and macrophages, lymphocytes, multinucleated giant cells, and plasma cell. Note the prominent eosinophilic infiltrates in the interstitium (original magnification x10). D, the wall of blood vessels is infiltrated by neutrophils and eosinophils. There is no fibrinoid necrosis. Dense lymphocytes and plasma cells infiltrates are present in the interstitium (original magnification x10)

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.[1] The presence of four or more criteria yields a sensitivity of 85% and a specificity of 99.7%.[2] 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.[3] 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.[3-5]

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.[4]

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.

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