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Case Report

Sarcoidosis of the Extra-Ocular Muscles: Case Report

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Abstract

Sarcoidosis is one of the leading causes of inflammatory eye disease. Patients with ocular sarcoidosis can present with a wide range of clinical presentations and severity. This case demonstrates an unusual presentation of unilateral infiltration of the extra-ocular muscles, sparing the uveal tissues and lacrimal glands. This case highlights the importance of screening for ocular involvement in sarcoidosis, and to consider sarcoidosis in the differential for peri-orbital swelling and diplopia.

Keywords: Sarcoidosis, Granulomatous Disease, Autoimmune Disease, Eye Sarcoid.

BACKGROUND

Sarcoidosis is a chronic multisystem inflammatory disease, most commonly affecting the lungs, but may also affects the skin, joints, lymph nodes and the eyes. It is common in adults between the ages of 20-40 years in Caucasians and African Americans [1]. This disease characterizes by pathological diagnostic findings of noncaseating granuloma, after other possible causes are excluded, including infectious diseases, such as tuberculosis. Eye involvement may occur with this disease and commonly presents with uveitis in 80% of cases. However, isolated extraocular muscle involvement is rare [2, 3]. This case illustrates an unusual presentation of unilateral infiltration of the extra-ocular muscles, presenting with a primary symptom of diplopia.

CASE PRESENTATION

A 62-year-old male presented with right sided proptosis and diplopia, worsening over a 7-month period. There was mild ptosis with swelling of the upper and lower eyelids. Visual acuity, slit-lamp examination, intra-ocular pressures, and fundi were normal. He was otherwise asymptomatic. Medical history was relevant for pulmonary sarcoidosis, diagnosed 8 years prior. At that time, the patient was experiencing dyspnea and chronic dry cough. CT chest showed mediastinal and hilar lymphadenopathy with several pulmonary nodules. Biopsy revealed non-caseating granulomas with negative staining for fungal or mycobacterial infection, or malignancy. His symptoms self-resolved and the lymphadenopathy and nodules decreased in size on follow-up chest x-rays.

At current presentation, blood work showed normal CBC and CRP, normal ACE and calcium, negative ANA and ANCAs. CT of the orbits showed thickening of the right superior and lateral rectus muscles with mild stranding of intra-orbital fat (Figure 1A and 1B). Imaging of the chest, abdomen and pelvis demonstrated hilar and mediastinal lymphadenopathy, decreased in size from previous, as well as numerous lymph nodes within the abdomen and pelvis. Orbitotomy with biopsy of the lateral rectus was performed, showing noncaseating granuloma. There was no evidence of IgG4 cell infiltration. Fungal culture and tuberculosis were negative. There was no evidence of malignancy. Based on his clinical presentation and biopsy finding in the setting of his previous diagnosis of sarcoidosis, he was diagnosed with ocular sarcoidosis.

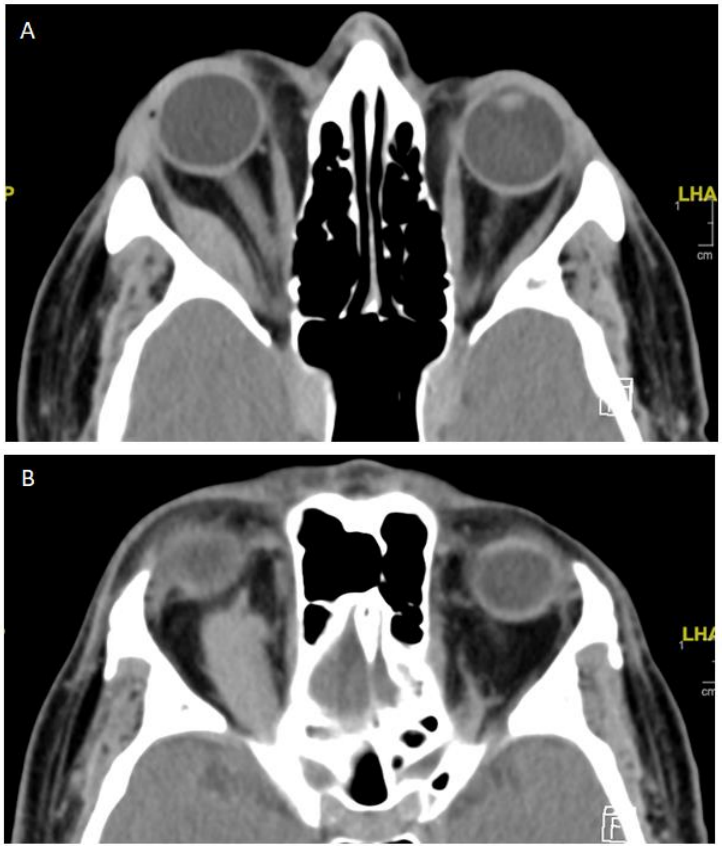

Figure 1: CT scans of the orbits with axial views showing (A) and (B) thickening of the right superior and lateral rectus muscles with mild stranding of intra-orbital fat

He was initiated on high dose prednisone with complete recovery of symptoms. However, due to his resistant disease after tapering down the dose of prednisone, he was started on steroid sparing agent, Methotrexate. His disease went into remission afterward and he was able to completely get off the steroids. His disease continues in remission at 2 years of follow-up.

DISCUSSION

Ocular sarcoidosis can affect any part of the eye and leads to a variety of clinical presentations ranging in severity. Most common manifestations include bilateral uveitis, lacrimal gland infiltration and conjunctival nodules [4]. Isolated extra-ocular muscle involvement without a mass is extremely rare. There are several studies of extra-ocular muscle sarcoid, most of which showed abnormal eye examination, inflammatory symptoms such as ocular pain, or systemic manifestations [5-11]. Our case presents a unique presentation of sarcoidosis with unilateral infiltration of the extra-ocular muscles, which lacks systemic inflammatory features. The diagnosis is usually evident with the presence of noncaseating granuloma in muscle biopsy. Baseline chest x-rays are important to rule out lung involvement. Calcium and ACE level could be elevated, although normal results don't rule out sarcoidosis [4].

Possible differentials for orbital sarcoidosis include infections such as tuberculosis, and Coxsackie virus B2. Others may include small vessel vasculitis such as Granulomatosis with Polyangiitis or orbital myositis and systemic lupus erythematosus. IgG4 disease with cell infiltration is a possibility. Lastly, Grave's disease and lymphoid tumours are also included in the list. It is always necessary to navigate the presence of systemic features associated with each of the previously named differential.

This case highlights the importance of screening for ocular involvement in sarcoidosis, and to consider sarcoidosis in the differential for peri-orbital swelling and diplopia. Early biopsy of orbital masses not responding to initial treatment is important to enable accurate diagnosis and appropriate management, and to reduce the likelihood of long-term complications from sarcoid.

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