Myositis Ossificans Progressive: A Case Report

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Abstract : Myositis ossificans progressiva is a rare autosomal dominant condition of ectopic ossification with primary involvement of the skeletal muscles associated with skeletal abnormalities. We presented a case of myositis ossificans progressiva, probably the first case to be reported in Malaysia.

CASE REPORT

A sixteen year-old Malay boy presented at the age of twelve with a painful swelling of the right scapular region. Over the next two months, multiple swellings developed over the right posterior triangle of neck, right axilla and right lumbar region. The right scapular swelling however was found to be smaller. Lymph nodes enlargement occurred over the right axilla and posterior triangle of neck. Both big toes were noted to be short but the diagnostic significance was not appreciated. Plain radiographs did not reveal ectopic ossification. A bone scan showed an increased uptake of the right of the tenth thoracic vertebra and medial to the mid-shaft of the right femur. The diagnosis was not established initially and the patient was lost to follow-up for two years. Clinical examination when the patient was seen again at the age of fourteen revealed bony swellings over the right paraspinal region from the neck to the lumbosacral region (Fig. 1). There was torticollis and a thoracolumbar scoliosis. The right shoulder was stiff and had 30 drgrees abduction, 20 degrees forward flexionand 20 degrees extension. The left shoulder had 100 degrees abductionand 120 degrees forwardflexion. The left hip had 10 - 110 degrees of flexion, 30 degrees abduction and 15 degrees abduction. The right hip, elbow and knee joints had normal movements. The thumbs had short distal phalanges and the little fingers were incuring. Both big toes were monophalangeal. Plain rediographs of the neck and thoracolumbar region showed ossification of the right paraspinalmuscles from the neck to the lumbosacral region (Fig. 2). The ossification was seen extending to the right scapula and axilla. Radiographsofthe pelivs revealed ossification around the left hip. He was encouraged to excerise regularly to maintain mobility. However when reviewed recently at age sixteen, stiffness of the neck and right shoulder had worsened.

DISCUSSION

Myositis ossificans progressiva starts during childhood and progresses duringthe first two decades of life. As in our patient, Connor and Evans[1] notedthat thesite of onset was usually at the neck or dorsal paraspinal region. Half of their patients had pain when new lumps appeared. Radiological evidence of ossification was usually not evident until four to six months after the appearance of a lump. The lumps caused stiffness which subsided as they shrank, but once bone had been formed, irreversible limitation of movement became evident. The diagnostic significance of the shortened big toes was not appreciated initially. Connor and Evans1 found abnormalities of the big toe in all their patients. This should enable physicians to recognize the disease earlier. The pathogenesis of this disorder is uncertain. Histologically, the pathologic material had features resembling those of periosteal grafts, with all stages of membranous bone formation and a tendency for more mature lesions later in the course[2]. Multiple forms of treatment using steroids, agents binding minerals or blocking calcifications (EDTA and EHDP) and high doses of vitamin A have been unsuccessful[3]. Surgical excision of the bony bridges to relieve the ankylosis and mobilisation of joints have been disappointing ;trauma appears to aggravate the condition[4].

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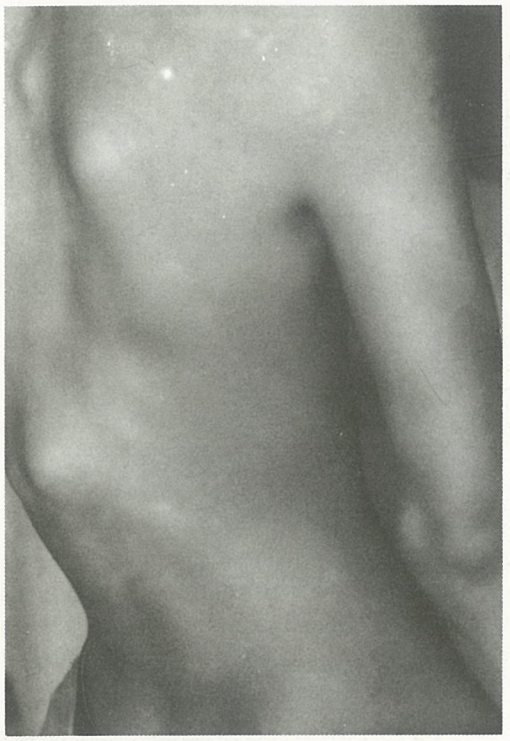

Fig. 1 Paraspinal ossifying lumps.

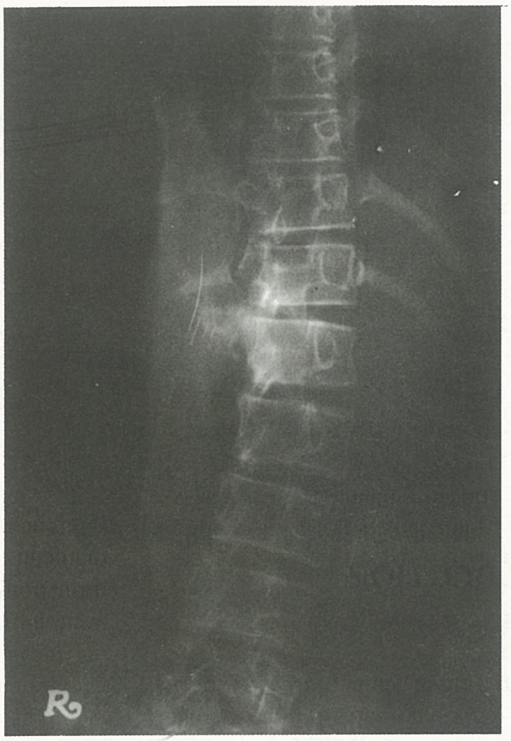

Fig. 2 Thoracolumbar radiograph showing paraspinal bony column.

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