OCULAR SARCOIDOSIS — A CASE REPORT

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SYPNOSIS

A case of sarcoidosis involving the eye is reported and its significance discussed.

INTRODUCTION

Sarcoidosis is a multisystem granulomatous disorder of unknown aetiology characterised by the presence in all affected tissues of epithelioid cell granuloma without caseation but occasionally may have some central necrosis.

This disease is relatively rare in Malaysia and Singapore. The first reported case in this region was by Tang et al in 1964 (1). Since then many other cases have been reported but there was no mention of any ocular involvement (2, 3, 4). We describe a case of sarcoidosis with classical ocular involvement.

CASE REPORT

A s/o M, an Indian male patient aged 38, was first seen in the Medical Unit of the University Hospital with complaints of three months duration of non-productive cough, weight loss, easy fatigability, aching sensation of joints and low backache.

Physical examination was essentially normal. Chest x-ray showed bilateral hilar lymphadenopathy. Erythrocyte Sedimentation Rate was 36 mm for 1 hr and other investigations viz Haemoglobin, Total White Blood Count, Packed Cell Volume, Urea, Glucose, Serum Sodium, Potassium, Calcium, and Serum Proteins were normal. Mantoux test was negative. Sputum staining and culture for acid fast Bacilli were also negative on three occasions. Two weeks after admission an open mediasternal lymph node biopsy was done. It showed multiple non-caseating granuloma with epithelioid cells and occasional multi-nucleated giant cells. These findings were compatible with tuberculosis or sarcoidosis. Staining for fungi was negative. Since tuberculosis is the commonest cause of granuloma in this country, he was treated with Rifampicin, INH and pyridoxine. A month later after admission he was referred to the Eye Department for complaints of blurred vision in both eyes. Clinically there was no response to anti-tuberculosis therapy.

OCULAR EXAMINATION

The patient could see 6/6 and read N5, and had normal colour vision in both eyes. Both eyes look quiet to the naked eye. however under slit lamp examination there was evidence of anterior uveitis with keratic precipitates on the corneal endothelium, and flare and cells in the anterior chamber. Both retrolental spaces showed inflammatory cells. Fundus examination under mydriasis showed evidence of candle wax dripping along the inferior temporal vessel with scattered choroidal nodules in the periphery (see Fig. 1). There were also numerous 'snow ball' opacities in the inferior vitreous.

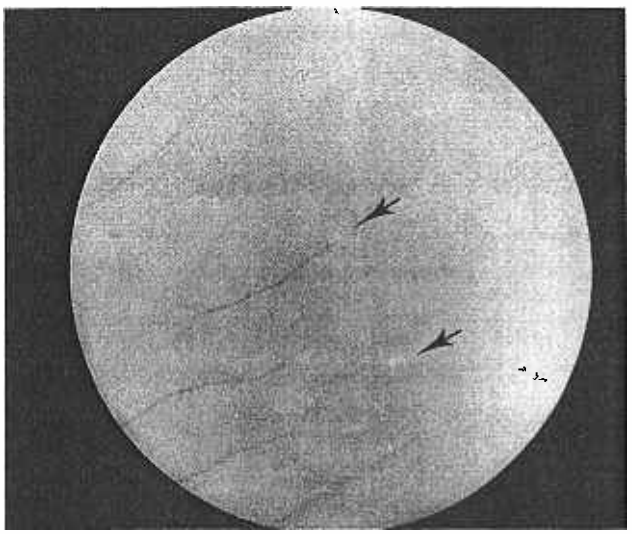

Fig. 1 CANDLE WAX EXUDATE (arrows)

Intraocular pressures were normal predilatation. However pressures were raised to 30 mm Hg after dilating both pupils. Conioscopy showed evidence of peripheral anterior synechiae. These findings were highly suggestive of sarcoidosis. He was treated with Dexametosone eye drops. No systemic steroids was given until the Kveim test was performed.

Results of other investigations were now available. 24 hours measurement for urine calcium was normal and lung function tests showed mild restrictive impairment. The Kveim test was positive. The diagnosis of sarcoidosis was established and he was commenced on oral prednisolone 40 mg daily.

Subsequent follow up showed clearing of the candle wax exudate, snow ball opacities and anterior uveitis.

DISCUSSION

The earliest description of sarcoidosis drew attention to the cutaneous manifestation of the disease. Sixty years were to elapse before it was realised that the eye could also be involved. Involvement of the eye occurs in 1/4 patients with multisystem sarcoidosis and sarcoidosis account for 4% of uveitis (5).

A recent retrospective survey of sarcodosis in 11 major cities of New York, London, Paris, Los Angeles, Tokyo, Novi Sad (Yugoslavia), Lisbon, Edinburgh, Reading, Geneva and Naples showed that sarcoidosis presents universally in 20-40 years age group and presents to 3 different disciplines - the chest physician, dermatologist and ophthalmologist (6). Ocular involvement may be anticipated in 15% of patients (6). From the ophthalmological point of view ocular sarcoidosis has two important implications. Firstly these characteristic ocular lesions often play a major role in the diagnosis of systemic sarcoidosis and secondly they may lead to blindness unless adequately controlled (7).

1. **Anterior Uveitis** is the most common lesion and can be granulomatous or non granulomatous in nature. Acute sarcoid iridocyclitis has an abrupt onset associated sometimes with erythema nodosum and bilateral hilar lymphadenopathy and responds well to local steroids. Chronic sarcoid iridocyclitis has an insidious onset and is normally complicated by secondary glaucoma and cataract as the disease progresses.
2. **Posterior uveitis** comprises choroidal nodules, papilloedema, haemorrhage, retinal perivasculitis, candle wax exudate and snowball opacities.
3. **Other** ocular involvement includes non specific conjunctivitis, conjunctival follicle, phylctenular conjunctivitis, keratoconjunctiva sicca, and scleral plaques.

Uveitis develops during the silent stage of the systemic sarcoidosis in about 80% of patients (8) as is seen in this patient who had a grumbling uveitis with extensive, peripheral anterior synechiae causing damage to his drainage angles. This may occur in eyes that look normal to the naked eye (white uveitis).

We feel that all patients with suspicion of sarcoidosis should be referred to an ophthalmologist for proper examination under the slitlamp and binocular indirect ophthalmoscopy.

The Kveim Slitzbach test is positive in 80% of patients with sarcoidosis. However this test can be suppressed by oral corticosteroid (6). Conjunctival granuloma if present is the best site to biopsy for sarcoid lesion (8).

As for treatment of ocular lesions, it is recommended to initially try patients on topical corticosteroids for iridocyclitis failing which local subconjunctival depot may be used. Oral steroids are indicated if local treatment does not lead to rapid response or if ophthalmoscopy reveals posterior uveitis.

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