

Orbital Sarcoidosis Presenting as Diffuse Swelling of the Lower Eyelid

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The author reports a case of orbital Sarcoidosis in a 70-year-old female that initially presented as diffuse swelling of the lower eyelid. The patient complained of painless swelling of the left lower lid without palpable mass, and a computerized tomography (CT) scan of the orbit was unremarkable. A serum angiotensin converting enzyme level was elevated, and hilar lymphadenopathy was noted on the chest CT. The patient underwent surgical debulking for histologic confirmation, which led to a final diagnosis of sarcoidosis involving the orbital fat. Unexplained chronic eyelid swelling without a mass should be considered a possible ophthalmic manifestation of orbital sarcoidosis.

Key Words: Chronic swelling, Lower lid, Orbital sarcoidosis

Sarcoidosis is a multisystem granulomatous disorder of unknown etiology. Ocular involvement of Sarcoidosis mainly includes uveitis and conjunctival granulomas, whereas orbital tissue involvement is rarely reported, varying from lacrimal gland infiltration to optic nerve sheath involvement [1-4]. Most lesions have granulomatous masses, but infiltration of the orbital adipose tissue without a mass lesion has not been reported. We managed a patient with sarcoidosis involving the orbital fat, and present the clinical features and outcomes here.

Case Report

A 70-year-old female presented with a history of painless swelling of the left lower eyelid for one year (Fig. 1A). She had no systemic complaints. On examination, her visual acuity was 20 / 20 (left eye), and a slitlamp examination of the left eye revealed no abnormalities except for corneal granular dystrophy. The lower eyelid of the left eye was diffusely swollen without any palpable mass. The

left upper eyelid showed a 3-mm ptosis with 10-mm levator function, without swelling, and the ptosis resolved after administration of 2.5% phenylephrine in the left eye. No exophthalmos was present, and the extraocular movements were normal. A fundus examination revealed no abnormalities. A computerized tomography (CT) scan of the orbit was unremarkable (Fig. 1B and 1C).

The patient's white blood cell count, chemistry panel, and thyroid function tests were normal. The erythrocyte sedimentation rate was 31 mm in the first hour, and serum angiotensin converting enzyme (ACE) levels were 58 units (normal, 8 to 52 units). A chest radiograph revealed an old tuberculous lesion without hilar lymphadenopathy (Fig. 2), and a *Mycobacterium tuberculosis* antigen-specific interferon gamma assay was negative. As these results were inconclusive, surgical debulking of the swollen lower eyelid via an inferior conjunctival fornical approach for diagnosis and treatment was performed, as well as a conjunctivo-mullerectomy for ptosis correction. The operative findings showed stiff and coarse orbital fat particles, and the histopathologic examination demonstrated non-caseating granulomas consisting of epithelioid histiocytes in the orbital fat, most likely caused by sarcoidosis (Fig. 3). Special stains for acid-fast bacilli and fungi were negative.

The patient was further reviewed by a pulmonologist who noted hilar lymphadenopathy on the chest CT. The patient was diagnosed with orbital sarcoidosis, and started

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Fig. 1. (A) Photograph of a 70-year-old female with diffuse swelling of the left lower eyelid. Computed tomographic scans, (B) axial and (C) coronal, showed no evidence of a mass lesion or soft tissue infiltration.

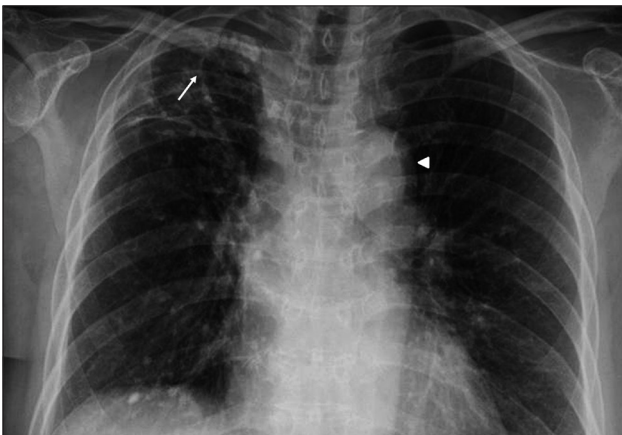


Fig. 2. Chest radiograph shows multiple confluent irregular opacities (arrow) in right upper lobe. There are no remarkable findings in hilar region with shadow of aortic arch (arrowhead).

on prednisolone (10 mg/day) in consideration of the significant improvement in eyelid swelling after surgical debulking (Fig. 4). The symptoms completely resolved within one month, and she has been followed for 12 months without a recurrence.

Discussion

The distinctive feature of our case is the initial presentation of diffuse eyelid swelling without mass lesions, even on CT examination. Despite the suspicion of sarcoidosis with a slightly elevated ACE level, inconclusive laboratory and radiologic results necessitated a biopsy to verify the diagnosis. In our patient with diffuse inflammatory lesions without any signs of a mass effect, surgical debulking of the lower lid was considered diagnostic and curative. The histologic examination showed non-caseating granulomatous infiltration in the orbital fat, which was consistent with sarcoidosis, and infectious etiologies were ruled out, as well as foreign body granulomatous disease. Subsequent investigations revealed hilar lymphadenopathy on a chest CT. A prompt response to systemic steroids also supported the diagnosis of sarcoidosis.

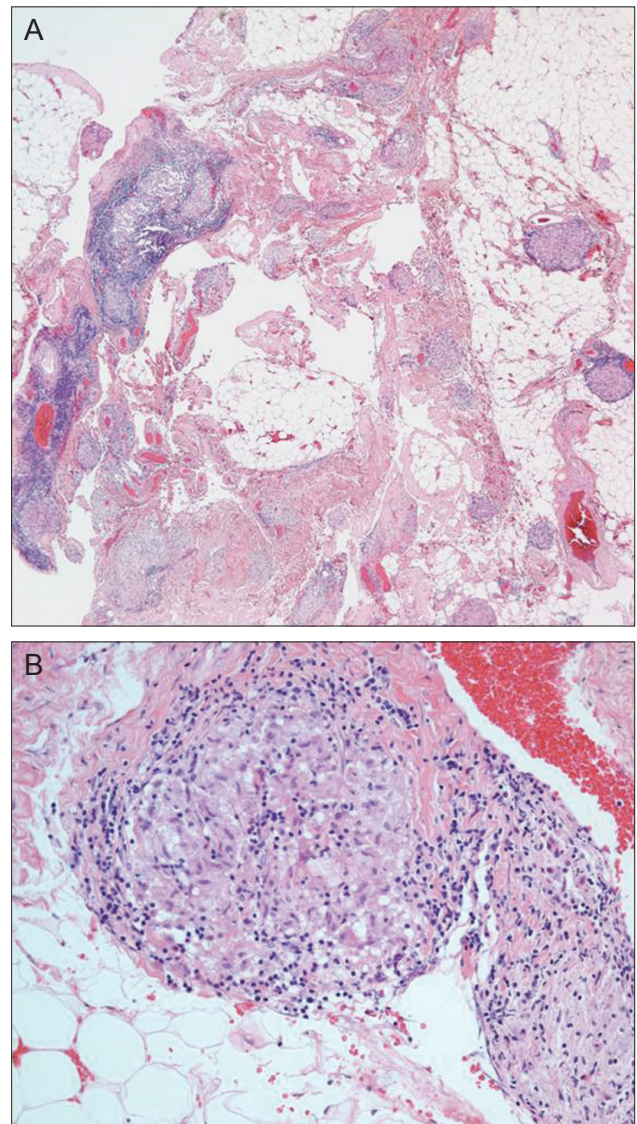


Fig. 3. (A) There are multifocal granulomas with lymphocytic infiltration in the fat (H&E, $\times 40$). (B) High magnification view shows a well-formed, non-caseating granuloma composed of epithelioid histiocytes with surrounding lymphocytes (H&E, $\times 400$).

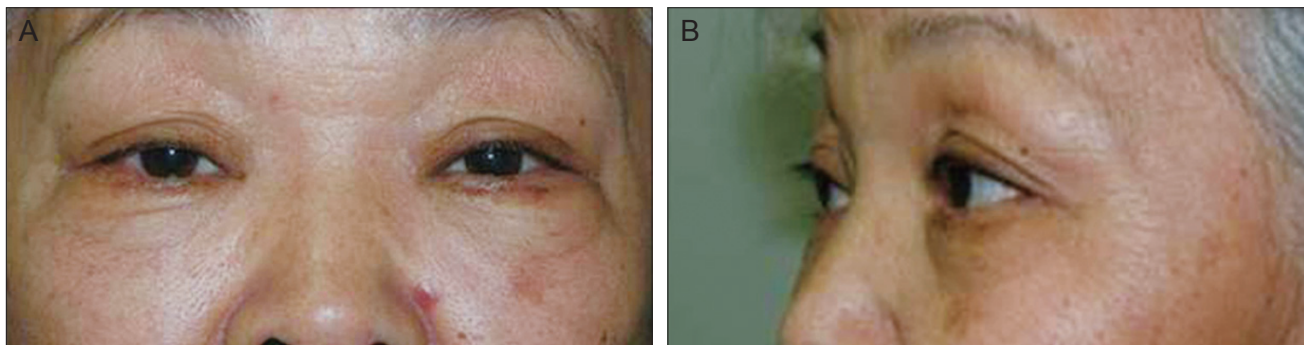


Fig. 4. (A) The patient showed significant improvement of the left lower lid swelling after surgery. (B) Left lower lid swelling was completely resolved after low dose steroid treatment.

Systemic corticosteroids are known to be effective in the treatment of orbital sarcoidosis. Although an initial dose of 1 mg/kg of body weight of oral steroids and gradual tapering is recommended [5], our patient was initially treated with 10 mg of oral prednisolone in consideration of the improvement in eyelid swelling after surgical debulking, and she responded promptly without a recurrence. Surgical debulking might reduce the required dose of steroids by eliminating the amount of inflammatory tissues and enhancing drug diffusion.

To the best of our knowledge, the case reported here is the first report of orbital fat infiltration of sarcoidosis without palpable mass in lower lid. A few examples of soft tissue involvement in orbital sarcoid have been reported, but most of them occurred within cutaneous or connective tissue, and isolated orbital fat infiltration has rarely been documented [5-7]. A case of retrobulbar fat infiltration of sarcoid mimicking a superior orbital fissure syndrome has been reported [8], our case is unique in that only the lower eyelid was involved, and there were no remarkable findings on the orbital CT scan. In conclusion, unexplained chronic eyelid swelling without clinical evidence of an orbital mass should be considered as a possible ophthalmic manifestation of orbital sarcoidosis.

Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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