

Childhood Sarcoid Myopathy Manifesting as Joint Contractures

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Sarcoidosis is a chronic multisystem disease of unknown cause that seldom occurs in children; however cases of pediatric sarcoidosis have been reported in India(1). There has been considerable progress in the understanding of this disease since Kendig(2) published an article in 1962. Most commonly the lungs, lymph nodes, skin, liver and spleen are involved, but sarcoid granulomata have been found in nearly every tissue and organ in the body. Such unusual locations as the pulmonary veins and the superior vena cava were reported by Powell(3) and nerve bundle by Ozer *et al.*(4). Asymptomatic granulomatous infiltration of the striated muscle as part of the generalized sarcoid disease was first described by Myers *et al.*(5). Muscle pain and tenderness may occur in the early stages of sarcoidosis(6) while palpable muscle nodules may present with pain, stiffness or muscle cramps(7). To our knowledge, 79 cases of sarcoid myopathy have been published of which 44% had an isolated

muscular involvement(8). This uniqueness of involvement may well be more apparent than real. We present an autopsy report of a case that demonstrates how the disease evolved in a child not suspected of having sarcoid myopathy.

Case Report

A 1 ½ year-old boy was first seen at the age of 4 months with fever, reduction in the movements of both extremities and swelling of both the knees, elbows and wrists. He became afebrile with treatment, but there was no improvement in the movements of the extremities. Massage of joints and extremities was painful. The mother noticed increasing flexion deformities of the knee, elbow and wrist joints, and thinning of the legs and forearms in the successive months. There was no history of rash over the body or redness of the eyes.

On examination, the child was irritable with fixed flexion deformities of both the elbow, wrist and knee joints. The extremities showed evidence of muscle wasting. The fingers and toes were spindly with loss of all reflexes. Liver was 2 cm palpable below the costal margin. Respiratory and cardiovascular system was within normal limits. Radiological investigations of the skeletal system revealed normal bones and joints with soft tissue swelling around elbow, wrist and knee joints. X-ray chest was unremarkable.

The peripheral blood picture showed microcytic hypochromic anemia and the WBC count was 10,000/cu mm with a differential count of neutrophils - 59%, lymphocytes - 35%, eosinophils - 4% and monocytes - 2%. The erythrocyte sedimentation rate by Westergren's method was 53 mm at the end of the first hour. Serum immunoglobulin levels were: IgG - 2.54 g/dl, IgA - 0.14 g/dl and IgM - 0.258 g/dl. The antinuclear antibody test, anti DNase test,

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rheumatoid factor, VDRL and Mantoux tests were negative. The clinical differential diagnoses entertained were juvenile rheumatoid arthritis, collagen vascular disease and Farber's disease. He was treated with salicylates. On the 27th day of hospitalization, he developed cardiac arrest, from which he could not be resuscitated. At necropsy the heart was enlarged and flabby. All other organs were normal. Microscopic examination revealed non-caseating sarcoid granulomas consisting of epithelioid cells and Langhans and foreign body type of giant cells in the heart, pharyngeal muscles, intercostal muscles, muscles of extremities and diaphragm (Fig. 1). Sarcoid granulomas were also seen in the liver, spleen, lymphnodes, bone marrow, lung, thyroid, parathyroid, kidney, tongue, esophagus, large intestine, meninges, sympathetic ganglion, nerve fibres, testes and pulmonary vein. The diagnosis of sarcoidosis was based on the exclusion of other granulomatous lesions like tuberculosis, fungal infections and syphilis. Special stains for acid fast bacilli, fungal bodies and spirochetes were negative. The characteristic histology of rheumatoid nodule was not seen, hence juvenile



Fig. 1. Section from thigh muscle showing destruction of muscle fibres by non caseating sarcoid granuloma (H & E \times 800).

rheumatoid arthritis was ruled out.

Discussion

The clinical syndrome of sarcoid arthropathy in childhood was first reported by Harris *et al.*(9) and later by North *et al.*(10). The clinical picture for children younger than 4 years usually consists of arthritis, uveitis and rash(11,12). As the disease progresses, the joints tend to be boggy, edematous and associated with minimal pain or limitation of motion. Usually there are no radiological abnormalities even after years of disease.

The pathogenesis of symptomatic muscle sarcoidosis is obscure. It is not known why sarcoid granulomas in muscle produce inflammatory symptoms in some patients, myopathy with weakness and wasting in others, while remaining completely asymptomatic in the great majority[^]. The extent of granuloma formation in the myopathic forms of muscle sarcoidosis can vary greatly and hence the diverse symptomatology(13). Myositis and arthritis were the first manifestations of sarcoidosis in these patients(14).

The average age of the patient with myopathic form of muscle sarcoidosis first seeking medical attention is slightly over 50 years of age(15). Our case seems to be the youngest patient presenting with myopathic symptoms. Weakness and muscle wasting, usually bilaterally symmetrical but more pronounced proximally, are the cardinal signs of myopathic forms of sarcoidosis. The deep tendon reflexes are usually diminished or absent.

In our case, the widespread sarcoidosis of voluntary muscles was responsible for the clinical features of muscle weakness, wasting and contractures. However, death in this case was due to cardiac arrest secondary to sarcoidosis of heart. The other organs

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though showing widespread involvement caused little disturbance, either constitutional or local. This type of generalized sarcoidosis of muscle with symptomless involvement of other organs appears to be a condition distinct from that in which clinically manifest systemic sarcoidosis is associated with concomitant muscle lesions. Childhood sarcoidosis may be confused with juvenile rheumatoid arthritis because of the similarity of eye and articular involvement(16). Nonsteroidal anti-inflammatory drugs and cortico-steroids are effective for most patients. Hydroxy-chloroquine, methotrexate and cyclosporine have been tried with success in patients with refractory sarcoidosis(16).

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