1964; 78: 684-93.
7. ROTHOVA A: Ocular involvement in sarcoidosis. *Br J Ophthalmol* 2000; 84: 110-6.
8. BOUCHER RM, GRACE J, JAVA DJ: Sarcoidosis presenting as multiple cranial neuropathies and a parotid mass. *Otolaryngol Head Neck Surg* 1994; 111:652-5.
9. JAMES DG, SHARMAOP: Parotid gland sarcoidosis. *Sarcoidosis Vasc Diffuse Lung Dis* 2000; 17: 27-32.
10. GLOCKER FX, SEIFERT C, LUCKING CH: Facial palsy in Heerfordt's syndrome:Electrophysiological localization of the lesion. *Muscle Nerve* 1999; 22: 1279-82.
11. WOLFE SM, PINALS RS, AELION JA, GOODMAN RE: Myopathy in sarcoidosis:Clinical and pathologic study of four cases and review of the literature. *Semin Arthritis Rheum* 1987; 16: 300-6.

Spondyloepiphyseal dysplasia tarda in a child with severe and an adult with mild clinical features

Sirs,

Spondyloepiphyseal dysplasia (SED) refers to heritable abnormalities in which the primary skeletal disturbance occurs in the spine and in the epiphyses of the long bones. SED may be categorised in 3 broad groups as follows:the congenital type (SED congenita), with abnormalities recognisable at birth; the pseudoachondroplastic type, which presents in infancy and early childhood; and the tarda type, recognised later in childhood (1). Two cases of spondyloepiphyseal dysplasia tarda (SEDT),one a child with severe and one an adult with mild clinical features, are presented here. The radiologic progression of the disease and the differential diagnosis are discussed.

The first case was a 12-year-old boy referred with progressive dorsal kyphosis, joint deformities, leg pain with activity, and gait disturbance. His physical development was normal up to age 8. His parents were first-degree cousins with no family history of a skeletal disease. His posture was hyperlordotic with an increased base width during gait. His dorsal kyphosis, lumbar lordosis and the antero-posterior diameter of the chest were increased. Moderate limitation of motion was present in both shoulders and hips. There were bony deformities in the medial aspects of the elbow joints, knees and ankles. A comparative radiographic examination at age 8 and 12 revealed progression and expansion of the platyspondyly towards the dorsal region with increasing anterior wedging, irregularities, subchondral sclerosis and Schmorl's nodes at the end-plates of the T8-11 vertebrae. The humeral and femoral heads were flattened and irregularities in the proximal epiphyses were noted.

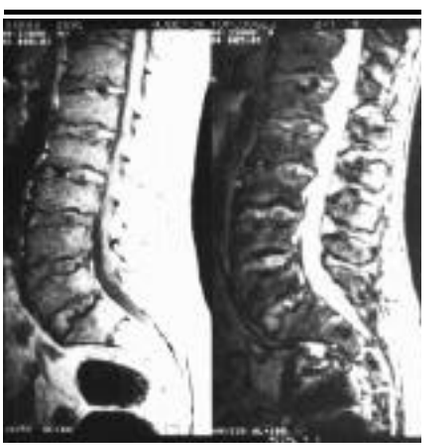

Fig. 1. T1 and T2-weighted sagittal magnetic resonance images of the lumbosacral spine in case 2, demonstrating platyspondyly, Schmorl's nodes and irregularity at the end-plates of the lumbar vertebrae with narrowing of the intervertebral disc spaces, emphasizing the findings in the plain radiographs.

The second case was a 29-year-old male who played soccer as a recreational activity, who was admitted with the complaint of pain in the right hip which became evident on physical activity. On examination he had a relatively short trunk with mild scoliosis and increased dorsal kyphosis. Paravertebral muscle spasm and pain in motion of the lumbar spine were observed, although the spinal mobility was not limited. The range of motion in the hips was limited and painful. The radiographic findings resembled those of the first case with increased degenerative changes in the spine (Fig. 1) and hips.

Spondyloepiphyseal dysplasia tarda, first described by Nilsonne in 1927 (2,3), is a developmental skeletal disorder that usually presents clinically late in the first or early in the second decade of life with progressive involvement of the spine and epiphyses. The diagnosis of SEDT is difficult in a sporadic case without a positive family history, but when suspected clinically radiographic findings are usually sufficient for the diagnosis. Hyperostotic new bone formation on the posterior two-thirds of the articular surfaces of the vertebral bodies is usually distinctive for the X-linked type (3, 4). Degenerative spine and hip disease characterize the later stages of the disorder (3). Dysplastic changes may be observed in other major joints (1,3).

Scheuermann's disease, juvenile lumbar osteochondrosis and spinal manifestations of the mucopolysaccharidoses resemble the spinal findings of SEDT (5). A subtype of pauciarticular juvenile chronic arthritis (JCA) with late onset may also affect the hips (6). JCA and ankylosing spondylitis might have been considered in the differential diagnosis of our cases. The results of the extensive laboratory workup were within normal ranges for the two cases.

We believe that case 1 represents a case of SEDT with severe and progressive features, beginning at an early age. The second case was a fairly mild one clinically. There was no back pain despite the striking radiographic findings. The late clinical presentation and the delay in diagnosis (at the age of 28),despite his physically active lifestyle seems unusual.

We would like to emphasize that SEDT should be considered in the differential diagnosis in patients who have complaints of back and hip pain and gait disturbances even in adulthood. After the diagnosis of SEDT, patients should be advised to engage in appropriate exercises and moderate activity and should be informed about the prognosis and genetic aspects. Overtreatment should be avoided.

A.O. SAHIN, *MD*
N. BÖLÜKBASI, *MD*
M. BEYAZOVA, *MD*

Gazi University Medical School, Dept. of Physical Medicine and Rehabilitation, Ankara, Turkey.

Address correspondence to: Dr. A. Oya Sahin, Marmara sokak No. 40/3, Yenisehir, Ankara 06420, Turkey. E-mail: osahin@tr.net

References

1. PRITZKER HL: Congenital dwarfism. *In:* MURRAY RO and JACOBSONHG (Eds.): *The Radiology of Skeletal Disorders.* Edinburg, Churchill Livingstone, 1977: 172-9.
2. WELLS III JA,ELLERINE NP, FERNHOFF PM, WARING III GO: Corneal opacities in spondyloepiphyseal dysplasia tarda. *Cornea* 1994; 13: 280-3.
3. WHYTE MP, GOTTESMAN GS, EDDY MC, MCALISTER WH: X-linked recessive spondyloepiphyseal dysplasia tarda. Clinical and radiographic evolution in a 6-generation kindred and review of the literature. *Medicine* 1999; 78: 9-25.
4. SCHANTZ K, ANDERSEN JR PE, JUSTESEN P: Spondyloepiphyseal dysplasia tarda. Report of a family with autosomal dominant transmission. *Acta Orthop Scand* 1988; 59: 716-9.
5. RESNICK D: Osteochondroses. *In:* RESNICK D (Ed.): *Diagnosis of Bone and Joint Disorders.* Philadelphia, W.B. Saunders Company 1995: 3595-601.
6. WHITE PH: Juvenile chronic arthritis. Clinical features. *In:* KLIPPEL JH and DIEPPE PA (Eds.): *Rheumatology*, vol. 2, 2nd ed., Mosby, London 1998, 5.18.1-10.

Pachydermodactily as a cause of painful swelling of the knuckles: Successful treatment with intralesional steroids

Sirs,

Pachydermodactily is a fibrosing condition characterized by symmetrical, diffuse swelling of the proximal interphalangeal (PIP) joints of the fingers (1-3). The lesions are usually painless, but may cause pain and stiffness of the PIP joints, which has in