Orbital Mass in A 9 Year Old Girl

S Deepak, K Warrier and S Rangaraj*

Department of Paediatric and Adolescent Rheumatology, UK
*Corresponding author: S Rangaraj, Department of Paediatric and Adolescent Rheumatology, UK

Received: December 18, 2018
Published: January 02, 2019

Introduction

Figure 1: Swelling and discoloration on the right upper eyelid.

Figure 2: The CT scan was suggestive of a mass lesion on the upper eyelid.

A 9-year-old girl presented with 4-month history of swelling and discoloration on the right upper eyelid and inability to open the eyes completely (Figure 1). She was previously fit and well with no past medical history of note. She was initially diagnosed with periorbital cellulitis at the local hospital and was treated with antibiotics and antihistamines for four weeks with no improvement. Initial investigations showed elevated platelet count of 508, and raised inflammatory markers - ESR of 67 mm/hr and CRP of 40 mg/L. The CT scan was suggestive of a mass lesion on the upper eyelid (Figure 2). Biopsy done by the ophthalmology team showed soft tissue fibro-inflammatory mass with reactive lymphoid aggregates and dense histiocytic and eosinophilic inflammation centred around variable calibre vessels. No atypical leukemic/lymphoid infiltrates were seen. Histiocytic cells were positive for CD160 and negative for Langerin. Hence a diagnosis of small vessel vasculitis was suggested. The child was then referred to our tertiary pediatric rheumatology service.

Figure 3: Erythematous swelling of the right upper eyelid and fullness around the right cheek.

On examination the child was apyrexial with normal vital signs. She had tenderness, erythematous swelling of the right upper eyelid and fullness around the right cheek (Figure 3). She had normal vision with blocked right nostril, reduced sensation of smell, and taste. The systemic examination was completely normal. The antibody testing showed negative ANA, and weakly positive p-ANCA, but MPO and PR3 were negative. An MRI of the orbit with contrast showed significant enlargement of the intra-orbital mass with enhancement in the infra-temporal and pterygopalatine fossa and
masticator compartment, and induration and enhancement of the fat of the right cheek (Figure 4). The possibility of IgG4 disease was discussed at the multi-disciplinary team (MDT) meeting involving pathology, oncology, and ophthalmology teams. The biopsy specimens were reviewed. This confirmed evidence of vasculitis with the presence of fibrinoid necrosis with mostly lymphocytes infiltrating the blood vessel wall. There were >10 IgG4 positive cells per high power field, but the total cells did not represent 40% of plasma cells. Her serum IgG levels were normal as well as the IgG subtypes. With a provisional diagnosis of localised IgG4 related disease, she received weaning course of steroids and weekly subcutaneous methotrexate (15mg/m²) with very good response.

**Conclusion**

IgG4-RD is a rare but significant disease in children that can lead to considerable morbidity. This case highlights the need to consider this as a differential diagnosis, thereby facilitating early treatment.

**References**


**Table 1:** Differential diagnosis for children presenting with orbital mass.

| Infiltrative – Rhabdomyosarcoma, Langerhans cell histicytosis, Leukaemia, Lymphoma, Neuroblastoma |
| Neuroblastoma |
| Benign lesions - Dermoid cyst |
| Vascular malformations & Vasculogenic tumours like Capillary haemangioma |
| Infection – Orbital cellulitis with abscess |
| Subperiosteal haemorrhage |

**Inflammatory lesions - IgG4-RD, vasculitis, sarcoidosis (rare in children)**

---

**Differential Diagnosis**

For children presenting with inflammatory lesions, a useful tool for diagnosis, staging and monitoring is the presence of autoantibodies negative. IgG4 level was elevated in 16/23 paediatric patients. Most of the patients undergo computed tomography (CT) or magnetic resonance imaging (MRI) scans as a part of investigations. 18F-fluorodeoxyglucose. Positron emission tomography (FDG PET)/CT, which can highlight active inflammatory lesions, is a useful tool for diagnosis, staging and