Melorheostosis of the Radius Bone: An Incidental Finding

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Abstract: We report a case of Melorheostosis of the radius in a middle-aged woman, who presented with pain over her forearm and was incidentally diagnosed based on the radiological findings. CT scan and MRI gave further information about the extent of the disease. Patient was treated conservatively with brace application, analgesics and injection zoledronic acid which led to early symptomatic improvement.

Key Words: Melorheostosis, Leri disease, Radius Bone.

Introduction:
Melorheostosis (Leri disease) is a benign sclerosing bone disease which is very rare and typically affects cortical bone along with soft tissue structures surrounding it. The name is derived from the Greek word "melos" means limb, "rhein" means to flow and "ostos" means bone. The long bones of the lower extremities are more commonly involved although this disease can affect any bone (1,2). The etiopathogenesis of Melorheostosis is not very clear and also there are variations in the clinical features (3). This disease can affect any age group, most commonly between 2-64 years and both sexes are equally affected. (4,5). The aim of this case report is to provide additional points to the literature about the clinical manifestation, imaging features and management of Melorheostosis in the radius.

Case Report
We report a case of 42-year-old woman of Andaman and Nicobar Islands, a housemaker who presented with dull aching pain over her left forearm of 4 months duration. Pain was present throughout the day which was more on exertional activities like lifting heavy objects and it relieved with analgesics. She did not give significant past clinical and family history. There was no history of trauma, fever and pain over other parts of the body. On examination, the skin over the forearm was soft, there was no swelling or colour change. Wrist and elbow movements including supination-pronation were normal. The movements were painless and there were no distal neurovascular deficits. Laboratory investigations included complete blood count, C-reactive protein, erythrocyte sedimentation rate, serum calcium, phosphorus, alkaline phosphatase (ALP) and rheumatoid factor, which were normal. The radiograph of the forearm showed hyperostosis of the radius bone which involved almost 75% of the bone extending from the distal articular margin to proximal third of the radius (Fig 1). The ulna and other bones were not involved. The dense hyperostosis of the radius resembled dripping candle wax down one side of the burning candle which is the characteristic feature of Melorheostosis on X-ray.

Fig 1: X-ray of the forearm showing characteristic sign of Melorheostosis.
On computed tomography (Fig 2), diffuse sclerotic cortical thickening of the radius was seen with narrowing of the medullary canal. The wrist and elbow joint spaces were normal and there was no evidence of pathological fracture. The consent was taken from the patient to do MRI for advanced imaging (Fig 3) and it revealed low signal intensity areas involving only the radius. There was minimal edema around the lesion without involvement of the surrounding soft tissue structures and adjacent joints.

The patient was prescribed wrist and forearm splint along with analgesics. Also, the patient was administered intravenous pantoprazole IV stat dose. The patient was advised for wrist and forearm brace along with analgesics accordingly as per her symptoms and presenting signs. She was advised for a regular follow up and there was significant improvement in her symptoms after 3 months.

Discussion

The most significant feature of Melorheostosis is sclerosis, dysplasia and hypertrophy occurring on one side of the long bone cortex akin to a ‘dripping candle wax’ (4). In present case scenario, it was incidentally diagnosed in the radius of a 42 years old lady based on the clinical and radiological signs mentioned in the literature. The long tubular bones of the lower limb are more commonly affected and it can also affect bones of hand and feet but rarely seen in axial skeleton (6,7). It can affect single or multiple bones but bilateral involvement is rare (8,9). We report a case of Melorheostosis of the radius which is being reported for the first time in the literature.

Many theories have been proposed about the origin of the Melorheostosis (10-15). Helleman et al suggested that the disease is due to the mutation in LEMD3 on 12q chromosome (11). This disease has also been associated with other sclerosing dysplasia like Osteopikliosis and Buschke-Ollendroff syndrome (12). One of the causes suggested was mosaicism as the basis for the occurrence of the dysplasia (13).

The segmental sensory lesion occurring during embryogenesis in the neural crest has also been hypothesized (14). Melorheostosis is sometimes associated with severe pain, swelling, bone deformity and joint stiffness. When there is surrounding soft tissue involvement, a scleroderma like features of the skin with edema, hypertrichosis, fibrosis and fibrolipoma has also been reported (15). Vascular anomalies (16), nerve compression syndromes like carpal tunnel syndrome and spinal nerve root compression (17) and serious defect leading to amputation has also been reported (15). The commonest symptoms reported are pain, deformity, limited range of movements at the joints, numbness and weakness (18). The lady presented to us with the only complaints of dull aching pain over her left forearm of four months duration which may be attributed to the early diagnosis of the condition. The radiological patterns of Melorheostosis may vary. The Xray features may resemble myositis ossificans, osteoma or mixed pattern (4). In a case series of 23 patients, only 5 patients had characteristic radiological signs (16). However, a classical sign of ‘dripping candle wax pattern’ on radiograph will easily distinguish it from the other conditions (17). CT scan and MRI are usually warranted in complex cases. CT scan will show the extent of undulating high density hyperplastic areas of the cortex similar to X ray. MRI will show invasion into the marrow and also the condition of the soft tissue surrounding the bone will be ascertained (19).

Melorheostosis have been treated mainly conservatively with NSAIDs, bisphosphonates, brace application, serial casting, physiotherapy, nerve blocks and sympathectomies (20, 21). Surgical treatment has been reserved for severe cases which include capulotomy, fasciotomy, tendon lengthening, bone resection although recurrences after surgical treatment are common (22). In the present case, we treated the patient accordingly as per her symptoms and presenting signs. She was advised for a regular follow up.

Conclusion

Melorheostosis is diagnosed incidentally in most of the cases. The characteristic sign on the radiograph will clinch the diagnosis. CT scan and MRI can be advised in complex cases. Treatment is mostly conservative. Injection zoledronate is much beneficial for immediate symptomatic control. Surgical methods are applied for advanced cases.

Acknowledgments

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References
