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OCULAR PAEDIATRIC SARCOIDOSIS- A RARE CHILDHOOD DISORDER

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Abstract : Sarcoidosis is a multisystem granulamatous disease that can affect almost every organ in the body in people of all racial and ethnic groups. We report a case of 9 year old girl who presented to us with complaints of pain, redness, and decrease in vision in both eyes for past 2 months, along with history of fever and painful knee swelling for past 1 month. History of recurrent fever, skin rash, and difficulty in walking since 5 years of age was present. On examination, the patient was febrile with right knee joint tender swelling, and on ocular examination she had a visual acuity of 624 in both eyes with bilateral circumciliary congestion, band shaped keratopathy, granulamatous keratic precipitates, anterior chamber cells-1 and fundus examination showed retinal granulomas. On investigations, ESR, C reative proteins, angiotensin converting enzymes are elevated, CT Thorax showed bilateral reticulo nodular opacities. With these clinical features and investigation reports, we arrived at the diagnosis of pediatric ocular sarcoidosis. The patient was treated with topical steroids and cycloplegics and immunosupperants was started under rheumatologist guidance. Thus ocular findings helped in the diagnosis of the disorder and to start a prompt treatment for the patient.

Keyword :ocular sarcoidosis, granulamatous disease, children

INTRODUCTION:

Sarcoidosis is a chronic inflammatory disorder with an unknown etiology characterized by noncaseating granulomas. The disorder is multisystemic and affects the lung, lymph node, skin, liver, heart and eye. The disease is rare in pediatric age group and present with varied clinical features in young children.1

CASE REPORT:

A 9 year old girl presented to us with complaints of pain, redness, and decrease in vision in both eyes for past 2 months. History of fever and painful swelling of right knee joint was also present for last 1 month. Patient had a past history of recurrent fever, skin lesions in the leg, and difficulty in walking since 5 yrs of age, for which she was diagnosed with juvenile idiopathic arthritis and treated with oral steroids and non steroidal anti inflammatory drugs. There was no relevant family history with normal antenatal and developmental history. No past history suggestive of primary complex. On examination patient was febrile with stunted growth, right knee joint tender swelling was present (fig 4) and no lymphadenopathy. Her visual acuity in both eyes was 6/24. Biomicroscopy revealed circumciliary congestion, band shaped keratopathy (fig 2), granulamatous keratic precipitates on the corneal endothelium, anterior chamber inflammation with1+ cells, posterior synechiae and peripheral anterior synechiae in both eyes. Fundus examination showed retinal granulomas(fig 1) in both eyes. Intraocular pressure in the right eye was 14 and left eye was 16mm of Hg.

Fig.1: Fundus Photo of RE showing Retinal Granuloma Fundus photo

Fundus photo of LE showing Retinal Granuloma

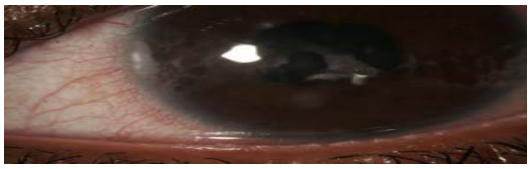

Fig.2: Band shaped Keratopathy

Fig.3: Arthritis

Blood investigations TC, DC,Hb, Urea and creatinine were within normal limits. ESR-40mm/hr (increased),ACE -59.5u/l(increased), Mantoux test was negative. Computed tomography of thorax showed bilateral reticulonodular opacities scattered in both lung fields and no hilar lympadenopathy. With these clinical features and investigation reports, we arrived at the **diagnosis of pediatric ocular sarcoidosis**. Patient was treated with topical steroid in tapering dose, and cycloplegics for ocular symptoms. The patient was referred to rheumatologist for systemic workup and immunosupperants was started under rheumatologist guidance, with T.Mycophenolate mofetil 1500mgm per day in divided doses, Methotrexate 10mg/week with folic acid and oral steroids (methylprednisolone)- 4mg/day. On follow up of the patient, the vision improved to 6/12 with decreased anterior chamber inflammation and fundus showed resolving retinal granuloma.

DISCUSSION

Childhood sarcoidosis appears to exist in two distinct forms. Older children usually present with multisystem disease similar to adults. In contrast early onset sarcoidosis, before 5 years of age, is a unique form characterized by the triad of rash, uveitis, and arthritis, without typical lung disease. The true incidence and prevalence of the childhood sarcoidosis is unknown because of rarity of the disease.2 The ocular presentations in childhood sarcoidosis may be iritis with granulomatous keratic precipitates, iris nodules, choroidal or retinal granuloma, peripheral multifocal choroiditis, conjuctival granuloma.1 Cutaneous manifestations may be erythematous rash, nodules, violaceous plaques, hyper or hypopigmentation and ulcers. Musculosketelal features of sarcoidosis include joint effusions, joint pain, and, rarely, osseous involvement. Multiple joints of both the upper and lower extremities may be involved. Laboratory evaluation may reveal elevated erythrocyte sedimentation rate (ESR) or other acute phase reactants, serum angiotensin-converting enzyme (ACE) is elevated in over 50% of children with late-onset sarcoidosis. The diagnosis of sarcoidosis is confirmed by demonstrating a typical noncaseating epithelioid cell granuloma on a biopsy specimen. 1 In our case, the child characteristically presented with uveitis, arthritis and skin lesion to diagnose as pediatric sarcoidosis. JIA which presents with similar features can be distinguished from sarcoidosis by nongranulamatous inflammation limited primarily to the anterior segment of the eye, absence of systemic symptoms and signs, and presence of positive ANA test result.4. Tuberculosis was excluded in this patient with no history of primary complex, and on examination absence of lymphadenopathy, Mantoux test was negative and CT thorax showed no fibrosis or evidence of tuberculosis. The current therapy of choice for childhood sarcoidosis with multisystem involvement are corticosteroids , immunosuppressive agents, especially low-dose methotrexate (MTX) have been used to treat adult patients with sarcoidosis who have steroid-resistant disease or in those with unacceptable adverse effects from glucocorticoids with good success. Sarcoidosis in very young children with involvement of the eyes, joints and skin have a guarded prognosis with the likelihood of a chronic progressive course.3 Ocular pediatric sarcoidosis is a diagnostic dilemma with a varied presentation in children. Prompt diagnosis and treatment helps to treat ocular and systemic symptoms of the child. Chronic course of the disease needs regular follow up of the patient.

REFERENCES

1. Shetty AK, Gedalia A. Childhood sarcoidosis: A rare but fascinating disorder. Pediatr Rheumatol Online J. 2008;6:16.

2. Drent M, Costabel U. European Respiratory Monograph 32: Sarcoidosis. European Respiratory Society; 2005. P. 357

3. Pediatric Sarcoidosis Treatment & Management. 2013 Aug 14 [cited 2014 Nov 27]; Available from: http:// emedicine.medscape.com/article/1003964-treatment

4. Nussenblatt RB, Whitcup SM. Uveitis: Fundamentals and Clinical Practice. Elsevier Health Sciences; 2010. p 257.

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