A rare case of eyelid sarcoidosis presenting as an orbital mass

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Intraorbital sarcoidosis presenting externally as a solitary eyelid mass has been described in the literature as isolated case reports. We describe a rare case of asymptomatic sarcoidosis with orbital mass as the presenting feature in a young woman. The lesion was excised with the clinical possibility of a thrombosed varix. On histology, the lesion was characterized by numerous nonnecrotizing epithelioid cell granulomas with several multinucleated giant cells containing abundant asteroid bodies and oxalate crystals. No tubercular bacilli were detected. A diagnosis of sarcoidosis was rendered and on further clinical work-up, she was detected to have hilar lymphadenopathy. Sarcoïdosis should be considered in the differential diagnosis of orbital mass as it could be the initial manifestation of the disease process.

Key words: Eyelid, orbital mass, sarcoidosis

Sarcoidosis is a systemic disease with an insidious onset most often affecting adolescents and young adults and presenting frequently with bilateral lymphadenopathy with or without pulmonary infiltration.[1] It is often diagnosed incidentally on routine chest radiography done as a part of evaluation for other diseases. The incidence varies according to age, sex, race, and geographic origin. In an appropriate clinical setting, the diagnosis of sarcoidosis is established based on the demonstration of noncaseating epithelioid cell granulomas which are negative for acid-fast bacilli.[2] Intraorbital sarcoidosis presenting externally as a solitary eyelid mass is extremely uncommon and reported in the literature as isolated single case reports.[2–5] We present an unusual case of asymptomatic sarcoidosis presenting with an orbital mass as the initial manifestation.

Case Report

A 33-year-old woman presented in the eye clinic with left lower eyelid swelling of 1-year duration without any accompanying systemic symptoms. There was no history of tattooing, trauma, or injection of any sort in the vicinity of the left eye. Examination revealed a bluish-black ill-defined elevated nodular lesion in the left lower eyelid associated with restricted down gaze [Fig. 1a]. Coronal contrast enhanced computed tomography (CT) image of the orbit showed a well-defined homogeneous soft tissue mass in the inferior extraconal space [Fig. 1b]. The fat plane with the inferior aspect of the globe was lost and no deformity of the globe or erosion of the orbital floor was seen. With a clinical possibility of thrombosed varix, inferior orbitotomy was done. Skin incision 4 mm below the lid margin was given and orbicularis muscle was dissected. A lobulated, dark bluish mass was seen which was adherent to the orbital septum and was arising from the periorbita of the orbital floor. The lesion was excised and submitted for histopathological examination. Gross examination revealed a globular, gray-white tissue measuring 1.5 cm in its greatest dimension. Microscopic examination demonstrated multiple, compact, well-formed, nonnecrotizing epithelioid cell granulomas devoid of any lymphocyte cuffing with numerous multinucleated giant cells containing abundant asteroid bodies and oxalate crystals [Fig. 1c and d]. Stain for acid-fast bacilli was negative, and a diagnosis of sarcoidosis was offered. A retrograde work-up for tuberculosis was done. Mantoux test was negative; however, contrast enhanced and high-resolution CT of the chest showed calcified hypodense right hilar lymph nodes largest measuring 1.1 cm × 0.8 cm. Her blood counts, thyroid function tests, serum calcium, 25(OH) Vitamin D₃, phosphate, angiotensin-converting enzyme (ACE), and albumin levels were normal. However, the autoimmune workup was significant as she had elevated thyroid peroxidase antibodies (63.76 IU/ml), speckled antinuclear antibody (ANA) and elevated C-reactive protein (6 mg/L). Her rheumatoid arthritis factor was negative. Hence, a diagnosis of subclinical sarcoidosis was rendered.

Discussion

Sarcoidosis is an immune-mediated multisystem granulomatous disease of unknown etiology with varied clinical manifestations affecting genetically and ethnically predisposed individuals.[1] While sarcoidosis presenting as eyelid nodules is uncommon, seen in 3% of patients with chronic sarcoidosis,[3] such a mass as the initial manifestation is exceedingly uncommon with single rare cases reported in the English literature.[3–5] In the index case, the histopathological diagnosis prompted further work-up, thereby asymptomatic pulmonary involvement and circulating autoantibodies were detected. This is not uncommon because the association of sarcoidosis with autoimmune diseases is well known.[7] The patient had a positive ANA test with a speckled pattern which is the most...
common but least specific pattern encountered. As only 60% of patients with sarcoidosis have raised ACE levels\(^8\) the normal ACE level does not argue against the diagnosis of sarcoidosis in this woman.\(^9\) Since she was completely asymptomatic, we decided to wait and watch instead of proceeding with further specific tests such as Smith antibody (anti-Smith), ribonucleoprotein antibody, Scl-70 kD kinetochore, (anti-topoisomerase I) and anti-La (anti-SSB). In addition, she had elevated thyroid peroxidase antibodies which is also well documented in various studies.\(^{[9,10]}\)

While the hypothyroid and euthyroid state in Hashimoto’s thyroiditis is well known and such a diagnosis has therapeutic and prognostic implications; complete thyroid function tests done were, however, normal. The patient was explained regarding her basic disease predispositions and advised close follow-up for the recurrence of primary disease and development of other autoimmune manifestations.

**Conclusion**

This case highlights the importance of considering sarcoidosis in the differential diagnosis of orbital mass and such a presentation could be the initial manifestation of the disease process.

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**Conflicts of interest**

There are no conflicts of interest.

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