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Case report

Successful reconstruction of an ocular defect resulting from granulomatosis with polyangiitis, following treatment with rituximab

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ABSTRACT

Purpose: To report a unique case of orbital inflammatory disease which was ultimately diagnosed as granulomatosis with polyangitis (GPA) and thus successfully treated.
Observation: A 47 year-old man presented with a rapidly progressive necrotic soft tissue mass within the medial antero-superior aspect of the right eyelid and orbit. He also had transient retinal vasculitis in the left. Serology, histology and imaging were atypical of, but consistent with, GPA. He was thus successfully treated with intravenous rituximab followed by reconstruction of the medial eyelid.
Conclusion and importance: A high index of suspicion of GPA is required in orbital inflammatory disease, especially when typical diagnostic findings are absent.

1. Introduction

Granulomatosis with polyangitis (GPA) is a systemic vasculitis which typically affects the renal and respiratory systems, demonstrates ANCA positivity in 80–90% of cases. It is typically characterised histologically by granulomatous inflammation, necrosis and vasculitis.[1] There is ocular involvement in approximately 50% of cases, but this is most often part of multisystem disease.[2] We report here a patient who presented with severe, focal, necrotic, orbital inflammatory disease and who represented a diagnostic and therapeutic challenge. Despite an atypical presentation, based on a working diagnosis of GPA, he was successfully treated with intravenous rituximab followed by eyelid reconstruction.

2. Case report

A 48 year old Caucasian male presented with a lesion at the medial portion of the right upper eyelid. Present for two months, it had arisen initially as a small white pustular lesion which progressively enlarged and became erythematous before discharging purulent material. Topical fusidic acid and subsequently chloramphenicol ointments had no effect on the lesion. It increased dramatically in size following an attempt at incision and curettage at another hospital.

This man was otherwise well and denied symptoms of fatigue, fever, night sweats, weight loss, arthralgia, haemoptysis, rash, or haematuria.

He had had a cholecystectomy and the family history was unremarkable. His career and hobbies provided no unusual exposures to fungi or other infections.

On examination, the right eyelid was erythematous and edematous with a palpable mass in the superomedial aspect of the right orbit. Snellen visual acuity was 6/9 in the right eye and 6/6 in the left. The movements of his right eye were limited on attempted elevation.

C-reactive protein (CRP) was 36 mg/L (normal < 7) and erythrocyte sedimentation rate (ESR) 25 mm/hr (normal 0–20). No other immunological or biochemical abnormalities were noted. In particular, serum IgG4, rheumatoid factor, perinuclear and cytoplasmic anti-neutrophil cytoplasmic antibody (p- and c-ANCA), anti-nuclear antibody (ANA), extractible nuclear antigens (ENA), and Quantiferon gamma were all negative. Complement levels, thyroid function tests and angiotensin converting enzyme were normal. No micro-organisms were visualized at microscopy of, or cultured from, a sample of the discharge from the lesion.

Magnetic resonance imaging (MRI) orbits revealed a soft tissue enhancing mass within the medial anterosuperior aspect of the right orbit with resultant mild deformity and downward displacement of the right globe (Fig. 1).

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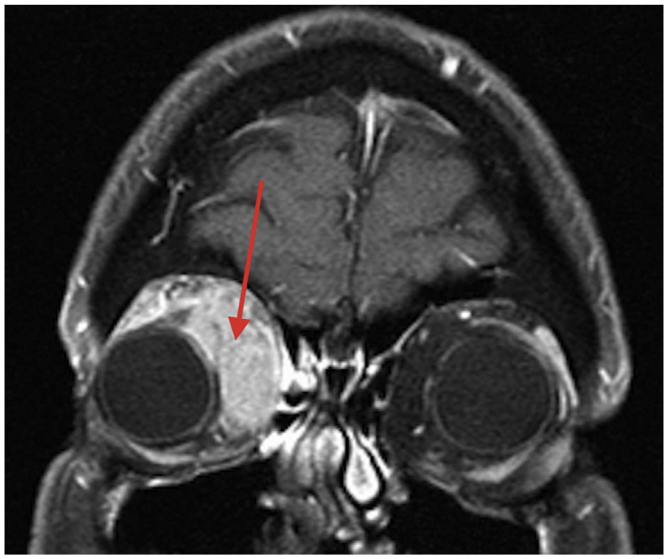

Fig. 1. Enhancing right periorbital mass on magnetic resonance imaging.

Considered in the differential diagnosis were infection (including atypical mycobacterial and fungal infection), orbital inflammatory disease (including GPA and immunoglobulin G_4 – related disease), sarcoid, lymphoma and other orbital malignancies including metastatic disease to the orbit.

CT thorax, abdomen and pelvis demonstrated no evidence of systemic disease.

The patient was empirically prescribed intravenous co-amoxiclav 1.2 g and metronidazole 500mg, both three times daily.

Biopsy of the lesion was performed via an upper eyelid skin crease incision. The tissues of the eyelid were found to be friable and the mass necrotic (Fig. 2A). Samples were sent for histological examination, microscopy and culture for bacteria, viruses and mycobacterium tuberculosis.

Histopathological analysis of the sample obtained demonstrated the presence sclerotic connective tissue and fat with very dense diffuse mixed inflammatory infiltrates of neutrophils, small and medium sized lymphocytes, macrophages, and plasma cells with neutrophilic micro-abscesses. The ratio of IgG4 vs. IgG positive plasma cells was 30%, with a maximum of 45 IgG4+ cells/high power field. There was no geographical necrosis, clear signs of vasculitis or well-formed granulomata. Immunohistochemistry revealed a mixed population of lymphocytes which marked primarily as T lymphocytes, and molecular analysis revealed polyclonal arrangements. This picture was deemed to be in keeping with a reactive inflammatory process. No micro-organisms were visualized at microscopy or cultured from the sample obtained at biopsy (Fig. 3).

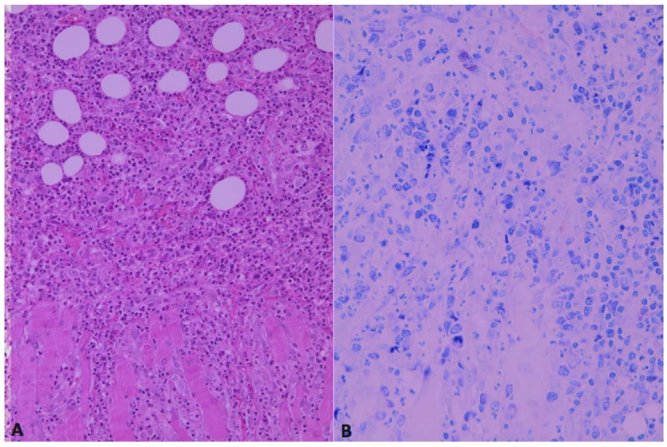

Fig. 3. A- Extremely dense inflammatory infiltrate of orbital fat (top) and ocular muscle (bottom) B-Mixed inflammatory cells: neutrophils, macrophages, plasma cells, and small lymphocytes with primary lymph follicle (right). No blasts, no granulomas or geographical necrosis.

The biopsy caused an inflammatory flare and subsequently the medial aspect of the eyelid became necrotic exposing the cornea.

Following biopsy of the lesion this gentleman's clinical condition deteriorated. The right upper eyelid became very painful. He described pain with ocular movement. He developed diplopia. The right eyelid swelling increased. Right relative proptosis of 3mm was measured. The movements of the right eye were restricted in all directions of gaze.

The lesion was found to have enlarged to involve the upper eyelid laterally and the orbit further medially upon repeat MRI scan of orbits. Treatment with intravenous vancomycin 1.5g twice daily and piperacillen and tazobactam 4.5 g three times daily had no effect. The patient's symptoms subsequently improved following three days treatment with intravenous methylprednisolone 1g daily.

With cessation of intravenous steroid therapy however this patient's condition again deteriorated. A second biopsy was performed. At attempted lid eversion the upper eyelid split at the junction of its medial third and lateral two thirds exposing the cornea. As previously, piecemeal removal of portions of the mass in an attempt to de-bulk it was completed.

This man's condition further declined thereafter. His vision deteriorated. Right proptosis and restriction of ocular movements further increased. Fundal examination showed choroidal folds and an inferior exudative retinal detachment. The ESR and CRP continued to rise.

He again received three days treatment with intravenous methylprednisolone 1g daily. Cyclophosphamide 100mg orally daily and prednisolone 60mg orally daily was subsequently prescribed. Cyclophosphamide was increased to 150 mg daily within 4 weeks. Improvement again occurred with reduced swelling and pain in the eye, and reduction in CRP and ESR.

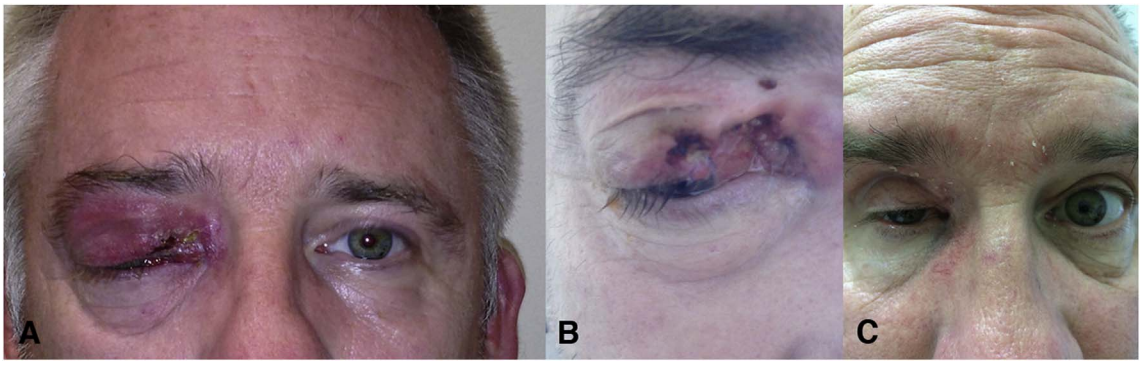

Fig. 2. A-Necrosis of eyelid post biopsy B- Reduction of inflammation post steroid and cyclophosphamide therapy C- Post operative repair.