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Paediatric Sarcoid - an Isolated Ocular Involvement, A Rare Case Report

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ABSTRACT

Childhood ocular sarcoidosis is an uncommon disease and recognition of this disease in children is often delayed because of lack of awareness and familiarity with its clinical features. 13 year old female child presented with redness, pain, photophobia. On clinical evaluation granulomatous uveitis was diagnosed, followed which tuberculosis was ruled out by mantoux and chest radiography. As Serum angiotensin converting enzyme level was grossly elevated, sarcoid was suspected for which she was started on oral steroids. Four months later she turned up with recurrent symptoms with pain on extraocular movements for which she was evaluated with MRI which showed bilateral lacrimal gland homogenous enchancement which was also suggestive of sarcoid with extraocular involvement. Hence she was started on steroids with methotrexate for which she showed remission of symptoms. We describe this case study because usually children with sarcoid presents with clinical triad of skin, joint and ocular involvement which is early onset type and late onset type children will have clinical picture similar to adults with predominant lung involvement, but our case presents with isolated ocular involvement.

KEYWORDS

Paediatric sarcoid, ocular sarcoid, granulomatous uveitis, serum angiotensin converting enzyme.

INTRODUCTION

Sarcoidosis is a systemic granulomatous disease, its characteristic histological finding of involved organs is the non-caseating granuloma. Childhood sarcoidosis is an uncommon disease and recognition of this disease in children is often delayed because of lack of awareness and familiarity with its clinical features. The ocular features of paediatric sarcoidosis have only infrequently described in the ophthalmology literature[1]. We describe this case study because usually children with sarcoid presents with clinical triad of skin, joint and ocular involvement which is early onset type and late onset type children will have clinical picture similar to adults with predominant lung involvement , but our case presents with isolated ocular involvement.

CASE REPORT

13 year old female child presented to us with the complaints of redness, pain and photophobia in left eye for past 6 months. No history of floaters and flashes. No history suggestive of tuberculosis, joint pain, backache, contact with pets, mouth ulcers and trauma. Patient consulted elsewhere and was treated with topical steroids for similar complaints. On examination her visual acuity measured by snellen's chart at 6 meters distance is 6/6. Anterior segment examination revealed mutton fat Keratic precipitates over back of cornea, Anterior chamber showed cells 2+ and flare 1+ with minimal vitreous activity. Fundus evaluation shows unilateral optic disc edema in left eye (fig-1)

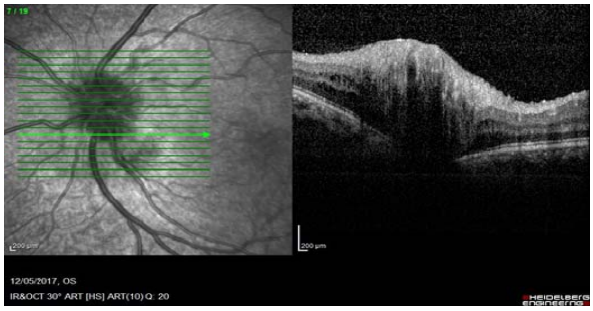

Fig -1 : Optical Coherence Tomography showing disc edema in left eye.

Her intraocular pressure is within normal limits in both eyes. She was investigated for granulomatous uveitis in which Serum angiotensin converting enzyme (ACE) was found to be elevated (105 units). Tuberculosis was ruled out as mantoux is negative with normal lung parenchyma in CT chest. Hence patient was started on oral steroids according to her body weight (prednisolone). Four months later patient presents with pain and redness in both eyes (Right >> left) for past 10 days. On examination her visual acuity measured by snellen's chart at 6 meters distance is 6/6. Anterior segment - Lid edema in both eyes, episcleral venous dilatation with congestion (fig-2) seen in right eye more than left eye.

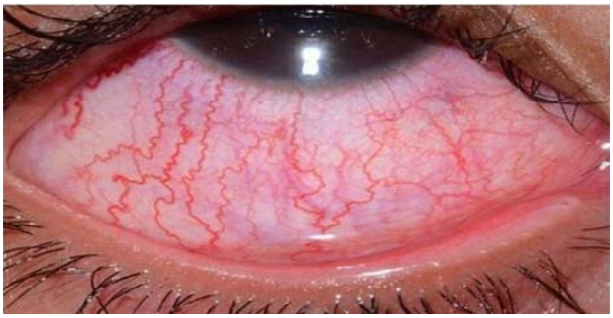

Fig-2 : Showing episcleral venous dilatation with congestion

Anterior chamber cells 2+ present in both eyes, painful eye movement in both eyes. Fundus shows hyperemic disc in both eyes. MRI brain shows bilateral diffusely enlarged and homogenously enhancing lacrimal glands suggestive of extraocular involvement in sarcoidosis (fig-3).

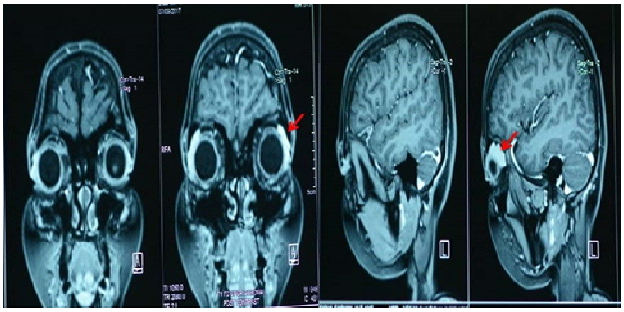

Fig - 3 : MRI brain with contrast shows bilateral diffusely enlarged and homogenously enhancing lacrimal glands.

Hence based on this clinical and radiological picture with elevated serum angiotensin converting enzyme (ACE), diagnosis of ocular sarcoid was made in this child. Patient was started on Intravenous methylprednisolone followed by tapering doses of oral steroids. One month later patient came for review; episcleral venous congestion was presented with bilateral hyperemic discs. So patient was started on immunosuppressive agent methotrexate weekly along with oral steroids for which patient showed improvement clinically (fig-4).

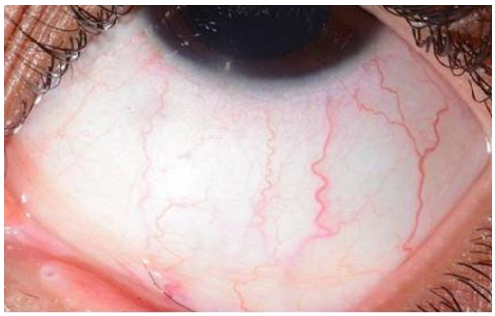

Fig-4 : showing reduced episcleral venous congestion after treatment.

DISCUSSION

Sarcoidosis is a chronic systemic granulomatous disease of uncertain etiology whose manifestations are protean and clinical course variable. The disease usually presents in adults between 20 and 40 years of age[2]. Two distinct forms of sarcoidosis exists in children. Early-onset sarcoidosis is a unique form of the disease characterized by the triad of rash, uveitis, and arthritis in children presenting before four years of age. Older children usually present with a multisystem disease similar to the adult manifestation, with frequent hilar lymphadenopathy and pulmonary infiltrations. The true incidence and prevalence of childhood sarcoidosis is unknown because of the rarity of the disease and the small number of reported cases in childhood. Childhood sarcoidosis is difficult to diagnose because of its variable presentation and ability to mimic other diseases[3].

Clinical manifestations of ocular Sarcoidosis

Anterior uveitis is the most common ocular manifestation of sarcoidosis in both younger and older paediatric age groups. It can present either as chronic granulomatous iridocyclitis with mutton fat keratic precipitates and iris nodules or acute iridocyclitis with fine keratic pericipitates. Conjuctival nodules and cysts have been reported in paediatric sarcoidosis[4] and biopsy of conjuctival specimen is often diagnostic. Scleral and episcleral involvement is rare in adults as well as in children. Lacrimal gland involvement considered to be rare under 15 years of age. If involved there is a significant enlargement of lacrimal gland and typical characteristic findings on CT are diffuse enlargement of gland with homogenous enchancement[5]

MANAGEMENT

In absence of known causative agent, diagnosis of sarcoid remains diagnosis of exclusion. Although non-caeseating granuloma in tissue sample remains gold standard for diagnosis but in children elevated serum Angiotensin converting enzyme is useful in confirming the diagnosis of Sarcoid[6] and is of greater value and as a sensitive indicator for following the course of the disease and the effectiveness of therapy. Radiographic evidence of bilateral hilar lymphadenopathy with or without parenchymal involvement is the hallmark of the disease in older children. The current treatment of choice for childhood sarcoidosis is corticosteroids[7], the patient may follow a course of chronic relapse and steroid dependence. Steroids sparing agents like methotrexate can be administered orally and has a fewer side effects in childhood[8]. Combination therapy with steroids and Methotrexate shows significant clinical improvement in addition to success tapering of oral steroid as in our patient. The prognosis and natural history of sarcoidosis in children are unclear because of rarity of disease but prognosis is better in children as compared to adults with few reported literature.

CONCLUSION

Here, we discussed a rare pediatric case with atypical presentation as isolated ocular involvement in sarcoid and the treatment with favorable clinical response. This case reminds us to include childhood sarcoid in differential diagnosis in paediatric patients presenting with granulomatous uveitis with or without systemic manifestations. Early diagnosis and regular ocular and systemic screening are important to avoid serious complications.

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