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Case Report

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Rare Case of Sarcoidosis Presenting As Bilateral Iridocyclitis

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Abstract

Background: *A case of acute onset bilateral iridocyclitis is reported in a previously undiagnosed and asymptomatic case of sarcoidosis.*

Case: *A 39 years old male presented with pain and redness in the right eye for one day. Slit lamp examination revealed granulomatous iridocyclitis in the eye with three hundred sixty degree posterior synechiae. While under investigation for the cause, on fourth day of initial presentation, left eye also developed similar granulomatous iridocyclitis, being symptomatic only overnight. All laboratory investigations, radiological evaluation and biopsy led to the final diagnosis of sarcoidosis.*

Conclusion: *Sarcoidosis is a multisystem granulomatous disorder of unknown etiology which may present as an acute uveitis or chronic form. As definite prevalence studies are not available, it should be considered in the differential diagnosis of all cases of uveitis in Nepal.*

Keywords: Sarcoidosis, Granulomatous, Iridocyclitis, Uveitis, Multisystem

Introduction

Sarcoidosis is a systemic disorder of unknown cause that is characterized by its pathological hallmark, the non-caseating granuloma [1]. Despite multiple investigations, the cause of sarcoidosis remains unknown. Currently, the most likely etiology is an infectious or noninfectious environmental agent that triggers an inflammatory response in a genetically susceptible host. Among the possible infectious agents, Propionibacter acnes, atypical mycobacteria and environmental exposures to insecticides and mold have been associated with an increased risk for disease [2]. Worldwide, the prevalence of the disease varies from 20-60 per 100,000 for many groups such as Japanese, Italians, and American whites [3]. Sarcoidosis often occurs in young, otherwise healthy adults with a second peak in incidence develops around age 60.

Clinical medicine reference states ocular involvement in sarcoidosis occurs in up to 29% of total cases with ocular symptoms being the presenting complaints in 12% of cases [4]. However, ophthalmology literature states that ocular involvement occurs in up to 78% of cases of sarcoidosis [5]. It has been stated that there are racial variations in the ocular involvement in cases of sarcoidosis with more involvement in Japanese (up to 70%) than in the US (20%) [3]. Ocular sarcoidosis is diagnosed based on results of the first international workshop on ocular sarcoidosis (IWOS criteria) [6].

There has been only a single study regarding epidemiology of sarcoidosis in Nepal. Hence, we report an interesting case of sarcoidosis with bilateral iridocyclitis with bilateral hilar lymphadenopathy without clinical thoracic complaints.

Case Description

A 39 years old male presented in the outpatient department with complaints of severe pain in the right eye for one day. Pain was associated with redness, watering, severe photophobia and blurred vision. There was no history of trauma to the eye, any foreign body entering the eye, no discharge and no foreign body sensation. Patient was a regular computer user with 6-8 hours on computer on average. One month ago, he had been diagnosed with dry eye disease (Schirmer 5mm OD, 2 mm OS in 5 minutes repeated after 15 days Schirmer 12mm OD, 8 mm OS in 5 minutes) and was on continuous treatment with lubricating eye drops for the same.

Patient had no history of similar illness in the past and no history of previous ocular trauma or surgery. There was no known history of any chronic disease. He had no history of joint pain, chronic cough, skin rashes, and ulcers in any site of the body or low grade fever. He also had no history of contact with patients of tuberculosis or leprosy.

He was married, manager of a technical institute by profession and commonly used motorbike for transportation. On examination: Vision 6/36 OD, 6/6P OS.

OD

Lids were mildly edematous, conjunctiva showed conjunctival congestion as well as circumcorneal congestion. Cornea showed

normal looking epithelium with mild stromal edema and small grey mutton fat keratic precipitates in Arlt's triangle. Anterior chamber was normal in depth peripherally with convex iris contour. Cell content of AC was 3+ (SUN classification). Iris was muddy in appearance with no specific nodules, no neovascularization. Pupil was round, 3mm in size, non-reactive to light or accommodation. Lens was apparently clear. Posterior segment was barely visible through constricted pupil.

OS

Lids were normal. Conjunctiva showed mild congestion. Cornea was clear, anterior chamber had normal depth and was quiet, iris contour was convex. Pupil was round, regular, reactive to light and accommodation. Lens was clear. Vitreous was clear and quiet. Retina was flat with normal color, shape, size and margin of optic disc and cup: disc ratio of 0.3:1. Foveolar reflex was present. With these initial findings, patient was advised for IOP measurement and dilatation and funduscopy. The IOP as measured by Goldmann tonometer was 12 mm Hg OD and 10 mm Hg at 10 AM.

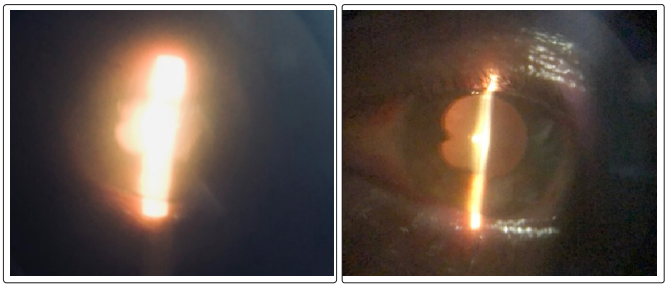

Figure 1: Slit lamp appearance of pupil (festooned pupil) after first dose of atropine

On dilatation with topical tropicamide and phenylephrine eye drops, though the congestion decreased and pain deceases, there was no dilatation as such. Patient was then applied atropine eye drops at 15 minute intervals for 3 attempts. While the left eye had fully dilated by this time, pupil of right eye did not dilate. Hence injection mydricaine 0.5ml was given subconjunctivally and patient was advised to rest for half an hour. After this interval, pupil was fully dilated in right eye with a synechiae remaining at 6 o'clock. There were iris pigments on anterior lens surface. Vitreous cells were 4+. Retina details were unclear with optic disc visible with normal colour shape size margin and a cup disc ratio of 0.3:1.

As the case was suggestive of granulomatous uveitis, as per international recommendations, search for systemic disease was done.

Systemic evaluation of respiratory, cardiovascular system and central nervous system were unremarkable.

The investigations advised were: complete haemogram, urine routine and microscopic examination, chest x-ray, Mantoux test, VDRL, RPR, ACE (angiotensin converting enzyme) and PCR-TB (highly sensitive test for tuberculosis).

Patient was referred to internist for evaluation of chronic ystemic disease. By that time, chest X-Ray (PA view) showed bilateral symmetric hilar lymphadenopathy with fibrotic changes in the lungs.

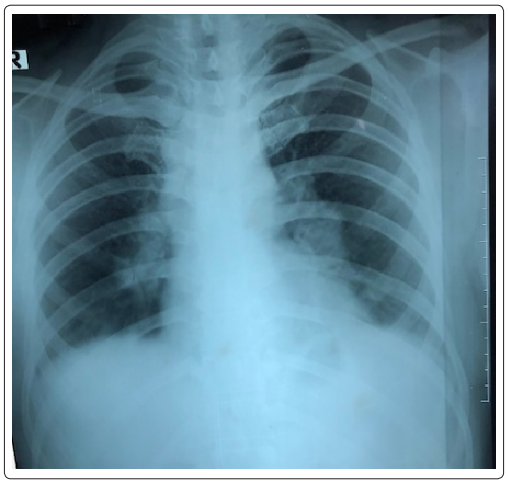

Figure 2: Chest X-ray shows bilateral lymphadenopathy

In the meanwhile, patient developed pain in the left eye overnight. Pain was constant dull aching type and was associated with redness of the eye, watering and blurring of vision. There was no history of colored haloes or discharge or foreign body sensation.

On Examination, OS, there was mild lid edema, conjunctival congestion and circumcorneal congestion. There was mild stromal edema in the cornea and multiple small grayish mutton fat KPs on the inferior corneal endothelium. The anterior chamber was of normal (VH grade IV) depth and anterior chamber reaction showed 2+ cells with no flare. Iris was muddy in appearance with multiple (5) Koeppe's nodules and Busacca's nodules (3) on anterior iris surface. Pupillary reaction was sluggish. However there was no formation of posterior synechiae. Lens was grossly clear.

With these findings, patient was advised to take IOP and then dilate OS. The IOP was 12 mm Hg in both eyes. After dilatation with Atropine, vitreous cells were seen to be 4+ (SUN classification). No vitreous snowballs or snow banking was observed. Retina was flat, with no evidence of vasculitis, no hemorrhages and no exudates.

The Results of Laboratory Examinations Showed Following

Parameter	Normal value/ Method	Patient report
Complete hemogram	------	Normal
ESR	0-9 mm in 1st hour	27 mm in 1st hour
VDRL	Flocculation method	Non-Reactive
Urine (Routine and microscopic examination)	------	Normal
RA factor	------	Non reactive
PCR-TB (specific test for Tuberculosis)	Not detected	Not detected
Angiotensin Convertin Enzyme(ACE)	8-52 U/L	71.30 U/L

First diagnosis with these investigation reports in mind was sarcoidosis. As the most suggestive sign of disease in the eye is inflammatory infiltrates at the AC angle and/or tent shaped peripheral

anterior synechiae, gonioscopy was mandatory. It revealed open angle (visible sclera spur) with dispersed pigments in trabecular meshwork. There were multiple inflammatory infiltrates in the angle but no PAS was seen. This finding further increased the likelihood of sarcoidosis being the final diagnosis.

Liver Function Tests (LFT) were Also Performed Which Reported as Follows

Parameter (LFT)	Normal value	Test value
Serum bilirubin	Upto 1.4 mg/dl	1.4 mg/dl
Direct bilirubin	Upto 0.3mg/dl	0.7mg/dl
SGPT(ALT)	5-45 IU/dL	65 IU/dL
SGOT(ALP)	5-40 IU/dL	45 IU/dL
S. Alkaline phosphatase	80-306 IU/dL	170 IU/dL
S. Total protein	6.4-8.3 gm/dL	8.1 gm/dL
S. Albumin	3.8-4.9 gm/dL	4.9 gm/dL

CT of the chest (non contrast and contrast) showed B/L hilar lymphadenopathy with minimal fibrotic changes in the right lung shown as below.

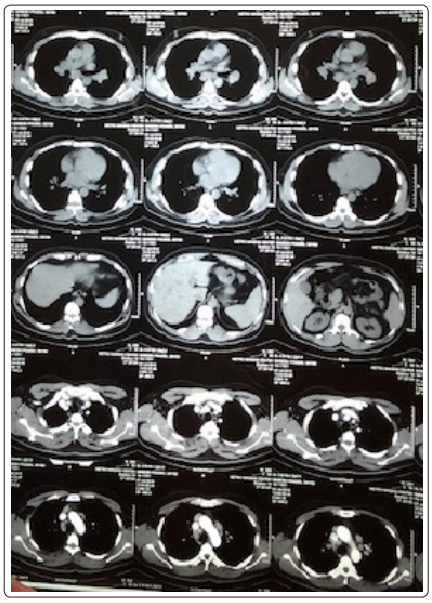

Figure 3: CT scan chest shows bilateral lymphadenopathy with fibrotic lung changes

Hence the diagnosis suggested by internist was Sarcoidosis stage IV. Pulmonologist's opinion was also the same and patient was started on oral steroids. Patient was followed up on regular intervals. His pain decreased and vision became gradually clear. Vision improved to 6/6P OD and 6/6 OS.

On examination, at 1 week of first presentation, patient had symptomatically improved and vision was stable. Retina consultation was done at vitreo-retina department of Bharatpur Eye Hospital for further opinion as soon as the media had cleared considerably.

The only significant findings were:
OD: AC reaction of 2+ and anterior vitreous cells 4+. Lens still had iris pigment at 11 o'clock and posterior synechiae at 6 o'clock. No retinal or choroidal involvement seen. IOP-8 mm Hg.

OS: AC reaction of 4+ and anterior vitreous cells 2+. No vitreous snowballs or string of pearls. Lens was clear with no posterior synechiae. Whitish scar <1 clock hour noted at 1 0'clock at ora serrata (indirect ophthalmoscopy with scleral indentation). IOP- 10 mm Hg.

Endobronchial Biopsy was done under USG guidance (EBUS). Biopsy reports stated: epithelioid cell granulomas in a background of polymorphous lymphoid cells, which further supports the diagnosis. Biopsy results were negative for Tuberculous bacilli by PCR technique (no TB bacilli DNA was detected).

Discussion

Although sarcoidosis is considered a rare disease in the South East Asia region, it is of considerable importance in terms of morbidity. A previous study in a tertiary hospital in Kathmandu shows it is a significant disease to consider, particularly noting the symptomatology closely resembles tuberculosis. Regarding ocular morbidity, a study had been done at a tertiary eye hospital in Kathmandu (November 2008-April 2010) to find systemic associations of uveitis [7]. This study reported sarcoidosis to be a cause of 1.6% of the total cases (308) investigated. There were 5 cases proven to be sarcoidosis and all presented as panuveitis.

Another study was performed at a tertiary care hospital in Kathmandu which retrospectively analyzed the charts of cases diagnosed as sarcoidosis in the interval from January 1st 2005 to December 31st 2010 [8]. In this interval, 42 cases were diagnosed as sarcoidosis of which 55% were females and 45% were males. Of the cases studied, only 2 patients (5%) had ocular symptoms.

The clinical presentation of this case was unusual as per previous data available from this region as the patient was not only previously asymptomatic and undiagnosed, but even though the onset of eye symptoms was acute, the signs were of chronic type. Also, this seemed to be an exacerbation of a disease that was present for a long duration as the second eye was involved with similar disease in less than a week time.

Conclusion

Sarcoidosis should be considered as one of the important diagnostic possibilities in all cases of granulomatous uveitis inpatients in Nepal.

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