Osteolytic Sarcoidosis of the Orbital Roof Masquerading as a Malignant Orbital Lesion

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Abstract. **A 51-year-old man without a significant past medical history presented with 4 weeks of progressive swelling and drooping of his left upper eyelid. A CT of the left orbit revealed an osteolytic mass lesion in the area of the lacrimal gland. A left orbitotomy with excisional biopsy was performed. The excised tissue was sent for infectious workup and histopathological examination, which revealed osteolytic sarcoidosis. The patient was treated with systemic and local injection corticosteroids, and followed over 10 months without evidence of recurrence. Systemic workup with CT of his chest, abdomen, and pelvis revealed no further evidence of sarcoidosis. To the best of the authors knowledge, this is the first report of an otherwise healthy patient presenting with isolated osteolytic sarcoidosis of the orbit and a negative systemic workup.**

Orbital masses in the region of the lacrimal gland with bony destruction classically are concerning for a malignancy, such as metastasis, multiple myeloma, or a lacrimal gland lesion. The most aggressive of these lesions is adenoid cystic carcinoma. Adenoid cystic carcinoma is the second-most common epithelial derived tumor of the lacrimal gland making up 25% to 30% of epithelial derived neoplasms in the lacrimal gland, and has an average age of presentation of 40 with a range of 12–72.[1] Due to the unencapsulated and aggressive growth of these lesions, patients typically present with symptoms, such as pain, diplopia, and paresthesias. Clinically, they can exhibit proptosis, ptosis, and a palpable mass in the superolateral orbit. Imaging often reveals a globular lesion with serrated borders owing to its infiltrative nature, and frequently, bony involvement.[1]

In contrast, orbital sarcoidosis of the lacrimal gland typically appears as a molded "V" shaped lesion on CT in the superotemporal orbit with extension beyond the orbital rim.[1] Sarcoidosis is a multisystem disease without a defined etiology that exhibits noncaseating granulomatous inflammation on histopathology. In the United States, the annual incidence is estimated at 11 per 100,000 white persons and 36 per 100,000 black persons.[2] While an estimated 90% of sarcoidosis patients have pulmonary involvement, osseous involvement is rare with a reported frequency of 3% to 13%.[3–6] Isolated erosive involvement of the bony orbit is a heretofore unreported presentation of sarcoidosis in this region. The authors present a case of orbital sarcoidosis with bony destruction that masqueraded most likely as adenoid cystic carcinoma on initial examination and studies.

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CASE REPORT

An otherwise healthy 51-year-old Caucasian man initially presented with 4 weeks of progressive discomfort, swelling, and ptosis of his left upper eyelid. He was initially treated with oral antibiotics and topical antibiotic-steroid eyedrops by his primary care physician with no improvement. He was then evaluated by an outside ophthalmologist who obtained a CT of the orbits which showed a mass lesion in the region of the lacrimal gland with bony destruction of the orbital roof (Fig. 1A, C).

On referral to the service, he had edema in the temporal portion of the left upper eyelid. He denied any associated symptoms, such as headache or visual disturbance. His corrected visual acuity was 20/40 on the right eye (OD) and 20/25 on the left eye (OS). His pupils were bilaterally reactive to light with no afferent pupillary defect in either eye. In addition, he had intact color vision, full motility, and no resistance to retropulsion. Hertel exophthalmometer measurements demonstrated 1 mm of proptosis on the left side. Given the CT scan, rapidity of onset and the concern for malignancy, the decision was made to perform a left orbitotomy with biopsy of the lesion.

During surgery, the lesion was noted to have eroded much of the superior orbital roof and frontal bone which raised concern for malignancy. Specimens were sent for aerobic culture to include Brucella, Francisella, mycobacteria, and fungi, all of which were negative. Acid-fast bacilli and Gomori-Methenamine Silver stains were also negative. Histopathologic examination of the remaining tissue revealed multiple areas of nodular-type granulomas without caseation, and several areas where the giant cells showed small asteroid bodies-consistent with sarcoidosis (Fig. 2A, B).

Given the diagnosis, he was placed on oral prednisone which was tapered over several weeks. At the conclusion of the taper, the eyelid edema returned. Given response to systemic steroid therapy, but desire to avoid further systemic glucocorticoids or steroid sparing agents, an intralesional injection of 20 mg of triamcinolone was favored. The injection was placed transcutaneously into the superolateral orbit into the lacrimal fossa. A systemic workup including CT of his chest, abdomen, and pelvis and pulmonary function testing was negative. At 10 months after presentation, his disease was clinically and radiographically stable without recurrence (Fig. 1B, D).

DISCUSSION

Sarcoidosis, first described by Hutchinson in 1878, is a multi-system disease without a defined etiology. Kreibich first described punched out osseous lesions attributed to sarcoidosis in 1904.[7] Since that time, bony involvement in sarcoidosis has been described in the literature, but remains a rare finding. Bony involvement usually occurs in advanced stages in patients with chronic multi-system disease.[3–6] To the best of the authors knowledge, this is the first report of osteolytic sarcoidosis of the orbit in a patient with a negative systemic workup.

Bony involvement in sarcoidosis is rare, and one of the landmark reviews of 29 patients who were followed for up to 43 years with bony involvement showed that 26 had involvement of the hands or feet, 3 had nasal bone involvement, and 1 patient each had involvement of the hard palate and temporal bones.[4] A more recent study reviewed 1,316 patients with sarcoidosis from 1994 and 2013 and found only 20 (1.5%) cases with bony disease, all but one of which had concomitant or prior systemic manifestations of sarcoidosis.[6] The lumbar spine and pelvis were most commonly involved; whereas only 3 patients had calvarial involvement, and all of which exhibited systemic

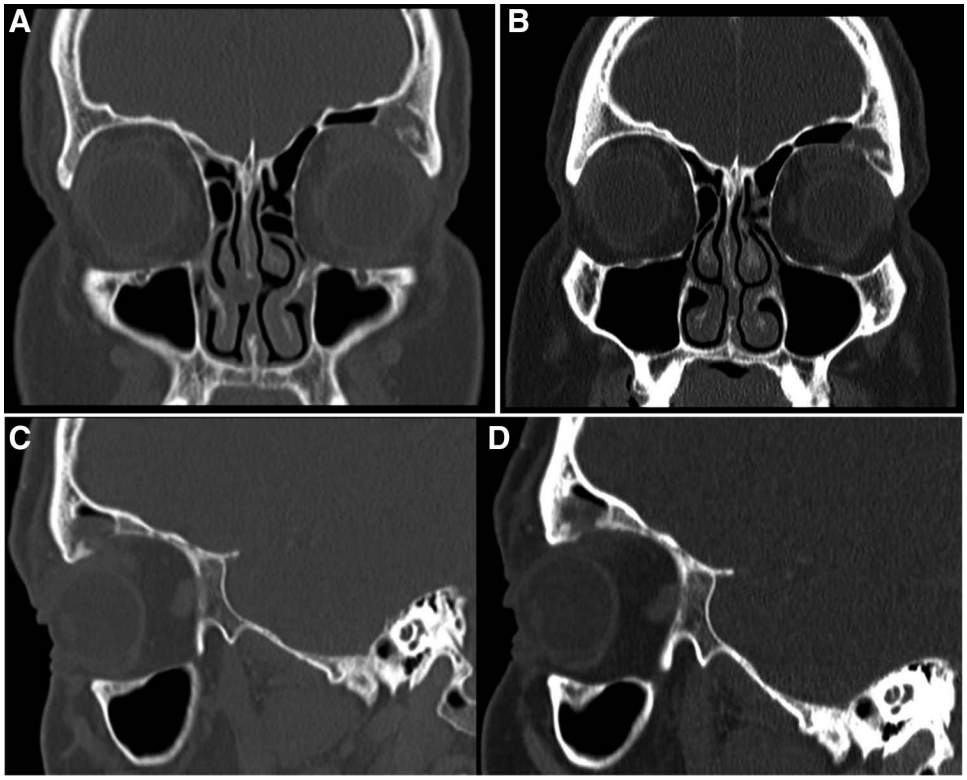

FIG. 1. **A**, Preoperative coronal CT image demonstrating an osteolytic mass lesion in the area of the left lacrimal gland with erosive destruction of the orbital roof. **B**, Postoperative coronal CT image demonstrating stability of the osteolytic changes, and no recurrence of the lesion. **C**, Preoperative sagittal CT image demonstrating an osteolytic mass lesion in the area of the left lacrimal gland with erosive destruction of the orbital roof. **D**, Postoperative sagittal CT image demonstrating stability of the osteolytic changes, and no recurrence of the lesion.

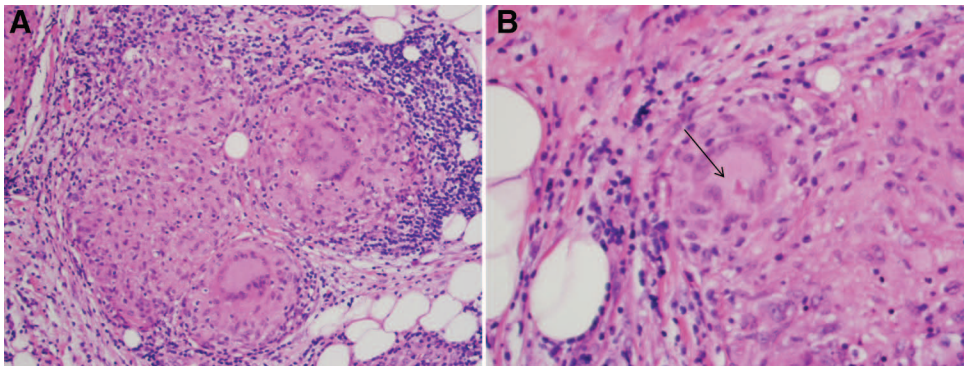

FIG. 2. **A**, Hematoxylin & Eosin slide of excised orbital tissue at 200× magnification demonstrating a mononuclear inflammatory cell reaction and multi-nucleated giant cells without evidence of caseation. **B**, Hematoxylin & eosin slide of excised orbital tissue at 400× magnification demonstrating a mono-nuclear inflammatory cell reaction and a central multi-nucleated giant cell with an asteroid body (*arrow*).

sarcoidosis. Fernandez-Ruiz et al.[3] recently described the case of a patient with a right supraorbital osteolytic sarcoid lesion, but a systemic work up revealed splenic and diffuse thoracic lymph node enlargement.

Given the patient's subacute presentation, it is impossible to determine if the sarcoidosis lesion originated in the lacrimal gland or as an osteolytic process that secondarily affected the lacrimal gland. The lacrimal gland is affected in an estimated 7% of sarcoidosis patients.[1] Two recent studies found that the lacrimal gland was the most frequently involved site in orbital sarcoidosis, with an incidence of 55% to 63% in that location.[2,8] None of the patients in either series had bony involvement. In addition, 1 study reported an 8% risk of developing systemic disease at 5-year follow up in patients with isolated orbital involvement.[2]

No consensus exists in the literature on how to treat bony involvement in sarcoidosis.[9] In the series above reporting 20 patients with bony involvement from a large cohort, 9 received treatment: 6 received prednisone, 4 were treated with methotrexate, and 3 patients each with hydroxychloroquine or tumor necrosis factor-alpha inhibitors.[6]

The case served as a lesson to maintain a broad differential diagnosis when encountering orbital lesions with bony

destruction, as well as a reminder that sarcoidosis may mimic many other disease processes. Finally, it remains important to follow these patients closely given the risk for development of systemic sarcoidosis.

REFERENCES

1. Spencer WH, Jakobiec FA, Font RL, et al. *Ophthalmic Pathology an Atlas and Textbook*. 3rd ed. Vol. 3. Philadelphia, PA: W. B. Saunders Company, 1986:2730–7.
2. Demirci H, Christianson MD. Orbital and adnexal involvement in sarcoidosis: analysis of clinical features and systemic disease in 30 cases. *Am J Ophthalmol* 2011;151:1074–1080.e1.
3. Fernández-Ruiz M, Guerra-Vales JM, Castelbón-Fernández FJ, et al. Sarcoidosis presenting as an osteolytic skull lesion: a case report and review of literature on skull sarcoidosis. *Clin Rheumatol* 2007;26:1745–8.
4. Neville E, Carstairs LS, James DG. Sarcoidosis of bone. *Q J Med* 1977;46:215–27.
5. James DG, Neville E, Siltzbach LE. A worldwide review of sarcoidosis. *Ann N Y Acad Sci* 1976;278:321–34.
6. Sparks JA, McSparron JI, Shah N, et al. Osseous sarcoidosis: clinical characteristics, treatment, and outcomes–experience from a large, academic hospital. *Semin Arthritis Rheum* 2014;44:371–9.
7. Young RC, Jr, Rachal RE, Cowan CL, Jr. Sarcoidosis–the beginning: historical highlights of personalities and their accomplishments during the early years. *J Natl Med Assoc* 1984;76:887–96.
8. Mavrikakis I, Rootman J. Diverse clinical presentations of orbital sarcoid. *Am J Ophthalmol* 2007;144:769–75.
9. Suri V, Singh A, Das R, et al. Osseous sarcoid with lytic lesions in skull. *Rheumatol Int* 2014;34:579–82.