

Invasive Orbital Xanthogranuloma

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Received: October 14, 2016; **Published:** October 27, 2016

Abstract

Adult orbital xanthogranulomatous disease is a condition which uncommonly manifests as an invasive mass. We discuss the clinical findings, radiographic studies, and the histopathological and immunological findings of a patient with an invasive xanthogranulomatous orbital mass.

Keywords: Orbit; Orbital Mass; Adult Orbital Xanthogranulomatous Disease; Touton Giant Cells; Non-Langerhans Cell Histiocytosis

Abbreviations

Igg: Immunoglobulin G; CBC: Complete Blood Count; AFB: Acid-Fast Bacilli; GMS: Gomori Methenamine Silver; CD: Cluster of Differentiation; Mg: Milligram; ml: Milliliter

Introduction

Adult orbital xanthogranulomatous disease is a type II non-Langerhans cell histiocytosis disorder which is characterized by the appearance of foamy histiocytes and Touton giant cells, as well as fibrosis [1-3]. The disease can be categorized into four recognized subtypes, which are listed in order of prevalence: necrobiotic xanthogranuloma, Erdheim-Chester disease, adult onset asthma and periocular xanthogranuloma, and adult onset xanthogranuloma [4]. Both clinical and histopathological criteria are utilized to diagnose the specific subtype. We discuss a patient who presented to the Emergency Department complaining of a severe headache and was found to have an invasive orbital mass, histopathologically determined to be a xanthogranulomatous inflammation.

Materials and Methods

A 45 - year old Caucasian female presented to the Emergency Department with a one month history of gradually worsening headache, changes in vision, and limited extraocular movement in her left eye. The patient's past medical history was significant for arthritis. She had a smoking history of 30 pack - years.

Ophthalmologic examination revealed intraocular pressures of 17 mmHg and 49 mmHg, with a corrected visual acuity of 20/25 - 2 and 20/40 - 2 in the right and left eye, respectively. Anterior chamber, lens, and cornea were normal. There was engorged vasculature medially in the conjunctiva of the left eye. Both pupils were equal, round, and reactive to light. Extraocular movement was normal in the right eye with inability to adduct the left eye. Visual fields revealed arcuate defects in both eyes. Funduscopic exam showed normal maculas, vasculature, and retinal periphery, with asymmetric cup-to-disc ratios of 0.4 in the right eye and 0.6 in the left eye.

The patient's lab values showed a normal CBC with automated differential and normal comprehensive metabolic panel. The patient was found to have a serum IgG4 level of 1.19 mg/mL, which was elevated but still within normal range. Computed tomography scan of

the brain without contrast and magnetic resonance imaging of the orbits with and without gadolinium revealed an infiltrative, intraconal mass of the left medial orbit, extending posteriorly to the optic nerve foramen. The mass was indenting the left globe, displacing the globe temporally and anteriorly, creating significant proptosis (Figure 1).

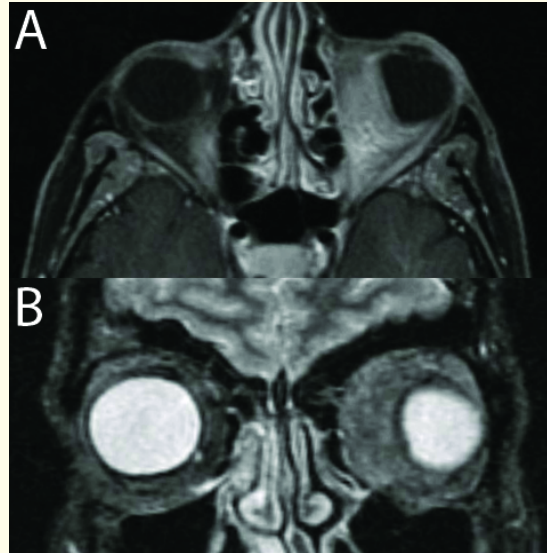


Figure 1: Axial (A) and coronal (B) magnetic resonance images of the orbit showing the mass pressing against the left globe and causing proptosis.

An orbitotomy with mass excision from the left orbit was performed and tissue samples from the mass were biopsied. AFB and GMS stains were negative for the presence of organisms. Histopathological findings revealed the mass to be a xanthogranulomatous inflammation, characterized by focally prominent foamy histiocytes, numerous Touton giant cells, and a fibrotic background (Figure 2). Immunoperoxidase stains for monoclonal keratin cocktail showed no evidence of lymphoma or carcinoma. The histiocytes stained positive for CD163.

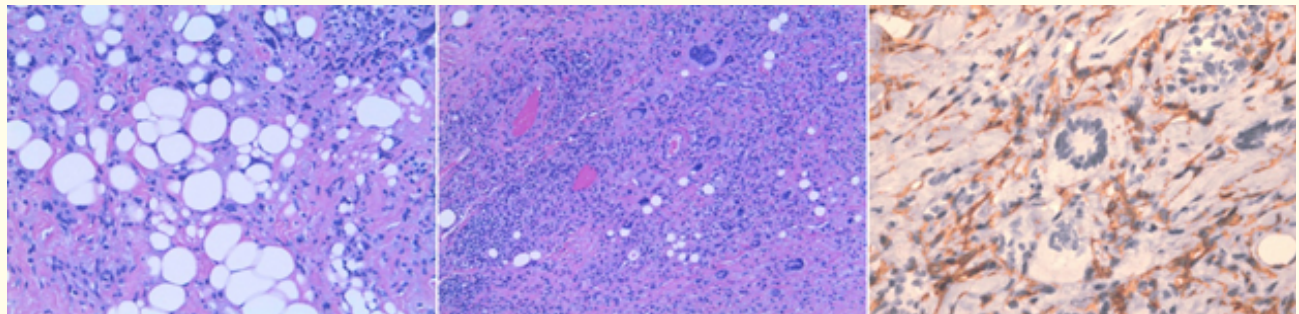


Figure 2: Hematoxylin and eosin stain (400x, 100x) and CD10 stain (400x) showing a xanthogranulomatous inflammation, characterized by a mixed chronic inflammatory infiltrate including focally prominent foamy histiocytes, along with numerous admixed multinucleated giant cells with Touton-type nuclei.

The patient was started on 80 mg of oral corticosteroids on a tapered course immediately following the orbitotomy. The patient's headache and visual problems were completely resolved. She continues to be monitored and shows no signs of recurrence.

Results and Discussion

A variety of processes can present as orbital masses such as infection, inflammation, neoplasm, trauma, and vascular disorders. Infiltrative xanthogranuloma of the orbit is uncommon, and the diagnosis can only be made after surgical biopsy and immunohistochemical tissue analysis.

Clinical correlations help in diagnosing the subtype of adult orbital xanthogranulomatous disease since all of the four subtypes have common histological findings of foamy histiocytes, Touton giant cells, and fibrosis. Necrobiotic xanthogranuloma has a distinguishing histopathological characteristic of necrobiosis in the tissues [1,3]. Erdheim-Chester disease is characterized by a systemic involvement of lymphocytes and histiocytes in the orbit as well as other organs, such as the heart, lungs, bones and other tissues; it has the worst prognosis of the four subtypes, and is often fatal [1,3]. The majority of patients with adult onset asthma with periocular xanthogranuloma have severe adult-onset asthma, sinus histiocytosis, and lymphadenopathy [1,5]. Adult-onset xanthogranuloma is characterized by a localized lesion that is usually self-limiting [1,3]. Based on the clinical and histological findings, this patient was determined to have adult-onset xanthogranuloma.

Paraproteinemia has a very common association with necrobiotic xanthogranuloma, and was recently noted as having a correlation with adult onset asthma with periocular xanthogranuloma [1-2]. As noted in the report, the patient was found with a serum IgG4 level of 1.19 mg/mL. The level is elevated but did not meet criteria for IgG4-related [2]. Additionally, the presence of IgG4 positive cells was only occasional in the IgG and IgG4 immunostained tissues.

Conclusion

The clinical presentation and radiographic findings of an infiltrating orbital mass makes it difficult to diagnose, and thus the necessity of a biopsy is needed for diagnosis. The unique histologic and immunologic features of orbital xanthogranuloma make it a distinct entity. It is crucial to follow up with patients such as these to ensure there is no relapse or emergence of new symptoms.

Conflict of Interest

The authors declare no financial interests or conflicts of interest.

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Volume 4 Issue 2 October 2016

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