

Churg-Strauss Syndrome Presenting with Conjunctival and Eyelid Masses: A Case Report

Ameli F, MD, Phang K S, MBBS, DCP, Masir N

Universiti Kebangsaan Malaysia Medical Centre, Jalan Yaacob Latif, Cheras, Kuala Lumpur 56000, Malaysia

SUMMARY

Churg–Strauss syndrome, a small and medium vessel vasculitis, was first described by Churg and Strauss in 1951. It is characterised by the presence of asthma, prominent tissue and blood eosinophilia, systemic vasculitis, and pulmonary and systemic necrotising allergic granulomas. Involvement of the skin, heart and gastrointestinal tract is well documented, but ocular presentation is unusual. We describe a 40-year-old lady who presented with recurrent upper eyelid swelling due to conjunctival lesions. Although she has chronic asthma, Churg–Strauss syndrome was never suspected. The diagnosis of Churg–Strauss syndrome was only made following histological examination of the conjunctival lesions.

KEY WORDS:

Churg–Strauss syndrome, conjunctival mass, eyelid swelling

INTRODUCTION

Churg-Strauss syndrome¹ is an uncommon disease with an estimated annual incidence of 1-3 per million. The disease can occur at any age. However, the occurrence in infants is unknown. The mean age of onset is 48 years with a female to male ratio of 1.2:1². The diagnosis of Churg–Strauss syndrome is usually confirmed histologically by the presence of vasculitis and eosinophilic granulomas in a patient with characteristic clinical manifestations such as asthma, mononeuritis multiplex, allergic rhinitis and sinusitis. Nonetheless, the diagnosis can be challenging since the disease may present with a variety of signs and symptoms³. In addition other entities may also provoke vasculitis similar to that found in this disease. In Churg-Strauss syndrome, the lesions may be found in multiple organs including lung, heart, skin, gastrointestinal tract and nervous system but ocular involvement is rare⁴. This may manifest as corneal ulcer, uveoscleritis, conjunctival granuloma, orbital inflammatory pseudotumour, retinal artery occlusion or ischaemic optic neuropathy⁵. In this report, we describe a case of Churg-Strauss syndrome in a 40-year-old lady who presented with recurrent upper eyelid swelling.

CASE REPORT

A 40-year-old Malay lady was referred to the Ophthalmology Department with a three-week history of left upper eyelid swelling. This was not associated with pain or visual disturbance. She gave a history of recurrent bilateral upper eyelid swelling in the past three years which was diagnosed as

pseudotumour. These lesions were successfully treated with intravenous methylprednisolone. In addition she has a past medical history of adult onset asthma but has been quiescent for the past two years and did not require any treatment. Patient also had history of primary infertility and failed attempted in vitro fertilisation (IVF). Systemic review of other systems including cardiovascular system, gastrointestinal tract, central and peripheral nervous system, and skin did not reveal any significant history.

Physical examination of the eye revealed a raised, non-tender, soft and smooth mass on the superior aspect of the left upper eyelid and an irregular firm, lobulated superior left conjunctival mass. In addition, the conjunctiva was mildly inflamed and edematous. The right eye was normal. A finding from a full, dilated ocular examination of both eyes was unremarkable. Examination of other systems did not reveal any abnormal findings to suggest systemic involvement.

Investigations showed elevated total eosinophil count (10.3 x 10⁹) and IgE levels (345 kU/l). The erythrocyte sedimentation rate and C reactive protein were within normal limits. Results of a urine analysis were negative for blood and protein. Tests for rheumatoid factor (RF), anti nuclear antibodies (ANA), circulating anti neutrophil cytoplasmic antibody (cANCA), perinuclear anti neutrophil cytoplasmic antibody (pANCA) as well as C3 and C4 were negative. Skin tuberculin test (PPD) was negative at 72 hours. Chest X-ray and echocardiography were normal.

A magnetic resonance imaging (MRI) scan of the orbits showed normal and symmetrical globe bilaterally, with unremarkable optic nerves and rectus muscles. There was mild enhancement of the superior aspect of the left sclera with no significant proptosis. In addition there were features of left maxillary and ethmoidal sinusitis. Clinical and radiological impression was a pseudotumour or lymphoma of the left eye. The conjunctival lesions were biopsied, and they were labelled superior fornix and superotemporal bulbar conjunctival masses.

The biopsies consisted of two tiny fragments of tissue measuring 3x2mm and 2x2mm respectively. Histopathology examination of both lesions showed similar histological features i.e., there were scattered granulomas surrounded by inflammatory cells, including eosinophils, lymphocytes and plasma cells with a few multinucleated giant cells (Figure.1). The granulomas were composed of a palisading rim of macrophages and a central collection of apoptotic

This article was accepted: 11 October 2011

Corresponding Author: Noraidah Masir, Universiti Kebangsaan Malaysia Medical Centre, Pathology, Jalan Yaacob Latif, Cheras, Kuala Lumpur 56000, Malaysia Email: Noraidah@ppukm.ukm.my

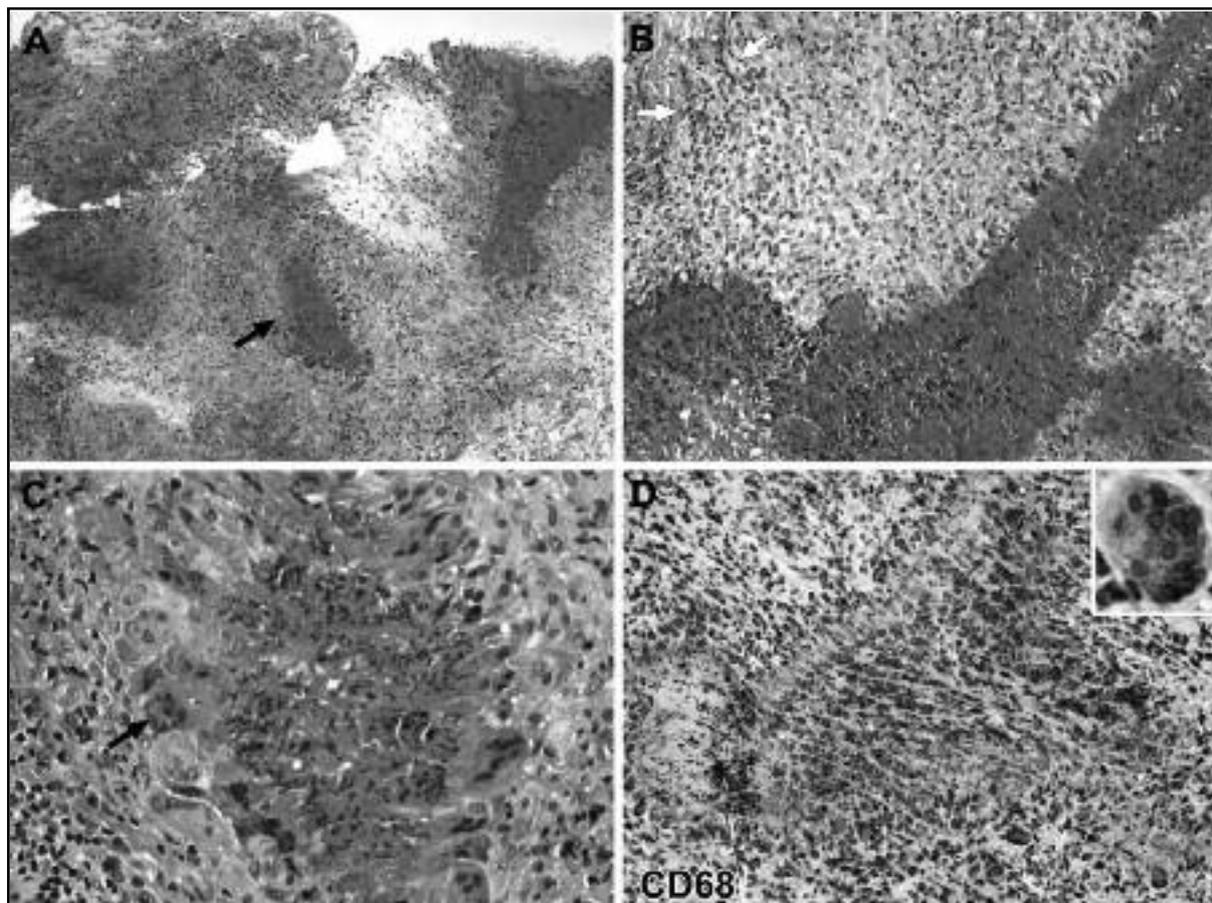


Fig. 1: A, Low-power view of the conjunctival mass revealed multiple eosinophilic granulomas with a central flare-like pattern (H & E, X10). B, This shows an eosinophilic granuloma surrounded by inflammatory infiltrates of eosinophils, lymphocytes and plasma cells (white arrow) (H & E, X20). C, The eosinophilic granuloma contains apoptotic eosinophils surrounded by palisading histiocytes and occasional multinucleate giant cells (arrow) (H & E, X40). D, Immunostaining for CD68 highlights the palisading macrophages. Inset: multinucleated giant cell positive for CD68 (IHC, X20).

eosinophils. In addition occasional areas showed angiitis with destruction of the vascular architecture and an intense inflammatory cell infiltrate. Special stain for masson trichrome revealed possible destruction of the vessel wall and Ziehl–Nielsen stain for acid fast bacilli was negative.

Based on the history of asthma, presence of peripheral blood eosinophilia, paranasal sinusitis as well as histopathological findings of eosinophilic granuloma and angiitis, a diagnosis of Churg–Strauss syndrome was made. The patient received daily dose of 1 mg/kg of prednisolone for three weeks which was later tapered to 8 mg/day. She was well after four months of follow-up with clinical and laboratory remissions.

DISCUSSION

Churg–Strauss is a rare systemic disorder and the underlying cause is unknown. It typically present with eosinophil-rich granulomatous inflammatory lesions involving mainly the respiratory tract, although other organs may also be involved.

This lesion is coupled with necrotising vasculitis affecting small- to medium-sized vessels, and is clinically associated with asthma and eosinophilia. Patients with Churg–Strauss syndrome often present with non-specific manifestations such as fever, malaise, anorexia and weight loss, characteristic of multisystemic disease. The diagnosis of Churg–Strauss syndrome can be challenging as the pathognomic features often do not occur simultaneously². There are different classification criteria for Churg–Strauss syndrome but more recently the diagnostic criteria have shifted from pathologic findings to clinical parameters.

It is recommended that to establish a diagnosis of Churg–Strauss syndrome, four of six criteria must be met i.e. (1) asthma, (2) eosinophilia greater than 10% on differential white blood cell count, (3) mononeuropathy (including multiplex) or polyneuropathy, (4) migratory or transient pulmonary infiltrates detected radiographically, (5) paranasal sinus abnormality and (6) extravascular eosinophilic infiltration in biopsy specimens⁴.

Our patient presented with four of these six criteria i.e., asthma, peripheral eosinophilia, paranasal sinusitis with eosinophilic granuloma and angiitis shown in the conjunctiva and eye lid masses. Thus a diagnosis of Churg-Strauss syndrome was made.

ANCA-positivity is seen in between 40-70% of patients with Churg-Strauss syndrome and its status subdivides the disease into two subgroups (ANCA-positive, ANCA-negative). ANCA-positivity is usually associated with small-vessel vasculitis, including necrotizing glomerulonephritis, mononeuritis and purpura, whereas ANCA-negative cases are more likely to have cardiac and lung involvement. However studies have shown that ANCA status does not influence patients survival or relapse rate⁶.

Takanashi *et al*⁵ classified the ocular manifestations into two types: orbital inflammatory pseudotumour and ischaemic vasculitis. They hypothesised that these two groups may represent the two essential characteristics of the disease processes: granulomatosis and angiitis. In their review the presence of ANCA was characteristic of the ischaemic type. They advised that this may be a risk factor for sudden visual loss and that prophylactic steroids may be advisable in these cases. Our case was of the pseudotumour type and was negative for ANCA.

The prognosis of untreated Churg-Strauss syndrome is poor, with a reported five- year survival of 25%². However for the majority of patients, glucocorticoids alone offer effective treatment with improved outcome. Myocardial involvement is the most frequent cause of death and is responsible for 39% of patient mortality². There was not cardiac involvement in our case at time of ocular manifestation.

CONCLUSION

Although Churg-Strauss syndrome is a disorder which is rarely encountered in the clinical practice, our case points to the importance of a detailed medical history and a high index of suspicion in patients with systemic illnesses who present with atypical lesions. Biopsy of the lesion and prompt medical work up may uncover an undiagnosed and potentially life threatening systemic illness.

REFERENCES

1. Churg J, Strauss L. Allergic granulomatosis, allergic angitis, and periarteritis nodosa. *Am J Pathol* 1951;27:277-301.
2. Fauci AS. *Harrison's Rheumatology* 2nd ed; 2010.
3. Rust RS and Worrell TE. Churg Strauss disease; January 20, 2010. <http://emedicine.medscape.com>
4. Weidner N, Cote R, Suster S, Weiss L. *Modern Surgical Pathology*. Vol 2. 2nd ed; 2009.
5. Takanashi T, Uchida S, Arita M, Okada M, Kashii S. Orbital inflammatory pseudotumour and ischaemic vasculitis in Churg-Strauss syndrome. *Ophthalmology* 2001; 108(6):1129-1133.
6. Sinico RA, Bottero P. Churg- Strauss Angiitis. *Best Practice Research & Clinical Rheumatology* 2009; 23:355-66.