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Extrapulmonary sarcoidosis: unusual cause of epiphora

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ABSTRACT

Sarcoidosis is a multisystemic granulomatous disease of unknown aetiology mainly affecting African-Americans, Scandinavians, and the Irish. However, individuals of other races and ethnicities are still not immune. The clinical presentations vary widely with most patients having some respiratory problems. Though extrapulmonary sarcoidosis is no longer rare, sarcoidosis involving the lacrimal sac is an infrequently-reported problem. We present a case of sarcoidosis involving the lacrimal sac in a 42-year-old Malay woman who presented with epiphora. She was successfully treated with steroids and dacryocystorhinostomy. There was no evidence of systemic involvement. It is suggested that in an unusual presentation of sarcoidosis, a thorough search should be made for localisation of other sites, lungs in particular, even in the absence of respiratory complaints.

Keywords: epiphora, lacrimal sac, nasolacrimal duct, sarcoidosis

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INTRODUCTION

Sarcoidosis is a chronic multisystemic granulomatous disease of unknown aetiology, believed to be due to exaggerated cellular immune response to a variety of self and non-self antigen.[1] The course of sarcoidosis ranges from asymptomatic to severe, and even lethal disease.[2] The clinical manifestations and severity of the disease vary widely, and are strongly associated with racial and ethnic factors.[2] Acute and more severe disease is typical of African American patients, whereas asymptomatic and chronic presentations are more frequent in caucasians.[3] Sarcoidosis affects people of all ages, with the peak age of onset in the third decade of life.[4] The disease affects predominantly the lungs and thoracic lymph nodes, with most of the patients having some respiratory problems.[4] Majority of the patients have constitutional symptoms such as fever, malaise, fatigue, and weight loss.[2] The most frequently affected extrapulmonary organs are the lymph nodes, eyes, and skin.[2]

Ocular involvement manifests in 25%–60% of patients with systemic sarcoidosis. However, ocular presentation can be the initial finding in sarcoidosis, which should initiate systemic screening.[2] Ocular sarcoidosis may present with a wide variety of ocular symptoms in all parts of the eye, and may be associated with chronic and progressive intraocular inflammation leading to visual deterioration.[2] The most common presentations are uveitis and conjunctival nodule.[2] Lacrimal sac involvement is still considered uncommon with less than 30 cases reported.[5-8] We report a case involving the lacrimal sac in a Malay woman.

CASE REPORT

A 42-year-old Malay woman presented to us a two-month history of left epiphora, without any indication suggestive of recurrent dacyrocystitis. She also complained of progressive eversion of her left lower lid. There were also noticeable small skin nodules, which were increasing in number without any signs of inflammation. She postulated that the presence of the nodules was the cause of her epiphora and was keen on the removal of the nodules. She denied any history of trauma or allergy. There was no history of respiratory problems. She was on treatment for ischaemic heart disease and hypercholesterolaemia for the past one year.

Her visual acuity was 6/6 in both eyes. There were multiple multinodular conjunctival lesions at the left lower fornix, without any sign of inflammation, which caused mild ectropion (Fig. 1). Several visible and palpable skin nodules were also noted scattered around the left lower eyelid (Fig. 1). There was no evidence of anterior or posterior uveitis. The fundus examination was unremarkable. On systemic examination, there were no evidence to suggest systemic sarcoidosis. Her lung function test was normal. There was a "hard stop" upon syringing of the left nasolacrimal duct, with failure of the fluid to reach the nostril, suggesting complete obstruction of the duct.

B-scan ultrasonography of the sac revealed thickened walls filled with mucous and a "floating" nodule inside the sac (Fig. 2). Computed tomography (CT) of the orbit did not show any intraorbital or

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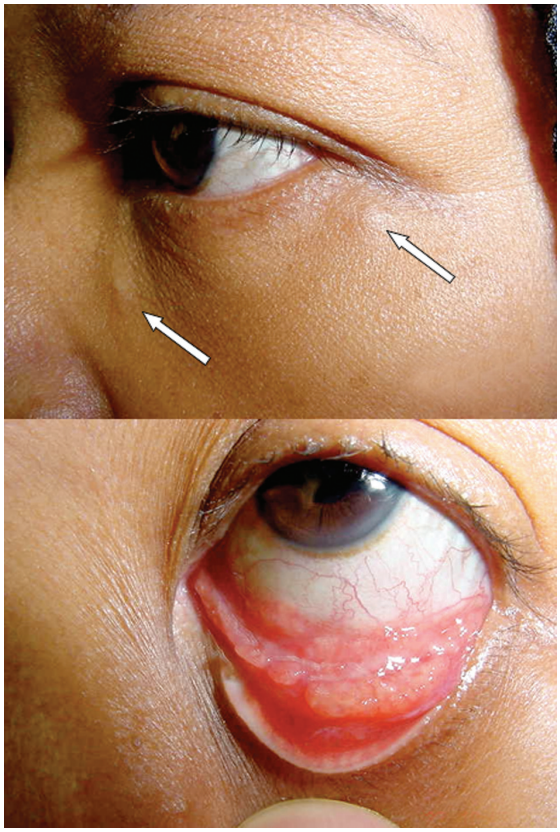


Fig. 1 Photographs of the patient at presentation show left palpebral conjunctival granuloma and eyelid skin nodules (arrows).

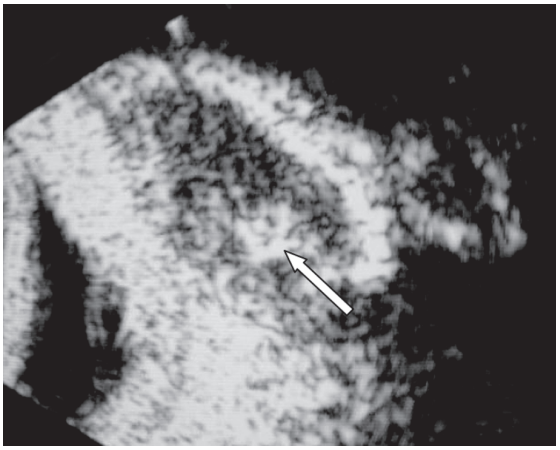
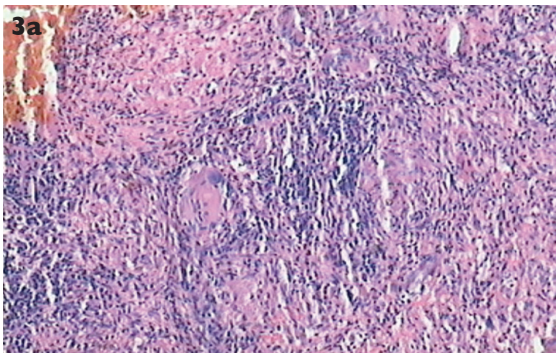


Fig. 2 B-scan ultrasonographical image of the left lacrimal sac taken in longitudinal plane (along the sac vertically) shows a thickened wall with a nodule floating inside (arrow).



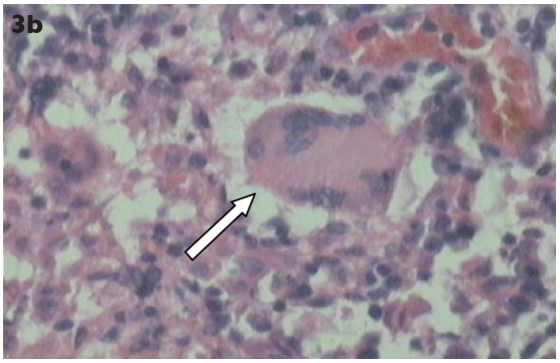


Fig. 3 Photomicrographs show (a) chronic inflammation with central noncaesating granuloma seen in the nodule biopsied from the lacrimal sac (Haematoxylin & eosin, x 200), and (b) the magnification of a multinucleated giant cell (arrow) surrounded by lymphocytes and plasma cells (Haematoxylin & eosin, x 400).

lacrimal gland involvement. Angiotensin converting enzyme (ACE), serum and urine calcium levels were not raised. Mantoux reading and erythrocyte sedimentation rate were also not significant. There was no evidence of hilar adenopathy or any interstitial lung disorder on chest imaging. However, excisional conjunctival biopsy revealed noncaseating, multinucleated giant cells with multiple epitheloid granuloma suggestive of sarcoidosis. Staining for acid-fast bacilli and bacteria proved negative.

Oral prednisolone was instituted in tapering dose over a period of three months. Although the conjunctival granuloma regressed tremendously and her ectropion improved, her epiphora persisted. Left conventional dacryocystorhinostomy (DCR) with stenting was performed after the completion of oral prednisolone. Intraoperatively, thickened lacrimal sac walls with two nodules were found inside the sac. Biopsy of the nodule also revealed similar evidence suggestive of chronic non-caseating granulomatous inflammation (Fig. 3). The stenting was removed after six months postoperatively. At 24 months follow-up, patient remained symptom-free with minimal residual conjunctival lesion.

DISCUSSION

Sarcoidosis involving the lacrimal sac may present with epiphora without evidence suggesting an underlying lacrimal sac tumour or disease. It may also present as recurrent dacryocystitis, which may mask the diagnosis and can only be confirmed by histological findings from a biopsy obtained intraoperatively.[5-7] Previous

reports found that there was no associated or simultaneous granulomatous uveitis in sarcoidosis of the lacrimal sac except one case by Harris et al.[7] In the absence of known causative agents, the diagnosis of sarcoidosis remains a diagnosis of exclusion.[2] Furthermore, without definitive classical clinical features and microscopical markers that are pathognomonic for the disease, the diagnosis of sarcoidosis depends on a combination of historical, clinical, laboratory, and histopathological evidence.[1] Elevated serum ACE level, which was believed to reflect the activity of the disease, may be affected by corticosteroid usage. However, it is not specific enough as a single definitive diagnostic tool.[2]

B-scan ultrasonography, which is noninvasive and relatively inexpensive, is useful to detect the presence of "floating" nodules in the lacrimal sac. However, this imaging technique is not highly specific. The B-scan findings in this case further heightened our suspicion of sarcoidosis involving the lacrimal sac. The sensitivity of the chest radiograph was reported to be 80% in active systemic sarcoidosis. However, chest CT and gallium-67 scanning have been reported to be more sensitive.[2] Gallium scans and transbronchial biopsies have been useful in providing evidence of pulmonary involvement in patients with sarcoidosis in the presence of clear chest radiographs.[7] Nevertheless, the tests are expensive, invasive and not readily available in certain places. Despite the absence of gallium scan in our set-up, we believe the evidence of noncaseating granuloma on tissue biopsy together with compatible clinical features is adequate proof of sarcoidosis. The histological findings may also aid in excluding other differential diagnosis, such as bacteria or fungi infection, malignancies and other inflammatory conditions, such as Wegener's granulomatosis.

DCR, using either an open or endoscopic technique, is recommended in the treatment of nasolacrimal duct obstruction. However, the open technique is preferred, due to reportedly higher success rates, especially in cases with high risks of recurrent obstruction such as sarcoidosis.[9] Chapman et al recommended the creation of a larger than usual rhinostomy and the placement of a stent to enhance the patency of the DCR.[5] Postoperative instillation of corticosteroid eye drops has also been suggested. Further treatment is aided by the presence of intranasal granulation tissue endoscopically.[5] We achieved satisfactory results with just the standard size of rhinostomy and stenting, without any additional steroid treatment postoperatively. We postulated the outcome was due to the absence of systemic sarcoidosis. Racial factors may also influence our outcome. Ophthalmic sarcoid lesions may remain as isolated, chronic presentations of sarcoidosis without physical, radiological, or even laboratory evidence of systemic disease. A high index of suspicion, and extensive investigations to rule out other exclusion diagnoses, is important to aid in the diagnosis and to reduce ocular morbidity.

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