Melorheostosis of Ring-finger: A case report.

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Citation

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Abstract

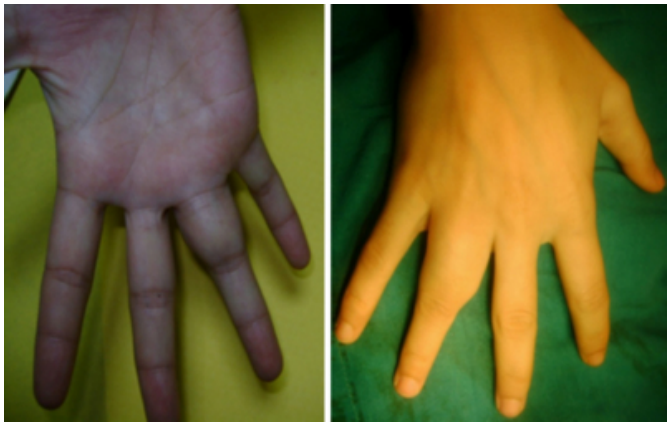
Melorheostosis, a rare mesenchymal dysplasia, is one of the sclerosing bone disorders. It is a developmental error, involving frequently one limb. It may be asymptomatic or symptoms like pain, stiffness, limitation of movement, leg length discrepancy, and limb deformity may occur. On plain radiograph, melorheostosis shows cortical hyperostosis resembling flowing candle wax. It occurs due to loss-of –function mutation in LEMD3 gene. We report a case of melorheostosis in a 16 year old girl presenting with a 4 year history of progressively increasing swelling along with intermittent pain in the ring-finger. Radiograph and computed tomography show findings characteristic of melorheostosis. An excisional biopsy along with bone grafting was done and patient is doing well after physiotherapy.

CASE REPORT

A 16 year old female with average socio-economic status presented to the out-patient department with a 4 year history of progressively increasing swelling and intermittent dull ache in the ring finger of the right hand. She had no similar swelling in any other part of her body. There was no history of paraesthesia or restriction of movement. There were no similar complaints in the family. On examination swelling was 3x2.5 cm. in size, firm to hard in consistency with no restriction of movements.(fig.1)

Figure 1

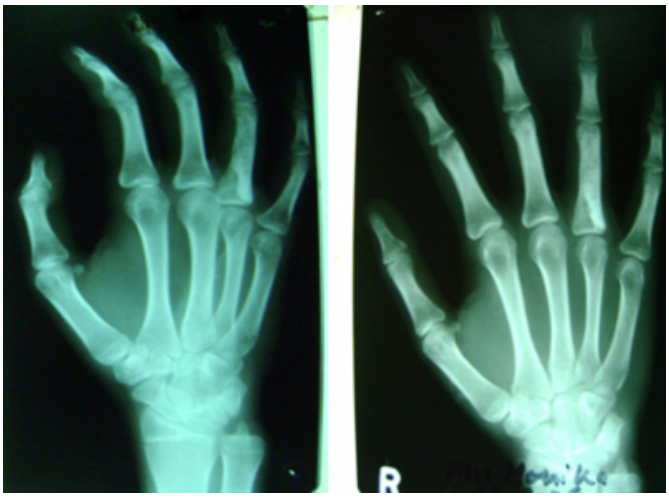
Photographs showing the swelling in the ring finger. Figure 1.



Antero-posterior and lateral radiograph show dense bone formation involving more than half of the proximal phalanx, with a flowing, slightly expansile margin along its ulnar aspect.(fig. 2)

Figure 2

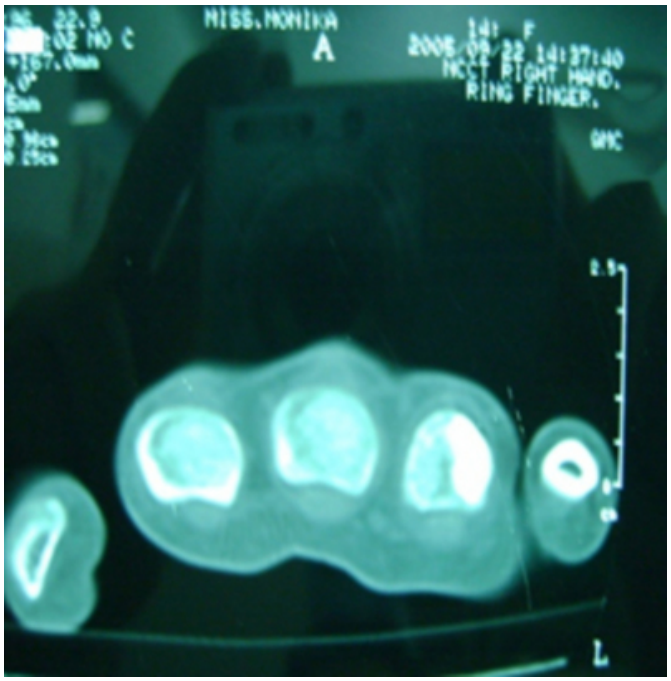
A.P. and lateral Radiographs showing candle wax appearance in the proximal phalanx of ring finger . Figure 2.



Computed tomography of hand shows findings identical to those on shown on radiograph i.e. cortical hyperostosis with thickening resembling candle wax.(fig 3)

Figure 3

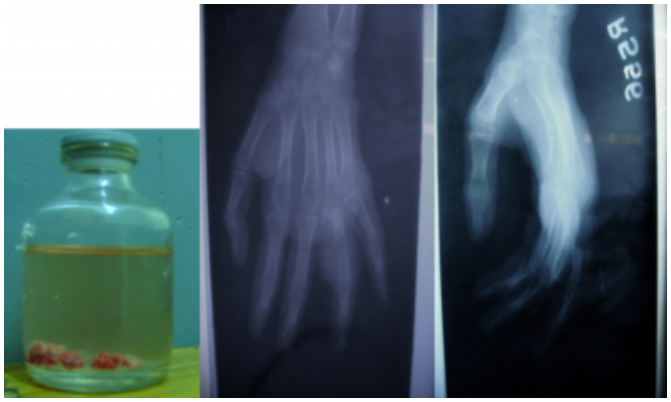
C.T. showing saggital section of hand figure 3.



Soft tissue shadows were unremarkable. The patient was admitted and all routine investigations were within normal limits like serum calcium and serum phosphorus. Patient was taken-up for biopsy and excisional biopsy done along with bone-grafting. Dense bone fragments were removed from the interior of the proximal phalanx.(fig4 and 5).

Figure 4

Biopsy jar with biopsy specimen alongwith post-operative A.P and Lat. Radiograph. figure 4.



Histological examination showed fragments of dense bone with fibrotic marrow consistent with melorheostosis. Patient is doing well after surgery.

DISCUSSION

Melorheostosis was first described by Leri and Joanny in 1922(1). This name comes from Greek Melos, meaning limb and rhein, meaning 'to flow'. The radiologic appearance has been likened to "melting candle wax"(2). Synonym for melorheostosis include Leri's disease and Flowing periosteal hyperostosis.

Untill very recently, the etiology of melorheostosis was unknown, but have recently been described as due to loss-of-function mutation in LEMD3 gene (also known as MAN1) which encodes for an integral protein of the inner nuclear membrane. The developmental error is at the site of both intermembranous and enchondral bone formation, predominantly the former. Its distribution often suggests a sclerotomal abnormality involving one or more sclerotomes (4).

Males and females are equally affected (2). Melorheostosis is a rare disorder, with a prevalence of about 0.9 per million (5). It is usually first diagnosed as an incidental finding on plain radiographs. Approximately half of diagnosed cases are found in patients 20 yrs of age or younger (2, 6).

Melorheostosis typically affects long bones of the limbs and sometimes small bones of the hand and feet. Extremely rarely, melorheostosis involves vertebral column (6,7). The characteristic hyperostosis of melorheostosis is distributed longitudinally along one or more sides of a single bone or set of contiguous bones (7). Plain radiographs of melorheostosis usually demonstrate hyperostosis of either the ossues cortex, the medullary space or both (8). C.T. and M.R. imaging can also be used to demonstrate the osseus and extraosseus findings of melorheostosis (8, 13). If radionuclide imaging is performed, it typically demonstrates multiple areas of increased uptake, corresponding to the areas of sclerosis noted on radiographs (11, 12). The most common complaints include dull pain, stiffness swelling and paraesthesias over the involved bones (14). While some patients are asymptomatic, others demonstrate severe deformities and pain (9). Other associated abnormalities have been reported including neurofibromatosis, arteriovenous fistula, cutaneous haemangioma, linear scleroderma and aneurysms (6, 15).

Treatment may include surgery for tendon lengthening and correction of deformities. In extremely rare cases, amputation is indicated in very painful limbs with contractures and ischemia (13). Most cases are benign and do not require operative intervention (14).

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