· Case report ·

Ocular and renal sarcoidosis in an Asian teenager: a case report

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Abstract

• Sarcoidosis typically affects young adults with bilateral hilar lympadenopathy, lung parenchymal disease and/or skin lesion. Childhood sarcoidosis is relatively a rare entity. Concurrent ocular and renal involvements are extremely uncommon especially in childhood sarcoidosis. We presented a case of an Asian teenager with bilateral recurrent anterior uveitis and features of renal failure due to childhood sarcoidosis. He showed remarkable clinical improvements after combination of oral corticosteroids and methotrexate. His final visual outcome was satisfactory following augmented trabeculectomy in both eyes due to intractable glaucoma. It is essential to make a prompt diagnosis and initiate proper management to avoid ocular mobidity in this unusual presentation.

• KEYWORDS: ocular; renal sarcoidosis; Asian; teenager
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INTRODUCTION

Sarcoidosis is a chronic granulomatous multi-system disease with the target organs being the lungs and lymph nodes. It is a disease of unknown etiology characterized by pathologic hallmark, the non-caseating granulomas [1,2]. This disease is relatively rare in paediatric population.

There are two distinct forms of sarcoidosis in children. Older chidren usually presented with a multisystem disease similar to the adult manifestation with frequent hilar lymphadenopathy and pulmonary infiltration. While, early onset childhood sarcoidosis which usually occurs in less than 4 years old is characterized by triad of arthritis, rash and uveitis.

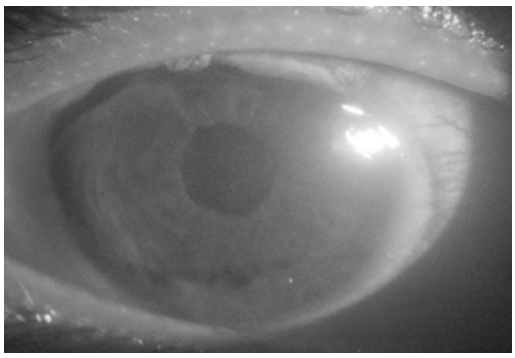

Figure 1 Anterior segment photograph showed a congested eye on presentation.

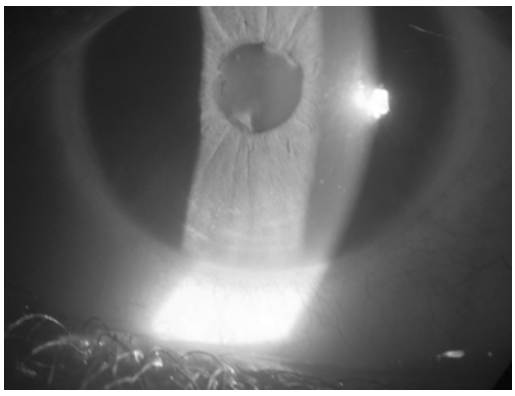

Figure 2 Slit-lamp photograph showed a relatively quiet eye with seclusio pupillae.

Uveitis is the most common ocular manifestation of sarcoidosis with occurrence up to 70% [3]. The ophthalmic involvement is in 25% of patients whereas renal involvement is relatively rare [4]. Meanwhile, ophthalmic and renal involvements are rarely found together [5]. This case illustrates a young Asian teenager with uncommon presentation of ocular and renal sarcoidosis and had been complicated by secondary glaucoma, in the absence of lymph nodes and pulmonary involvement.

CASE REPORT

A 14-year-old Indian teenager consulted a private ophthalmologist with recurrent attack of bilateral red and painful eyes for 2 months. It was associated with reduced vision bilaterally. He denied symptoms of joint swelling or pain, skin or genital lesions and exposure to tuberculosis patients. His past medical and family history were not significant. His presenting visual acuity was 6/6 on both eyes. Both conjunctivas were generally injected (Figure 1). Slit-lamp biomicroscopy releaved minimal flare and occasional cells, with seclusio pupillae bilaterally (Figure 2). The cornea was

clear with no keratic precipitate or iris nodules in both eyes. The intraocular pressure, lens, vitreous and funduscopic examination were normal respectively. He was treated initially with topical corticosteroid and cycloplegia.

His systemic examination was within normal. The lung was clear, with no palpable lymph nodes or visible skin lesions. His blood tests including serum angiotensin converting enzyme, serum calcium, serum creatinine, antinuclear antibody, anti-neutrophil cytoplasmic antibody, and chest radiograph were initially unremarkable.

One month later, he was admitted for severe bilateral ankle and foot pain. It was associated with giddiness, generalized lethargy and anorexia. His blood investigations showed markedly elevated serum urea (20mmol/L), serum creatinine (406mol/L), serum calcium (3.09mmol/L) and globulin (53g/L).

He was then referred to the nephrologist for further assessment. Ultrasonography of his urinary system was normal. A renal biopsy performed subsequently revealed granulomatous interstitial nephritis which was consistent with sarcoidosis.

He showed a remarkable clinical improvement with oral prednisolone 60mg daily and oral methotrexate 5mg daily, without the need of dialysis. The urea and creatinine fell to almost normal level within 3 months. During the tapering dose of oral prednisolone, he developed another low grade attack of bilateral uveitis. The uveitis subsequently resolved with short course of topical corticosteroid.

Six months later, his visual acuity deteriorated to 6/36 in both eyes. The eyes were quiet except for elevated intraocular pressure bilaterally. The gonioscopic findings showed occludable angle due to presence of extensive peripheral anterior synechiae. The intraocular pressure remained persistently high despite maximum topical and systemic antiglaucoma medications. The oral corticosteroid was discontinued with low dose of oral methotrexate.

He underwent bilateral augmented trabeculectomy at the end of the battle. His best-corrected visual acuity remained 6/24 in the right eye and 6/12 in the left eye. The intraocular pressure was within normal with cup to disc ratio of 0.6 bilaterally. His eyes were quiet with faint posterior subcapsular cataract in both eyes. He remained asymptomatic with stable kidney function after 2 years follow-up. The oral methotrexate was finally terminated.

DISCUSSION

Sarcoidosis is uncommon in Asian population especially Southeast Asia region. It is even rarer among Asian children with only a few reported cases [6-8]. Our patient, an Asian teenager presented with extremely rare manisfestation of eye and renal involvement due to childhood sarcoidosis.

Early-onset sarcoidosis with involvement of eyes and joints suggest a guarded prognosis with the likelihood of a chronic progressive course. Progressive ocular disease may produce severe disability with secondary glaucoma resulting in blindness. Our patient was very fortunate to restore satisfactory visual outcome in both eyes at the end of his battle. However, he needs to be monitored closely in future.

Renal involvement is infrequently seen in childhood sarcoidosis [9,10]. Renal disease in sarcoidosis can be attributed to abnormal calcium metabolism in majority of the cases, and granulomatous interstitial infiltration which is extremely rare [9,10]. The reported changes included proteinuria, hematuria, hypertension, membraneous nephropathy, interstitial nephritis and renal failure [11,12].

Our patient presented with renal failure due to presence of hypercalcemia. His renal failure reversed with appropriate treatment and restored to normal status after 3 months of the acute attack. Favourable responses to oral corticosteroids can be expected in both abnormal calcium metabolism and granulomatorus interstitial infiltration provided permanent damage has not taken place yet [13,14].

The first line of treatment for childhood sarcoidosis is corticosteroids with individualized dose and duration [15,16]. Gedalia *et al*[17] reported their experience with low dose of oral methotrexate for 6-months in 7 children with biopsy-proven sarcoidosis. Unlike in adult, the use of other immunosuppressive agents such as azathioprine, cyclophosphamide, chlorambucil and cyclosporine has never been reported in childhood sarcoidosis.

Management of this rare presentation is a great challenge to all the ophthalmologists, paediatricians, and nephrologists. Symptom of bilateral recurrent anterior uveitis with concurrent renal failure should raise the possibility of sarcoidosis in our population, even in the absence of lympadenopathy, skin lesion and normal chest radiograph. It is mandatory to diagnose and manage promptly to avoid serious ocular mobidity especially in young patient.

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亚洲少年眼和肾结节病1例

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摘要
结节病通常导致年轻人患双侧肺门淋巴腺病,肺实质疾病和/或皮肤损伤等。儿童结节病是一种相对罕见的疾病,尤其是眼和肾脏同时受累的儿童患者。我们这里呈现了一个双侧复发性前葡萄膜炎伴发肾功能衰竭的结节病的少年亚裔病情。口服类固醇与甲氨喋呤后临床症状得到明显改善。因顽固性青光眼行小梁切除术后双眼视力令人满意。因此,给予及时的诊断及迅速适当的处理能避免这类罕见疾病的眼部发病率。

关键词:眼;肾结节;亚洲;少年