CASE REPORT

Childhood Sarcoidosis

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Abstract

A case report of a young child who had presented with erythema nodosum, arthritis, uveitis, segmental lung lesion and was diagnosed as sarcoidosis has been projected, in which the role of fine needle aspiration biopsy has been highlighted.

Key words

Uveitis, Fine needle aspiration biopsy (FNAB), Sarcoidosis

Introduction

Sarcoidosis is a multisystem granulomatous disease of unknown etiology. It is a rare disease among children. Children four years and younger, present with the triad of symptoms; uveitis in 60% to100%, skin nodules or rash in 30% and joint symptoms in 75% (1). Children above four years and adolescents present as in the adults with weight loss, fatigability, uveitis, skin manifestations and enlargement of lymph nodes, lung manifestations and milder joint disease. Here we report a child who presented with cutaneous nodules, arthritis involving large joints, lung and eye manifestations, to the out patient department of Rheumatology.

Case report

A child aged 3 years and 6 months presented with skin lesions over both legs, painful swelling of knee and ankle joints , recurrent respiratory tract infection and with redness of the left eye of six months duration.There was no history of prolonged fever, loss of weight or contact with known case of tuberculosis. On examination there were multiple, tender, erythematous nodules over the anterior aspect of both legs suggestive of erthyema nodosum along with arthritis involving knee and ankle joints. She also had hepatomegaly. Opthalmologist suggested that the left eye had circumcorneal congestion, irregular pupil , sluggish reaction to light, few keratic precipitates on the cornea, Bussaca nodule and posterior synechiae suggesting pan iridocyclitis. Right eye showed cells in the vitreous, confirming posterior uveitis (Fig. 1).

Investigations revealed a total count of 7200/cumm, differential count-P68, L30, E2, erythrocyte sedimentation rate-42 mm/hour, hemoglobin-13.2 gm% and platelets-1.65lacs/cumm. Mantoux was negative. SGOT 40IU, SGPT 39IU, SAP 75IU, bilirubin-0.6mg, creatinine 0.6mg, calcium-10 mg and 24 hours urinary calcium-48mg. The serum angiotensin converting enzyme (ACE) level was135u/L (range 8-52). Rheumatoid factor, anti-nuclear antibody and antistreptolysin O titre were negative. The C-reactive protein was 12 mg/L. Ultrasound of the abdomen revealed hepato-splenomegaly. X-Ray chest showed bilateral hilar lymphadenopathy, with collapse

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consolidation of the middle lobe on the right side. Fiber-optic bronchoscopy done by a pediatric pulmonologist revealed the bronchial tree of the middle lobe blocked by purulent secretions, with the remaining areas being normal. The bronchial lavage showed chronic inflammatory cells. A muscle biopsy was planned but her mother was not willing for an invasive procedure. Fine needle aspiration biopsy (FNAB) of the quadriceps muscle was done and it showed muscle bundles infiltrated with small groups of lymphocytes and histiocytes with lobulated nuclei, with no foci of necrosis in the centre (Fig. 2).

She was started on oral prednisolone 15mg daily and tablet methotrexate 7.5 mg once weekly along with calcium and folic acid supplementation. She gradually improved and is on regular follow up at our rheumatology out patient department.

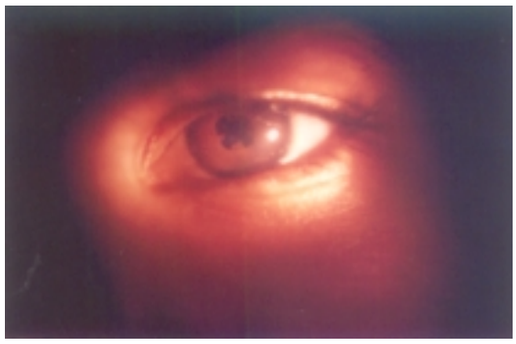

Fig. 1. Left eye showing panuveitis.

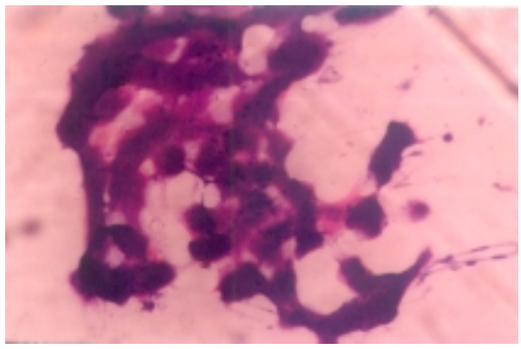

Fig. 1. FNAB of muscle showing histiocytes.

Discussion

Sarcoidosis in children presents differently in various age groups (2). Uveitis, arthritis and skin manifestations are usually seen below 4 yrs of age, but they can also have features which occur in older children, as was seen in this child. In our patient, skin, lungs, joints and eyes were involved clinically and muscle involement was asymptomatic.

Lymphadenopathy is seen in 40-70% and hepato-splenomegaly is seen in 40% of children with sarcoidosis but elevated liver enzymes and portal hypertension are rare among children. Our patient had hepato-splenomegaly with normal level of liver enzymes with no significant lymphadenopathy. In childhood sarcoidosis the bones, kidneys, parotid glands testes and brain can also be involved. Pattishall reported one patient with involvement of eight different organs (3). Sarcoid arthropathy involves the larger joints in 45- 58 % of children, which may mimic Juvenile idiopathic arthritis (JIA) (4).

Bone involvement is rarely detected in the absence of skin lesions and pulmonary disease. Soft tissue swelling and cutaneous lesions of the hands and feet can accompany osseous disease. Hand is the predominant site of skeletal sarcoidosis wherein acro osteosclerosis is a charecteristic finding in the radiograph.

Cutaneous manifestation occurs in 77% of younger children and 24-40 % in older children. Skin manifestations include erythematous papules and lichenoid lesions seen over the extremities and trunk Erythema nodosum presenting with ankle arthritis and uveitis is pathognomic of sarcoidosis, as was seen in this child.

Panuveitis is common but features of anterior uveitis and intermediate uveitis may be the only presenting signs. Anterior uveitis is seen in 58%-90 % in children below 4 years and 24% to 54 % in older children. It may mimic juvenile idiopathic arthritis (JIA) but subtle features like the synechia being focal and firm cellular aggregates differentiate it from JIA. In this case, there was pan uveitis in the left eye and posterior uveitis in the right eye.

Muscle involvement is of the granulomatous type and may be symptomatic or asymptomatic. Muscle is clinically involved in 1.4% of children and in 40 % of adults (5).

FNAB is a useful tool in the diagnosis of sarcoidosis, involving the liver, lungs, muscle, lymph nodes and salivary

glands (6, 7). It is cost effective and less traumatizing. The cytology usually shows epitheliod histiocytes, multi nucleated giant cells, without necrosis, consistent with non caseating granuloma.In our case FNAB revealed similar findings.

In this case, the child was initially diagnosed to have JIA and when she developed segmental lung lesions, tuberculosis was suspected. Subsequent evaluation did not suggest tuberculosis and the child was investigated further at rheumatology care clinic. Later as the child developed uveitis and erythema nodosum and with corraborative evidence of raised ACE levels and FNAB of muscle suggestive of granulomatous disorder, the diagnosis of childhood sarcoidosis was confirmed.

FNAB being minimally invasive can be routinely performed to diagnose sarcoidosis as an alternate investigative tool to muscle biopsy.

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