

A Rare Presentation of Solitary Ocular Sarcoidosis

Kafil Akhtar¹, Suhailur Rehman¹, Sadaf Haiyat¹, Abdul Waris²

¹Department of Pathology, Jawaharlal Nehru Medical College, Faculty of Medicine, Aligarh Muslim University, Aligarh, India

²Institute of Ophthalmology, Jawaharlal Nehru Medical College, Faculty of Medicine, Aligarh Muslim University, Aligarh, India

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Corresponding author:

Dr. Kafil Akhtar,

Professor,

Department of Pathology,

Jawaharlal Nehru Medical College,

Aligarh Muslim University,

Aligarh (U.P)-India

Email: drkafilakhtar@gmail.com

ABSTRACT

Ocular sarcoidosis leads to inflammation of the eye, manifesting as uveitis, scleritis and conjunctivitis with granuloma formation. It can involve any part of the eye and its adnexal tissues, and may result in complications like glaucoma and cataract. Ophthalmic manifestations of sarcoidosis can be isolated, or it may be associated with other organs of the body. The clinical presentation in ocular sarcoidosis is quite varied. Multi-disciplinary modality of treatment is required for both ocular and systemic manifestations of the disease. We report a rare presentation of solitary ocular sarcoidosis in a 42-year-old woman, who presented with complaints of painless left inner canthus swelling and blurry vision for 1 month.

KEYWORDS: Eye, Inner Canthus, Solitary, Sarcoidosis, Uveitis

INTRODUCTION

Sarcoidosis is a chronic idiopathic granulomatous inflammatory disease that was first described by Sir Jonathan Hutchinson in 1878, as a dermatological disorder [1]. It was later in 1909 that Heerfordt, a Danish ophthalmologist, reported for the first time the uveoparotid-fever syndrome (“Heerfordt syndrome”), thus introducing ocular involvement as a clinical manifestation of sarcoidosis.² The eye is involved in approximately 13.0-79.0% cases of systemic sarcoidosis [2, 3]. Sarcoidosis may involve any part of the eye, orbit or the lacrimal system [3, 4]. Uveitis is the most common ocular manifestation of the disease, presenting clinically as eye congestion [3, 4].

Sarcoidosis is a multiorgan granulomatous disorder that most often affects the lungs. Although sarcoidosis has unknown etiology, the association of sarcoidosis with specific occupations, genetic susceptibility and various infectious pathogens has been described [5]. The common clinical manifestations are

shortness of breath, erythematous skin nodules, uveitis, hilar lymphadenopathy and/or parenchymal infiltrates on chest radiography [5, 6].

Biopsy of the affected tissue aids in the diagnosis. Sarcoidosis is histologically characterized by discrete non-caseating granulomas comprising of epithelioid histiocytes. Multinucleated giant cells may be present, occasionally bearing Asteroid bodies or Schaumann bodies [7]. We report a rare occurrence of solitary sarcoidosis of the inner canthus of the eye in a 42-year-old woman, who presented with complaints of painless left inner canthus swelling and blurry vision for 1 month.

CASE PRESENTATION

A 42-year-old woman presented to the Ophthalmic Clinics with complaints of painless left inner canthus swelling and blurry vision for 1 month. She had no systemic diseases and no recent trauma history. Family and social history was non-contributory.

On examination, her visual acuity was 6/18 in both eyes and intraocular pressure was within the normal limits in both eyes. Slit-lamp and fundus examinations revealed no abnormalities. The inner canthus of the left eye was mildly erythematous, with a firm, non-tender, subcutaneous mobile mass of size 3x2 cm (Figure 1). Her extraocular movement showed a slight limitation of supraduction and abduction. Her pupils were isocoric and reacted promptly to light stimuli. A chest radiograph revealed no hilar lymphadenopathy.



Figure 1: The inner canthus of the left eye shows mildly erythematous, firm, non-tender, subcutaneous mobile mass of size 3x2 cm.

Orbital computerized tomography showed a heterogeneous mass without any bony defect or muscle involvement around the left inner canthus (Figure 2). An incisional biopsy was performed. Grossly, the tumor was firm, grayish white and measured 2.3x1.2 cm in size. Histopathological examination showed multiple noncaseating epithelioid granulomas with presence of scattered multinucleated giant cells and asteroid bodies within a giant cell (Figure 3). Foci of asteroid bodies within a langhan's giant cell was also seen (Figure 4). Ziehl-Neelsen stain for acid-fast bacilli was negative. PAS stain performed to rule out any fungal granuloma was negative. Based on the histopathologic findings, a diagnosis of sarcoidosis of inner canthus of eye was given. She was treated with 10 mg of prednisolone per day for 1 month. The eyelid swelling and limitation of extraocular motion resolved. Our patient is doing well after 6 months of follow up without any evidence of recurrence.



Figure 2: Orbital computerized tomography shows a heterogeneous mass without any bony defect around the left inner canthus.

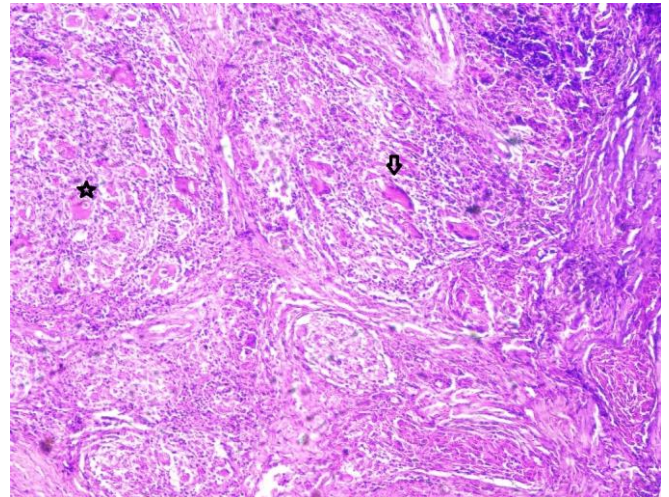


Figure 3: Histopathological examination showed multiple noncaseating epithelioid granulomas with presence of scattered multinucleated giant cells (black arrow). Also seen are asteroid bodies (black star) within a giant cell. Hematoxylin and Eosin, 100X

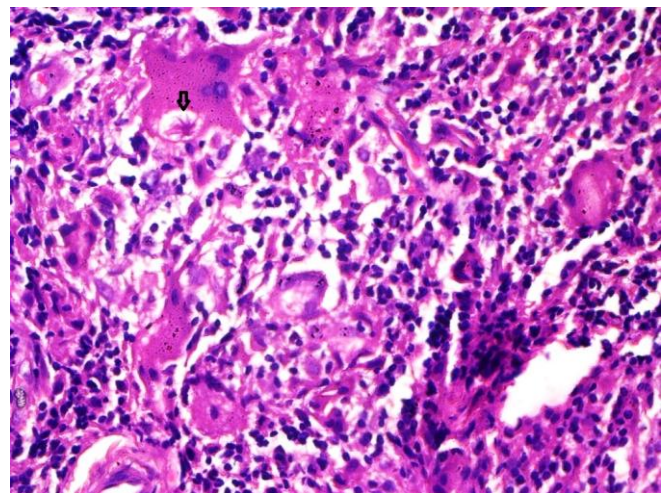


Figure 4: Higher magnification showing Asteroid bodies (black arrow) within a Langhan's giant cell. Hematoxylin and Eosin, 400X.

DISCUSSION

Sarcoidosis is a chronic inflammatory disease, which affects multiple organ-systems of the body, characterized by non-caseating granulomatous lesions of unknown etiology [5]. Ocular sarcoidosis is seen in about 40.0% individuals, with uveitis as the most common clinical presentation [3, 4]. Orbital sarcoidosis predominantly occurs in older persons, with a mean age of 55.9 years, and is more common in the lower lid of females [8, 9]. No racial predilection of orbital sarcoidosis has been documented [10].

Ocular sarcoidosis constitutes one of the leading causes of inflammatory eye disease, exhibiting a wide range of clinical manifestations and can affect the sclera, cornea, conjunctiva, uvea, eyelids, orbit, lacrimal system and the optic nerve [11, 12]. An estimated 20%–30% of patients present with *de novo* ophthalmic findings compatible with sarcoidosis but lack of evident extraocular disease [13, 14]. Cases of extraocular muscle involvement may be seen with orbital lesions [15]. Cutaneous sarcoidosis of the eyelids presents in the form of millet-seed nodular lesions [16]. The lacrimal gland is reported to be affected in 15.8% of ocular sarcoidosis [17]. The diagnosis of lacrimal gland involvement is based on clinically enlarged gland, presence of dry eye and microscopic presence of chronic non-caseating granulomatous inflammation of the nasolacrimal duct mucosa [7, 17]. Our case of isolated orbital sarcoidosis presented with left inner canthus swelling, without any systemic disease.

In 2009, the International Workshop on Ocular Sarcoidosis (IWOS), published criteria for diagnosing ocular sarcoidosis, based on the presence of seven clinical signs suggestive of ocular sarcoidosis: 1. Mutton-fat keratic precipitates/small granulomatous keratic precipitates and/or iris nodules (Koeppe/Busacca), 2. Trabecular Meshwork nodules and/or tent-shaped peripheral anterior synechiae, 3. Vitreous opacities displaying snowballs/strings of pearls, 4. Multiple chorioretinal peripheral lesions (active and/or atrophic), 5. Nodular and/or segmental periphlebitis (\pm candle-wax drippings) and/or retinal macroaneurysm in an inflamed eye, 6. Optic disk

nodule(s)/granuloma(s) and/or solitary choroidal nodule, 7. Bilaterality [18].

Known to be a systemic disorder, sarcoidosis affects multiple major organ systems, primarily the lungs in more than 90% of cases, which tend to be in the spotlight of clinical attention [19]. Extrapulmonary disease frequently involves the lymph nodes, skin, eye, cardiovascular, musculoskeletal, gastrointestinal, renal, and central nervous systems [20, 21]. Despite it being known for more than 100 years, sarcoidosis remains an enigmatic disease, the etiology of which has still to be resolved, demonstrating a heterogeneous clinical course that often poses a diagnostic and treatment challenge for the treating physician. Our case highlights the rare clinical manifestation of a primary ocular sarcoidosis.

Though a prior history of sarcoidosis is rare, the chest x-ray usually reveals enlarged hilar lymph nodes. High-resolution computed tomography of the chest may be a sensitive investigative modality, in suspicious cases of sarcoidosis, with a normal chest radiograph [22].

The management of orbital sarcoidosis depends on the exact site of disease, degree of functional loss, with or without active systemic disease. Oral corticosteroids are the mainstay in the treatment, with a good therapeutic response [23]. Oral prednisolone, in a dosage of 1 mg/kg body weight, tapered over 3 months is the initial therapy, in cases without active systemic disease. Cytotoxic drugs such as methotrexate may be used in steroid resistant cases. Methotrexate and mycophenolate mofetil, a selective suppressor of T- and B-lymphocyte proliferation are favored as first-line steroid-sparing agents [23]. In localized orbital disease, 1-ml injection of triamcinolone acetonide in dosage of 40 mg/ml may be considered [23]. TNF inhibitors like subcutaneous adalimumab and Infliximab, a chimeric IgG1 monoclonal antibody against TNF α , have mainly been used in sarcoidosis-related uveitis [24, 25]. Surgical excision is an effective treatment for localized orbital mass lesion [26]. Our patient tolerated well the steroidal treatment of 10 mg prednisolone per day for 1 month. Her eyelid swelling and limitation of extraocular motion resolved and she is doing well after 6 months of follow up without any evidence of recurrence.

CONCLUSION

Ocular disease is an important manifestation of sarcoidosis, and can lead to significant visual disability or blindness if not adequately or promptly treated. A wide variation in clinical presentation renders this entity one of the most challenging to diagnose and manage, usually requiring a multidisciplinary approach. It is a rare occurrence in the eye, but is known to recur with systemic disease. So clinicians should be aware of the common sign and symptoms of ocular sarcoidosis. Long-term follow-up is recommended in all cases, for early diagnosis of active systemic disease.

Conflict of Interest

Authors declare none.

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Author's contribution

KA performed the reporting of the case and wrote the paper; SR collected the data, SH did the literature search and AW did the clinical inputs of the case.

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