ISSN: 2573-9573

Case Report

Journal of Ophthalmology & Clinical Research

An Orbital Mass Partially Responsive to Steroids

Tyler M Kaplan[1*], Bruce H Kaplan[1] and Kathryn Winkler[2]

[1]*Department of Ophthalmology, Chicago Medical School, Rosalind Franklin University of Medicine and Science, North Chicago, IL, USA*

[2]*Des Plaines Eye Physicians and Surgeons, Ltd. Des Plaines, IL, USA*

[*]Corresponding author

Tyler M Kaplan, Department of Ophthalmology, Chicago Medical School, Rosalind Franklin University of Medicine and Science, North Chicago, IL, Address: 2 Burr Oak Trail, Riverwoods, IL, 60015,USA,Tel: 847-338-7600

Submitted: 24 Apr 2021; **Accepted**: 04 May 2021; **Published**: 20 May 2021

Citations: *Tyler M Kaplan, Bruce H Kaplan and Kathryn Winkler (2021) An Orbital Mass Partially Responsive to Steroids. J Ophthalmol Clin Res 5(2): 86-88.*

Abstract

A 68 year old woman presented with a progressively enlarging orbital mass. MRI of her brain and orbits was consistent with an orbital pseudotumor. Although there was some improvement in the patient's pain and the size of the mass, it did not fully resolve.The patient had a biopsy demonstrate non-caseating granulomatous inflammation. A chest X-ray and CT chest demonstrated bilateral hilar lymphadenopathy. Further examination and interviewing of the patient revealed several months of joint paints and lower extremity nodules. The patient was diagnosed with orbital sarcoidosis and was started on methotrexate by a rheumatologist.

Keywords: Orbital masses, Oculoplastics, Pseudotumor, Rheumatology, Sarcoidosis

Case Report

A 68 year old black woman with a history of type 2 diabetes mellituspresented with a 6 weekhistory of a progressively enlarging, painful mass located near her left lateral canthusand left lower eyelid. Upon presentation, her vision was 20/25 in the right eye and 20/40 in the left eye, her pupils were equally round and reactive to light with no afferent pupillary defect. The patient's extraocular movements and color vision were full. The mass was mass solid, tender to palpation, and associated with periorbital edema. It was approximately 30mmx15mm in size across the left lower eyelid to the left lateral canthus extending to the left zygomatic arch. In the central lower lid, the mass was felt to be extending into the anterior orbit. There was2 mm of relative enophthalmos and retraction of the left lateral canthal angle related to the mass. An MRIwas ordered which demonstrated a non-enhancing, lobulated mass in the subcutaneous region of the left and lateral cheek extending into the lateral and inferior part of the left orbit. The paranasal sinuses were unremarkable. The MRI was consistent with an orbital pseudotumor. Apresumptive diagnosis of idiopathic orbital inflammatory syndrome was given, and the patient was started on40 mg oral prednisone.Over the course of 3 weeks, the patient experienced partial improvement in tenderness and periorbital edema, however there was no significant improvement in size of the mass.

What Would You Do Next?

A. Begin parenteral or oral antibiotics
B. Increase the dosage of steroids
C. Biopsy of the lesion
D. Repeat MRI of the brain and orbits

Diagnosis

Orbital Sarcoidosis

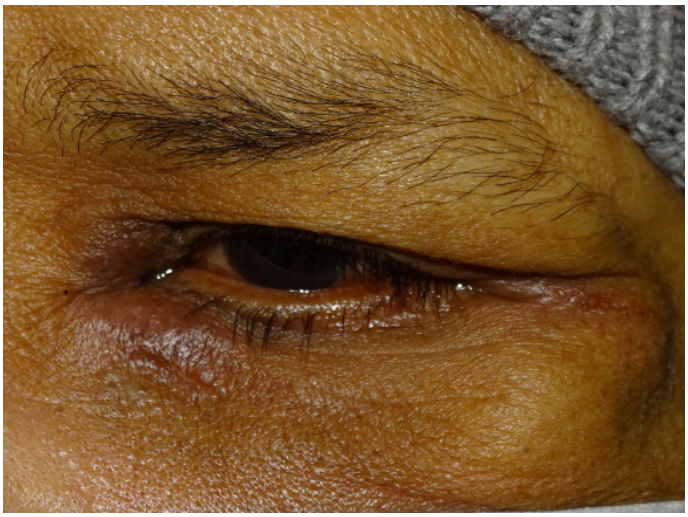

Figure 1A: External photograph of the patient's left eye demonstrating a mass located in the inferior and temporal portion of the patient's orbit

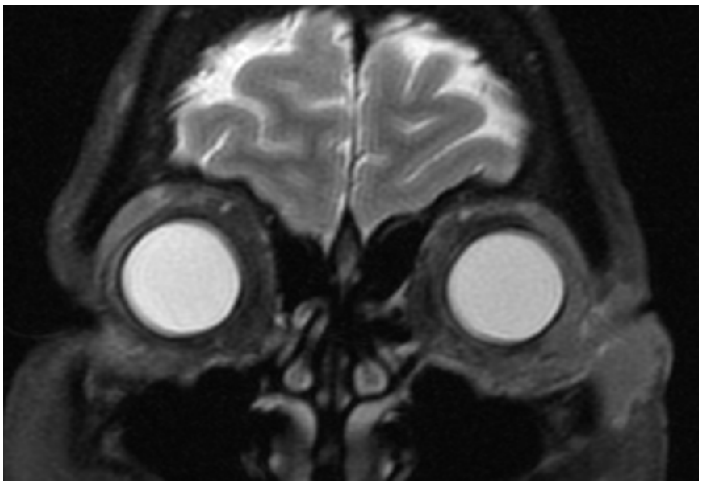

Figure 1B: T2 MRI in a coronal section demonstrating a mass lesion in the subcutaneous region of the left cheek and lateral to the left eye that is extending into the lateral and inferior part of the left orbit

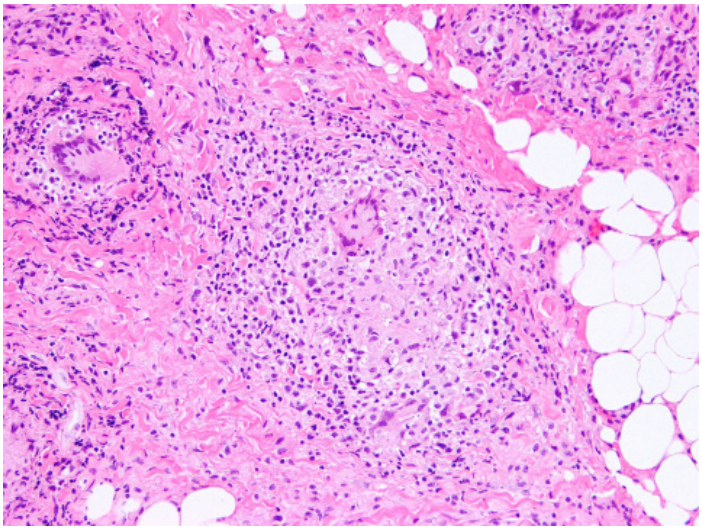

Figure 2: Hematoxylin and Eosin stain 10x micrograph of non-caseating granulomas with giant cells, present in fibroadipose tissue

Discussion

The differential diagnosis of a rapidly progressive, painful, andenlarging orbital mass is vast and includesinflammatory, infectious, malignant, and vascular etiologies. Orbital inflammatory syndrome (OIS), also known as orbital pseudotumor,is a benign, non-infectious, inflammatory condition, oftentimes idiopathic in nature.OIS is the third most common orbital disorder in adults, following Graves' disease and lymphoproliferative processes [1]. It is commonly diagnosed based on history andimaging studies. Further imaging of the mass is unnecessary since the size did not dramatically increase (D). OISis often presumptively treated with systemic glucocorticoidswith a dramatic response seen in more than 75% of patient within 48 hours [1,2]. When responsive, this often confirms the diagnosis without need for further investigation.In this case, intensification of the patient's steroid regimen was decided against due to minimalresponsivenessof the initial regimen (B). When an orbital mass is only partially responsive to steroids, a biopsy should be performed due to concern for the underlying etiology of the mass (C) [1]. In addition, an inflammatory work up should be considered. Antibiotics would not be indicated sincethe patient's MRI scan was not consistent with an infectious process,such as orbital cellulitis,and the patient did not demonstrate infectious symptoms (A).

Pathology demonstrated fibrous tissue with multiple non-caseating granulomas. Acid-Fast Bacilli (AFB) and Gomori- Silver (GMS) stains were negative. Flow cytometry was unremarkable for a lymphoproliferative process. On further interview, the patient noted several months of joint pains and lower extremity nodules, which resolved with the addition of oral steroids. A chest X-Ray and CT demonstrated mediastinal and hilar lymph nodes. The clinical picture and pathology was most consistent with systemic sarcoidosis. Although granulomatous OIS mimicking sarcoidosis has been described, the imaging and presence of systemic symptoms made sarcoidosis the most likely diagnosis [2]. In addition, the granulomatous subtype of OIS is generally more responsive to steroid therapy than in this case [1].

Sarcoidosis affects multiple organ systems and is histologically characterized by the presence of non-caseating granulomas along with typical clinical features such as hilar lymphadenopathy, erythema nodusum, lupus perino, and restrictive lung disease [3,4]. Ocular involvement is observed in nearly 25% of patients, while orbital involvement is uncommon [4,5]. Although sarcoidosis is most frequently diagnosed in the second or third decade of life, sarcoidosis presenting with orbital involvement is more common in women greater than 50 years of age [4,5]. Orbital sarcoidosis generally involves the lacrimal gland, soft tissues, the optic nerve, and extraocular muscles [4,5]. Orbital sarcoidosis can be diagnostically challenging to distinguish from isolated orbital non-caseating granulomatous disease if the presence of systemic symptoms are not discussed with a patient. It is crucial to inquire about systemic symptoms inpatients with orbital masses containing non-caseating granulomas to make the proper diagnosis. In addition, close rheumatology follow-up is required as the treatment modalities differ. Although steroids are often the mainstay of treatment for sarcoidosis, immunosuppressants such as methotrexate are often required [4]. When a patient presents with an orbital mass, rheumatologic processes should beconsidered as part of the differential diagnosis.

Patient Outcome

The patient was referred to a rheumatologist for treatment of systemic sarcoidosis and was started on methotrexate therapy.

Declarations of Interest

The authors report no commercial or proprietary interest in any product or concept discussed in this article.

Funding

None

References

1. Chaudhry I, Shamsi F, Arat Y, Riley F (2008) Orbital pseudotumor: Distinct diagnostic features and management. Middle East Afr J Ophthalmol 15:17.
2. Yuen SJA (2003) Idiopathic Orbital Inflammation. Arch Ophthalmol 121:491.
3. Harris GJ (2006) Idiopathic Orbital Inflammation: A Pathogenetic Construct and Treatment Strategy. Ophthalmic Plast Reconstr Surg 22:79-86.
4. Prabhakaran VC (2007) Orbital and Adnexal Sarcoidosis. Arch Ophthalmol 125:1657.
5. Kobak YS, Topaloglu A, Oncel G, Karaarslan A (2015) Sarcoidosis presenting as orbital pseudotumor. Reumatismo 67: 78-81.