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Case Report

An erosive lesion in the orbital apex as the presenting sign of sarcoidosis

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Summary

Sarcoidosis is an autoimmune granulomatous disease that can affect any organ system in the body. Ocular and orbital manifestations are relatively common. Osseous involvement is rare and usually involves bones of the appendicular skeleton. We present an unusual case of an erosive sarcoid granuloma in a 48-year-old woman that involved the orbital apex. This case highlights diagnosis, treatment, and the importance of full systemic workup to determine the extent of the disease.

Case Report

A 48-year-old, African American woman with a past medical history significant for nonischemic cardiomyopathy presented to the cardiology clinic at the University of Kansas Hospital for follow-up examination. She complained of a 1-week history of parieto-occipital headache and progressive vision loss in the right eye. She denied eye pain, flashes/floaters, diplopia, or changes in vision in her left eye. Neurology was consulted, and the patient was transferred to the emergency department for a stroke workup. Computed tomography (CT) of the head without contrast showed no abnormalities.

On ophthalmological examination, her best-corrected visual acuity was 20/30 in the right eye and 20/20 in the left eye, with no afferent pupillary defect (APD). There was a mild temporal restriction on confrontation visual field testing in the right eye; the visual field was full in the left eye. The dilated posterior segment appeared unremarkable in each eye. The patient was discharged with a diagnosis of an atypical migraine and was instructed to follow-up at the outpatient ophthalmology clinic the following week.

Two days later, the patient presented to the emergency department with worsening of her symptoms. On examination, visual acuity in the right eye was hand motions; in the left eye, 20/20. There was a new APD in the right eye. Intraocular pressure (IOP) by Tono-Pen (Reichert Technologies, Depew, NY) was 14 mm Hg in the right eye and 13 mm Hg in the left eye. Visual field by confrontation was restricted in all quadrants except superotemporally in the right eye and full in the left eye. Dilated fundus examination again revealed an unremarkable-appearing optic nerve head, macula, vessels and periphery in each eye.

With no abnormalities seen on the dilated fundus examination, workup for a posterior optic neuropathy was initiated. A CT of the brain without contrast was repeated, and no abnormalities were noted. A CT of the orbits without contrast showed an abnormal soft tissue lesion with an aggressive erosive process involving the posterior right orbit and ethmoid sinus (Figure 1). CT of the chest without contrast showed mediastinal and bilateral hilar lymphadenopathy as well as multiple bilateral lung nodules concerning for sarcoidosis or lymphoma (Figure 2). Positron emission tomography scan showed increased fluorodeoxyglucose uptake within the region of the right ethmoid sinus and metabolically active supraclavicular, mediastinal, hilar, periportal, and inguinal lymphadenopathy with diffuse increased uptake of the spleen. Lumbar puncture revealed no abnormalities. Erythrocyte sedimentation rate was moderately elevated at 34 mm/hour. Serum and cerebrospinal fluid angiotensin-converting enzyme levels were normal. Syphilis, Histoplasma, Coccidioides, and Blastomyces antibodies were negative. Lymphoma panel and autoimmune panels were also negative.

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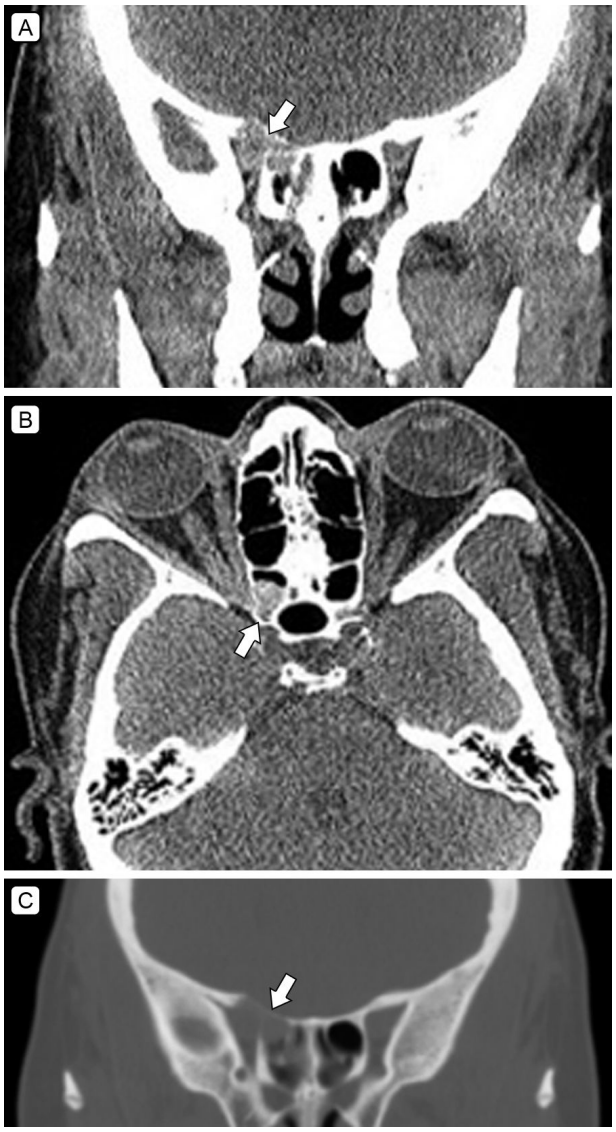

Figure 1. Computed tomography of the orbit without contrast showing an erosive mass (arrow) involving the posterior right orbit and ethmoid sinus. A, Coronal soft tissue window. B, Axial soft tissue window. C, Coronal bone window.

With the assistance of the otolaryngology team, endoscopic sinus surgery was performed, with biopsy of the soft tissue lesion involving the posterior right orbit and ethmoid sinus. Hematoxylin and eosin staining revealed abundant noncaseating granulomas consistent with sarcoidosis, with no evidence of infection or malignancy (Figure 3). The patient was started on 60 mg prednisone daily. After 1 month of treatment, the patient's symptoms had substantially improved. Repeat examination revealed visual acuity of 20/20 in the right eye and no APD. Humphrey automated visual field testing showed

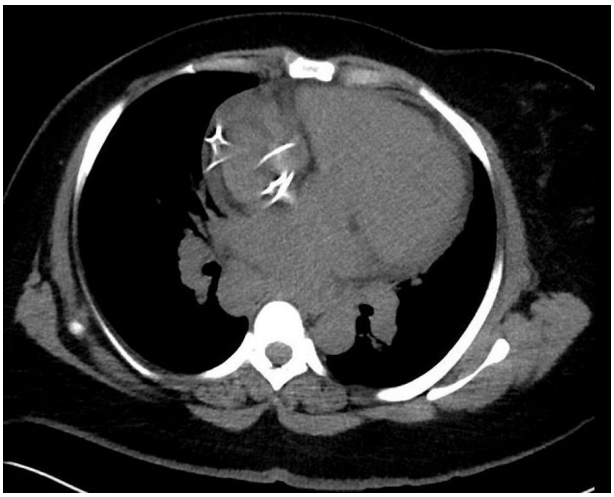

Figure 2. Computed tomography of the chest without contrast showing an enlarged right hilar lymph node.

moderate improvement with marginal false positives. The optic nerve head remained normal in appearance. She was seen in rheumatology clinic and started on a slow prednisone taper. She experienced no recurrence of ocular symptoms.

Discussion

Sarcoidosis is a granulomatous disease that can affect many organ systems. Although it can occur at any age, it is often seen in adults 20–40 years old.[1] The most common involvement is found in the lung, accompanied by a complaint of fatigue by the patient. Ocular involvement occurs in approximately 12% of patients with sarcoidosis.[2,3] Anterior uveitis is the most common intraocular manifestation of the disease.[4] Orbital sarcoid is much less common and can present in a variety of ways, including lacrimal gland infiltration, soft tissue mass, and extraocular muscle involvement.[5] Osseous involvement in sarcoidosis is uncommon and most frequently involves the middle and proximal phalanges.[6] To our knowledge, this is the first report of an aggressive sarcoid granuloma eroding into the orbital apex causing rapid onset, severe visual loss. A similar case of orbital apex sarcoidosis has been reported; however, the patient described presented with progressive proptosis, diplopia, and optic disc edema.[7]

In a case where visual acuity is severely affected and an APD is seen with no abnormality noted on dilated fundus examination, a posterior orbital process should be considered and imaging of the orbit should be performed. In the case of sarcoidosis, biopsy of involved

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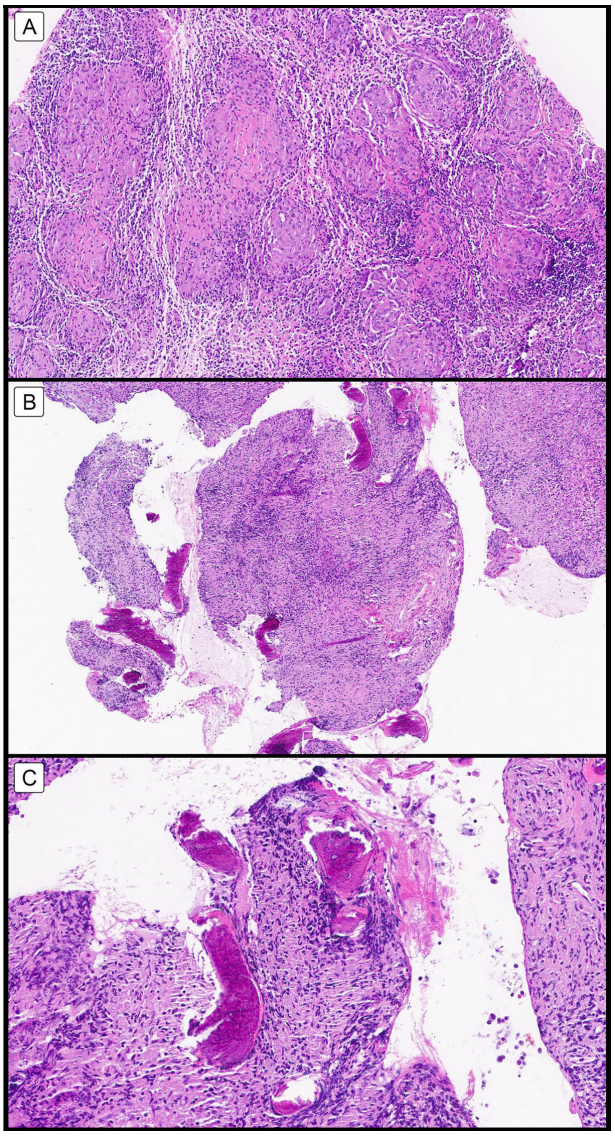

Figure 3. A, Biopsy of lesion indicating abundant non-necrotizing granulomas (hematoxylin and eosin, original magnification ×4). B, Granulomatous process involving bone (original magnification ×2). C, Granulomatous process involving bone (original magnification ×20).

tissue provides the most accurate diagnosis, but further imaging and laboratory tests should be obtained in order to assess the extent of the disease. Imaging of the lungs by CT scan should be used to stage pulmonary involvement. Serum angiotensin converting enzyme (ACE) can also be helpful for diagnosis if elevated.[8]

The mainstay of treatment for orbital sarcoidosis is oral steroids. This generally results in a good response and resolution of symptoms. A short course of oral prednisone is recommended, starting at 1 mg/kg and tapering over 3 months. If this treatment fails, more aggressive cytotoxic agents such as methotrexate may be used. In localized orbital sarcoid, a 1 ml injection of 40 mg/ml triamcinolone acetonide can be considered.[9] Patients with sarcoidosis should have long-term follow-up with a rheumatology or pulmonology specialist in order to manage active disease or relapse.[10]

Literature Search

PubMed was searched for English-language articles on June 30, 2015, using the following terms: *sarcoid posterior orbit*, *osteolytic sarcoidosis*, *eye manifestations of sarcoidosis*, and *sarcoidosis.* Sources in retrieved articles were cross-referenced.

Acknowledgments

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