



Ocular manifestations during sarcoidosis: about 13 cases. Experience of the Military Hospital Mohamed V of Rabat

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Keywords

Ocular Manifestations; Sarcoidosis; Dyspnoea; Hypercalcaemia

Introduction

Sarcoidosis, or Besnier-Boeck-Schaumann disease, is systemic granulomatosis of an unknown etiology characterized by the formation of immune granulomas in the affected organs. The clinical presentation can be very variable [1]. The diagnosis is made on a bundle of clinical, paraclinical and anatomopathological arguments when a biopsy is performed. The most commonly affected organs are the mediastinal lymphatic system, lungs, skin, and eyes.

Ocular manifestations of sarcoidosis could take very different forms. We could find it in one in four patients and it may be revealing of sarcoidosis in 19% of cases [1]. All the tunics of the eyeball and the ocular adnexa can be involved.

Patients and Methods

We report the results of a retrospective study conducted in the ophthalmology department of the Military Hospital Mohamed V of Rabat during a period of 3 years.

We have included the patients treated in the

ophthalmology department of the Military Hospital Mohamed V of Rabat between January 2016 and January 2019. The diagnosis of sarcoidosis was retained on the presence of a bundle of clinical and paraclinical proofs, including at least anatomopathological evidence and/or a hyperlymphocytosis with a CD4/CD8 ratio exceeded 5 in the bronchoalveolar lavage. Furthermore, the diagnosis of ocular sarcoidosis was based on a compatible ophthalmic presentation and histological proof of granulomatosis.

We have collected the clinical, paraclinical, therapeutic and evolutionary characteristics of the patients including:

- I. Clinically: the age and the sex of the patient, functional signs, visual acuity, biomicroscopy examination of the anterior segment and the fundus, eye tone measured with Goldman tonometer, gonioscopy and the results of fluorescein angiography and optical coherence tomography.
- II. Paraclinically: the dosage of the angiotensin-converting enzyme (ACE) and corrected calcemia, chest radiograph, thoracic computed tomography scan, cardiac ultrasonography, brain magnetic

Case Report

resonance imaging, tuberculin intradermal reaction, electrocardiogram, bronchoalveolar lavage (to precise the percentage of lymphocytes and the CD4/CD8 ratio), the location and the number of biopsy.

- III. The therapies are given to the patients and their dosage: corticotherapy, biotherapy (Infliximab), synthetic antimalarials (hydroxychloroquine), topical and systemic glaucoma medication and the possible recourse of cataract surgery, vitrectomy or trabeculectomy.
- IV. Evolutionarily: the overall duration of follow-up, duration of oral corticosteroid therapy, the free interval before recurrence at discontinuation of corticosteroids, degree of recovery, the appearance of macular edema during follow-up and/or the persistence of refractory macular edema.

In total, 13 patients were included in the study with a follow-up of 25 eyeballs (one patient had unilateral ocular involvement).

Results

The average age at the diagnosis was 51 years with extremes ranging from 19 to 79 years. The majority were women (77%). The median free interval between the diagnosis of sarcoidosis and the first ophthalmologic sign was six years [2-10]. The median follow-up of the patients was 36 months (between 7 and 130 months). Nine patients (69%) had initially a systemic involvement associated with the ocular manifestations of the sarcoidosis, while the disease was strictly ocular at the time of diagnosis in

five cases.

The main ocular functional signs were: visual acuity decline in 84% of the cases and eye pain in 53% of the cases. Meanwhile, the systemic signs were as follows: arthralgia in 38% of the cases, myalgia in 23%, dyspnoea in 15% of the cases and nervous system involvement in 15% of the patients (Table-1).

Number of patients		13
Sex	Women	77%
	Men	23%
Average age at the diagnosis (years)		51
Median follow-up (months)		36 (7-130)
Median free interval between the diagnosis and the first ophthalmologic sign (years)		6 (2-10)

The ophthalmological involvement was bilateral in 92% of the cases. One patient presented hypertrophy of the lacrimal gland with ptosis. The slit lamp examination found 69% of anterior uveitis with 56% of Keratic precipitates (Fig-1), 38% of posterior synechiae between the iris and the capsule of the lens (Fig-2), iris nodules in 15% (Koeeppe in all the cases).

The eye tone measured with Goldman tonometer showed initially a median of 17 mmHg (with extremes between 12 and 50 mmHg). Ocular hypertonia over than 25 mmHg was found in 30% of the cases.

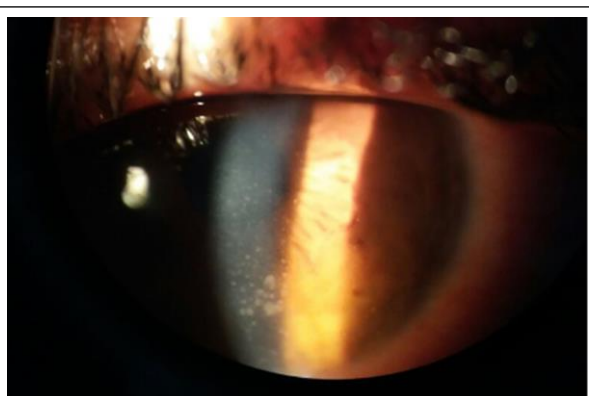


Fig-1: Granulomatous mutton-fat keratic precipitates

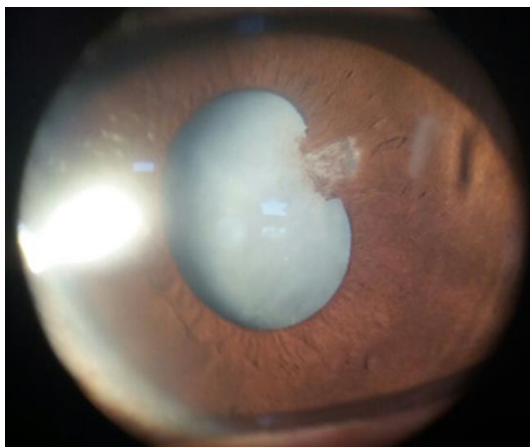


Fig-2: Posterior synechiae between the iris and the capsule of the lens

The examination of the fundus showed 30% of vitritis and 15% of papillary edema and 15% of macular edema that was confirmed by the macular optical coherence tomography. Otherwise, the fluorescein angiography revealed 38% of retinal vasculitis that was associated with retinal ischemia in 7.6% of the cases and with multifocal choroiditis in 23% of the cases.

The biological investigation found an elevated level of the angiotensin-converting enzyme in 61% of the patients and a hypercalcaemia (albumin-corrected serum calcium) in 7.6% of the patients while the tuberculin intradermal reaction was negative for all the patients.

In lungs, chest radiography was abnormal in 46% of cases, thoracic computed tomography scan found abnormalities in 15% of the cases and the bronchoalveolar lavage found an hyperlymphocytosis with a median of 0.4 [0.2-0.7] in 61% of the patients with a positive CD4/CD8 ratio in 53% of the patients.

The biopsies was made in the lungs and found an epithelioid granuloma in 85% of the patients. Regarding the therapeutic side, we prescribed eye drops containing corticosteroids in 85% of cases and hypotonizing eye drops in 30% of cases (50% of whom received oral or intravenous acetazolamide to control high hypertension). 30% of the patients benefited of one or more boluses of Solumedrol and 70% of them benefited of oral corticosteroids at the dose of 1 mg / kg per day followed by a gradual decline. The median duration of oral corticosteroid therapy was 12 months.

During follow-up, 23% of the patients underwent cataract surgery and 7% underwent trabeculectomy. As a result of the initiation of anti-inflammatory therapy, 85% of the patients have had a rapid recovery. Recovery was considered partial in 53% of cases, and total in 30% of cases. The recurrences concerned 4 patients.

Discussion

Sarcoidosis is a chronic systemic granulomatous disease of unknown origin that can reach all the organs (lungs, lymphatic system, skin, and eye). The ophthalmological involvement is relatively frequent and occurs in 25% of the cases [2]. It is the inaugural manifestation in 20% of the cases and may precede pulmonary manifestations by several years [3].

Almost all elements of the eyeball, ocular adnexa and orbit may be affected by this disease. Although the involvement of the anterior segment is the most common, it's the involvement of the posterior segment that endangers the visual prognosis [4]. The disease is usually bilateral. The conjunctival and lacrimal gland disorders are relatively frequent but they do not significantly affect the visual functional prognosis. In contrast, ocular complications such as retinal ischemia with neovascularization and isolated optic nerve damage are relatively rare but affect significantly the functional prognosis. The most common ocular manifestations observed during sarcoidosis are uveitis. All types of uveitis can be seen, from simple unilateral anterior uveitis to severe bilateral panuveitis. Otherwise, the uveitis presents a

Case Report

difficult management problem due to its chronic evolution and its functional repercussion (**Table-2**) [4].

The first symptoms of ocular sarcoidosis occurred in patients over 40 years old in 61% of the cases. Classically, pulmonary sarcoidosis occurs on average before 40 years old [5]. However, the mode of occurrence of the disease is different according to the

ethnic origins of the patients. Sarcoidosis is a more common in black population with a female-predominant [6], suggesting genetic susceptibility.

Many studies have been done on the genes of the HLA system. The results are in favor of susceptibility for HLA typing A1, B8 and DR3 [7].

According to the data of the literature, the

Table-2: Clinical characteristics of the patients	
Conjunctiva	Conjunctival nodules, Cicatricial symblepharon or entropion, dry syndrome
Sclerotic	Scleritis, episcleritis
Cornea	Interstitial keratitis
Uvea	Anterior, intermediate or posterior uveitis, panuveitis
Lens	Cataract
Retina	Retinal ischemia with neovascularization, vasculitis, macular edema
Choroid	Choroidal granulomas, neovascularization
Optic nerve	Clinical papillary edema, optic atrophy, optic nerve granuloma and prepapillary neovascularization
Lacrimal gland	Keratoconjunctivitis sicca, lacrimal hypertrophy
Eyelids	Granuloma, ptosis
Orbit	Diffuse unilateral orbital inflammation

paraclinical tests performed for the diagnostic have variable degrees of sensitivity and specificity and none of them has 100% sensitivity or specificity.

The 2017 Revised criteria of the International Workshop on Ocular Sarcoidosis (IWOS) for the diagnosis of ocular sarcoidosis [8] identifies seven clinical intraocular signs and five ancillary tests suggestive of ocular sarcoidosis. In our serie, two of the seven clinical signs were found frequently: mutton fat keratic precipitates and/or iris nodules (Koeppel/Bussaca) in 56% of the cases and bilateral inflammation in 92% of cases). Goniosynechiae (38%) and choroidal nodules (23%) were also present.

Four of the five ancillary tests were represented: tuberculin anergy (100%), Elevated angiotensin-

converting enzyme (61%), bilateral hilar lymphadenopathy showed in Chest X-ray (46%) and only found in Chest tomography (15%). Liver-enzyme tests was normal in all the patients of the study.

Conclusion

Sarcoidosis is a multisystemic inflammatory disease. Its expression seems to result from a complex interaction between environmental factors and genetic susceptibility with many different genes [9,10]. The genes of the HLA system have been studied. The results showed a correlation between the HLA phenotype and the risk of occurrence of the disease in one side, its mode of presentation or its evolutionary profile in another side. These associations seem to vary according to geography and ethnicity [9,10]. The diagnostic approach requires a lot of clinical and paraclinical investigations [11], involving a

Case Report

collaboration between internists, ophthalmologists and pulmonologists.

If we suspect ocular involvement of sarcoidosis, the first line of tests should be: tuberculin intradermal reaction, angiotensin-converting enzyme, chest CT scan, brain MRI and biopsy of the accessories salivary glands.

The tests that could be discussed in a second-line, if the clinical examination is highly suggestive is the bronchoalveolar lavage (to precise the percentage of lymphocytes and the CD4/CD8 ratio). In our study, lungs biopsies that had been performed during the bronchoalveolar lavage had found histological evidence in 11 out of 13 cases (85%).

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