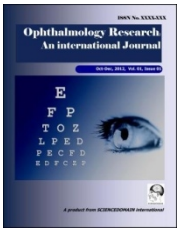

Ophthalmology Research: An International Journal
2(6): 268-274, 2014, Article no. OR.2014.6.002

SCIENCEDOMAIN *international*
www.sciencedomain.org

Asymptomatic Uveitis as Primary Manifestation of Sarcoidosis

F. Nilüfer Yalçındağ[1], Özge Yanık[1*] and Özlem Özdemir Kumbasar[2]

[1]Department of Ophthalmology, Ankara University, Faculty of Medicine, Ankara, Turkey.
[2]Department of Pulmonary Diseases, Ankara University, Faculty of Medicine, Ankara, Turkey.

Authors' contributions

This work was carried out in collaboration between all authors. Author ÖY designed the study, wrote the first draft of the manuscript, and managed the literature searches. Authors FNY and ÖÖK revised it critically for intellectual content. All authors were involved in the assessment and treatment of the patient. All authors read and approved the final manuscript.

Case Study

Received 2nd April 2014
Accepted 1st May 2014
Published 24th May 2014

ABSTRACT

Aim: To emphasize that ocular involvement in sarcoidosis may be the initial manifestation of the disease and can be completely asymptomatic.
Presentation of Case: A 36 year-old female was referred to uvea service with detection of keratic precipitates and anterior chamber cells during routine eye examination. She did not show any symptoms of uveitis. A slit lamp examination revealed bilateral medium-sized, round, centrally located granulomatous keratic precipitates on corneal endothelium and +1 cell reaction in anterior chamber. Dilated fundus examination showed bilateral vitreous cells and small vitreous opacities in the right eye. Serum levels of angiotensin-converting enzyme were increased. Her chest X-ray study was significant for bilateral hilar adenopathy, and transbronchial needle aspiration biopsy confirmed the diagnosis of sarcoidosis. Patient responded well to systemic steroid therapy with complete disappearance of the signs.
Discussion: Ocular involvement in sarcoidosis may occur as the disease process, it also can be the first clinical manifestation of the disease. Although the incidence of pulmonary involvement is nearly one and half times higher than ocular involvement, the most frequent initial symptom of sarcoidosis is eyesight disorder. The reason for that is late emergence of symptoms caused by lung involvement such so symptoms related to ocular involvement generates the major complaints at the first application.

**Corresponding author: E-mail: oyanik05@hotmail.com;*

Conclusion: Early diagnosis of sarcoidosis is important for avoiding ocular complications as well as prevention of other system involvements. Ophthalmologists have a critical role in the diagnosis and follow-up of the disorder because of increasing incidence of the ocular involvement.

Keywords: Granulomatous uveitis; indocyanine green angiography; sarcoidosis.

1. INTRODUCTION

Sarcoidosis is a systemic disease with unknown etiology and characterized by non-caseous granulomas [1]. It mainly affects individuals between the ages of 20-50 and slightly more common in women [2]. The disease process is caused by accumulation of T lymphocytes and macrophages forming epitheloid granulomas in target tissues [1]. Most common side of involvement is lung and includes 90% of the cases [2]. Ocular involvement was reported to be seen in 30-60% of the patients [3]. Sarcoidosis may also affect lymph nodes, skin, joints, central nervous system and heart [3].

Ocular involvement usually includes anterior or posterior uveitis, also can include lacrimal gland, conjunctiva and optic nerve [4]. The incidence of the ocular involvement is increasing according to recent studies [5,6] and early detection of ocular findings is gaining importance to prevent disease progression. The aim of this report is to emphasize that ocular involvement in sarcoidosis may be initial manifestation of the disease and can be asymptomatic.

2. PRESENTATION OF CASE

A 36-year-old female was referred to our uvea service due to detection of keratic precipitates and anterior chamber cells during routine eye examination. She did not show any symptoms of uveitis. Best corrected visual acuity in both eyes was 20/20. A slit lamp examination revealed bilateral medium-sized, round, centrally located granulomatous keratic precipitates on corneal endothelium (Fig. 1) and +1 cell reaction in anterior chamber. Dilated fundus examination showed bilateral vitreous cells and vitreous opacities below the right eye. In the fluorescein angiography, there was fluorescein leakage from peripheral vessels in the right eye (Fig. 2).

Indocyanine green angiography showed infrequent hypofluorescent small round spots in the right posterior pole (Fig. 3). The results of laboratory examinations for syphilis and tuberculosis were negative, including venereal disease research laboratory (VDRL) test, rapid plasma regain (RPR) test and Quantiferon test. Serum level of angiotensin-converting enzyme (ACE) was raised to 67U/L (8-52U/L). Serum calcium level was normal. A posteroanterior chest x-ray showed bilateral hilar lymphadenopathy (Fig. 4). With the pre-diagnosis of asymptomatic uveitis associated with sarcoidosis, pulmonary disease consultation was requested. Pulmonary disease department performed endobronchial ultrasound-guided transbronchial needle aspiration biopsy and diagnosis of sarcoidosis was confirmed by detection of non-caseous granulomatous lymphadenitis. Patient was completely asymphtomatic in terms of pulmonary involvement and pulmonary function tests were totally normal. Therefore treatment was not needed for pulmonary disease. 30mg prednisolone (Deltacortril 5mg tablets ®) treatment was initiated due to ocular involvement and tapered 5mg per week. After six weeks, anterior chamber inflammation, keratic

precipitates and vitreous opacities were completely disappeared. Indocyanine green angiography was repeated and it showed initially hypofluorescent small round spots turned to isofluorescent spots three months later (Fig. 5).

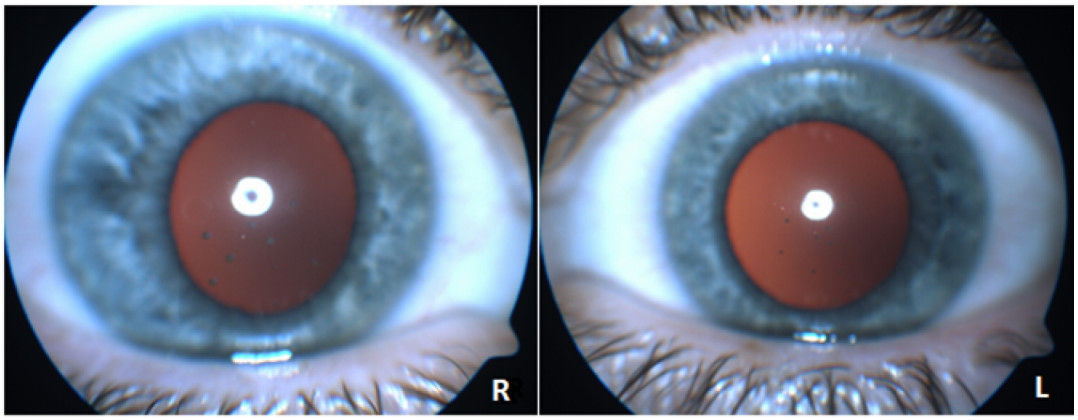

Fig. 1. Bilateral medium-sized, round, centrally located granulomatous keratic precipitates. R, right eye; L, left eye

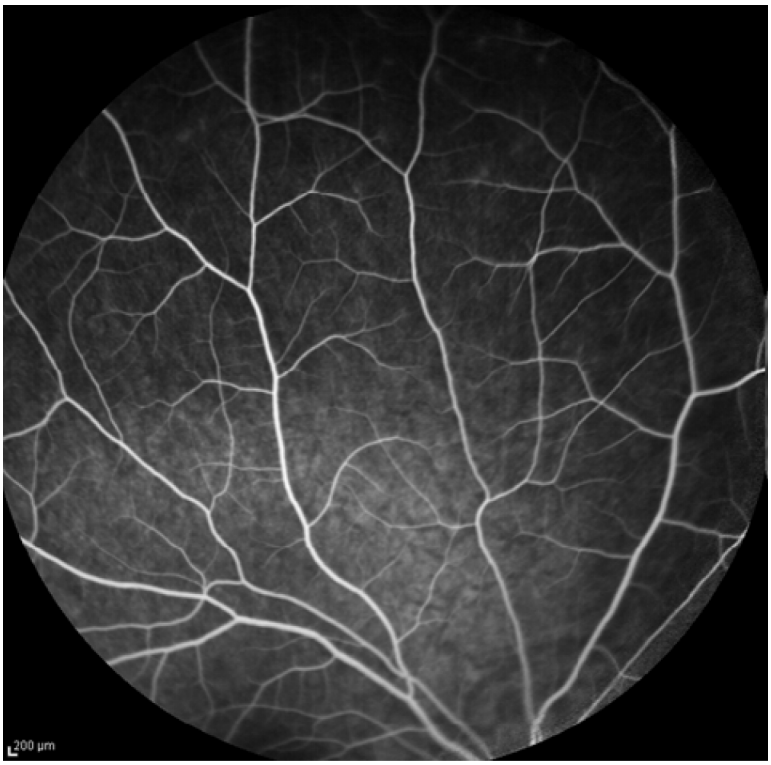

Fig. 2. Fluorescein leakage from peripheral vessels of the right eye on early phase of fluorescein angiography

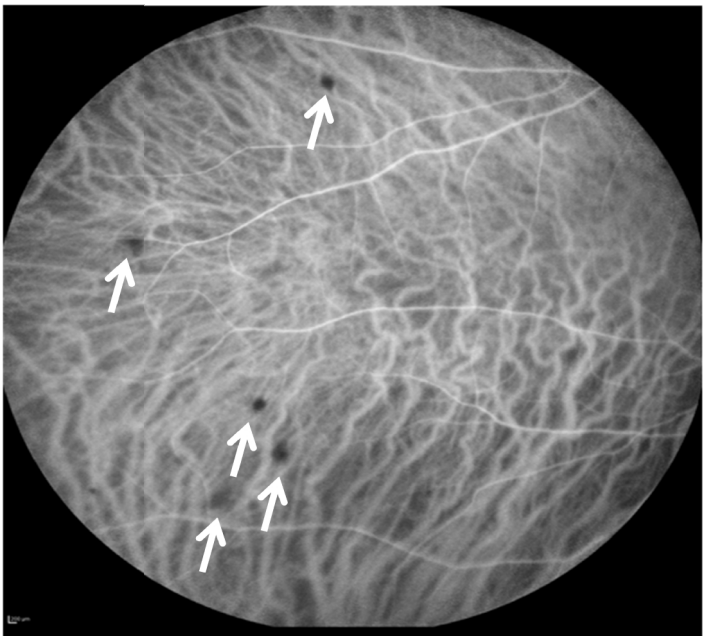

Fig. 3. Hypofluorescent small round spots (arrows) on the right posterior pole on indocyanine green angiography

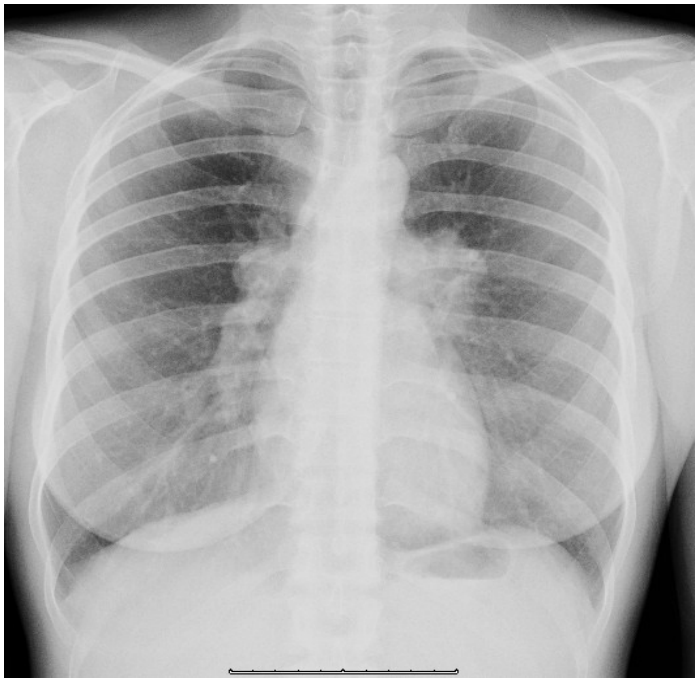

Fig. 4. Posteroanterior chest x-ray showing bilateral hilar lymphadenopathy

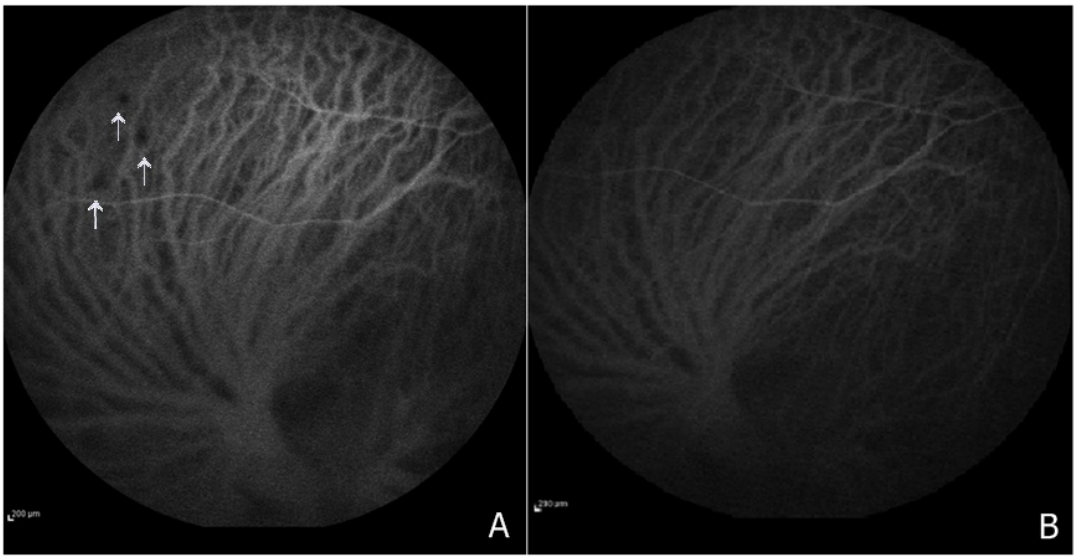

Fig. 5. Hypofluorescent small spots (arrows) on the inferior mid-periphery of right eye on indocyanine green angiography (A), three months later hypofluorescent spots became isofluorescent (B)

3. DISCUSSION

Ocular involvement in sarcoidosis may occur as the disease process, but it also can precede the non-ocular manifestation of the disease [7]. Hunter et al. reported that the first sign of sarcoidosis was ocular granulomatous lesions in 2% of the patients [8]. However, an epidemiological study in Japan showed that the most frequent symptom was eyesight disorder (28.8%) [6]. The same study also reported that incidence of pulmonary and ocular involvements were 86.0% and 54.8%, respectively and eye involvement was the most common extrapulmonary manifestation of sarcoidosis [9]. Although the incidence of pulmonary involvement is nearly one and half times higher than ocular involvement, the most frequent initial symptom of sarcoidosis is eyesight disorder. The reason for this situation is late emergence of symptoms caused by lung involvement such as dyspnea, so symptoms related to ocular involvement generates the major complaints at the first application. Heiligenhaus et al. studied 1800 sarcoidosis patients and indicated that ocular disease was commonly the first manifestation of sarcoidosis (21.2%) [5].

Ocular involvement of sarcoidosis includes intraocular inflammation, adnexal and orbital lesions [10]. Heiligenhaus et al. reported that the most frequent form of eye involvement was anterior uveitis with a rate of 76.4%, followed by intermediate uveitis, 17.3% [5]. Anterior uveitis may either presents as acute iridocylitis or chronic granulomatous uveitis [3]. Large 'mutton fat' type keratic precipitates are most characteristic findings of chronic uveitis. Iris and trabecular meshwork nodules and tent-shaped peripheral anterior synechiae could also be seen. Chronic involvement may also lead to band keratopathy, glaucoma and cataract formation [11]. In our case, chronic granulomatous uveitis was the first manifestation of sarcoidosis proved by biopsy later. We would like to emphasize the importance of our case as being completely asymptomatic until it was incidentally detected.

Considering the intermediate uveitis, snowballs and string of pearls vitreus opacities are suggestive of granulomatous process but differentiation from idiopathic pars planitis and uveitis related to multiple sclerosis is required [12]. Posterior uveitis involves multiple chororetinal lesions, periphlebitis, granulomas of retina and optic nerve. Periplebitis is associated with perivenous cuffing, sheating and exudates referred as 'candle wax drippings' [3]. The present case did not show abnormality on dilated fundus examination and fluorescein angiography showed dye leakage from peripheral vessels which was nonspecific for differential diagnosis. However, indocyanine green angiography revealed infrequent hypofluorescent small round spots in the right posterior pole and midperiphery resembling the type 1 hypofluorescent lesions on early phase that Wolfensberger and Herbort had defined [13]. These spots were invisible on dilated fundus examination and fluorescein angiography. After six weeks, these lesions became isofluorescent as Wolfensberger and Herbort had reported in the late phase of sarcoidosis [13]. Therefore, indocyanine green angiography have a critical role for diagnosis and follow-up especially when the posterior segment involvement could not be detected on routine ophthalmoscopy and fluorescein angiography.

4. CONCLUSION

In conclusion, ocular involvement in sarcoidosis may occur as the first clinical manifestation. Early diagnosis of the disease is important for avoiding ocular complications as well as prevention of other system involvements. For this reason, ophthalmologists have a critical role in the diagnosis and follow-up of the sarcoidosis.

CONSENT

All authors declare that 'written informed consent was obtained from the patient for publication of this case report and accompanying images.

ETHICAL APPROVAL

Not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Newman LS, Rose CS, Maier LA Sarcoidosis. N Engl J Med. 1997;336:1224-1234. Doi: 10.1056/NEJM199704243361706.
2. Dempsey OJ, Paterson EW, Kerr KM, Denison AR. Sarcoidosis. BMJ. 2009;339:b3206. Doi: 10.1136/bmj.b3206 bmj.b3206 [pii].
3. Herbort CP, Rao NA, Mochizuki M. International criteria for the diagnosis of ocular sarcoidosis: Results of the first International Workshop On Ocular Sarcoidosis (IWOS). Ocul Immunol Inflamm. 2009;17:160-169. Doi: 10.1080/09273940902818861 912965083 [pii].
4. Umur KA, Tayfun B, Oguzhan O. Different ophthalmologic manifestations of sarcoidosis. Curr Opin Ophthalmol. 2012;23:477-484. Doi: 10.1097/ICU.0b013e328358c7a6.

5. Heiligenhaus A, Wefelmeyer D, Wefelmeyer E, Rosel M, Schrenk M. The eye as a common site for the early clinical manifestation of sarcoidosis. Ophthalmic Res. 2011;46:9-12. Doi: 10.1159/000321947 000321947 [pii].
6. Sungur G, Hazirolan D, Bilgin G. Pattern of Ocular Findings in Patients with Biopsy-proven Sarcoidosis in Turkey. Ocul Immunol Inflamm. 2013;21:455-461. Doi: 10.3109/09273948.2013.775311.
7. Kaye GB, Salmon JF, Isaacs S. Risk factors for uveitis in sarcoidosis. S Afr Med J. 1993;83:342-344.
8. Hunter DGFC. Ocular manifestations of sarcoidosis. Saunders, Philadelphia; 1994.
9. Morimoto T, Azuma A, Abe S, Usuki J, Kudoh S, Sugisaki K, Oritsu M, Nukiwa T. Epidemiology of sarcoidosis in Japan. Eur Respir J. 2008;31:372-379. Doi: 09031936.00075307 [pii] 1183/09031936.00075307
10. Bradley D, Baughman RP, Raymond L, Kaufman AH. Ocular manifestations of sarcoidosis. Semin Respir Crit Care Med. 2002;23:543-548. Doi: 10.1055/s-2002-36518.
11. Baughman RP, Lower EE, Kaufman AH. Ocular sarcoidosis. Semin Respir Crit Care Med. 2010;31:452-462. Doi: 10.1055/s-0030-1262213.
12. Rothova A. Ocular involvement in sarcoidosis. Br J Ophthalmol. 2000;84:110-116.
13. Wolfensberger TJ, Herbort CP. Indocyanine green angiographic features in ocular sarcoidosis. Ophthalmology. 1999;106:285-289. Doi: S0161-6420(99)90067-2 [pii] 10.1016/S0161-6420(99)90067-2.

Peer-review history:
The peer review history for this paper can be accessed here:
http://www.sciencedomain.org/review-history.php?iid=523&id=23&aid=4677