Radiology Case Reports 19 (2024) 2988–2991

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journal homepage: www.elsevier.com/locate/radcr

Case Report

Bilateral swelling of the lacrimal glands as first manifestation of systemic sarcoidosis in a patient with breast cancer ☆

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ARTICLE INFO

Article history:
Received 21 March 2024
Revised 30 March 2024
Accepted 1 April 2024

Keywords:
Sarcoidosis
Lacrimal gland
Non-caseating granulomas
MRI
CT

ABSTRACT

Sarcoidosis, a multifaceted granulomatous disease primarily affecting the lungs, occasionally presents in atypical locations. Lacrimal gland involvement, though rare, poses distinct diagnostic challenges. This case report details a 52-year-old female with bilateral lacrimal gland swelling initially suggestive of metastatic tumor due to a history of breast cancer. Subsequent investigations, including CT and MRI, unveiled pulmonary sarcoidosis. Discussion emphasizes the diverse ocular manifestations of sarcoidosis, with lacrimal gland participation potentially indicating early stages. Diagnostic complexities involve differentiation from other lacrimal pathologies, including neoplasms, lymphoproliferative disorders, Sjögren's syndrome, Wegener's granulomatosis, tuberculosis, and IgG4-related disease. In summary, while lacrimal gland involvement in sarcoidosis is infrequent, it should be considered in orbital masses, necessitating a comprehensive approach for accurate diagnostic orientation in such cases.

Introduction

Sarcoidosis is a multisystemic granulomatous disease whose etiology remains poorly understood. It primarily affects the lungs and mediastinum, although the orbit may be involved in up to 79% of cases. This incidence varies in different regions, such as 13% in a Turkish study and 79% in a Japanese study [1]. Lacrimal gland involvement is uncommon and ranks third in ocular manifestations of sarcoidosis, after uveitis and conjunctivitis. In this case report, we outline the possible diagnoses of bilateral lacrimal gland swelling and emphasize the importance of radiological reasoning in the search for etiology.

Case report

We present a case involving a 52-year-old female, previously diagnosed with breast cancer and currently under

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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https://doi.org/10.1016/j.radcr.2024.04.010

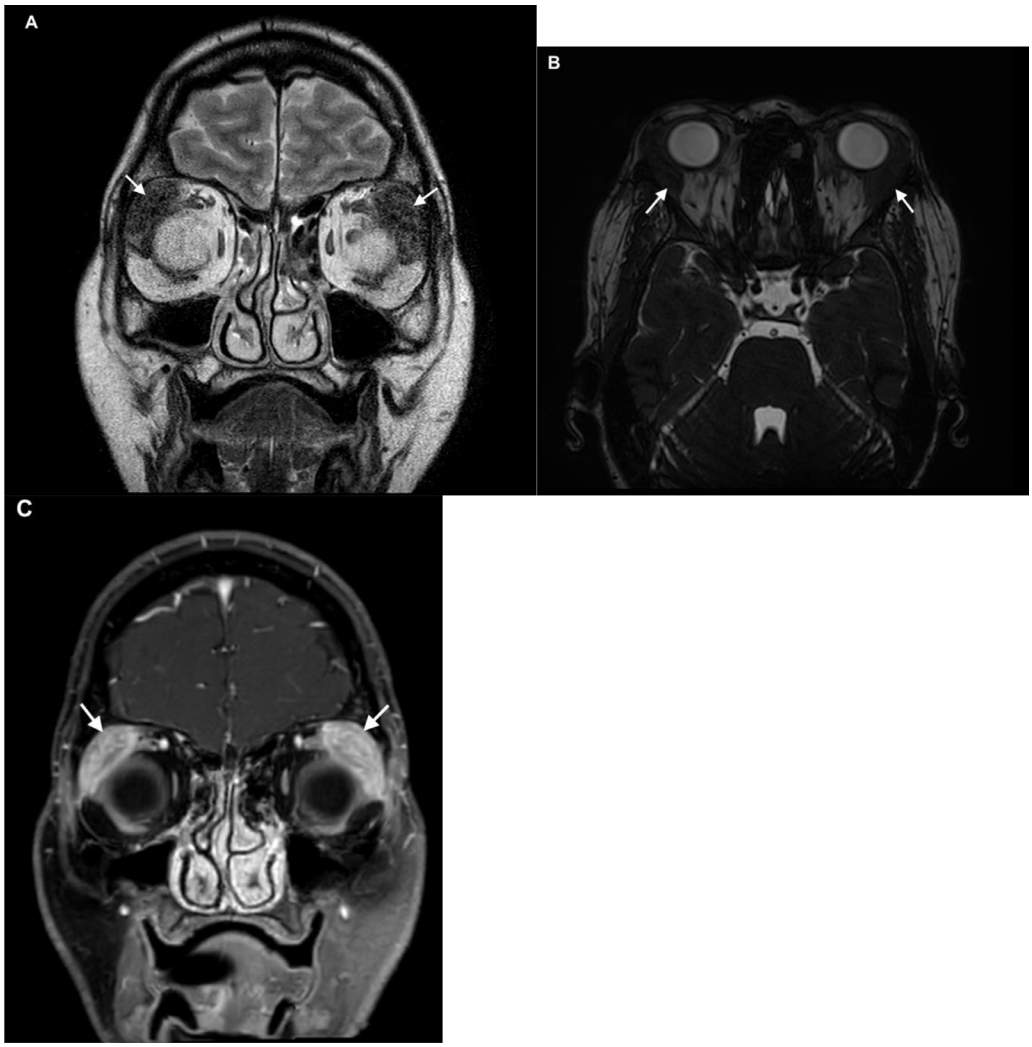

Fig. 1 – Orbital MRI with coronal T2-weighted sequence (A), axial T2 FIESTA (B), and coronal T1 FAT SAT after contrast injection (C), revealing bilateral and asymmetric enlargement of the lacrimal glands (yellow arrows). The glands exhibit an isosignal compared to the muscles, extending to the lateral rectus muscle and orbit without affecting them. There is subtle infiltration of extraconal fat, and intense enhancement is observed after contrast injection.

chemotherapy, exhibiting rapid-onset bilateral upper eyelid oedema accompanied by a decline in visual acuity and headaches. An initial cerebral and orbital CT scan revealed a diffusely enlarged and infiltrated appearance of both lacrimal glands, displaying enhancement post-injection, along with extraconal fat infiltration. Subsequent orbital MRI (Fig. 1) demonstrated asymmetric infiltration of the lacrimal glands, exhibiting an isosignal compared to the muscle, no diffusion restriction, and intense enhancement post-injection. Oculomotor muscles, the globe, and bony structures remained unaffected. While the initial suspicion leaned towards a rare metastatic tumor origin due to the clinical context, the bilateral nature and sparing of adjacent structures prompted additional investigations. A thoraco-abdomino-pelvic CT scan was performed to explore potential alternative locations, revealing pulmonary involvement consistent with sarcoidosis (Fig. 2). Histological confirmation of the diagnosis was obtained through a biopsy of the lacrimal gland, which revealed the presence of non-caseating granulomas, comprised of epithelioid histiocytes and lymphocytes. The patient demonstrated a favorable response to corticosteroid therapy.

Discussion

Sarcoidosis, a multifaceted disease of unknown origin affecting individuals across all age groups, is frequently asymptomatic, with a predilection for lung parenchyma and hilar lymph nodes. The ocular manifestations encompass all eye components, with uveitis emerging as the predominant presentation in 67% of cases [2]. Two extensive studies report incidence rates of 15.8% and 7% of lacrimal gland involvement.

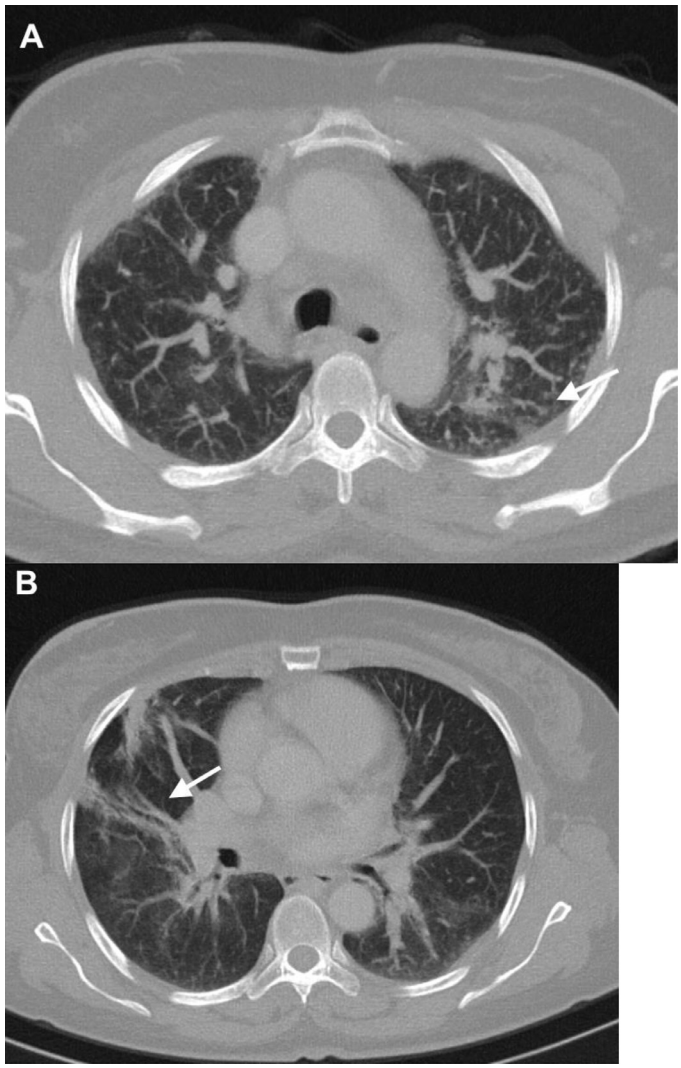

Fig. 2 – Thoracic CT in axial section, in lung window, demonstrating perilymphatic distribution of micronodules (A) along with peribronchovascular thickening and pulmonary architectural distortion (B).

Notably, lacrimal gland participation can exceptionally serve as the inaugural indication of sarcoidosis [3].

Lacrimal gland involvement usually presents as chronic painless bilateral gland enlargement that rarely involves extraocular muscles or leads to diplopia, associated with dry eyes symptoms [3,4]. The substantial enlargement of the lacrimal gland results in observable effects in patients, such as eyelid swelling, ptosis, and globe displacement [5]. Additionally, up to 26% of individuals demonstrate simultaneous enlargement of the salivary and lacrimal glands, recognized as the Mikulicz syndrome, also known as Mikulicz-Radecki syndrome [3]. This syndrome typically involves bilateral and symmetrical swelling of these glands without signs of inflammation or neoplasia. Patients commonly experience dry eyes and dry mouth due to involvement of the lacrimal and salivary glands.

The diagnostic process involves imaging studies of the orbits, although there is no specific imaging features. CT studies reveal a diffuse, homogeneous enlargement of the glands with moderate postcontrast enhancement. Additionally, subcentimeteric nodules (granulomas) may be observed [1]. On MRI, the glands typically demonstrate both T1 and T2 hypointensity [6].

The treatment for lacrimal gland sarcoidosis typically involves corticosteroids, such as oral prednisone or topical eye drops, to reduce inflammation. In cases where corticosteroids are insufficient, immunosuppressive medications like methotrexate or biologics such as infliximab may be considered. Surgical intervention is rare but may be an option for complications. The prognosis varies, with many individuals experiencing a good outcome, including spontaneous remission, while others may have a chronic course or permanent organ damage leading to symptoms such as dry eyes, eye pain, and vision problems.

The lacrimal gland is susceptible to a diverse array of diseases, encompassing isolated or systemic inflammatory conditions as well as neoplastic processes.

Differentials of lacrymal neoplasm include adenocarcinomas arising in the lacrimal duct, pleomorphic adenomas, lymphomas, and metastatic carcinoma. Metastatic carcinoma to

the lacrimal gland is uncommon but can be observed, particularly with breast and lung carcinoma. [7]. Malignant lacrimal gland tumors typically manifest symptoms six months post-development, although in metastatic cases, the onset is notably rapid [8].

On the other hand, the bilateral and symmetrical chronic enlargement of lacrimal glands should alert both clinicians and imaging specialists to consider a systemic disease pattern, with a differential diagnosis including lymphoproliferative disorder, Sjögren's syndrome, sarcoidosis, and Wegener's granulomatosis.

As the lacrimal glands naturally harbor lymphocytes, the differential diagnosis includes lymphoproliferative diseases, exhibiting a spectrum ranging from lymphoid hyperplasia to lymphoma. Lymphoproliferative disorders can also manifest as bilateral lacrimal gland enlargement, either as a primary condition or secondary to systemic lymphoma, often accompanied by common mediastinal and hilar lymphadenopathy seen in lymphomas.

Sjögren's syndrome typically induces diffuse enlargement of the parotid glands, resulting in a honeycombing appearance. While bilateral lacrimal gland enlargement is occasionally observed, the primary manifestation involves the parotid glands.

Wegener's granulomatosis, being a multisystem disease, exhibits orbital involvement in 40%-50% of cases, usually concomitant with paranasal sinus disease.

Tuberculosis, though rare, may present with bilateral symmetrical lacrimal gland enlargement.

Immunoglobulin (Ig) G4-related disease, an inflammatory condition characterized by the infiltration of Ig-G4 positive cells into various organs such as the pancreas, lungs, kidneys, hepatobiliary system, salivary glands, and lymph nodes, may also involve the ocular adnexa, referred to as IgG4-related ophthalmic disease. This typically leads to lacrimal gland enlargement. Diagnosis relies on localized swelling in one or more organs, elevated serum IgG4 concentrations, and specific histopathological findings [1].

Sarcoidosis is distinguished histologically by the presence of non-caseating granulomas, comprised of epithelioid histiocytes and lymphocytes, often accompanied by multinucleated giant cells. In contrast, tuberculosis, also characterized by chronic granulomatous inflammation, tends to exhibit coalescent granulomas with necrosis. The identification of atypical lymphocytes in lymphoma, IgG4-positive plasma cells in IgG4-related Mikulicz's disease, periductal and perivascular inflammation of lymphocytes, and intralobular fibrosis in Sjögren's syndrome are crucial factors aiding in the differential diagnosis of sarcoidosis [2].

This report details a case involving the diffuse enlargement of the lacrimal glands in a patient undergoing treatment for breast cancer, prompting consideration of metastasis. While orbital involvement is not a typical feature of sarcoidosis, it is essential to include this condition in the differential diagnosis of orbital masses, particularly given its potential as an initial manifestation of systemic sarcoidosis.

Conclusion

In the case of sarcoidosis, involvement of the lacrimal glands represents a frequently overlooked differential diagnosis when presented with an orbital mass. A meticulous analysis of epidemiological and clinical data, coupled with a thorough understanding of radiological semiotics, facilitates accurate diagnostic orientation.

Patient consent

Written informed consent for the publication of this case report was obtained from the patient.

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