# Melorheostosis - Scintigraphic And Histopathological Features: A Case Report

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**Citation**

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**Abstract**

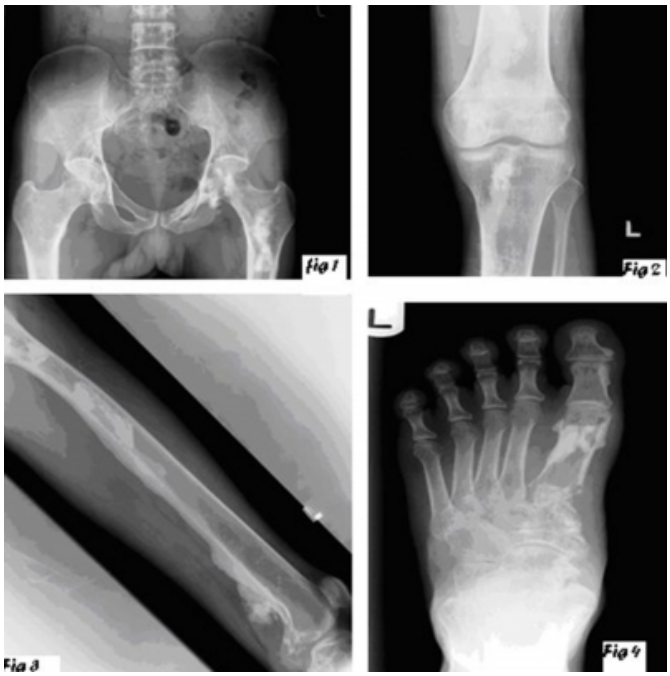
Melorheostosis is a rare bone disorder of unknown etiology with linear hyperostosis. We describe the scintigraphic, radiologic and histopathological findings in a 40 year old gentleman who presented with pain and swelling in the left knee. Scintigraphic appearance is characteristic, with asymmetric cortical activity that may cross joints to involve contiguous bones. Bone scintigraphy eliminates the need for multiple radiological images. Histology shows marked endosteal sclerosis with marrow fibrosis consistent with melorheostosis

## CASE REPORT

A 40-year-old gentleman was evaluated for pain and swelling in the left knee with difficulty in walking. Plain radiographs of pelvis and left lower limb showed peripherally located hyperostosis with ‘flowing candle wax dripping ‘ appearance in left hip and bones of left lower limb, suggestive of melorheostosis (Fig. 1-4).

**Figure 1**

Figure 1-4: Plain radiographs of pelvis and left lower limb showing peripherally located hyperostosis with ‘flowing candle wax dripping ‘ appearance - features suggestive of melorheostosis



Bone scintigraphy with Tc99m methylene diphosphonate (MDP) revealed abnormal increase in radionuclide uptake and asymmetric cortical activity along the cortex of entire length of left femur, left tibia, left tarsal and metatarsal bones. The activity was found to be crossing the left hip and knee joints (Fig. 5). Histopathological examination of the lesion from femur showed marked endosteal sclerosis with

marrow fibrosis consistent with melorheostosis (Fig. 6).

**Figure 2**

Figure 5: Bone scintigraphy with Tc99m methylene diphosphonate (MDP) showing abnormal increase in uptake and asymmetric cortical activity along the cortex of entire length of left femur, left tibia, left tarsal and metatarsal bones , and crossing left hip and knee joints.

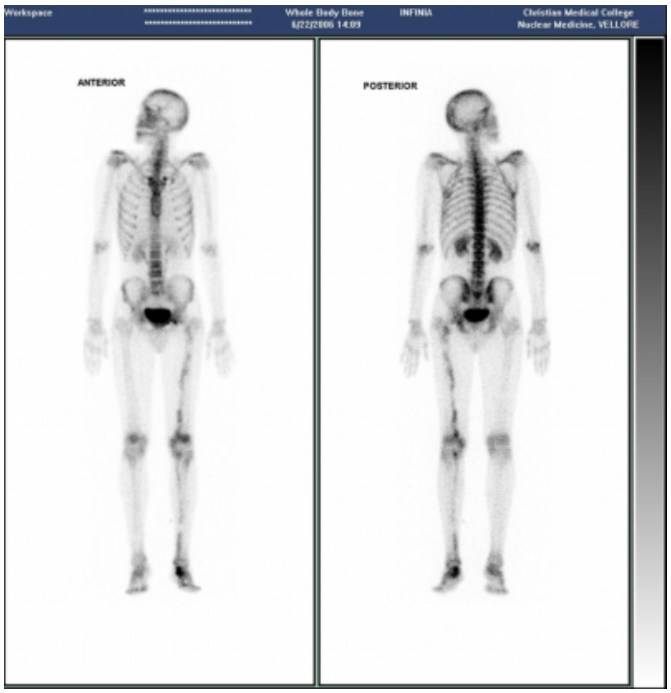
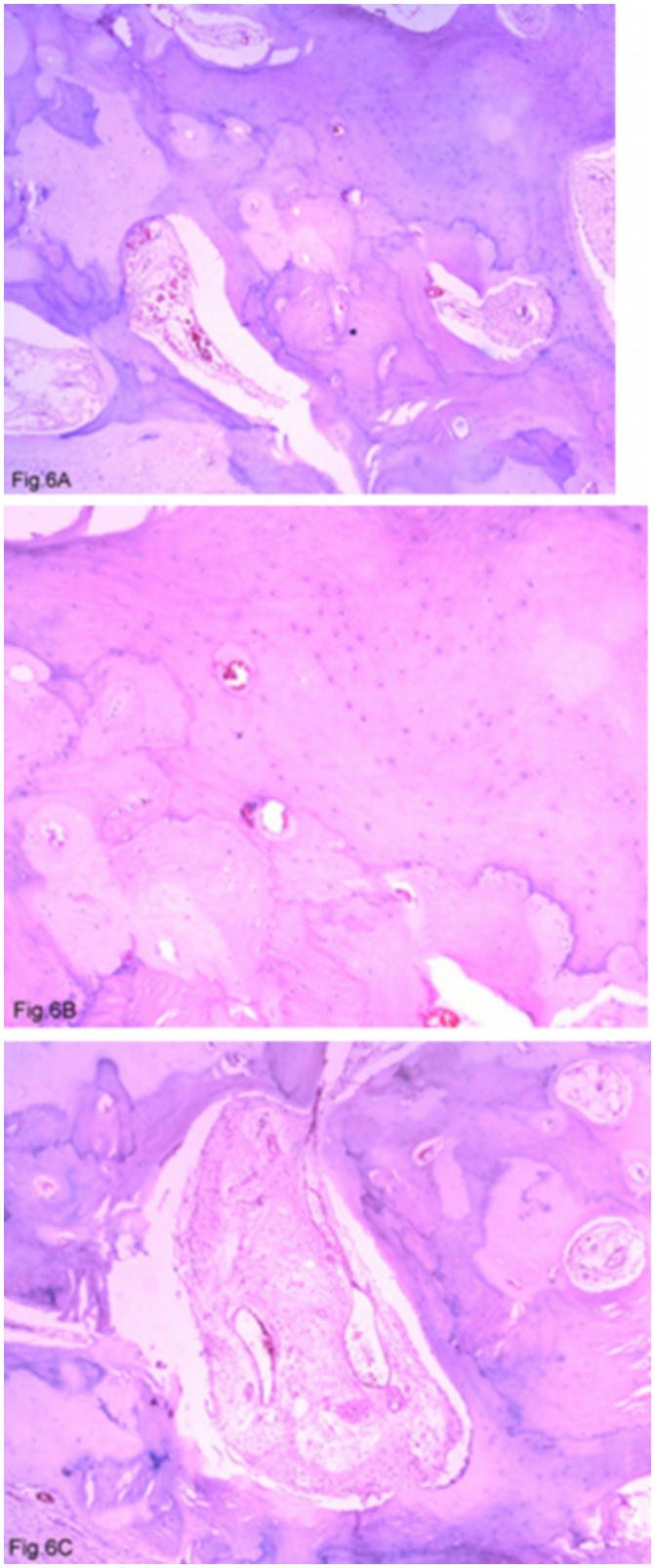



**Figure 3**

Figure 6abc: Biopsy of the lesion from femur showing marked endosteal sclerosis with marrow fibrosis consistent with melorheostosis



## DISCUSSION

Melorheostosis is a rare benign bone sclerosing dysplasia of unknown etiology with linear hyperostosis which may be monostotic, polyostotic, or monomelic with a prediliction for the diaphyseal and epiphyseal regions of the lower limbs and may also be associated with soft tissue changes. Patients may be asymptomatic or may experience symptoms such as stiffness, pain (typically worsened by activity), impaired joint mobility, altered limb length, deformity, and various other orthopedic, dermatologic, vascular, lymphatic, and soft-tissue abnormalities.

Other sclerosing lesions of bone such as osteopathia striata and osteopoikilosis, which usually involve a single bone, have overlapping radiographic features[1]. Distinguishing melorheostosis from these lesions is difficult by radiography alone. As demonstrated in this case, the interpretation is made easy by correlation of radiographic images with scintigraphic images[2,3], the latter showing typical features. Scintigraphic appearance is characteristic, with asymmetric cortical activity that may cross joints to involve contiguous bones.[4] Bone scintigraphy also gives distribution of the lesions including asymptomatic lesions in a single planar whole body view, thus eliminating the need for multiple radiological images.

Plain radiography typically demonstrates lesions which are confined to one limb. The lower extremity is more often affected than the upper extremity. Lesions can also affect the bones of the skull, face and clavicle. Within the long bones lesions are characterized as peripherally located hyperostosis. Lesions tend to have a ‘flowing' appearance and have been compared to a ‘candle wax dripping.’ Endosteal lesions may partially or completely obliterate the medullary cavity. In lesions of the carpal and tarsal bones, the lesions tend to be more discrete and somewhat rounded which may appear similar to those seen in osteopoikilosis. In flat bones, the lesions may be localized, patchy and sclerotic. Soft tissue calcifications are not infrequent, especially in the para-articular regions. These calcifications may progress to joint ankylosis. Bone scintigraphy with Tc99m MDP demonstrates abnormal increase in tracer uptake and asymmetric cortical activity along the cortex of bones, with the activity often crossing the joints.

Histologic findings include variable degrees of cortical thickening consisting of chondroid islands surrounded by mature lamellar and woven bone, as well as adjacent zones of fibrocartilage with irregular surface fibrillation .

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### References

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