· Case report ·

# Childhood ocular sarcoidosis: a case report

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## Abstract

• Sarcoidosis is a rare and potentially disabling disease in the paediatric age group. Clinical management of such cases is difficult and requires long-term monitoring. We report a case of a 9 years old Caucasian girl who presented initially with bilateral chronic anterior uveitis, cataracts and glaucoma secondary to sarcoidosis. She was treated with Guttae Levobunolol, topical and systemic steroids over the last 2 years but subsequently required cataract extractions with foldable intraocular lens implants (Alcon Laboratories, Inc. Acrysof® IOL Model: MA60AC). Her uveitis was well controlled with both topical steroids and low dose methotrexate over the last 2 months following cataract surgery. Her full blood counts and renal functions are monitored regularly. There was no adverse effect from the methotrexate reported so far. Sarcoidosis is a multisystem disease and requires multidisciplinary input from ophthalmologists, neurologists and paediatricians. Medical and surgical treatment of such ocular manifestations is challenging. This case highlights both the safety of low dose methotrexate in the management of childhood chronic uveitis and the need for prompt treatment in such cases to avert significant morbidity from this disease.

• KEYWORDS: sarcoidosis; cataract; uveitis
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## CASE REPORT

A 9 years old Caucasian girl presented initially with an acute onset of bilateral red and painful eyes associated with reduced vision. She had no significant respiratory or musculoskeletal symptoms but admitted getting tired on exertion. Her medical and family histories were unremarkable. There was no evidence of any exposure to TB. On examination, her visual acuity was 6/60 on the right and 6/36 on the left which improved with pinhole to 6/12 and 6/9 respectively. She had bilateral incomplete band keratopathy. Slit-lamp biomicroscopy revealed mild flare and odd cell in her anterior chambers with absence of keratic precipitates. She had few posterior synaechiae, but the pupils dilated reasonably well. The intraocular pressures were 20mmHg on both eyes. There were signs of early lenticular haze (Figure 1). Her vitreous showed a few cells on both sides but otherwise the optic discs, retinal vessels and fundi were normal. She was commenced on topical steroid therapy and was investigated further with preliminary blood tests and a chest X-ray. Blood results revealed a raised angiotensin converting enzyme level of 100U/L, C-reactive protein of 12mg/L and corrected calcium of 2.58mmol/L. Chest X-ray revealed focal scoliosis at the level of T4/5 with concavity to the left side. In light of her investigative blood results and radiological findings, both the ophthalmologist and rheumatologist diagnosed bilateral chronic anterior uveitis secondary to childhood ocular sarcoidosis.

She was then started with low dose oral methotrexate (MTX) 10mg and folic acid 5mg weekly. There was mild abdominal upset at the commencement of MTX therapy but it resolved quickly when the dose was reduced to 5mg per week. Her uveitis was well controlled with both topical corticosteroid and MTX. Subsequently, she developed raised intraocular pressure which was treated with guttae levobunolol. Her cataracts had progressed within the last 2 years and required cataract extractions. She was given prophylactic intravenous methylprednisolone and remained on MTX therapy before cataract surgery of her right eye. She underwent routine phacoemulsification cataract surgery and a foldable intraocular lens (Alcon laboratories, Inc. Acrysof® IOL Model: MA60AC) was implanted in the capsular bag (Figure 2). Both the lens aspirate and anterior capsule were sent for histopathology analysis.

Her post-operative visual recovery was uneventful and her best-corrected visual acuity had improved to 6/9. Seven months later, the visual acuity of her right eye remained stable. Microscopy of the anterior capsule revealed abnormal lens epithelial cells exhibiting pleomorphism with microvacuolation of the cytoplasm (Figure 3). Lens aspirate revealed typical cataract degenerative changes characterized by

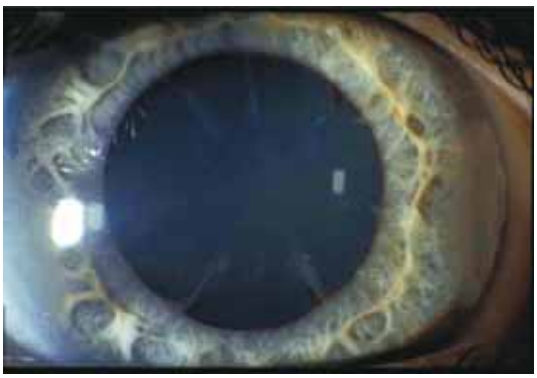

**Figure 1 Early band keratopathy, posterior synaechia and cortical cataract.**

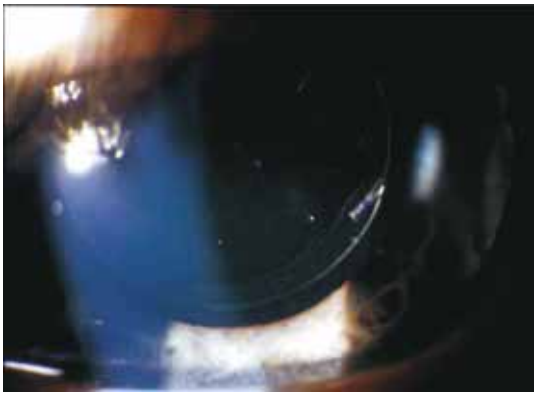

**Figure 2 Three months post cataract showing quiet anterior chambers with clear intraocular lens implant.**

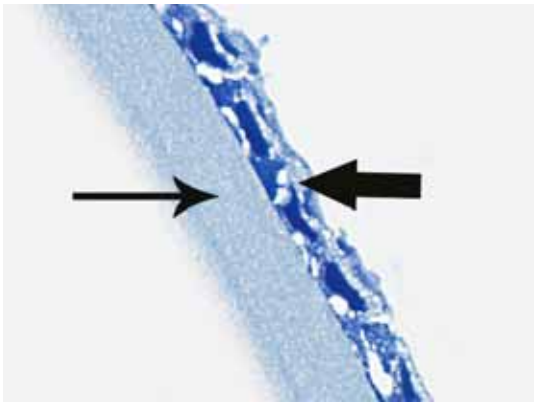

**Figure 3 Anterior lens capsule, lens basement membrane (thin arrow). Attenuated, vacuolated degenerate lens epithelial cells (thick arrow) (toluidine blue).**

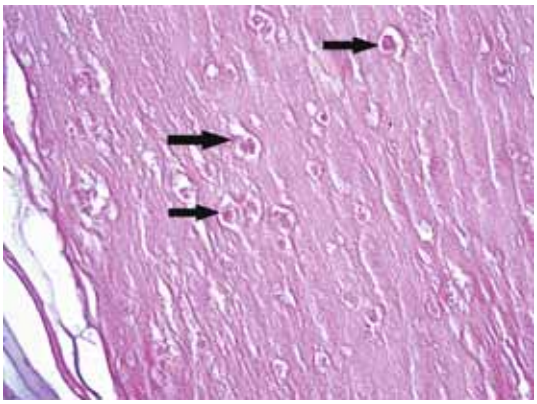

**Figure 4 Lens aspirate showing granular eosinophilic degenerate lens epithelial cells (arrows) amongst viable epithelial fibres (hematoxylin and eosin).**

cell membrane lysis with exudation of granular eosinophilic proteinaceous bodies (Figure 4). Unfortunately, no granuloma formation was identified. She remains treated with both topical steroids and low dose 7.5mg MTX. Her full blood counts, renal and liver functions are monitored regularly. There was no adverse effect from the MTX reported so far. She is awaiting cataract surgery and lens implant on her fellow eye.

## DISCUSSION

Sarcoidosis is a chronic multisystem disease with variable clinical manifestation. In children 5 years and older, the most common clinical findings include lymphadenopathy, pulmonary abnormalities, and uveitis[1]. Neurological involvement from the disease is not uncommon. Therefore, management of childhood sarcoidosis requires input from ophthalmologists, neurologists and paediatricians. Ocular manifestation of childhood sarcoidosis can result in visual impairment from chronic uveitis, band keratopathy, cataracts, persistent macular oedema and secondary glaucoma. Corticosteroids are the mainstay of treatment for chronic anterior uveitis but long-term treatment in any route of administration is associated with adverse ocular and systemic side-effects[2-4]. The combined use of a steroid sparing medication can reduce the dependence on high dose steroids to control ocular inflammation. MTX is a folate analogue that inhibits dihydrofolate reductase which is necessary for DNA synthesis. This antiproliferative effect on rapidly dividing immune cells is the basis of its immunosuppressive properties. Low dose MTX is well tolerated in the paediatric age group. Its use in the treatment of chronic uveitis secondary to juvenile idiopathic arthritis (JIA) and sarcoidosis has been well documented[5-7]. The side-effects of MTX include cytopenia, pneumonitis and hepatotoxicity.

Secondary cataracts in such cases are commonly attributable to chronic uveitis or long-term use of topical corticosteroids. There are several factors to consider in the timing of cataract surgery, which include the risk of amblyopia, quiescence of uveitis and child/parents' decision. The Acrysof® MA60AC acrylic foldable intraocular lens implant was chosen because of its stability in the capsular bag, size of its optic diameter and more inert compared to other intraocular lens implant material. The concurrent use of corticosteroids and low dose MTX in this case for cataract surgery had reduced the risk of severe post-operative uveitis. We found two reported case series with the use of MTX in the treatment of refractory uveitis for ocular sarcoidosis in the paediatric age group (16 years old and below) in English literature using the Medline and EMBASE search. Shetty *et al*[6] assessed the use of oral MTX in 4 patients with uveitis related to both JIA and sarcoidosis not adequately controlled by corticosteroids. All 4 patients tolerated MTX therapy without any adverse effects. Malik *et al*[7] reported the use of a low dose of oral MTX in conjunction with topical corticosteroids to control ocular inflammation in 10 children with idiopathic uveitis and presumed sarcoidosis. All children received folic acid 2.5-5mg per day upon commencement of MTX therapy. Mild

nausea was reported in 2 patients but there was no other adverse events reported to discontinue the use of MTX. None of these previous published case series described that any one of their subjects had undergone uncomplicated cataract surgery. In conclusion, sarcoidosis is a multisystem disease and requires multidisciplinary input from ophthalmologists, neurologists and paediatricians. Medical and surgical treatment of such ocular manifestations is challenging. We recommend the use of an ultraviolet absorbing, biconvex, acrylic intraocular lens with a large haptic (10-11mm) and optic diameter (5.75-6.0mm) for cataract surgery in children. The Acrysof® MA60AC has a complex haptic which can prevent decentration and backward movements of the lens, ensuring very good stability inside capsular bag. Its hydrophobic properties reduce the incidence of both anterior capsular phimosis and posterior capsular opacification[8]. This case report and previous case series also reinforce the safety of low dose MTX in the management of childhood chronic uveitis. Serum electrolytes, full blood count and liver function tests (especially serum alanine or aspartate aminotransferase) should be measured monthly to monitor MTX toxicity. Its concurrent use with corticosteroids can reduce the risk of post-operative cataract surgery complications such as augmented uveitis and persistent macular oedema. It is effective and well tolerated as a corticosteroid sparing agent and should be considered early in the management of such cases to avert significant morbidity from this disease.

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## 儿童眼结节病1例

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**摘要**

结节病是一种罕见的和潜在的致残儿科疾病。此病临床治疗困难,需要长期监测。我们报告了1例9岁的白人女孩的情况,就诊最初表现为双侧慢性前葡萄膜炎、白内障和继发于结节病的青光眼。在过去的2a,患者接受左布诺洛尔滴剂,局部和全身类固醇治疗,随后要求折叠式人工晶状体植入。在白内障手术后的2mo,患者葡萄膜炎以局部类固醇和低剂量甲氨蝶呤一直控制良好。她全血计数和肾功能定期监测,没有甲氨蝶呤不良影响的报告。结节病是一多系统疾病,需要眼科学家,神经学家和儿科医师多学科的投入。内科和外科治疗本病富于挑战。本例强调了低剂量甲氨蝶呤在儿童慢性葡萄膜炎治疗中的安全性和对此病及时治疗以防止显著发病的需要。

**关键词**:结节病;白内障;葡萄膜炎