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Myositis Ossificans Progressiva

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Myositis ossificans progressiva is a rare disease characterized by skeletal abnormalities particularly of toes and fingers and ectopic ossification in connective tissues of the muscles, tendons, fascia and aponeurosis. Since the 17th century, 550 cases have been reported in world literature(1), the largest series being reviewed by Lutwak in 1964(2). In all 12 cases have been reported in the Indian Literature(3-5). We report one such case due to its rarity.

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Case Report

An eight-year-old girl was referred with the presenting complaints of swellings over preauricular region and neck for two years and inability to move right upper limb for two months. Over a period of two years, similar swellings appeared in the sternocleidomastoid and upper cervical regions, left scapular, right scapular, occipital, paravertebral and right shoulder regions, associated with limitations of movements of various joints. There was no history of trauma, arthritis or any medication. Her perinatal events and subsequent milestones were normal. No other family members were affected.

General examination revealed a short child with a height of 107 cm and weight of 17.5 kg. Her vital signs were normal. There was clinodactyly, hallux valgus and short great toes, the latter having been noticed by the mother since birth. Stony hard swelling of variable sizes (0.5 to 2.5 cm) were observed in all the areas described above. Skin over the swellings was normal. There were marked restrictions of the movements of neck, flexion of hip and opening of mouth. However, there were no feeding difficulties.

Laboratory investigations revealed a normal hemogram and normal serum levels of calcium, phosphorus, alkaline phosphatase and creatinine phosphokinase. Her skeletal survey showed ectopic ossification in paravertebral, subscapular and neck regions (*Fig.*). Muscle biopsy

revealed cartilaginous and bony cells, proving the suspected diagnosis. This condition has to be differentiated from calcinosis universalis, scleroderma, polymyositis, Ollier's disease and hypervitaminosis D.

Discussion

A classical case of myositis ossificans is being reported. This disorder may be transmitted by a dominant gene or may be due to autosomal dominant mutations exposed to strong selection pressure, *i.e.*, almost all cases are new mutants(6). In more than 90% of cases, the onset is in infancy or childhood and almost always before 20 years(2). Sainaba *et al.* reported that swellings appear in muscles, associated with heat and pain(7). However, there was no history of the latter complaint in our case. Other deformities associated are hallux valgus, short big toes, baldness and deafness(1). Progression of the disease leads to calcified bridges across joints causing scoliosis and limited thoracic expansion. The tongue and smooth muscles are spared. Death occurs as a result of respiratory embarrassment, pneumonia or starvation due to fixation of jaw.

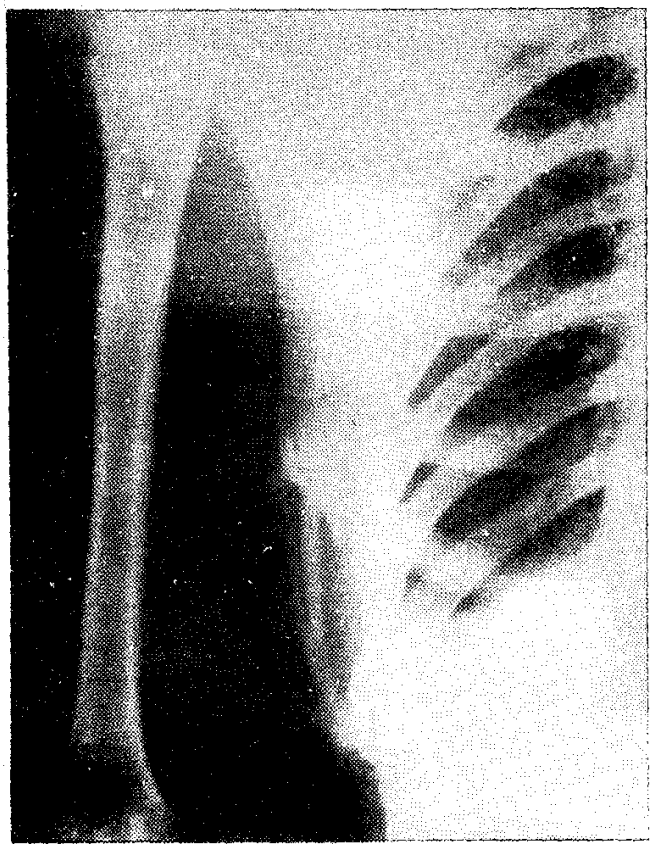

Fig. Radiograph showing ectopic ossification in the subscapular region.

The aim of treatment is to prevent the formation of active bone causing fixation of and progressive immobility of joints and to increase mobility in joints already crippled. The known remissions and exacerbations of the disease make the assessment of treatment a matter of great difficulty.

Treatment with disodium etidronate has been currently investigated. It prevents the formation and dissolution of calcium apatite crystals and retards the mineralisation of ectopic bone matrix after surgical removal(8,9). Complications like hyperphosphatemia, diarrhea and hypersensitivity may occur and monitoring should be done in patients with decreased GFR. Serial determinations of serum alkaline phosphatase and radiographs of wrists and knee should be done on follow-up. Treatment with cortico-steroids have shown extremely variable reports(10). Surgical removal of plaques is only justifiable if their removal will permit movement in a joint which has otherwise become immobile due to disease.

Due to non-availability of disodium etidronate, prednisolone 2 mg/kg was started in the present case and the patient was advised a regular follow-up.

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