**Case Report**

**Melorheostosis: Report of a New Case with Linear Scleroderma**

M. Birtane, M. Eryavuz, H. Ünalan and F. Tüzün
Department of Physical Medicine and Rehabilitation, Cerrahpaşa Faculty of Medicine, Istanbul University, Istanbul, Turkey

**Abstract:** Melorheostosis is a very rare bone disease of unknown etiology characterised by linear hyperostosis and associated with fibrosis of soft tissues and the skin. This uncommon sclerosing bone dysplasia was first described by Leri and Joanny in 1922, and since then, until 1993, approximately 300 cases were reported in the literature. Linear scleroderma is a localised proliferation of connective tissue and has rarely been associated with melorheostosis.

In this paper, we present a new case of melorheostosis with linear scleroderma which, to the best of our knowledge, is the first case reported in Turkey.

**Keywords:** Linear scleroderma; Melorheostosis

**Introduction**

Melorheostosis is an uncommon bone disorder that shows highly characteristic radiographic alterations. The disease generally becomes manifest during childhood and affects females and males equally. The aetiology and pathogenesis are unknown; no hereditary features have been established and the treatment is empirical [1,2].

In the early stages, the patient may present with swelling of the joints and pain. The disease tends to be slowly progressive, subsequently, muscle atrophy and joint contractures may become evident. Changes commonly involve one limb and the lower limbs are involved more frequently than the upper limbs [1,2]. Ippolito et al. [3] suggested that 300 cases have been reported in the literature since 1993.

Melorheostosis is also associated with other abnormalities of mesodermal origin, one of which is linear scleroderma. It was suggested that linear scleroderma and melorheostosis are manifestations of a congenital disorder of mesodermal development. Linear scleroderma has very rarely been associated with melorheostosis [1,4].

In this paper, we present a new case of melorheostosis with two uncommon features: co-existence of linear scleroderma and bilateral involvement of the lower extremities.

**Case Report**

A 48-year-old woman was admitted to our hospital with the complaints of pain and stiffness in both hips, the left knee and the ankle. She also had difficulty in walking. A history revealed that the symptoms started as pain in both feet when she was 9 years old. At the age of 22 years she had swelling, pain and limitation of movement in both hips, knees and ankles for 3 months. She reported that she recovered after aspirin and steroid treatment and began to walk comfortably again. After a few months she again developed pain in her left hip, left knee and ankles, worsening during standing and walking. After this attack she felt that her left leg was shorter than the other. The disease process had reportedly progressed since then and 3 months before admission she had another attack of swelling, erythema, pain and stiffness in her left knee and ankle. After a period of irregular analgesic treatment she was admitted to our hospital.

Inspection revealed an antalgic and waddling gait. Erythema was observed on the left knee and ankle, both of which were also swollen and quite tender to palpation. Physical examination revealed limited internal rotation of the left hip and hypertrophy of the left knee. There
Fig. 1. Prominent hyperostotic involvement in the right acetabulum, left ramus pubis and the femoral head.

Fig. 2. Characteristic ‘wax flowing down the side of a candle’ appearance involving the right tibia and left fibula.

Fig. 3. Sclerosis in the ankle and foot bones.

Examination of the leg revealed skin thickening, which was evaluated as linear scleroderma after rheumatological referral.

Routine laboratory studies including complete blood count, erythrocyte sedimentation rate, fasting plasma glucose, urea, uric acid, creatinine, aspartate aminotransferase, alanine aminotransferase, alkaline phosphatase, serum calcium and phosphorus were all within normal limits. C-reactive protein, rheumatoid factor and anti-DNA were all negative.

Radiographs revealed characteristic sclerotic bands resembling wax flowing down the side of a candle, more prominent in both knees, but also present in the pelvis and ankles. These characteristic views were evident in both limbs, beginning from the pelvis and extending down to both ankles (Figs 1–3).

A diagnosis of melorheostosis was made based on these characteristic radiological findings.

Discussion

Melorheostosis is a rare sclerosing bone dysplasia, which was originally described by Leri and Joanny in 1922 [1,5]. In a report published in 1993, the number of cases in the literature was reported to be only 300 [3].