

CASE REPORT

Open Access



Low dose corticosteroid in association with methotrexate for therapy of ocular sarcoidosis: report of a case

Grasiane Nunes Mayer^{1*} , Morgana Longo², Bárbara Brandi Gomes³ and Mário Junqueira Nóbrega⁴

Abstract

Sarcoidosis is a multisystem granulomatous disease of unknown etiology. Ocular involvement can be the initial manifestation, occurring by itself, or it can be associated with other systemic signs of sarcoidosis. A 31 years-old caucasian female presented a 10-day history of decreased vision and pain in OS. Biomicroscopy revealed fine keratic precipitates, a mild reaction in the anterior chamber and in the vitreous cavity in both eyes and a small posterior synechiae in OS. Intraocular pressure was 12 mmHg in the OD and 9 mmHg in OS. Fundoscopy disclosed mild swollen and hyperemic optic discs and some subretinal yellowish nodules of 1/3 to 1 disc diameter, partially delimited, located in the posterior pole and midperiphery in both eyes. The Chest X-ray disclosed the suspicion of hilar lymphadenopathy, especially on the left-side. The positron emission tomography (PET-scan) showed increased 18 F-fluorodeoxyglucose uptake in the paratracheal and bilateral hilar lymph nodes, indicating high local cellular metabolism. A mediastinal lymph node biopsy was performed afterwards, which depicted non-caseating granulomas with multinucleated giant cells and absence of acid-alcohol resistant bacilli. The patient was initially treated with oral prednisone 60 mg/day with slow tapering over the next months. A decreased papillitis and an increased size of the choroidal granulomas was observed after a 6-month therapy (Figs. 1B and 2B). Then, a combination of methotrexate 20 mg/week and prednisone 10 mg/day was given over the following months, which led to a gradual reduction in size and thickness of the choroidal granulomas. A weak response to steroid monotherapy was initially observed in this patient. The introduction of a non-steroid immunosuppressant drug (methotrexate) was associated with significant anatomic improvement in the following months. This report raises the suggestion that an early introduction of a nonsteroidal immunosuppressive drug to systemic steroids may be beneficial for a rapid remission of the uveitis.

Keywords: Ocular sarcoidosis, Sarcoidosis in Ophthalmology, Diagnosis of sarcoidosis

Background

Sarcoidosis is a multisystem granulomatous disease of unknown etiology [1]. Ocular involvement can be the initial manifestation, occurring by itself, or it can be associated with other systemic signs of sarcoidosis [2]; eyes are affected in 25 to 50 % of patients [3, 4]. Early and effective treatment of ocular sarcoidosis is critical because of the high risk of progression to irreversible vision loss, especially for glaucoma and cystoid macular edema [5].

Case presentation

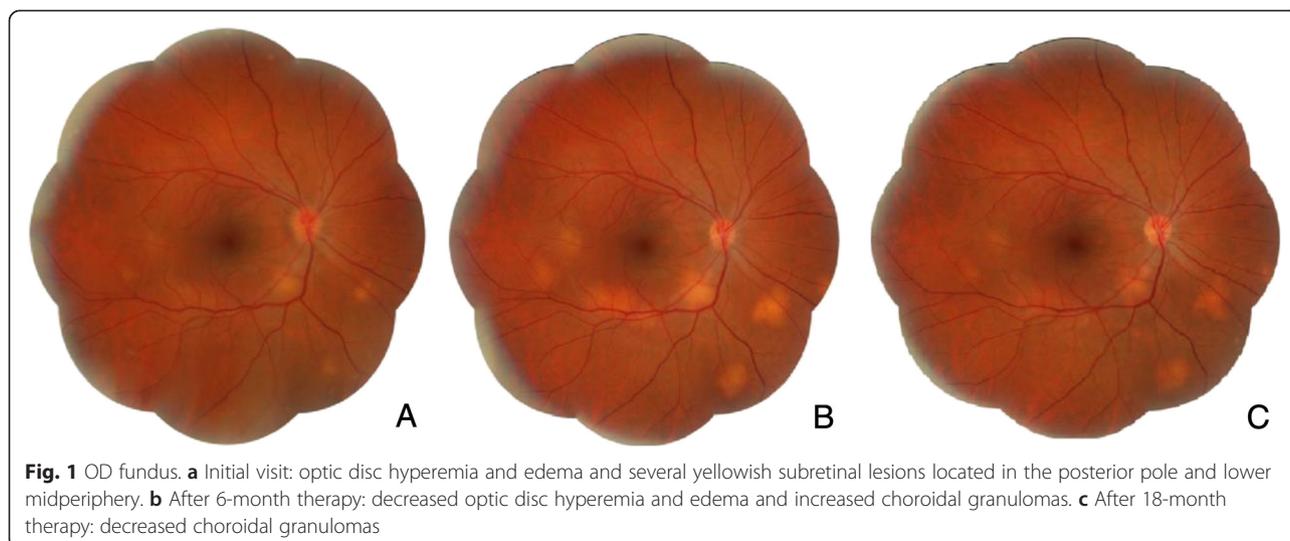
A 31 years-old caucasian female presented a 10-day history of decreased vision and pain in the OS. Examination showed visual acuity of 20/20 in OD and 20/25 in OS. Biomicroscopy revealed fine keratic precipitates, a mild reaction in the anterior chamber and in the vitreous cavity in both eyes and a small posterior synechiae in OS. Intraocular pressure was 12 mmHg in OD and 9 mmHg in OS. Fundoscopy disclosed mild swollen and hyperemic optic discs and some subretinal yellowish nodules of 1/3 to 1 disc diameter, partially delimited, located in the posterior pole and midperiphery in both eyes (Figs. 1a and 2a).

Serologic tests for toxoplasmosis, syphilis and acquired immunodeficiency syndrome were negative as well as skin test for tuberculosis (purified protein derivative or

* Correspondence: grasinmayer@hotmail.com

¹Resident in Ophthalmology, Sadalla Amin Ghanem Eye Hospital, Rua Desembargador Nelson Nunes Guimarães, 811, CEP 89203060 Joinville, SC, Brasil

Full list of author information is available at the end of the article



PPD). Serum lysozyme was 14.0 mg/L (laboratory normal range 9,6 to 17,1 mg/L) and angiotensin-converting enzyme was 87.0 IU/L (laboratory normal range 35 to 90 IU/L). The Chest X-ray disclosed the suspicion of hilar lymphadenopathy, especially on the left-side (Fig. 3). The PET-scan showed increased 18 F-fluorodeoxyglucose uptake in the paratracheal and bilateral hilar lymph nodes, indicating high local cellular metabolism (Fig. 4). A mediastinal lymph node biopsy was performed afterwards, which depicted non-caseating granulomas with multinucleated giant cells and absence of acid-alcohol resistant bacilli (Fig. 5).

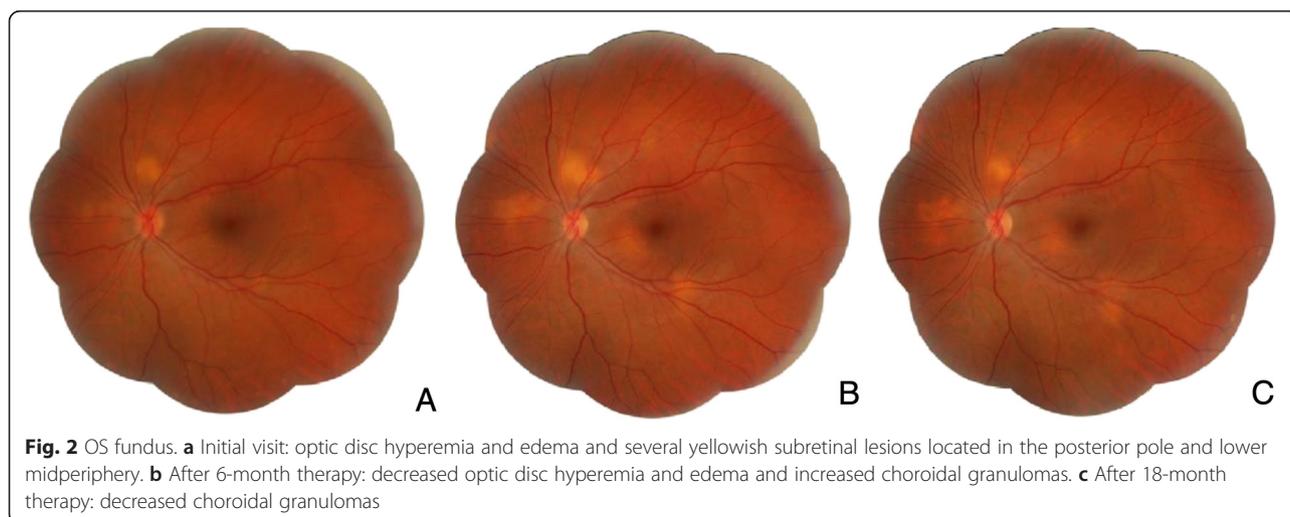
The patient was initially treated with oral prednisone 60 mg/day with gradual reduction. Six months after the treatment began the prednisone intake was 20 mg/day, when there was a decrease in papillitis and an increase in the choroidal granuloma size (Figs. 1b and 2b). At that visit a combination of methotrexate 20 mg/week and prednisone 60 mg/day was prescribed. During the

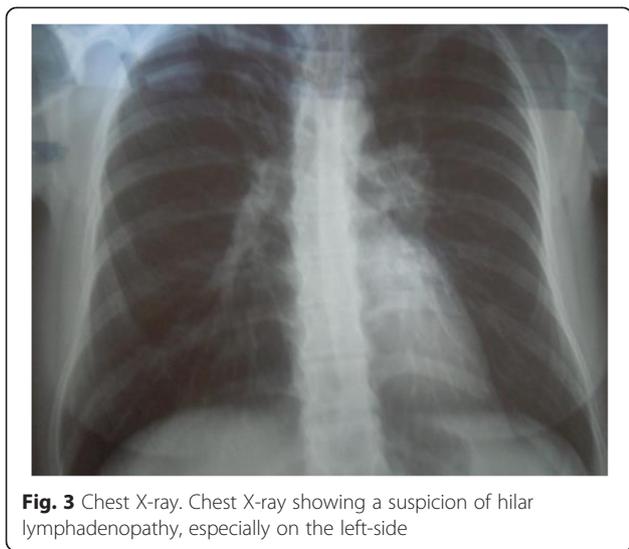
following 12 months the prednisone daily dose was decreased 10 mg each visit. By the 18th month of treatment the intake of prednisone was 10 mg/day, leading to a gradual reduction in the choroidal granulomas' size and thickness and stabilized uncorrected visual acuity of 20/20 in both eyes (Figs. 1c and 2c).

Discussion

Ocular sarcoidosis is still rarely diagnosed in Brazil. In 2004, Gouveia et al. observed a prevalence of 2.29 % among of the causes of uveitis among all age groups in São Paulo (SP) [6].

The clinical suspicion of sarcoidosis requires a biopsy to confirm the diagnosis. When the disease causes only ocular manifestations, the diagnosis is more difficult and may be based on ophthalmologic evaluation and eventual findings of a systemic granulomatous inflammation, such as abnormal serologic tests and chest imaging [7]. Intraocular biopsy is usually not performed in these

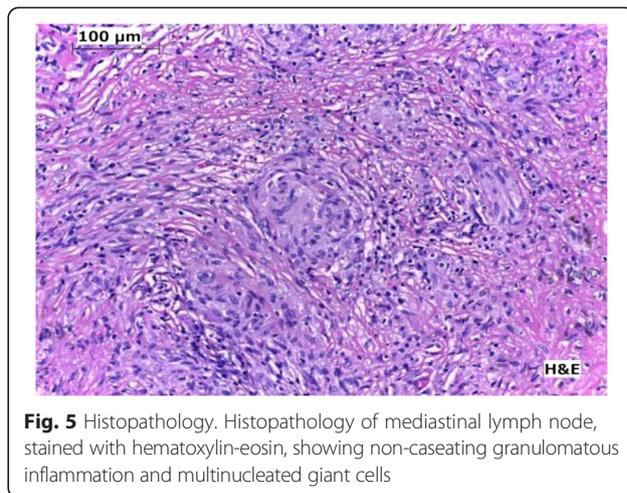




situations, due to its inherent risk of causing severe visual complications.

In the present case, ocular sarcoidosis was not accompanied by other clinical complaints. In addition, laboratory evaluation displayed normal values of serum lysozyme and angiotensin-converting enzyme. However, patients with active ocular sarcoidosis may be in systemic remission already, justifying a possible decrease in angiotensin-converting enzyme values [8]. Moreover, a suspected chest radiography image motivated additional workup and a definite diagnosis of sarcoidosis through mediastinal lymph node biopsy.

A weak response to steroid monotherapy was initially observed in this patient. The introduction of a non-steroid immunosuppressant drug (methotrexate) was associated with significant anatomic improvement in the



following months. This fact drew attention because systemic steroids are the first line of therapy for sarcoidosis and provide faster and better outcomes even before the combination with other pharmacologic agents [9].

Conclusion

Although sarcoidosis is a well-known disease, it rarely causes ocular abnormalities. The present case illustrates that ocular sarcoidosis may be unaccompanied by other clinical complaints or by elevated serum lysozyme and angiotensin-converting enzyme. Otherwise, complementary chest imaging and mediastinal lymph node biopsy were critical to make the diagnosis and start prompt therapy. This report raises the suggestion that an early introduction of a nonsteroidal immunosuppressive drug to systemic steroids may be beneficial for a rapid remission of the uveitis. Further studies with bigger number of cases are needed to draw a definite conclusion.



Consent

Written informed consent was obtained from the patient for publication of this Case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Abbreviation

PET-scan: Positron emission tomography.

Competing interests

The authors have no financial or non-financial conflicts of interest to disclose.

Authors' contributions

MN held medical consultation of patient and obtained the consent of the patient, reviewed the article and made consistent criticism. GM, ML and BG reviewed the bibliographic reference on the subject, wrote the article, made contributions to the design and structure of the article. All authors read and approved the final manuscript.

Author details

¹Resident in Ophthalmology, Sadalla Amin Ghanem Eye Hospital, Rua Desembargador Nelson Nunes Guimarães, 811, CEP 89203060 Joinville, SC, Brasil. ²University of Joinville, Rua Saí, 305, apto 704, Anita Garibaldi, CEP 89202-170 Joinville, SC, Brazil. ³University of Joinville, Rua Alexandre Schlemm, 233, apto 301, Bucarein, CEP 89202-417 Joinville, SC, Brazil. ⁴University of Joinville, Sadalla Amin Ghanem Eye Hospital, Rua Camboriú, 35 (Eixo Marquês de Olinda), CEP 89216-222 Joinville, SC, Brazil.

Received: 25 February 2015 Accepted: 24 May 2015

Published online: 26 June 2015

References

1. Rothova A. Ocular involvement in sarcoidosis. *Br J Ophthalmol.* 2000;84(1):110–6.
2. Rizzato G et al. Uveitis as a presenting feature of chronic sarcoidosis. *Eur Respir J.* 1996;9(6):1201–5.
3. Wu W et al. Overexpression of IL-17RC associated with ocular sarcoidosis. *J Transl Med.* 2014;12(1):152.
4. Capella MJ, Foster CS. Sarcoidosis. In: Foster CS, Vitale AT, editors. *Diagnosis and Treatment of Uveitis.* Philadelphia: W. B. Saunders; 2002. p. 951–72.
5. Lazarus A. Sarcoidosis: epidemiology, etiology, pathogenesis, and genetics. *Dis Mon.* 2009;55(11):649–60.
6. Gouveia EB et al. Causes of uveitis in a tertiary center in São Paulo City, Brazil. *Arq Bras Oftalmol.* 2004;67:139–45.
7. Birnbaum AD et al. Clinical features and diagnostic evaluation of biopsy-proven ocular sarcoidosis. *Arch Ophthalmol.* 2011;129(4):409–13.
8. Dimantas MAP et al. Anterior uveitis associated to systemic diseases. *Arq Bras Oftalmol.* 2003;66:235–8.
9. Desai UR et al. Choroidal granulomas in systemic sarcoidosis. *Retina.* 2001;21(1):40–7.

Submit your next manuscript to BioMed Central and take full advantage of:

- Convenient online submission
- Thorough peer review
- No space constraints or color figure charges
- Immediate publication on acceptance
- Inclusion in PubMed, CAS, Scopus and Google Scholar
- Research which is freely available for redistribution

Submit your manuscript at
www.biomedcentral.com/submit

