Bilateral Exophthalmos caused by Systemic Sarcoidosis: a case report

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ABSTRACT

Background: Ocular involvement, although common in sarcoidosis, granulomatous infiltration of the orbital tissue remains very rare.

Case Presentation: We describe the case of a 55-year-old man who presented bilateral protrusion of eyeballs that had been evolving for 1 year. Clinical examination found an axial and non-pulsatile bilateral exophthalmos with cervical lymphadenopathy. CT and MRI showed infiltration of oculomotor muscles. Biopsy of salivary glands showed chronic inflammatory granulomatous infiltration without caseous necrosis. There was a significant rapid improvement with the use of a steroid as primary therapy.

Conclusion: Sarcoidosis may be a part of exophthalmos causes, after ruling out the main causes which include: Graves' disease, carotid-cavernous fistula, tumors, and infectious causes.

Keywords: Exophthalmos, systemic sarcoidosis, orbital involvement, corticosteroids.

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Background

Sarcoidosis is a systemic granulomatous inflammatory disease. Granulomatous orbital infiltration causing exophthalmos is unusual. We report the case of a systemic sarcoidosis revealed by orbital localization in a Moroccan man.

Case Presentation

A 55-year-old man was admitted for the management of bilateral exophthalmia. He had no particular medical history of goiter, weight loss, thermophobia, or tumor. No members of his family had thyroid disease, sarcoidosis, tuberculosis, or any other pulmonary diseases.

The beginning of the symptomatology goes back to 1 year by the appearance of a progressive protrusion of both eyeballs without any complaint of a decrease in the visual acuity.

On admission, vital signs showed a heart rate at 80 beats per minute, blood pressure at 135/90 mmHg, respiratory rate at 19 breaths per minute, and temperature at 37°C. The examination of the lymph nodes areas found a mobile, painless cervical lymphadenopathy interesting jugulo-carotid and submandibular lymphatic nodes. The thyroid gland was not enlarged and no bruit was audible. The pulmonary exam did not find rules on auscultation. The cardiovascular exam was normal and abdominal exam did not find hepatomegaly or splenomegaly.

Ophthalmologic examination showed bilateral, axile, non-pulsatile, and partially reducible exophthalmos associated with conjunctival hyperemia (Figures 1 and 2). The fundus was normal. Exophthalmometric measurements confirmed the proptosis of both eyes.

The biological assessment showed slightly elevated liver enzymes: Aspartate Aminotransferase (ASAT) at 40 UI/l (N: 8–30 UI/l) and Alanine Aminotransferase (ALAT) at 38 UI/l (N: 8 à 35 UI/l). Blood count showed thrombocytopenia at 122,000 elements/mm^3. The erythrocyte sedimentation rate, C-reactive protein, and procalcitonin were in normal range. Thyroid function was normal. Phosphocalcic blood assay was normal but the urine exam showed hypercalciuria at 560 mg/day (N: 100 à 300 mg/day). The angiotensin-converting enzyme (ACE) level was very high at 230 ($N < 40$). Antinuclear, antiphospholipid, anticytoplasmic neutrophil antibodies, and rheumatoid factor were normal. Anti-thyroperoxidase and antithyroglobulin antibodies were negative.

The orbital CT scan showed a thickened and infiltrated aspect of the lower and oblique external ocular muscles without reaching the sphericity of the globe and the MRI confirmed the infiltration of oculomotor muscles.

Thoracoabdominal CT scan showed bilateral mediastinal lymphadenopathy without pulmonary parenchymal involvement, coelio-mesenteric, retroperitoneal, and bilateral inguinal lymph nodes. Lung function test results were normal.

Cervical lymphadenopathy biopsy demonstrated non-specific lymphadenitis. Minor salivary glands biopsy

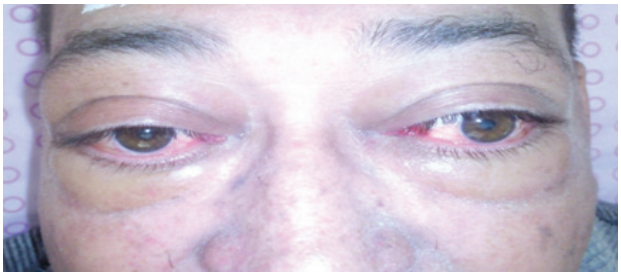

Figure 1. *Front view showing bilateral and axial exophthalmos.*

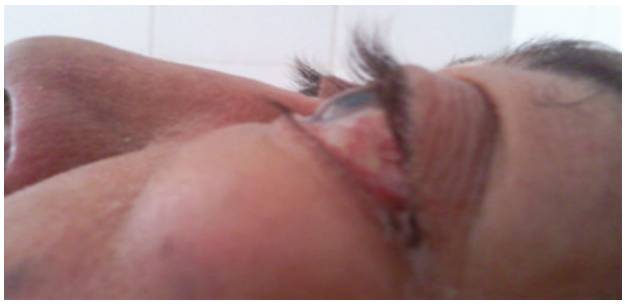

Figure 2. *Exophthalmos in tangential view.*

found polyclonal inflammatory infiltration and noncaseating granuloma.

The diagnosis of sarcoidosis was retained in view of the above-mentioned clinical data, especially the typical hilar adenopathy and the microscopic demonstration of noncaseating granuloma. The patient was put on oral corticosteroids, prednisone at a dosage of 1 mg/kg/day, and oral potassium supplementation. The evolution was marked by a clinical improvement with a regression of the exophthalmia and disappearance of lymphadenopathies.

Discussion

Sarcoidosis is an inflammatory disease of unknown cause that affects one or more organs but most commonly affected are the lungs, hilar and paratracheal lymph nodes, the skin, and the eyes [1]. The histopathologic hallmark of the disease is noncaseating granulomas. It is usually presenting in adults in their third through fifth decades [2–3].

Ophthalmological involvement in sarcoidosis (30% of sarcoidosis) may be characterized by granulomatous inflammation which can affect any part of the eye and its adnexa [4]. It is mainly represented by granulomatous anterior uveitis and intermediate uveitis. Palphalic sarcoids, lacrimal gland involvement, conjunctival nodules, posterior uveitis, and optic nerve involvement are also common.

Orbital disease in sarcoidosis is rare, it may be the initial manifestation of patients with sarcoidosis, and may cause severe visual impairment. The isolated orbital disease is uncommon but it is possible and generally limited to the lacrimal gland [2]. When it occurs in this context, it is usually unilateral and is the initial symptom. It can be bilateral—as in our case—only in rare cases [3,5–8].

Exophthalmos can be seen in various circumstances. It is confirmed by an exophthalmometry or the calculation of the oculo-orbital index on imagery [9]. Even in the absence of evocative ophthalmological involvement, sarcoidosis may be a part of exophthalmos causes, after ruling out the main causes which include: Graves' disease, carotid-cavernous fistula, tumor, and infectious causes.

The morphological exploration of exophthalmia is essentially based on CT scan and magnetic resonance imaging, ruling out a possible orbital tumor. Color Doppler ultrasound is used to study the expansive processes, as well as the circulatory velocities in the vessels of the optic nerve head. In case of suspicion of inflammatory or tumoral pathology, the histological study makes it possible to affirm it by means of biopsies.

In our case, orbital/brain CT scan and MRI eliminated an intracranial tumor, the normal thyroid function excluded Graves' disease. The absence of inflammatory syndrome, the negative procalcitonin, and the good response to prednisone ruled out the hypothesis of infectious causes.

The results of a histological study of minor salivary glands biopsy, the elevated ACE level, the hypercalciuria, and typical hilar adenopathy were strong arguments to conclude that the presence of sarcoid granulomas in the retro-orbital tissues produced the patient's exophthalmos.

Visual prognosis of ocular sarcoidosis may vary depending upon severity and chronicity of eye inflammation. When orbital inflammation occurs, the treatment is based on oral corticosteroids and/or immunosuppressive agents. Systemic corticosteroids are rapidly effective. A high dose (1–1.5 mg/kg/day) of prednisone should be used for a limited duration to avoid side effects and tapered gradually to avoid relapse. Systemic immunosuppressive agents are indicated in patients who are corticosteroid-dependent or -intolerant [4].

Conclusion

Ocular involvement in sarcoidosis is an integral part of extra-thoracic locations. Sarcoidosis can involve almost any structure within or around the eye. Granulomatous infiltration of the orbital tissue during sarcoidosis remains very rare and exceptionally reported. Clinicians need to be aware of atypical ocular manifestations of sarcoidosis in order to make the diagnosis quickly and start corticosteroid therapy to avoid irreversible complications especially blindness.

Acknowledgement

None.

List of abbreviations

ACE	Angiotensin-converting enzyme
ALAT	Alanine Aminotransferase
ASAT	Aspartate Aminotransferase

Consent for publication

Written informed consent was obtained from the patient's next of kin for publication of this case report and the accompanying images to publish this case.

Ethical approval

Ethical approval is not required at our institution for publishing a case report in a medical journal.

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Summary of the case

Patient (gender, age)	1	Male, 55-year-old
Final Diagnosis	2	Systemic sarcoidosis
Symptoms	3	Exophthalmos
Medications	4	Corticosteroid
Clinical Procedure	5	Prednisone: 1 mg/kg/day
Specialty	6	Internal Medicine/Ophthalmology