Orbital tumor presented systemic sarcoidosis

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ABSTRACT

Sarcoidosis is a granulomatous, multisystemic disorder of unknown origin usually affecting young Black-American adults. Bilateral hilar lymphadenopathy and skin or eyelid lesions are the most common symptoms noted. Except for lacrimal gland enlargement, orbital involvement with sarcoidosis is rare and is usually unilateral when it occurs. The aim of this article is to report an isolated case of sarcoidosis that initially presented as an orbital tumor, and to document the CT and MR appearance of the lesions.

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Sarcoidosis is a granulomatous systemic disease of unknown etiology characterized by subacute or chronic inflammation involving multiple systems, including orbital and ocular structures. It is an immunologically mediated disease effecting delayed hypersensitivity. It is much more common in Black Americans. Virtually any part of the globe or orbit may be involved in sarcoidosis. Uveitis and chorioretinitis, keratoconjunctivitis, and conjunctival inflammatory nodules may be seen.[1-3] The most common form of orbital involvement in sarcoidosis is chronic dacryoadenitis, which is often unilateral.[1] Orbital sarcoid occurs predominantly in women older than 50 years.[2] In this article, we report a systemic sarcoidosis case presenting with orbital involvement at a younger age with respect to other cases in the literature.

Case Report. An 11-year-old Caucasian female was referred to the eye clinic with swelling, blurred vision, and redness of the left eye. External examination revealed a 1.5 mm left sided exophthalmos with obvious limitation of looking laterally. There was no history of nystagmus, ptosis, or neurologic deficits. An axial postcontrast CT scan showed a homogenous, hypodense retroorbital mass invading the lateral and superior rectus muscles. The mass that has caused exophthalmos abuts the superior ophthalmic vein. There was no bone destruction. T1- and T2-weighted MR scans showed the mass on the superolateral aspect of the left bulbus oculi invading the lacrimal gland, and superior and lateral rectus muscles. The bulbus oculi was displaced anteriorly. The posterior aspect of the bulbus oculi was irregular on T1-weighted images, and the mass was iso-minimal hyperintense compared to the rectus muscles. On T2-weighted images, the mass was hypointense compared to the rectus muscles. There was pathologic enhancement in the mass, which contains small amounts of calcifications and measured 3.7 x 3.5 x 3.2 cm in size (**Figures 1 & 2**). Thorax CT showed bilateral hilar lymphadenopathy. A biopsy specimen of the orbital wall showed fibro adipose tissue with multiple noncaseating granulomas.

Discussion. Seven to 19% of patients with systemic sarcoidosis present with ophthalmic symptoms, most commonly uveitis. Initial orbital involvement is rare. In 1978, Obenauf et al[3] reviewed 532 patients with sarcoidosis and reported that of 101 patients who presented with ophthalmic symptoms, 2 had unilateral proptosis caused by orbital granulomas. In 1956, Stein and Henderson[4] stated that only 2 cases of unilateral orbital involvement had been seen in more than 200 pathologically proven cases of generalized sarcoidosis. Pseudotumor, lymphoma, and other granulomatous diseases should be considered in the differential diagnosis.

Sarcoidosis usually appears in the second and third decades of life and most commonly in the Black American population. In 23 cases of orbital sarcoidosis, Henderson[5] described the mean age of the patients to be 57 years. Orbital sarcoid occurs predominantly in women older than age 50. Unlike the literature findings, our patient displayed the clinical manifestations at 11 years old.

In 1986, Collison, Miller and Gren[6] reported on 15 patients with biopsy-proven orbital sarcoidosis involving the lacrimal gland, orbital fat and connective tissue, or both.[6] Of the 12 patients not previously known to have sarcoidosis, systemic evidence of the disease was found upon evaluation in 11. One

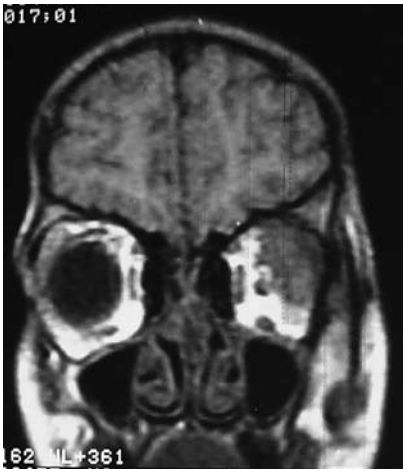

Figure 1 - Coronal T1A weighted MRI showing a soft tissue mass invading the lateral and superior rectus muscles in the lacrimal gland localization.

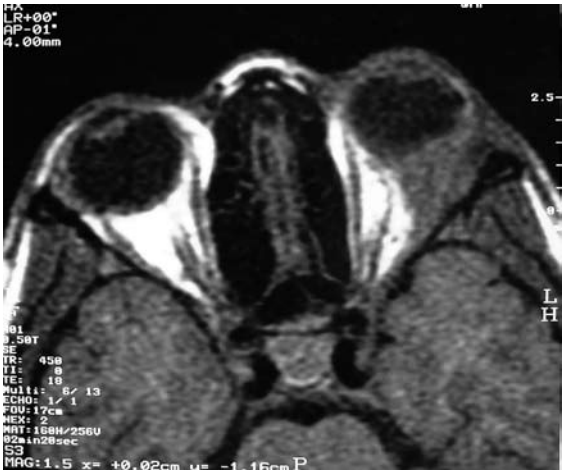

Figure 2 - Axial contrast enhanced T1A weighted MRI image showing homogeneous enhancement of the lesion.

patient, with unilateral lacrimal gland involvement only, had no evidence of systemic sarcoidosis. As demonstrated by our patient, systemic sarcoidosis rarely may present initially with unilateral orbital involvement.

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