Ocular Sarcoidosis Masquerading as Acute Frontal Sinusitis

Joshua Greene MD[1], Christian George MD[2], Robert Deeb MD[1]

[1]Department of Otolaryngology-Head and Neck Surgery

[2]Department of Ophthalmology

Henry Ford Health System, Detroit, Michigan

Abstract

Objectives: 1) Describe an unusual presentation of ocular sarcoidosis which was initially mistaken for classic acute frontal sinusitis. 2) Recognize that ocular sarcoidosis is rare and may mimic traditional sinonasal diseases.

Methods: Case report and review of the literature. A PubMed search using terms sarcoid, ocular, sinusitis, and proptosis was performed

Results: A 65-year-old female presented with 2 weeks of nonpainful, right eye proptosis with right forehead pressure. A CT scan was performed which showed right frontal sinus opacification as well as orbital proptosis. The patient was treated with antibiotics and steroids for the presumed diagnosis of orbital cellulitis secondary to frontal sinus infection. After initial improvement of symptoms, her proptosis returned with additional eyelid swelling within several days of completing the course of medications. Follow-up CT scan showed improved aeration of frontal sinus, but she continued to suffer from intermittent proptosis which only improved with intermittent steroids. Transcutaneous orbitotomy with incisional biopsy of pre-aponeurotic fat revealed noncaseating granuloma, consistent with sarcoidosis.

Conclusion: Extrapulmonary involvement by sarcoidosis is observed in ~30-40% of patients however ocular sarcoidosis presents as the initial manifestation in only 1.5-12.4% of patients. The presentation is often confounded by sinonasal symptoms, which may delay the diagnosis. Sarcoidosis should be considered in all patients who present with ocular and sinus symptoms.

Introduction

Sarcoidosis is a multisystematic granulomatous disease of unknown etiology. Head and neck involvement has been reported in 10-15% of patients with the most common sites being the skin, nose, paranasal sinuses, larynx, salivary glands, cervical lymph nodes and middle ear[1,2]. Additionally, 35-55% of these cases will present with ophthalmologic complaints, including enlargement of the lacrimal gland, masses in the upper eyelid or uveitis[3]. Research on the prevalence of sinonasal involvement in sarcoidosis is generally limited to small case series. As such, involvement of the sinonasal tract has been shown to have a wide ranging prevalence from ~1% to 61%[2,4]. We present a unique case of a patient with presumed fontal sinusitis, ultimately found to have ocular sarcoidosis.

Materials/Methods

Institutional review board approval was obtained. A retrospective chart review was performed highlighting the clinical presentation, work-up, intraoperative findings, and final diagnosis. Literature review of relevant research concerning paranasal sinus and orbital involvement of sarcoidosis was performed. A multidisciplinary approach with Otolaryngology and Ophthalmology was used to formulate the diagnostic plan.

Case Description

A 65 year old healthy female presented with a two week history of non-painful right eye proptosis. She denied any significant ophthalmoplegia or vision changes. She received a CT scan which revealed right frontal sinus opacification concerning for sinusitis as well as thickening and enlargement of the superior rectus/levator complex with minimal prominence of the superior preseptal soft tissues. There was also noted to be some demineralization and questionable dehiscence of the right lateral frontal sinus wall. She was treated for a presumed orbital cellulitis secondary to frontal sinusitis with Augmentin and methylprednisolone (Figure 1). Evaluation by Otolaryngology the following day included nasal endoscopy which was essentially normal. She had near resolution of her symptoms within 1-2 days after initiating treatment.

She then presented 5 days later with recurrent right sided proptosis and eye pressure. She was again given Augmentin and methylprednisolone. Her symptoms progressed to included eyelid swelling and headache over the next week. Repeat CT scan showed improved aeration of the right frontal sinus, however the orbital findings were unchanged (Figure 2). Given that her sinus opacification improved on her subsequent CT scan, a suspicion of a primary orbital process was raised. As such, she underwent right transcutaneous orbitotomy with incisional biopsy of the mass. Her preaponeurotic fat was noted to be firmer than usual with a rubbery putty colored consistency. This tissue in addition to aponeurosis from the levator was sent for pathology, which returned as noncaseating granuloma.

Discussion

Sarcoidosis is a chronic idiopathic systemic disease, that may involve the head and neck region in 10-15% of the cases.[5,6] Those with sinonasal involvement typically present with symptoms of chronic crusting rhinitis, nasal obstruction, and anosmia[7]. A recent review by Dr. Lawson et al proposed a new system of classification which would subtype sinonasal sarcoidosis into 1 of 4 categories: atrophic, hypertrophic, destructive or nasal enlargement.[8] This classification could then act as a guide to diagnosis and treatment.

The challenge in the diagnosis and management of sinonasal sarcoidosis is that the presentation is often indistinguishable from other conditions such as chronic rhinosinusitis, Wegener's granulomatosis, allergic rhinitis, atrophic rhinitis and NK/T cell lymphoma. The diagnosis of sarcoidosis is based on clinical exam, radiographic studies, biopsy and laboratory analysis.[9] Histopathology shows non-caseating granulomas. ACE levels may reflect disease activity, being elevated with active disease and normal when quiescent.

The data regarding sinonasal sarcoidosis is mostly limited to case reports and case series. As such, it is difficult to ascertain the frequency with which both sinonasal and orbital involvement occur in the same patient. It is generally believed to be a rare finding. Bronson and Fisher reported the first case of sinonasal sarcoidosis with extension to the orbit in 1976.[10] *Baughman et al*. found that patients with both sinonasal and ocular sarcoidosis, often have lacrimal gland involvement and present with dacrocystitis[11]. This case highlights the importance of maintaining a broad differential diagnosis in those patients presenting with isolated sinus and ocular signs and symptoms.

Figures

Figure 1. CT sinus showing right frontal sinus opacification

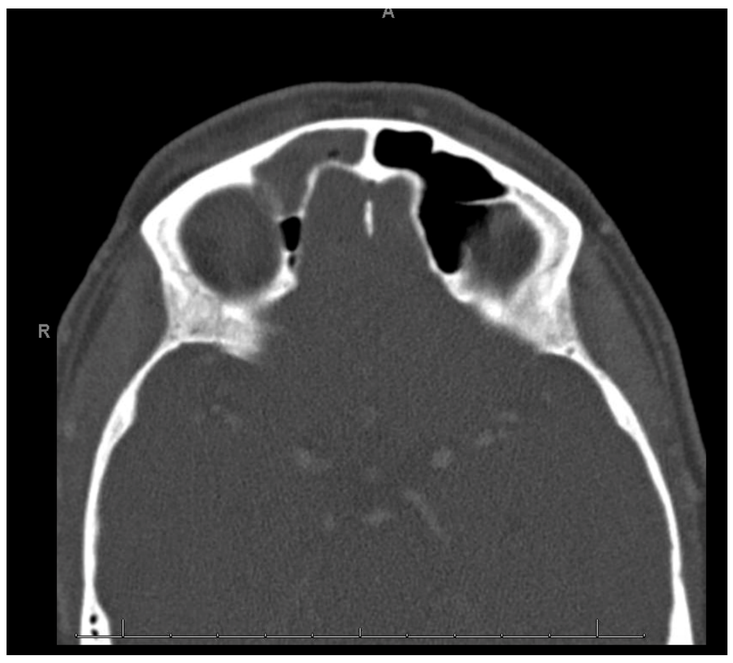
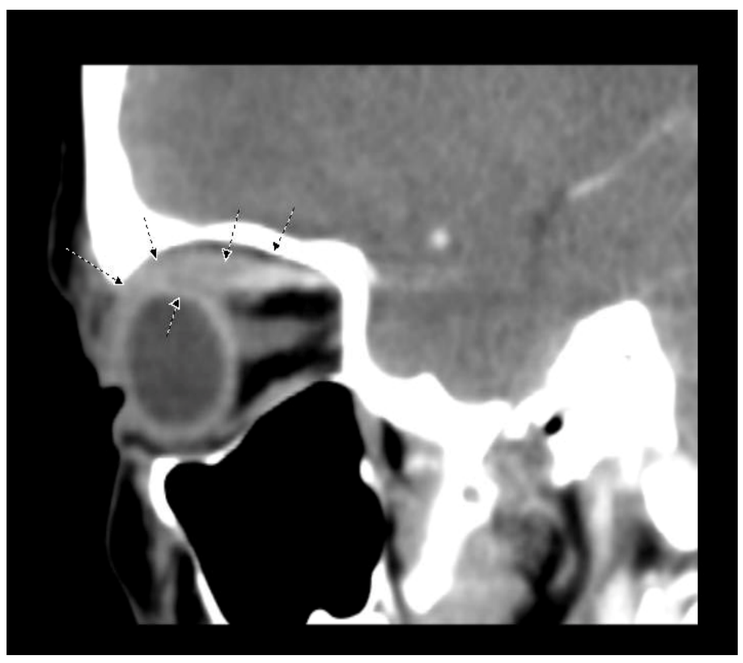


Figure 2. Follow up CT scan showing soft tissue density within the right orbit.



Conclusion

Sarcoidosis is a common granulomatous disease with rare extrapulmonary involvement of the eye or sinonasal tract. Although it's presentation may be confounded by more common disease processes, it is important to consider sarcoidosis in patients who present with recurrent isolated sinonasal and ocular symptoms.

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