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Orbital Granuloma Annulare as Presentation of Systemic Sarcoidosis

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Abstract

Purpose: To report a unique case of an orbital mass with atypical histopathologic features that although overlap with other granulomatous disorders, likely represents orbital involvement of systemic sarcoidosis.

Methods: Observational case report.

Results: A 51-year-old woman presented with an orbital mass and was found to have mild proptosis and restriction of ocular motility. The remainder of her ophthalmic exam was unremarkable. Excisional biopsy revealed ill-defined granulomas with central necrosis, chronic inflammation and sclerotic collagen; several features consistent with granuloma annulare. Further systemic workup including laboratory and radiologic studies were compatible with a diagnosis of systemic sarcoidosis.

Conclusion: Our case emphasizes the importance of a complete systemic workup with the detection of an orbital granuloma to ensure accurate diagnosis and appropriate management.

Keywords

Granuloma annulare; orbital granuloma; sarcoidosis

Sarcoidosis is multisystem granulomatous disorder of unknown etiology. We describe a patient with an orbital mass with unique and unusual histopathologic features that presented as the initial manifestation of systemic sarcoidosis.

CASE REPORT

A 51-year-old African American female was referred for evaluation of an orbital mass found on imaging for orbital pain. Her medical history was significant only for hypertension. Examination revealed a tender mass near the supraorbital rim, 2 mm of right-sided axial

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DECLARATION OF INTEREST

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

proptosis and restriction of motility by 75% on upgaze without associated hypoglobus. The remainder of the ophthalmic examination was normal. Review of CT brain and MRI of brain and orbits demonstrated a poorly circumscribed mass ($19 \times 16 \times 13$ mm) in the right superomedial orbit (Figure 1).

An anterior orbitotomy and excisional biopsy was performed. Histopathology revealed fibroconnective tissue with sclerotic collagen, chronic inflammation, and ill-defined granulomas with central necrosis (Figure 2). Increased stromal mucin confirmed by colloidal iron stain was present. AFB, Fite and GMS stains for acid-fast bacilli and fungi as well as flow cytometry for lymphoma were negative. The morphologic and histochemical features suggested a diagnosis of granuloma annulare. Laboratory work up was initiated to rule out possible associated systemic disease. Abnormal laboratory results included elevated angiotensin converting enzyme (91 U/L), lysozyme (23 mcg/ml), erythrocyte sedimentation rate (89 mm/hr), C-reactive protein (2.8 mg/dl) and a positive quantiferon gold.

Other tests including FTA-Abs, ANCA, ANA, anti-DS DNA, anti-SM, anti-SSA, anti-SSB, anti-SCL-70 and rheumatoid factor were negative. CT chest revealed significant hilar and mediastinal lymphadenopathy as well as numerous hypodensities within the liver and spleen, suggestive of sarcoidosis. Pulmonary function test values showed moderate obstructive disease. A rheumatology consult was obtained and the patient was started on methotrexate for systemic sarcoidosis. On follow-up, 7 months after initial presentation, she had no recurrence of her orbital disease.

COMMENT

We present unique and unusual histopathologic findings of a patient with an orbital mass. Based on initial pathology results, a tentative diagnosis of granuloma annulare was made. A benign granulomatous condition that affects the skin and less commonly the subcutaneous tissues, granuloma annulare classically shows necrotic collagen surrounded by a zone of histiocytes and fibroblasts with increased amounts of mucin on pathology.¹ The subcutaneous form is hypothesized as an isolated finding without any systemic association according to recent literature.^{1,2} However, as it has been previously linked to various disorders, we initiated a systemic work up to rule out any associated disease.

From the results of our systemic review, our patient's clinical picture was compatible with a diagnosis of systemic sarcoidosis, although the histopathological features of the orbital mass (central necrosis, sclerotic collagen, increased mucin) are atypical for the disease. Central necrosis within sarcoid granuloma, although uncommon, has been previously described but it is crucial to rule out other causes of granulomas that may present with compatible features.^{3,4} In addition, our case was initially diagnosed as granuloma annulare due to similar overlapping histopathologic characteristics and interestingly, these diseases have been previously associated.⁵⁻⁷ However, it is unclear if these cases may represent histopathologic variants of sarcoid, as focal necrobiosis associated with granulomas was found in 14% and increased dermal mucin was present in 18% of cutaneous biopsies of patients with confirmed systemic sarcoidosis.⁴ Further investigation is needed to better understand if there is a common pathogenetic mechanism between these entities or if

sarcoidosis may histologically mimic other granulomatous disorders such as granuloma annulare.

As our case illustrates, it is of great importance to conduct a complete systemic workup with the finding of an orbital granuloma as management will differ based on the underlying etiology. Granuloma annulare has a favorable prognosis as it is a benign process and treatment is most commonly observation.¹ In contrast, 50–2% of patients with orbital sarcoidosis and no prior history of systemic disease were found to have evidence of systemic sarcoidosis upon further investigation.^{3,8,9} Thus, sarcoidosis should be considered in the evaluation of an orbital mass, even if the patient is asymptomatic at the time of diagnosis. Corticosteroids are typically used and are effective in the treatment of orbital and systemic disease.⁹ For localized orbital disease, intralesional injection of corticosteroids or surgical excision or debulking have also been used with good results and may avoid systemic immunosuppression.^{8,9}

In conclusion, we describe a case of an orbital mass with unique and unusual histopathologic features that overlap with other granulomatous disorders. Our case emphasizes that a thorough systemic workup should be done to ensure accurate diagnosis and determine appropriate further management.

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FIGURE 1.
(A–D) Computerized tomography scan of the brain without contrast, axial section, demonstrating the extent of the mass in the right superomedial orbit.

FIGURE 2.

Orbital biopsy of deep orbital mass. (A) Representative section of the specimen showing a granuloma with central necrobiosis, chronic inflammation and sclerotic collagen (Hematoxylin-eosin, 200 × magnification). (B) Colloidal iron stain showing increased stromal mucin in area of collagen degeneration (Colloidal iron, 100 × magnification).