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A case of oculocutaneous sarcoidosis

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ABSTRACT

Purpose: To present a case of extrapulmonary sarcoidosis presenting with ocular and cutaneous involvement.
Observations: We report a 54-year-male who presented with bilateral redness of eyes, photophobia, and diminished vision for a week. The best corrected visual acuity in the right eye was 6/60 and the left eye was counting fingers close to face (CFCF). He also had multiple brown plaques on the nape of the neck, chest, back, and arms. Furthermore, he was on multiple antipsychotic drugs for schizophrenia for 3 years. Uveitis investigation workup revealed raised serum angiotensin converting enzyme (ACE), negative Mantoux, and other serological tests. The patient was treated for acute anterior uveitis secondary to sarcoidosis. Clinical improvement was seen after a few days following treatment. The patient presented a year later with multiple yellowish conjunctival nodules in the superior bulbar conjunctiva associated with hyperemia. A biopsy of the plaque like skin lesions was done, which suggested cutaneous sarcoidosis. Involvement of the skin and the eyes raised suspicion that the persistent psychotic episodes despite multiple antipsychotic drugs could be attributed to neurosarcoidosis. However, magnetic Resonance Imaging (MRI) of the brain and orbit showed normal findings. After treatment with corticosteroids and immunosuppressives (methotrexate), the conjunctival nodules as well as skin lesions drastically improved, and the psychosis also responded well to clozapine.
Conclusion: A high index of suspicion is needed in cases presenting with granulomatous uveitis with multisystem involvement. Long-term follow-up is crucial to monitor the disease progression and adverse effects of medications.

1. Introduction

Sarcoidosis is a chronic multisystemic granulomatous disorder characterized by the formation of non-caseating granulomas in affected organs. It can affect virtually every organ of the body, pulmonary involvement being the most common occurring in around 90% of patients. Other organs affected are skin, eye and liver. Ocular involvement is common and has been reported in up to 78% of patients. Skin can be affected in around 25–35% of patients.[1] Here, we present a case of 54-year-old male with oculocutaneous sarcoidosis without pulmonary involvement.

1.1. Case report

A 54-year-old male with a known history of psychotic illness presented with redness of both eyes associated with photophobia and blurred vision for one week. There was no history of fever, cough, joint pain, or past history of tuberculosis.

On examination, the best corrected visual acuity in the right eye was 6/60 and the left eye was counting fingers close to face (CFCF). There was bilateral circumcorneal congestion and mutton-fat keratic precipitates (KPs) on the corneal endothelium (Fig. 1). There were 4 +cells and 2+ flare in the anterior chamber of both eyes. The posterior segment could not be evaluated properly due to marked anterior segment inflammation. The intraocular pressure was 16 mm and 18 mm of Hg in the right and left eyes, respectively.

He also had multiple, non-painful, non-itchy brown plaques on the nape of the neck, chest, back, arm and forearm (extensor aspect)(Fig. 2). These were present two years before the onset of ocular symptoms. Dermatology consultation was advised but the patient deferred

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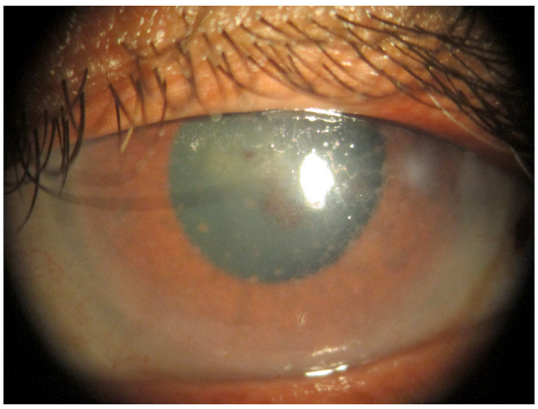

Fig. 1. Circumcorneal congestion and mutton fat KPs.

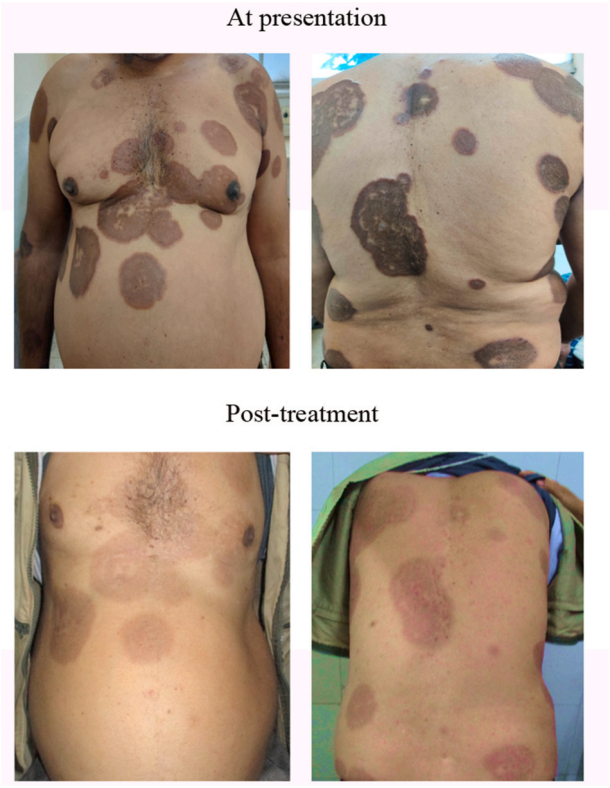

Fig. 2. Multiple, brownish plaques on chest, back at presentation and resolving lesions after treatment.

consultation.

A diagnosis of bilateral granulomatous anterior uveitis was made and routine uveitis investigations were sent. On investigation, the routine haemogram, blood sugar and urinalysis were normal, and the serological tests for venereal disease research laboratory (VDRL), human immunodeficiency virus (HIV),and toxoplasma antibody test were negative. Mantoux's test was also negative. Serum angiotensin converting enzyme (ACE) level was 85 U/L (normal range:12–68 U/L).The serum calcium level was normal (2.20mmol/L). Cardiac workup, renal function tests and liver function tests were normal.

To exclude other causes of autoimmune uveitis, anti-nuclear antibodies (ANA), antineutrophil cytoplasmic antibodies (c-ANCA, p-ANCA), antibodies to double-stranded DNA (anti dsDNA), anti SS-A(Ro) antibodies, and anti SSB(La) antibody were sent and was found to be negative.

Based on the presence of bilateral granulomatous anterior uveitis, a negative mantoux test, and a raised serum ACE level, the patient was suspected to have uveitis secondary to sarcoidosis. Pulmonary function tests were normal. High-resolution computed tomography (HRCT) of the chest showed emphysematous changes in both lungs with bilateral pleural thickening. This could be attributed to his history of smoking and sequelae of coronavirus disease (COVID) pneumonia, as analysis of bronchoalveolar lavage fluid showed a CD4/CD8 ratio of less than 2.

He was treated with topical (1% predacetate every 2 h) and oral (prednisolone 1mg/kg/day) steroids in a tapering dose and cycloplegics (1% tropicamide). The inflammation subsided and the visual acuity improved to 6/18 in the right eye and 6/24 in the left eye after three weeks. Posterior segment examination was found to be normal in follow-up visits.

One year later, the patient presented with bilateral fullness of the lids. On examination, there were multiple, nontender, elevated, yellowish conjunctival nodules in the superior bulbar conjunctiva associated with hyperemia (Fig. 3). The visual acuity was 6/60 in the right eye and 6/36 in the left eye. There was no restriction of extraocular movements. no diplopia, proptosis, or ptosis. There was mild anterior segment inflammation with 0.5+ cells. Nuclear sclerosis grade 2 (LOCS III) was present bilaterally. The posterior segment examination was normal.

The skin lesions on the chest, back, and nape of the neck were unchanged from the original presentation. A dermatologist consultation was done, and the histopathology of the biopsy of the skin lesions showed multiple epithelioid cell granulomas separated by band of lymphocytes with no necrosis suggestive of granulomatous dermatitis (Fig. 4). In light of a skin biopsy suggesting cutaneous sarcoidosis and granulomatous uveitis suggestive of ocular sarcoidosis, we suspected that the psychotic episodes could be related to neurosarcoidosis. Thus, neurological consultation was done, and magnetic resonance imaging (MRI) of the brain and orbit was advised. A plain and contrast enhanced MRI of the brain and orbit showed nonspecific changes secondary to aging but no findings consistent with neurosarcoidosis.

MRI findings not being suggestive of neurosarcoidosis and the persistence of psychotic symptoms despite multiple antipsychotics, our patient was diagnosed with treatment resistant schizophrenia and was started on clozapine by the psychiatry team.

Patient was started on oral steroids (prednisolone 1mg/kg/day), methotrexate 15 mg once a week, and folic acid 5 mg twice weekly after all the baseline investigations were found to be normal. On follow-up examination, skin lesions were improving (Fig. 2) and conjunctival nodules resolved (Fig. 3).

After using immunomodulators for four months, the patient underwent bilateral cataract surgery (phacoemulsification with foldable intraocular lens implantation). The procedure was scheduled following a two-month period of quiescence, coupled with preoperative and postoperative oral steroid administration. The early postoperative period was uneventful. However, in the follow-up visits, he had raised intraocular pressure (IOP of 30 mmHg) in the left eye, probably due to steroid use. The steroid was tapered and antiglaucoma medications (timolol and brimonidine) was prescribed. IOP was not controlled despite the use of two drugs; thus, Brimatoprost was added. After a few weeks, patient developed an ocular surface disorder associated with brimatoprost in the left eye. Thus, Brimatoprost was stopped and the associated keratopathy was treated. Following two weeks, the corneal findings improved, the IOP was under control, and the vision in the right eye and left eye was improved to 6/9 and 6/24, respectively. There was a nebular scar in the inferior cornea of left eye, with scleral thinning superiorly and no cellular activity in the anterior and posterior segment. No retinal/choroidal lesions were present. Cupping of 0.2 in the right eye and 0.8 in the left eye was present with normal IOP. The patient is being treated with artificial tears, low dose oral steroid (2.5mg every other day), methotrexate 15 mg, folic acid, and antipsychotic drugs.

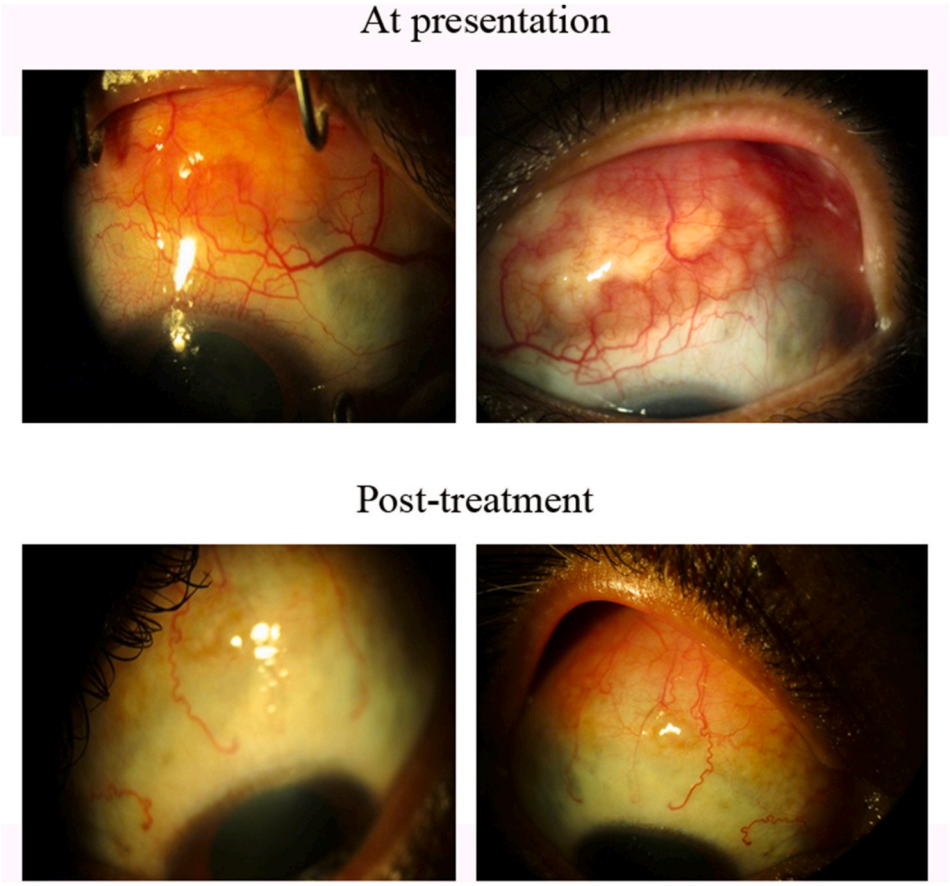

Fig. 3. Multiple yellowish conjunctival nodules infiltrating the superior bulbar conjunctiva and extending to the fornix in both eyes at presentation and resolved lesions after treatment.

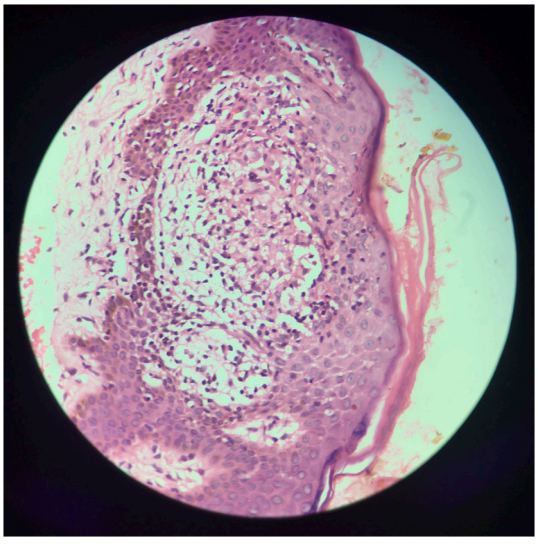

Fig. 4. Histopathology showing noncaseating granuloma.

2. Discussion

Extrapulmonary involvement in sarcoidosis is quite common, but isolated cases of extrapulmonary sarcoidosis are a rare entity with less than 10% of cases reported in the literature. The diagnosis depends on a compatible clinical presentation and radiographic findings; however, a definitive diagnosis can only be established with the histopathological detection of epithelioid noncaseating granuloma of the affected organ and the exclusion of other diseases that may have similar presentations.[2]

Ocular involvement may be the initial manifestation of sarcoidosis, and anterior uveitis is the most common presentation.[3] The uveitis is characteristically granulomatous with medium to large-sized, mutton--fat KPs. Other ocular manifestations of sarcoidosis include dacryoadenitis, conjunctival nodules and granulomas, scleritis, episcleritis, non-specific conjunctivitis, interstitial keratitis, intermediate uveitis, posterior uveitis, and optic neuritis.

Cutaneous involvement in sarcoidosis may occur at any stage of the disease, but is most often present at the onset. Skin lesions may appear at any stage of the disease, and specific skin lesions may occur in 9%–37% of the patients.

Specific skin lesions associated with sarcoidosis are lupus pernio (LP), infiltrated plaques, maculopapular eruptions, subcutaneous nodules and scars, and rare morphologies such as alopecia, ulcers, hypopigmented patches, and ichthyosis.[4]

Neurosarcoidosis occurs in approximately 5%–15% of cases. Cranial neuropathy (most commonly of the optic nerve and facial nerve) is the most common neuro-ophthalmic presentation of sarcoidosis.The patient may also present with papilledema, nystagmus, visual field defects, abnormal eye movements, visual hallucinations, encephalopathy, seizures, aseptic meningitis, and psychiatric symptoms.[5] The typical imaging finding in neurosarcoidosis is enhancement of leptomeninges and thickening with a predilection around the base of the brain. Other imaging findings include periventricular and white matter lesions, sellar and suprasellar involvement, involvement of cranial nerves, cavernous sinus, orbit lesions, and intramedullary lesions. No such findings were seen in our patient.

Our patient presented with cutaneous and ocular manifestations

without pulmonary involvement, which responded well to steroids and immunosuppressives. A similar case of sarcoidosis has been reported by Aggarwal H.K(1).To our knowledge, this is the second report of isolated skin and ocular involvement in sarcoidosis. In the report by Aggrawal H. K, the patient presented with granulomatous iridocyclitis similar to our case but without conjunctival nodules. The cutaneous involvement included multiple erythematous skin lesions on the dorsum of hand, whereas in our case, much widespread plaques were present. Serum ACE was raised in both cases.

3. Conclusion

Ocular disease may be the initial presenting sign of sarcoidosis. Early diagnosis and prompt treatment can prevent profound visual loss. Because of the multisystem involvement of the disease, a multidisciplinary approach is often required. Furthermore, optimal control of the systemic disease has been shown to play an important role in controlling eye inflammation as well. Immunosuppressive agents and biologic anti-TNFα drugs have revolutionized the management of chronic disease and should be considered as steroid-sparing therapy to minimize steroid related complications and achieve long-term reduction of ocular and systemic morbidity. Regular follow-up is crucial, as the patients may later progress to have multi-system involvement.

Patient consent

Written informed consent was obtained from the patient for publication of case report and any accompanying images.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

All authors have no financial disclosures.

Acknowledgement

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