



## Orbital Sarcoidosis Masquerading as Acute Frontal Sinusitis

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### Abstract

Sarcoidosis is a chronic systemic disease that rarely manifests within the sinuses or orbit. A 65-year-old female presented with 2 weeks of eye proptosis and forehead pressure. A computed tomography scan revealed frontal sinus opacification. The initial diagnosis of orbital cellulitis secondary to frontal sinus infection was made but symptoms returned after medical therapy. Biopsy of preaponeurotic fat revealed non-caseating granuloma, consistent with sarcoidosis. Extrapulmonary involvement by sarcoidosis is observed in ~30-40% of patients; however, orbital 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.

### Introduction

Sarcoidosis is a multisystemic granulomatous disease of unknown etiology. Sarcoid is a term used to describe the isolated presentation of the disease. 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]. Additionally, 35-55% of these cases will present with ophthalmologic complaints, including enlargement of the lacrimal gland, masses in the upper eyelid or uveitis [2].

A study of 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% [3,4]. We present a unique case of a patient with presumed frontal sinusitis, ultimately found to have orbital sarcoid.

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### Case Presentation

A 65-year-old healthy female presented with a 2 week history of non-painful right eyelid swelling and proptosis. She denied diplopia and vision changes. There was firmness to palpation in the orbit. She underwent prompt sinus computed tomography scanning that revealed right frontal sinus opacification concerning for sinusitis as well as hyperdensity of the orbital soft tissues adjacent to the frontal sinus extending to the anterior superior rectus/levator palpebrae muscle complex. Demineralization was also noted, as well as possible small dehiscence of the right lateral frontal sinus wall. She was treated for a presumed orbital cellulitis secondary to frontal sinusitis with amoxicillin/clavulanic acid (Figure 1). Evaluation by otolaryngology included nasal endoscopy, which was essentially normal. She had near resolution of her symptoms 4 days after initiating treatment, which included methylprednisolone.

She presented 2 days after completing her 10-day antibiotic course with recurrent right upper eyelid swelling and subjective facial pressure. She was again given amoxicillin/clavulanic acid and methylprednisolone. Her symptoms progressed to include eyelid swelling and headache over the next week. A repeat computed tomography 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 computed tomography scan, a primary orbital process was implicated. 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 putty colored rubbery consistency. This tissue and a section of the levator palpebrae muscle was found to consist of non-caseating granulomas. Ziehl-Nielsen stain for acid fast bacilli, immunostain for mycobacteria and Grocott stain for fungi were all negative. Flow cytometry and gene rearrangement studies for lymphoma were negative. The findings were consistent with sarcoid.



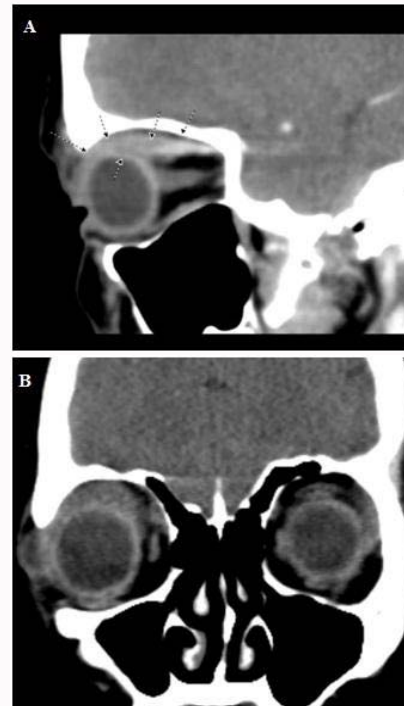
**Figure 1:** (A) Computed tomography of the sinus showing right frontal sinus opacification. (B) Adjacent superior orbital hyperdensity on the right side.

## Discussion

Sarcoidosis is a chronic idiopathic systemic disease, that may involve the head and neck region in 10%-15% of cases [5,6]. Those with sinonasal involvement typically present with symptoms of chronic crusting rhinitis, nasal obstruction, and anosmia [7]. A recent review by Lawson et al. [7] proposed a new system of classification that subtypes sinonasal sarcoidosis into 1 of 4 categories: atrophic, hypertrophic, destructive or nasal enlargement. This classification can 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, granulomatosis with polyangiitis, allergic rhinitis, atrophic rhinitis and natural killer/T-cell lymphoma. The diagnosis of sarcoidosis is based on clinical examination, radiographic studies, biopsy and laboratory analysis [8]. Histopathology shows non-caseating granulomas. Angiotensin-converting enzyme levels may reflect systemic 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 [9]. Baughman et al. [10] found that patients with both sinonasal and ocular sarcoidosis often have lacrimal gland involvement and can present with dacryocystitis. This case highlights the importance of maintaining a broad differential diagnosis in those patients presenting with sinus and orbital inflammation and having an atypical response to treatment.



**Figure 2:** Follow-up computed tomography scan showing soft tissue density within the right orbit. (A) Sagittal, (B) Axial view.

## Conclusion

Sarcoidosis is a common granulomatous disease with rare extrapulmonary involvement of the eye or sinonasal tract. Although its 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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