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# Sarcoidosis presenting as lacrimal gland enlargement: Eyes speak the truth

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### Abstract:

Bilateral lacrimal gland enlargement is uncommon; however, its presence induces brainstorming process and intensive discussion between a clinician and an imaging specialist, leading to exploration of multiple systemic disease patterns such as lymphoproliferative disorders, sarcoidosis, Sjögren's syndrome, and tuberculosis. Thoughtful analysis and diagnostic work-up are required to confirm the diagnosis. Sarcoidosis is a rare systemic disease, with ocular involvement being still rarer. Here, we report a case of a young male presenting with nodular swelling over lateral aspects of both the eyes. The imaging study revealed bilateral lacrimal gland enlargement. Further work-up revealed mediastinal and hilar lymphadenopathy with pulmonary nodules which along with biochemical tests lead to the diagnosis of sarcoidosis. The case highlights the ocular symptoms in sarcoidosis and clinicroadiological approach to bilateral lacrimal gland enlargement.

### Keywords:

Inflammation, lacrimal gland, sarcoidosis

## Introduction

Systemic sarcoidosis may show ocular involvement in up to 79% of the cases;<sup>[1]</sup> however, this incidence varies in different regions such as 13% in a Turkish study and 79% in a Japanese study.<sup>[2]</sup> It may involve all parts of the eye, with uveitis being the most common presentation. Lacrimal gland involvement in sarcoidosis can rarely be the initial manifestation of the disease, with no specific imaging features. Diagnosis of sarcoidosis in tubercular-endemic regions is a challenge both for a clinician and for a radiologist and requires detailed background knowledge of various diseases considered in the differential diagnosis.

We present a rare case of systemic sarcoidosis, presenting with lacrimal gland enlargement, wherein the emphasis is on the radiological manifestations of the disease. The case also describes the imaging and clinical features of causes of lacrimal gland enlargement.

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## Case Report

A 28-year-old male presented with nodular swelling over lateral aspects of both the eyes. The swelling was present for the last 4 months and was occasionally tender. No other ocular or extra-ocular symptoms were present. The clinical and ophthalmological examination revealed no other abnormality. The patient was advised computed tomographic (CT) study for the orbital swelling which revealed mild bilateral symmetrical enlargement of the lacrimal glands with mild enhancing subcentimeter-sized nodules [Figure 1]. No other lesion was noted in the intraconal or extraconal compartments of the orbits. Extra-ocular muscles and optic nerve were unremarkable. No other nodular-enhancing thickening was noted elsewhere. A systemic cause for the bilateral symmetrical enlargement of the lacrimal glands was suspected, and the patient was evaluated accordingly. Detailed history was inconclusive.

A chest radiograph (CXR) and an abdominal ultrasound were done. CXR showed bilateral

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hilar prominence with central reticular pattern [Figure 2]. Abdominal ultrasonography was within normal limits. High-resolution CT (HRCT) of the chest subsequently revealed bilateral symmetrical mediastinal and hilar lymphadenopathy without calcification. Multiple small irregular nodules were also noted along the central bronchovascular bundles as well as along the interlobar fissures [Figures 3 and 4]. These findings were typical of pulmonary sarcoidosis. Few nodules were also noted along the skin of the chest wall indicative of sarcoid granulomas [Figure 3].

A further diagnostic work-up for sarcoidosis was done where elevated serum calcium and angiotensin-converting enzyme levels were found. Negative Mantoux test was added to the diagnosis. Confirmation was done by fine-needle aspiration cytology (FNAC) which revealed epithelioid cells with nonnecrotizing granulomatous pattern diagnostic of sarcoid granulomas. The patient was put on corticosteroids and immunosuppressive medication, and on follow-up, there was clinical reduction in the ocular swelling.

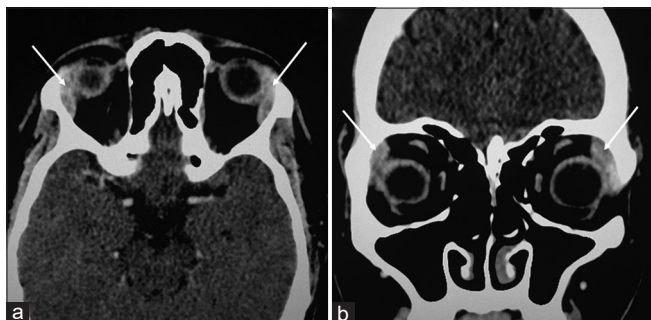
### Discussion

Sarcoidosis is a multisystem disease of unknown etiology affecting all age groups.<sup>[1]</sup> The disease is often asymptomatic, with lung parenchyma and hilar lymph

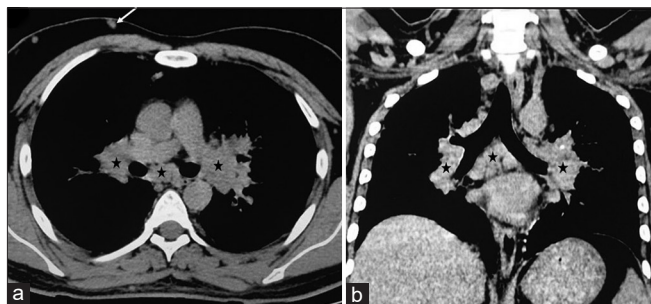
nodes being the most common sites of involvement.<sup>[3]</sup> Because the disease shares its features with tuberculosis and lymphoproliferative disorders, it is considered as a diagnosis of exclusion.

Uveitis is the most common manifestation of ocular sarcoidosis followed by conjunctival follicles, lacrimal gland enlargement, dry eye, dacryocystitis, and retinal vasculitis.<sup>[4]</sup> Lacrimal gland involvement can be the initial manifestation of the disease with no specific imaging features. CT study shows diffuse, homogenous enlargement of the glands with moderate postcontrast enhancement. Few subcentimetric nodules (granulomas) may be visualized. Bilateral symmetrical enlargement of the lacrimal glands should sensitize the clinician and the imaging specialist toward a systemic disease pattern with differential diagnosis of lymphoproliferative disorder, Sjögren's syndrome, sarcoidosis, and Wegener's granulomatosis.

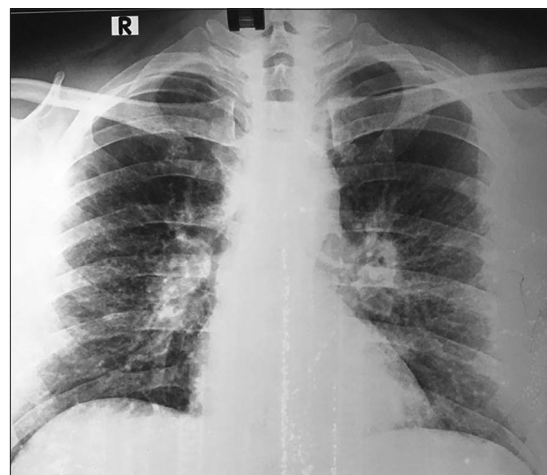
Because the lacrimal glands have native lymphocytes, the primary differential diagnosis in this scenario is lymphoproliferative disease where the spectrum can vary from lymphoid hyperplasia to lymphoma.<sup>[5,6]</sup> Lymphoproliferative disorders also manifest as bilateral lacrimal gland enlargement, which may be primary or



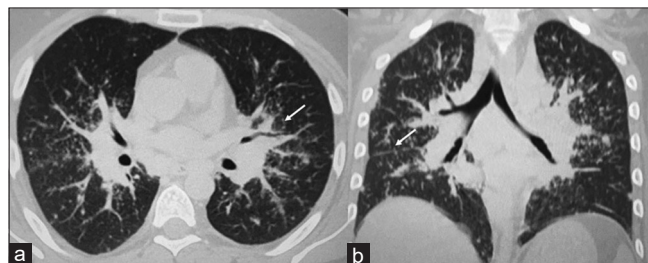
**Figure 1:** Contrast-enhanced computed tomography showing bilateral lacrimal gland enlargement (white arrows) on both axial (a) and coronal (b) planes



**Figure 3:** Computed tomography chest in mediastinal window showing multiple enlarged, homogenous lymph nodes in bilateral hilar and mediastinal regions (asterisks), in axial (a) and coronal (b) planes. Note was made on small subcutaneous nodules in the right anterior chest wall (white arrow)



**Figure 2:** Chest radiograph showing bilateral hilar enlargement with reticulonodular pattern in parahilar regions



**Figure 4:** (a) Axial and (b) Coronal sections of CT chest in lung window showing multiple bilateral parahilar peribronchovascular and perifissural nodules (white arrows)

secondary to systemic lymphoma. Mediastinal and hilar lymphadenopathy are common in lymphoma. However, the presence of pulmonary nodules and FNAC were not compatible with the diagnosis in this case.

Sjögren’s syndrome usually causes diffuse enlargement of the parotid glands giving honey-combing appearance. Bilateral lacrimal gland enlargement may also be seen occasionally. Pulmonary disease manifests as lymphocytic interstitial pneumonia which shows interstitial pattern with small lung cysts on HRCT chest.<sup>[2]</sup> Histopathology shows diffuse lymphocytic infiltration. These findings were absent in our case.

Wegener’s granulomatosis is also a multisystem disease. Orbital involvement is seen in 40%–50% cases and is usually accompanied with paranasal sinus disease.<sup>[7]</sup> Pulmonary nodules in Wegener’s disease are larger and cavitary with involvement of the upper respiratory tract. Elevated c-ANCA levels are found in Wegener’s disease, however; it was not done in this case due to cost constraints.

Tuberculosis may very rarely present with bilateral symmetrical lacrimal gland enlargement.<sup>[8]</sup> Pulmonary tuberculosis shows upper lobe predominance with branching nodules and areas of consolidation. Necrotic mediastinal lymph nodes are frequently seen. These features were absent in our case. Mantoux test and sputum for acid-fast bacillus were also found to be negative; hence, this diagnosis was excluded.

Immunoglobulin (Ig) G4-related disease is an inflammatory disease characterized by infiltration of Ig-G4 positive cells into one or more organs such as pancreas, lung, kidneys, hepatobiliary system, salivary glands, and lymph nodes. It may also involve ocular adnexa better known as IgG4-related ophthalmic disease, typically leading to

lacrimal gland enlargement.<sup>[9]</sup> The diagnosis is based on localized swelling in one or more organs, elevated serum IgG4 concentrations, and histopathological findings of lymphoplasmacytic infiltration and storiform fibrosis. None of these features except for lacrimal gland enlargement were present in our case.

Sarcoidosis is grossly under-reported in the tubercular-endemic regions and the true burden of the disease cannot be truly estimated as there are no reliable epidemiological data. In earlier reports, it was considered to be commonly seen in the west with rare occurrence in the developing countries.<sup>[10]</sup> The clinical course varies from asymptomatic to severe disease. Because the disease closely resembles tuberculosis in its clinical and imaging features, its diagnosis is always a challenge. It is a multisystem disease with lung being the most common site of involvement.<sup>[3]</sup> Pulmonary manifestations may range from Stage 0 to Stage IV disease better called as Siltzbach classification.<sup>[11]</sup> Stage 0 is devoid of any radiographic findings, Stage I shows mediastinal and hilar lymphadenopathy, Stage II disease shows both nodal and parenchymal manifestations, while Stage III is restricted to only parenchymal disease. End-stage fibrosis is noted in Stage IV disease. The typical nodal pattern of the disease shows bilateral hilar and right paratracheal lymph nodes enlargement. The lung disease shows upper and middle lobe predominance of the peribronchovascular, interlobular septal nodules with fibrosis ranging from reticular opacities, septal thickening, and tractional bronchiectasis.<sup>[12]</sup>

It may involve other organs such as lymph nodes, skin, eyes, salivary glands, liver, spleen, heart, as well as nervous system (neurosarcoid).<sup>[10]</sup> There has been an increase in the incidence of sarcoidosis which can be attributed to increasing awareness about the disease pattern and advances in diagnostic modalities.

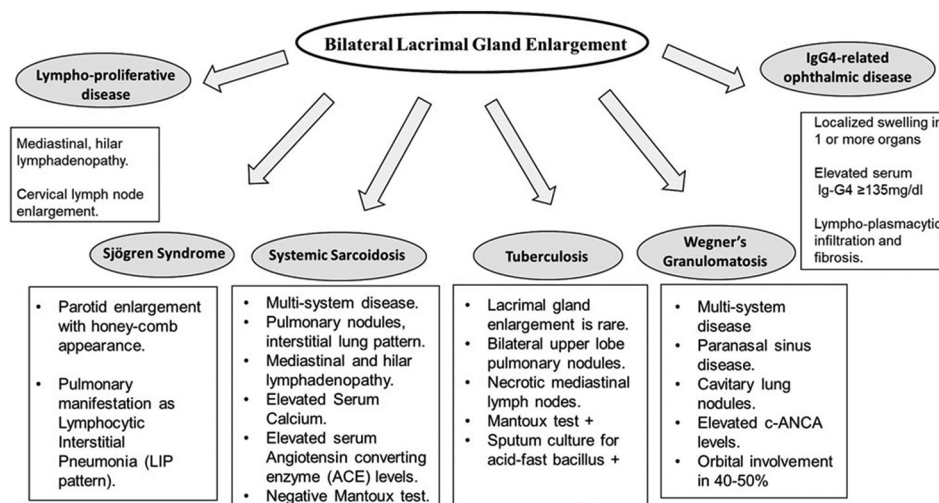


Figure 5: Differential diagnosis of bilateral lacrimal gland enlargement in a tabulated form with their key features

Evaluation of a case of bilateral lacrimal gland enlargement is important as it may be tell-tale sign of an undiagnosed systemic disease. The differential conditions and their important features are presented in Figure 5. Every disease entity related to lacrimal gland enlargement should be given an equal consideration, and one should not be biased by the common condition which may lead to unnecessary management.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initial will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Nil.

### Conflicts of interest

The authors declare that there are no conflicts of interest in this paper.

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