

Demonstration of melorheostosis on bone scan

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ABSTRACT

Background: Melorheostosis has been discussed infrequently in the nuclear medicine literature. A 25-year-old female patient presented with a one-year history of bunion pain, sclerotic change and skin pigmentation on the left big toe interphalangeal joint. Ulceration, poor healing and hyperkeratoses of the wound were noted post debridement. The plain film of the left foot incidentally showed melorheostosis. **Materials and Methods:** Radiographs revealed cortical hyperostosis of big toe. After further evaluation by X-ray and bone scan of the left foot, the radiologic findings still indicated melorheostosis. A further Tc-99m MDP bone scan revealed extensive bone involvement in the left side of the pelvis and entire left lower extremity. Moreover, plain film whole body bone scan and MRI also revealed melorheostosis. **Results:** The clinical symptoms of her left big toe included interphalangeal joint callus formation, debridement, ulcer and hyperkeratoses. X-rays often reveal a pattern of thickened bone that resembles dripping candle wax, with periosteal cortical thickening, confined to sclerotomes; and can be seen apparently flowing across joints to the next bone. In the nuclear medicine image, an increase in radiotracer uptake is usually present on late phase bone scans. **Conclusion:** There was excellent correlation between the scintigraphic and radiographic distribution of these lesions in the following radiographs of pelvis, left femur, lower leg and MRI of the left foot. This study reported a rare case of melorheostosis affecting the big toe and reviewed the role of various imaging diagnosis of this rare bone dysplasia.

Keywords: Bone scan, melorheostosis, radiography, MRI.

► Case report

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INTRODUCTION

Melorheostosis is an uncommon, non-

hereditary, benign, sclerosing mesodermal disease that affects the skeleton and adjacent soft tissues, with an incidence of 0.9 cases per

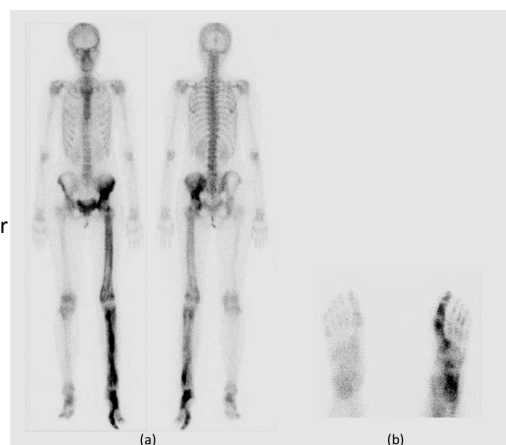
million (1-3). The etiology and pathogenesis of this disorder remains unclear. Various theories have been forwarded to explain the etiology of this condition (4). It is a rare, benign, sclerotic bone dysplasia, demonstrating dense bone formation, typically along the cortex of tubular bones, occasionally in hands or feet, but rare in the axial skeleton (5,6). It is a chronic, sometimes debilitating disease and rarely is associated with other bone malignancy such as giant cell tumor or osteosarcoma. Onset is usually insidious, frequently diagnosed in childhood or young adulthood, but may manifest at any time. The onset rate in both genders is equal. The most common signs/symptoms often present with scleroderma-like skin change, pain, vascular change and swelling of affected joints. Other signs/symptoms include deformity from muscle contractures and atrophy, and growth disturbance from abnormal growth plate fusion or contractures. The image findings are well-circumscribed, found incidentally with undulating areas of cortical sclerosis on radiographs. This appearance is often referred as a "dripping candle wax" pattern on plain films. It may be regional, crossing joints in an X-ray, or following a sclerotome. Usually radiographic appearances are sufficiently characteristic to make diagnosis. The bone scintigraphy, MRI or CT aids the findings on the plain radiography.

CASE REPORT

We present the case of a 25-year-old female

who was diagnosed with melorheostosis affecting her left foot. The diagnosis was suspected to be melorheostosis, but the patient was previously well until a year ago when she first mentioned bunion pain with ulceration in her right lower leg and foot. Sclerotic change and skin pigmentation were also noted. The clinical symptoms of her left big toe included interphalangeal joint callus formation, debridement, ulcer and hyperkeratoses. Examination of the left foot revealed skin thickening on the medial aspect and multiple scars were present. Limited rotation of motion, decreased dorsiflexion of the left big toe and poor healing of the wound were noted. Radiographs revealed cortical hyperostosis of big toe. After further evaluation by x-ray and bone scan of the left foot, the radiologic findings still indicated melorheostosis. Therefore, she was referred to our hospital for the impression of the pressure sore of her left big toe; and for further survey and treatment (figure 1 and 2). The subsequent plain film whole body bone scan and MRI also revealed melorheostosis (figure 3). She received left proximal interphalangeal joint osteotomy of the left big toe and the pathological report was compatible with melorheostosis. Because the patient did not come back to our hospital after treatment, the follow-up situation was unclear. This is rarely considered as a disease and the treatment opinions are either to take pain relievers or to have an amputation. The bone scan technique was used to guide the surgical resection range. Unlike the usage in cancer treatment, it can be used to diagnose and track the efficacy in stages.

Figure 1. Bone scintigraphy with Tc99m MDP showing abnormal increase in uptake. (a) The whole body bone scan shows intensely increased uptake in skeletal structure of left lower limb and left side of pelvic bones. (b) The plantar view shows radiotracer accumulation in left foot, especially in 1st metatarsal bone and big toe.



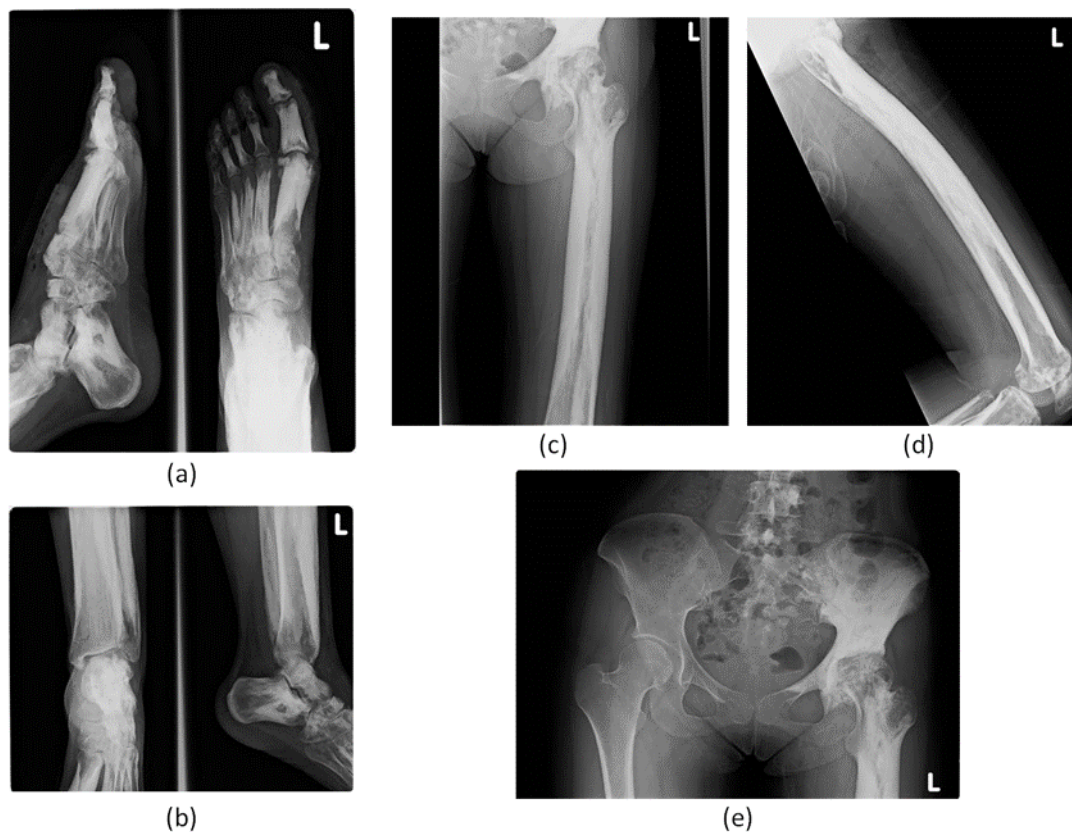


Figure 2(a). Left foot AP and Lateral Views: Classical appearances of melorheostosis affecting the left foot: Anteroposterior and lateral radiograph demonstrates dense cortical and extra-cortical bone formation.-“the dripping candle wax appearance” Deformity of the 1st metatarsal head is present. **(b)** Ankle AP and Lateral Views: There are ossified/calcified nodules at the dorsal side of the 1st and 2nd metatarsal base. **(c and d)** Lt. Femur AP & Lat: Plain radiographs of pelvis and left lower limb showing peripherally located hyperostosis with ‘flowing candle wax dripping’ appearance - features consider as melorheostosis. **(e)** AP View of Pelvis: Diffuse dense osteoclerotic change is shown in the left femur, the left aspect of pelvis, and lumbar spine.

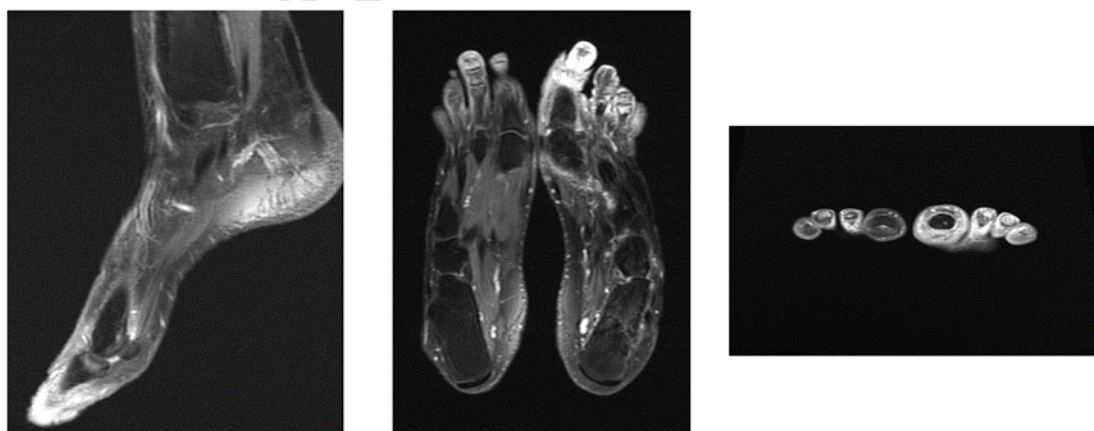


Figure 3. MRI imaging of left foot: Cortical thickenings with candle wax dripping in the tarsal bones, metatarsal bones, and phalanges is noted with low signal intensity on T2WI. Axial coronal and sagittal images showing thickened cortices which were hypointense all imaging sequences confirming the sclerotic and hyperostotic nature of the tissue. However, with enhancement, the left big toe and midfoot show increased infiltration of the soft tissue and high signal intensity on T2WI.

DISCUSSION

MRI findings of the sclerotic bone show low signal in T1W1, T2W2, but no marked marrow edema or destruction typical of malignancy. High-signal, soft tissue mass appears adjacent to affected bone (7). MRI is useful when clinical change raises the possibility of malignant degeneration, but is not required for initial evaluation of a lesion discovered on radiograph.

Melorheostosis causes a positive bone scan, mild to moderate avid Tc-99m MDP accumulation, but localization only to areas of sclerosis closely corresponding to radiographic abnormality distribution (8). Unlike melorheostosis, these other benign sclerotic etiologies such as osteopoikilosis, or osteopathia striata do not generally show increased radioactivity on bone scan. Bone scan is helpful when superimposed osseous disease must be differentiated from these rare sclerosing bone disorders for an accurate diagnosis, to prevent senseless treatment or lost therapeutic opportunities (9-11). It can complement MRI, by allowing a whole body survey and possibly better tumor margin detection, discriminating signal change from edema. Detailed radiographs should be performed when a bone scan shows abnormal uptake in a linear pattern in long bones or a sclerotome distribution (12).

Although melorheostosis is a benign dysplasia it can cause significant morbidity. The morbidity of melorheostosis appears to be mostly due to pain, contractures, limitation of joint motion, limb length discrepancies, and osseous deformities. Treatment of melorheostosis includes medical therapy for symptom control, and surgical treatment of soft tissue complication (13). Infrequently, advanced cases require joint replacement or amputation. Treatment options have been largely surgical in the past, including tendon lengthening, excision of soft tissue masses, release of joint contractures, and osteotomies, but recurrence is common. Antiresorptive agents, such as bisphosphonates, are commonly used for pain relief in conditions associated with increased bone turnover. Nitrogen-containing

bisphosphonates inhibit osteoclast-mediated bone resorption by direct and indirect actions on osteoblasts and macrophages (4). Melorheostosis is a benign bone dysplasia affecting predominantly the appendicular skeleton and adjoining soft tissues (14). The diagnosis can be established on plain radiographs alone and advanced imaging modalities can be avoided (15). The bone scintigraphy, MRI or CT helps to accurately delineate the extent of the disease and assists the surgeon in further management of these patients.

CONCLUSIONS

Melorheostosis has been discussed infrequently in the nuclear medicine literature. Bone scintigraphy is known to show mild to moderately increased activity in all areas involved on radiographs, which may be due to increased turnover of bone or increased bone mass in the affected area. The overall diagnosis of the clinical manifestation, scintigraphic correlation and radiographic findings distinguished the rare entity from other causes of osteosclerosis. In melorheostosis, bone scans appear to be markedly positive. However, on magnetic resonance imaging (MRI) there is usually a low signal on all imaging sequences, with no enhancement. X-ray imaging is the preferred diagnostic tool for melorheostosis. X-rays often reveal a pattern of thickened bone (sclerotic bone lesions) that resembles dripping candle wax, periosteal cortical thickening (endosteal with intramedullary extension may be also present), confined to sclerotomes, and can be seen apparently flowing across joints to the next bone. In the nuclear medicine image, an increase in radiotracer uptake is usually present on late phase bone scans. This study reported a rare case of melorheostosis affecting the big toe and reviewed the role of various imaging diagnosis of this rare bone dysplasia.

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