ous hemangioma, CHF and hepatic bruit with diagnosis of hemangioendothelioma with jaundice these were present in 44.4, 55.5 and 16.6% cases, respectively(4). It is further stressed that absence of these clinical features does not rule out the presence of hepatic hemangioendothelioma. The infant reported from India had an enlarged firm nodular liver with single cutaneous hemangioma(6). These infants should be investigated further. Non-invasive techniques like ultrasonography and CT scan of the liver can delineate the tumor(4,6). Selective celiac arteriorgraphy can confirm the diagnosis of hemangioendothelioma(4). Close liver biopsy is contraindicated as the tumor is vascular. Open liver biopsy confirms the diagnosis(6). Ultrasound guided fine needle aspiration cytology can be attempted.

A majority (82.3%) of cases of hepatic hemangioendothelioma associated with hyperbilirubinemia had a fatal outcome(4). The present patient also succumbed. Our earlier experience shows that if diagnosed in life, it can be treated effectively with steriods(6). Associated CHF can be treated with concomitant decongestive therapy(2). Radiation of the tumor and surgical ligation of hepatic artery can facilitate the involution of the tumor(5,6). The prognosis can be improved upon by early diagnosis and by institution of appropriate medical treatment with steroids(6). It is suggested that as a differential diagnosis of neonatal hepatitis, possibility of hepatic hemangioendothelioma should also be thought of.

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Spondylo-Epiphyseal Dysplasia Tarda with Progressive Arthropathy

S. Sood
A.K. Gupta
M. Berry

Spondylo-epiphyseal dysplasia tarda with progressive arthropathy is a rare inherited dysplasia with clinical resemblance

From the Department of Radio-Diagnosis, AIIMS, New Delhi 110 029.

Reprint requests: Dr. A.K. Gupta, Department of Radio-Diagnosis, All India Institute of Medical Sciences, New Delhi 110 029.

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to rheumatoid arthritis and radiographic changes in spine similar to those of spondylo-epiphyseal dysplasia tarda. It was first described in 1982 by Wynne *et al.*(1). To the best of our knowledge no other case has been reported after the initial report of fifteen patients by Wynne *et al.*(1). We are reporting a case with classical findings of the above mentioned dysplasia.

Case Report

A ten-year-old male child, of non-consanguinous marriage was referred to our institution with 5 year history of progressive limitation of movement and swelling of small joints of hands. On examination the patient was a thinly built boy of normal height. He had swelling around the small joints of hands with limitation of movement. There was no redness or tenderness associated with swellings. The child also had scoliosis of lumbar spine and genu recurvatum of the left lower limb. Routine laboratory investigations (hemoglobin, TLC, DLC, ESR) were within normal limits. Serum for rheumatoid factor was negative.

Radiological features: The skull and chest radiographs were normal. Spine showed platyspondyly (*Figs. 1a & b*), throughout with varying degree of end plate irregularity and wedging. Mild scoliosis was present in the lumbar region. Radiographs of hands (*Fig. 2*) showed enlarged epiphysis of metacarpals with widening of metaphysis of all the phalanges. There was evidence of soft tissue contractures at the interphalangeal joints with periarticular osteoporosis. Radiographs of both knees showed evidence of irregularity of the articular surfaces. Radiographs of pelvis showed irregularity of articular surface of the heads of femur and acetabulum.

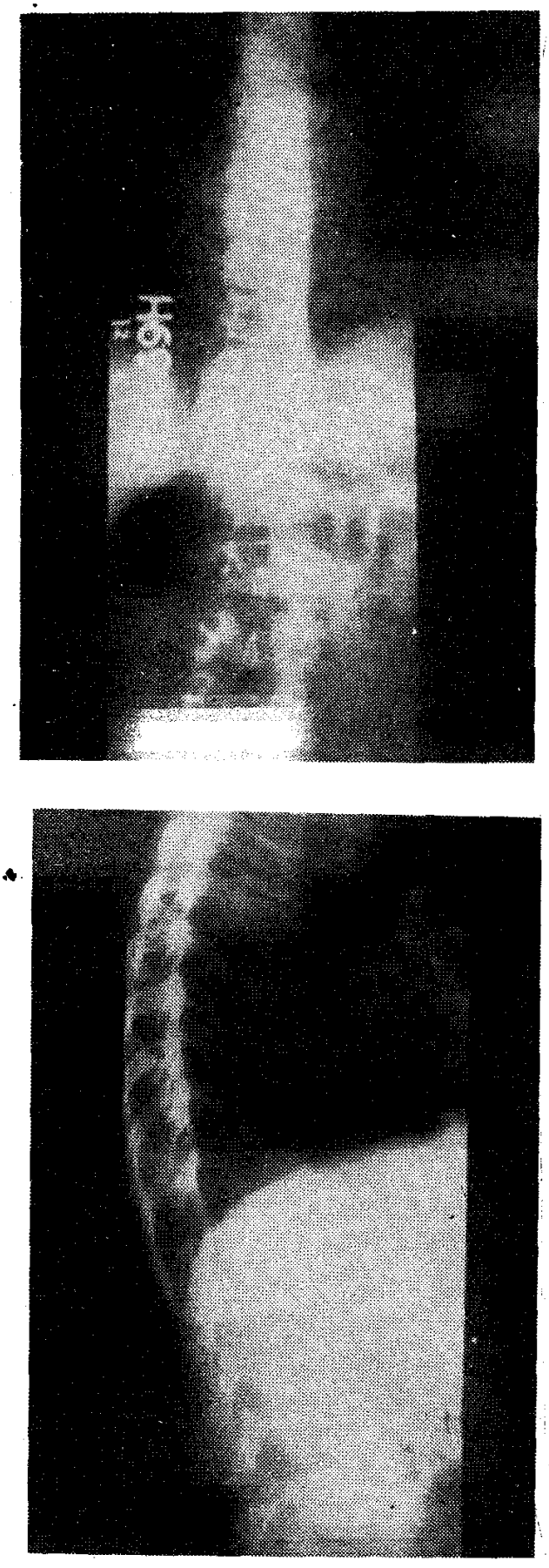

Fig. 1a & b. Radiograph of spine showing platyspondyly throughout with varying degrees of end plate irregularity and wedging.

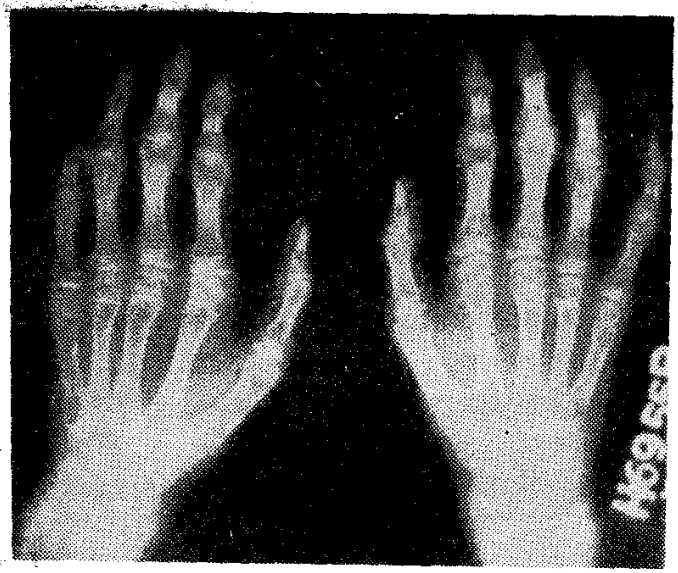

Fig 2. Radiographs of hands showing gross widening of the metaphyses of the phalanges with particular osteoporosis. Epiphyses of the metacarpals are enlarged. Evidence of soft tissue contracture seen at proximal interphalangeal joints. Note absence of soft tissue swelling around the joints.

Discussion

Involvement of hands in this disorder has several features which differentiate it from inflammatory arthritis involving joints, *i.e.*, lack of bony erosions, no periosteal reaction and absent soft tissue swelling around the joints (Note that the enlargement of small joints of hands is due to widening of metaphyses and not due to soft tissue swelling) (*Fig. 2*). Clinically absence of pain and tenderness around the joint is a pointer against the diagnosis of inflammatory arthritis.

This disorder also has several distinguishing features from the more frequently encountered spondylo-epiphyseal dysplasia tarda: symptoms of this disorder begin at an early age (3 to 8 years versus 12 to 13 years in case of spondylo-epiphyseal dysplasia tarda), typical involvement of hands with enlargement of epiphyses and metaphyses, and soft tissue contractures of small joints of hands with particular osteoporosis. The major clinical difference is that the patient of this disorder presents with involvement of hands unlike patients of spondylo-epiphyseal dysplasia tarda who present with shortness of stature.

We believe that the differential diagnosis of spondylo-epiphyseal dysplasia tarda with progressive arthropathy should be suspected in all cases presenting as atypical juvenile arthritis.

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Pattern of Pediatric Malignancies in Rajasthan

N. Mangal
N. Miglani

Various Welfare Programmes for children in our country; aimed at reducing morbidity and mortality due to communicable and nutritional diseases, are bound to

From the Department of Pediatric Medicine, SPMCHI, SMS Medical College, Jaipur.

Reprint requests: Dr. Narendra Mangal, C-145, Dayanand Marg, Tilak Nagar, Jaipur 302 004.

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