Systemic Sarcoidosis With a Pseudo-Tumoral Phenotype

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Abstract- Sarcoidosis is a systemic disease histologically characterized by the presence of non-caseating granulomas. Granulomas can affect all structures of the body, giving heterogeneous manifestations and making the diagnosis of this disease a real challenge. We report the case of a 72-year-old woman who presented with two rare manifestations of sarcoidosis: an orbital and a pulmonary pseudotumor. The orbital tumor revealed the disease. Clinically, the patient had palpebral swelling. Orbital MRI showed an orbital pseudotumor hypointense on T1, and hyperintense on T2, heterogeneous and enhanced after gadolinium injection. The thoracic localization was asymptomatic, revealed by the chest Computed Tomography (CT) scan. Histological evidence of granuloma was obtained at both locations. The level of angiotensin-converting enzyme was high. All the other systemic granulomatous diseases were eliminated. We started a systemic corticotherapy with good clinical results.

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Keywords: Systemic sarcoidosis; Orbital tumor; Pulmonary pseudotumor; Angiotensin converting enzyme; Corticotherapy

Introduction

Sarcoidosis is a systemic granulomatosis of unknown etiology. Bilateral hilar lymphadenopathy, skin and ocular involvement are the most common symptoms (1).

We present an unusual form of systemic sarcoidosis with a pseudotumoral phenotype, associating an orbital and a pulmonary pseudotumor.

Case Report

We report the case of a 72-year-old woman who presented with a hard, adherent, and painless mass of the upper eyelid progressively evolving for 3 months, without any decrease in visual acuity. She had no history of tuberculosis. She was a non-smoker woman, and she had no occupational exposures to molds, gases, or fumes. No other clinical manifestations were objectified apart from a xeropthalmia with a short break-up time. This mass was initially explored by a CT scan of the head and face, which revealed a soft tissue mass in the upper-internal angle of the right orbit closely related to the medial rectus muscle with no detectable bone invasion (Figure 1A). Magnetic Resonance Imaging (MRI) of the orbit revealed an intra and extra-conical expansive process of the inner wall of the right orbit hypointense in T1, hyperintense in T2, and heterogeneously enhanced after gadolinium injection. The process infiltrates the medial rectus muscle as well as the retroconical fat (Figure 1B). This orbital pseudotumor was biopsied, revealing confluent tuberculoid granulomas without caseous necrosis.

Laboratory tests showed no inflammatory syndrome, normal blood count, calcium level, and liver function. The tuberculin intradermal reaction (IDR) and the BK test in sputum and urine were negative. The level of angiotensin-converting enzyme (ACE) was high. The anti-nuclear antibodies (ANA) and anti-neutrophil cytoplasmic antibodies (ANCAs) were also negative.

Looking for systemic granulomatosis, a chest CT scan was performed revealing a right hilar parenchymal process, associated with two right postero-basal spicular condensations (Figure 2).

A lung tumor (primary or secondary) or a lymphoma

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with intra-orbital infiltration were first suspected. A computed tomography-guided biopsy of the hilar mass revealed a granulomatous inflammation without caseous necrosis. Respiratory function tests and blood gases were normal. Biopsy of accessory salivary glands was also performed, showing no granulomatous lesions.

The diagnosis of systemic sarcoidosis with orbital and pulmonary pseudotumor was made. Oral corticosteroid therapy was prescribed starting at 1 mg/kg/d of prednisone equivalent. The initial evolution was good with the disappearance of the palpebral mass. An orbital MRI and a chest CT scan were considered to control the lesions, but the patient was lost to follow-up.

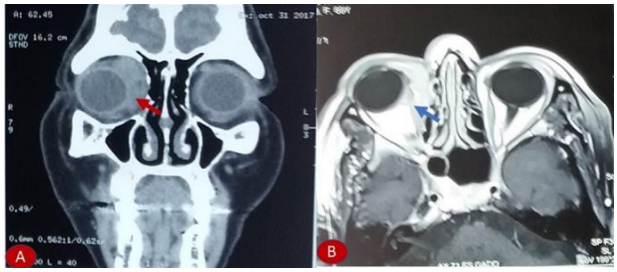

Figure 1. A: Facial scan: tissular mass in the supero-internal orbital angle (red arrow); Figure 1 B: Orbital MRI: expansive process of the inner wall of the intra and extra-conical right orbit (blue arrow) hypointense on T1, enhanced after gadolinium injection

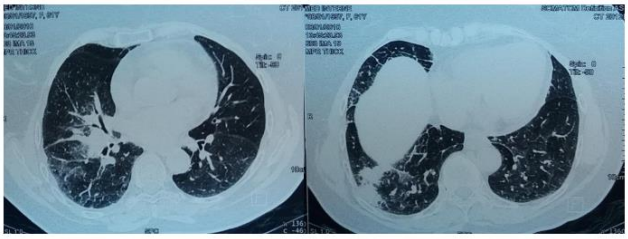

Figure 2. Chest CT scan: a right hilar mediastino-parenchymal process, associated with peri-bronchovascular thickening and two right posterior-basal spiculated condensations

Discussion

We describe an unusual presentation of sarcoidosis revealed by an orbital tumor. Ocular damage is noted in 25%-60% of patients with uveitis being the most prevalent manifestation (2). However, orbital sarcoidosis is rare. Only few cases of orbital tumor revealing sarcoidosis are reported in the literature (3,4). The lachrymal glands are the most affected orbital site (42 to 63%) (5). Orbital involvement during sarcoidosis mainly affects old women with a mean age of 55,9 years (range: 27-85 years) and a sex ratio 4:3 (2). In concordance with literature, our case concerns a 72-year-old woman. Orbital pseudotumor may manifest clinically by periocular mass, ptosis, xerophthalmia, diplopia and even decreased vision. Our patient presented xerophthalmia and a palpable palpebral mass. Biopsy with histological analysis confirms the orbital non-caseating granulomatosis, which was the case of our patient. It is necessary to look for other systemic damages in order to define systemic sarcoidosis. In fact, isolated orbital granulomatous disease is often confused with the sarcoidosis in ophthalmic literature (2). A chest X-ray must be performed and ideally completed by a high-resolution CT of the chest. In our case, the chest CT scanner revealed a pulmonary pseudotumor.

Nodular lung disease is a rare manifestation of sarcoidosis (2,4 to 4%), with limited data in the literature. McCord and Hyman reported in 1952 the first case of nodular sarcoidosis with multiple bilateral nodules mimicking pulmonary metastasis. Nodular sarcoidosis mostly affects African American women aged between 20 and 40 years (6).

It can be manifested by a single or multiple lesions (7). Histological evidence is necessary because of the variability in radiological presentation and in order to eliminate other etiologies, particularly malignant tumors (8). Pulmonary lesions were multiple in our case (one hilar parenchymal mass and two postero-basal spicular condensations). Histological confirmation of a non-caseating granuloma was obtained from a biopsy of the hilar mass. Malignant etiologies were thus eliminated. Besides, the mismatch between the absence of clinical symptoms and the extent of radiological lesions was

another argument for the benignity of the process. Tuberculosis was also ruled out by the absence of general signs and the negativity of the IDR and the BK tests. The diagnosis of systemic sarcoidosis was made, supported by the high level of ACE.

The management of orbital sarcoidosis depends on the site of the mass, its functional impact, the activity of the disease, and the presence of other systemic damage. General corticotherapy remains the cornerstone of the treatment, with good response. Methotrexate may be used as second-line treatment in cases of corticoresistance or intolerance to corticoids. In the case of localized orbital disease, local injections of steroids are proposed. Chirurgical excision is also a good alternative in this case (4). The evolution of pseudotumoral pulmonary sarcoidosis may be spontaneously favorable. General corticotherapy is indicated in cases of progressive and symptomatic pulmonary disease or the presence of other severe damages (8). Our patient was treated with oral corticosteroids with a good clinical outcome. She was unfortunately lost to follow-up, and radiologic images have not been checked.

In conclusion, we reported an atypical presentation of systemic sarcoidosis revealed by an orbital pseudotumor, with a particular and extremely rare pulmonary manifestation consisting of multiple unilateral pulmonary nodules. Histological proof of non-caseating granuloma was mandatory to confirm sarcoidosis and to exclude other diagnoses, especially malignant tumors.

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