A case report of Intraorbital Inflammatory Myofibroblastic Tumor

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INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) is a rare, mesenchymal neoplasm that can affect any soft tissue or viscera but usually affects the lung, abdomen, and retroperitoneum. IMTs are predominately found in children and young adults. They are histopathologically composed of spindle cells with an inflammatory infiltrate background [1]. More than half of extrapulmonary IMTs have gene rearrangement in the anaplastic lymphoma kinase (ALK) gene [2].

OBJECTIVE

1. We discuss a rare case of intra-orbital IMT presenting as orbital cellulitis
2. Discuss the clinical presentation, histopathology and treatment of IMT

CASE DESCRIPTION

14-year-old female with history of allergies presented to the emergency room with right eye pain without vision changes and eyelid swelling for 3 weeks. Her eye pain worsened with extraocular movement. She had a history of intermittent eyelid swelling that was suspected to be allergies but was significantly worse at the time of presentation. No family history of autoimmune disease.

Exam:

VA 20/20 OU, IOP normal, External Exam: marked proptosis of the right eye. Slit lamp exam: swelling and erythema of the right upper and lower eyelid with conjunctival injection and chemosis. Motility: restricted motility of the right eye in all directions. Fundus exam: right optic nerve had 360-degree disk edema. Normal eye exam of left eye.

Initial treatment:

Patient was started on IV antibiotics and subsequently taken to the OR for exploration of the orbit and possible abscess drainage. A transconjunctival approach was used to explore the medial orbit and intraconal space. No abscess or purulence was found, rather, a firm thickened mass of the orbital tenon's fascia in the medial and retrobulbar space was encountered. An incisional biopsy was performed and sent to pathology.

LAB AND IMAGING RESULTS

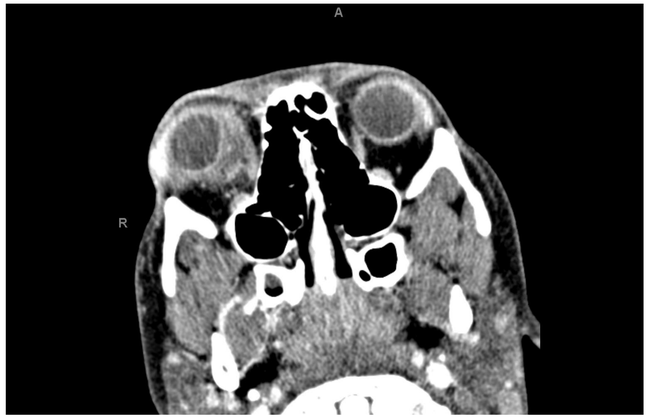
Lab results: WBC-wnl, CRP-wnl, ESR-mildly elevated

Imaging: A CT orbit with contrast was obtained which was read as right preseptal and postseptal cellulitis with crescent-shaped low-density rim-enhancing fluid collection consistent with evolving abscess in the right intraconal space. (fig 1)

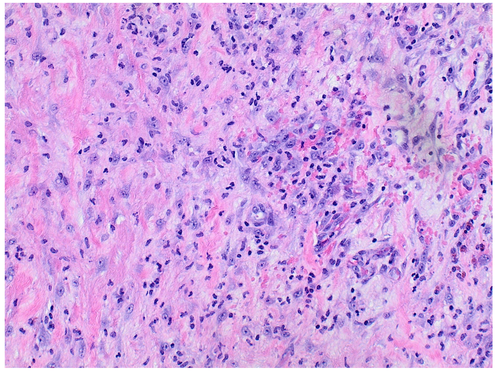
Pathology: Cells of interest were SMA positive and negative for ALK1 and Desmin . The final diagnosis was determined to be primary orbital inflammatory myofibroblastic tumor. (fig 2)

CLINICAL IMAGES

(Figure 1)



(Figure 2)



CONCLUSION

Post Operative treatment: Patient started on IV steroids with resulting improvement in eyelid swelling, proptosis, eye pain, and motility restriction. She was discharged on an extended taper of oral steroids.

DISCUSSION

The etiology of inflammatory myofibroblastic tumor is unknown and is not limited to any particular location [1], however, the lung, mesentery, and omentum are the three most common sites that IMT is found. IMT is usually benign but there are rare occurrences of metastasis to other locations [3,4]. There have only been a handful of cases of primary orbital IMT. Based upon our literature review gender does not seem to influence the occurrence of orbital IMT[1,3,5,6,7].

Orbital IMT can have various clinical presentations such as painless mass, eye pain, diplopia, progressive vision loss, ptosis, conjunctivitis, invasion of the lacrimal gland, extraocular muscle movement restriction, and/or edema. To confirm a diagnosis of IMT, histopathological evaluation is needed. Histopathological findings include proliferating myofibroblasts, spindle cells, and inflammatory cells that include plasma cells, lymphocytes and eosinophils. 50% of IMT are ALK-1 positive on immunochemistry and is useful to help differentiate this tumor from other types of mesenchymal tumors. Literature review has not shown clinical distinction between ALK-1 positivity and ALK-1 negativity to clinical outcome, recurrence, regression, or metastasis [8,7,9,10]. Our patient was ALK-1 negative and had complete resolution of symptoms after treatment with systemic steroids.

Management of IMT varies depending on the clinical presentation and location but may include surgical excision, systemic steroids, and/or radiotherapy. Systemic steroids are usually the first line treatment with partial to complete regression in 31-78% of patient within 48 hours but has a high rate of recurrence [7]. Low dose radiation therapy is used in refractory cases with local regression in 67-87% of patients [7]. Some cases are more amenable to surgical resection and in rare cases of large orbital IMT, enucleation or exenteration may be necessary for complete remission [11, 12]. Metastasis is rare, <5% of cases, but the most common sites are brain and liver [3].

Overall, this was a rare case of IMT involving the orbit masquerading as orbital cellulitis. Although rare, this case illustrates the importance of having a broad differential in a patient presenting with orbital inflammation and proptosis. Response to systemic steroids for IMT is usually rapid with partial to complete resolution of symptoms but a prolonged taper is often necessary to prevent recurrence of these lesions.

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