

Orbital Tumor Revealing a Systemic Sarcoidosis

Samia Hannanachi Sassi¹, Rim Dhouib¹, Fatma Khanchel¹, Raoudha Doghri¹,
Nadia Boujelbène¹, Hedi Bouguila², and Karima Mrad¹

¹ Department of Pathology, Salah Azaeiz Institute, Bab Saadoun, 1006 Tunis, Tunisia

² Department of Ophthalmology, Hedi Rais Institute of Ophthalmology, Bab Saadoun, 1006 Tunis, Tunisia

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Abstract- Ocular involvement is seen in approximately 25% of patients with sarcoidosis. Uveitis is the most common ocular manifestation, but sarcoidosis may involve any part of the eye. Orbital manifestations of sarcoidosis are uncommon with few series in the literature. A 65-year-old woman presented with redness of the right eye and painless, unilateral eyelid swelling. Orbital scanning revealed mass infiltrating the soft tissue of the inferior right orbital quadrant. Biopsy results showed nodular, non-caseating granulomas consistent with sarcoidosis. The complete systemic workup revealed systemic manifestations of sarcoidosis at the time of examination with hilar and mediastinal lymphadenopathies noted on CT scan. Systemic prednisone therapy followed the orbital surgical treatment with good response. Although rare, orbital sarcoidosis must be considered in the evaluation of orbital tumors in elderly patients. A search for systemic findings should be undertaken, and appropriate therapy should be instituted.

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Introduction

Although the lung is the primary target of involvement, sarcoidosis can involve any organ in the body, including the eye (1). Ocular involvement may also be the initial feature of systemic sarcoidosis in many patients. In these patients with purely orbital involvement, where there is no evidence of systemic disease, the diagnosis of the orbital sarcoidosis may be difficult. Here we describe a patient with systemic sarcoidosis revealed by an orbital tumor.

Case Report

A 65-year-old woman presented with recent redness of the right eye, a slowly progressing mass of the inferior right eyelid associated with joint pain and weight loss. The patient had no prior history of tuberculosis or occupational lung diseases. Magnetic resonance imaging (MRI) of the orbit (Figure 1) revealed a soft tissue mass involving the inferior right orbital quadrant. There was neither osteolytic lesion nor invasion of the right optic nerve. The chest roentgenogram was interpreted as normal. Based on these clinico-radiological findings, right orbital tumor

was highly suspected. The patient underwent a surgical excision of the mass. The specimen consisted of a gray firm nodule of 3 cm in great axis (Figure 2).

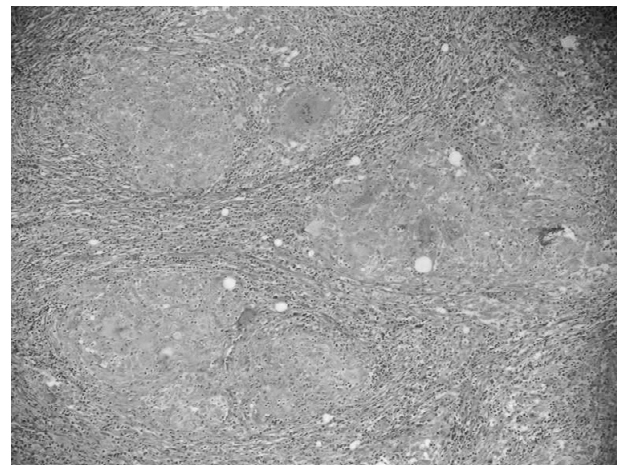


Figure 1. Magnetic resonance imaging is showing a soft tissue mass involving the inferior quadrants of the orbit. The orbital fat exhibits an ill-defined infiltration

Histologically, it showed granuloma embedded in orbital fat. The granuloma was composed predominantly of epithelioid histiocytes with scattered giant cells and lymphocytes (Figure 3). None demonstrated caseating

Corresponding Author: F. Khanchel

Department of Pathology, Salah Azaeiz Institute, Bab Saadoun, 1006 Tunis, Tunisia
Tel: +216 98 337553, Fax: +216 71 150747, E-mail address: fatma.khanchel@gmail.com

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necrosis, and special stains for fungal and tuberculous infection showed negative results. Polarization was performed to rule out presence of foreign material. Thus, the specimen was consistent with Orbital soft tissue sarcoidosis.

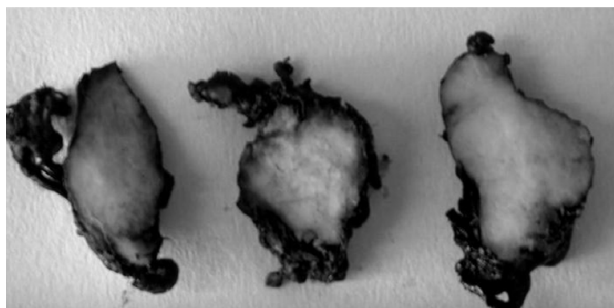


Figure 2. The resected specimen consisted of gray firm nodule of 3cm in great axis

A second review of the chest roentgenogram showed bilateral hilar adenopathies. Thoracic CT scan revealed multiple bilateral hilar and mediastinal adenopathies without parenchymal involvement. Laboratory studies showed an inflammatory syndrome and intradermal tuberculin was negative. Oral steroids followed the surgical treatment with a good response. The patient is followed-up by a specialized medical department.



Figure 3. Photomicrograph showing a characteristic sarcoidal granuloma (hematoxylin-eosin, original magnification X400). The granuloma is composed predominantly of epithelioid histiocytes with scattered giant cells (arrow). The granuloma is surrounded by lymphocytes (so-called naked granuloma, typical of sarcoidosis)

Discussion

Sarcoidosis is a granulomatous, systemic disorder of unknown origin usually affecting young adults in the second and third decades of life. Bilateral hilar lymphadenopathy and skin or eyelid lesions are the most common symptoms noted. Ophthalmologic involvement in sarcoidosis is common (25%-60% of patients) with anterior uveitis being the most common manifestation (2). However, involvement of the orbit and adnexal structures is much less common. Orbital sarcoid has a diverse clinical presentation varying from the lacrimal gland infiltration, soft tissue orbital mass, intraorbital and extraorbital optic nerve sheath and dural involvement to extraocular muscle involvement. The orbital site most commonly involved was the lacrimal gland (3).

Approximately half of all ocular sarcoidosis involvement occurs in patients who were not previously given the diagnosis and leads to findings diagnostic of systemic sarcoidosis (4).

Although sarcoidosis usually affects young patients, review of the literature focuses that the cases of sarcoidosis with orbital involvement occur predominantly in old patients (mean age, 55.9 years, range, 27-85 years) and is more common in women (ratio, 4:3) (5).

Clinically, the most common feature at first examination was a palpable periocular mass followed by discomfort, proptosis, ptosis, dry eye, diplopia, and, decreased vision (5). The orbital mass presents as a rapidly progressing, usually nontender pseudotumor (6).

Regarding the location, the inferior orbit was involved in 60% of reported patients (5).

Ocular tumor biopsy generally provides the diagnosis, but a systematic search for sarcoidosis involvement in other sites, lungs in particular, should be carried out. As a roentgenogram might be interpreted as normal when hilar lymph nodes are only slightly enlarged, we consider that CT scan is useful and more sensitive for confirmation of thoracic localizations (4). These data are consistent with current findings. Serum angiotensin converting enzyme (ACE), if elevated, may be helpful in diagnosing sarcoidosis.

Management of orbital and adnexal sarcoidosis depends on the extent and site of disease, degree of functional impairment, and presence or absence of active systemic disease. Oral steroids have been the mainstay of treatment with a good response. In patients without active systemic disease, a short course of oral prednisolone (starting at 1 mg per kilogram of body

weight and tapering over three months) may be considered as initial therapy. In those who fail to respond or are steroid intolerant, cytotoxic agents such as methotrexate may be used. In localized orbital disease, periocular steroids (1-ml injection of triamcinolone acetonide 40 mg/ml) may be considered (7). Surgical debulking or excision appears to be an effective treatment and may be considered for localized orbital disease and especially for eyelid disease. Current patient underwent a surgical excision because the clinico-radiological findings suspected an orbital tumor. A long-term follow-up of these patients by a pulmonary specialist is recommended to focus a relapsed or active systemic disease (8).

In conclusion, orbital sarcoidosis must be considered in the diagnosis of orbital tumors in elderly patients particularly woman and a search for systemic findings should be undertaken. Furthermore, an appropriate therapy should be instituted, and a long-term follow-up is recommended.

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