

Correspondence

Melorheostosis

Melorheostosis or Leri disease is a genetic disease, caused by a loss-of-function mutation in LEMD3 gene(1). It frequently involves a single limb characterized by hyperostotic linear bone densities(2). We describe here a girl with a rare generalized form of Melorheostosis.

A 9-year-old girl born to non consanguineous parents presented with progressive painful deformities of both elbows and right knee. She had short stature and grade III PEM. Fixed flexion deformities of bilateral hip, elbows, ankles and right knee were noted. Her left lower limb was shorter than the right. Pectus excavatum and

lumbar lordosis were present. A simian crease was present in the right hand and both palms showed wasting of small muscles. All the fingers showed evidence of early contracture and the second and third fingers were thickened bilaterally. There was over riding of toes in her left foot. The skin was shiny but no fibrosis was noted. The tone and deep tendon reflexes were normal. A skeletal survey revealed generalized endosteal sclerosis which appeared as symmetric streakiness of long bones and patchy hyperostosis of the pelvis, scapula and small bones of the hands and feet (*Fig.1*). She had symptomatic relief of pain with Ibuprofen and is on follow up.

Soft-tissue contractures causing severe and rigid joint deformities, inequality of limb length,



Fig. 1. Clinical picture showing lumbar lordosis and joint deformities and X-ray leg showing endosteal sclerosis of long bones.

woody thickening of the skin with tethering of the underlying fascia are commonly noted in Melorheostosis. Growth disorders of the limbs are often the first signs in the affected children(3). It is usually associated with pain in adults, but not in children. In contrast to the typical extra-osseous sclerosis flowing like molten wax in adults, endosteal sclerosis is noted in children(2). The clinico-radiological features in this girl were consistent with Melorheostosis and the polyostotic involvement with symmetric radiological findings makes this case interesting. The simian crease as noted in this child has not been reported with Melorheostosis but could be just coincidental. The differential diagnoses include Scleroderma, Post Poliomyelitis paralysis, Rheumatic fever, Enchondromatosis, Diaphyseal dysplasia, Osteopathia striata, Osteopoikilosis and osteopetrosis. It may co-exist with osteopoikilosis, osteopathia striata and malformations of blood vessels or lymphatics(4). Bone scintigraphy is invariably positive in Melorheostosis revealing increased uptake of tracer predominantly in the cortex. Surgical procedures such as tendon lengthening, excision of fibrous and osseous tissue, fasciotomy, capsulotomy and amputation of severely affected and painful limbs have been described. However, recurrences are common(5).

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Tips to Treat Asthma

I read with interest the article titled "Adherence issues in Asthma" published in the December issue of Indian Pediatrics and would like to share our experiences over the past one and half decade at our Asthma clinic at Bangalore.

1. The successful adherence to inhaled medications by the parents and the child will mainly depend upon the time spent by the physician during the first visit to explain, convince and support them.

2. The concept of Blue (reliever) and Brown (controller) and their differing yet contributory effects on Asthma control needs to be explained clearly during the first visit itself. Otherwise, as the Blue inhalers are less expensive and the Brown inhalers relatively 4 times the cost, and the effect of the Blue inhalations being evident within fifteen minutes in contrast to the brown inhalers which take weeks to perceive their effects, most often the Blue inhalers replace the Brown ones very shortly after initiation.
3. At each followup visit, the physician should always check, reinforce the technique of