Complementary Role of Skeletal Scintigraphy in a Case of Polyostotic Melorheostosis

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ABSTRACT

Melorheostosis is an atypical sclerosing bone dysplasia also known as "candle bone disease," "melting wax syndrome," or "Leri disease," with an incidence rate of less than one in every million. The diagnosis is usually made by the radiographic appearance of "dripping candle wax" on the affected bone cortex. We describe the case of a 35-year-old male patient who presented with a long history of different joint swelling, notably in the small joints of the right hand, left foot, and ankle. He was thoroughly investigated to rule out all other possible causes of swelling before being diagnosed with melorheostosis with osteopoikilosis based on radiological patterns. The patient was referred to our institute for a bone scan, which not only showed correlation with X-ray findings but also revealed involvement of the axial skeleton, which is a rare incidence in this disorder.

Keywords: Melorheostosis, Bone scan, Candle bone disease.

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INTRODUCTION

Melorheostosis is a benign bone disorder first described in 1922 by Leri and Joanny, which derives its name from the Greek words melos-limbs, rhein-to flow, and osteon-bone as a tribute to the pathognomonic radiological appearance resembling hardened wax dripping down the side of a candle (1). It can affect any age group with no gender predilection and an incidence rate of 0.9 in every million (2). This disease commonly affects unilateral long bones, sometimes hands or feet, but involvement of multiple sites or the axial skeleton is rare. Patients may present with pain, bony swelling, or joint deformities, with a history of insidious onset (3). Plain radiography is often sufficient to make the diagnosis, with bone scintigraphy, a computed tomography (CT) scan, or magnetic resonance imaging (MRI) having complementary roles. Sometimes X-ray findings do not conform to the classic pattern, and there may be considerable overlap with other bone sclerosing conditions like osteopoikilosis, osteopathia striata, and various hyperostoses (1). Melorheostosis can be fairly debilitating despite its benign nature, and management is mainly symptomatic through a multidisciplinary approach (4).

CASE REPORT

A 35-year-old Asian male presented with painful joint swelling since adolescence, mostly involving small joints of the right hand, left foot, and left ankle. It had been gradually worsening over the last few years. He had no history of fever, trauma, or any other abnormality, and there was no family history of a similar illness. His right hand and left foot appeared swollen with multiple non-tender hard nodules on physical examination (Figure 1). He exhibited a slightly limping gait but no discernible limb length discrepancy. There was no change in skin color or temperature. All of his blood parameters, including serum calcium, alkaline phosphatase, parathyroid hormone, fetoprotein, carcinoembryonic antigen, C-reactive protein, serum TSH, serum uric acid, and erythrocyte sedimentation rate, were within normal limits.

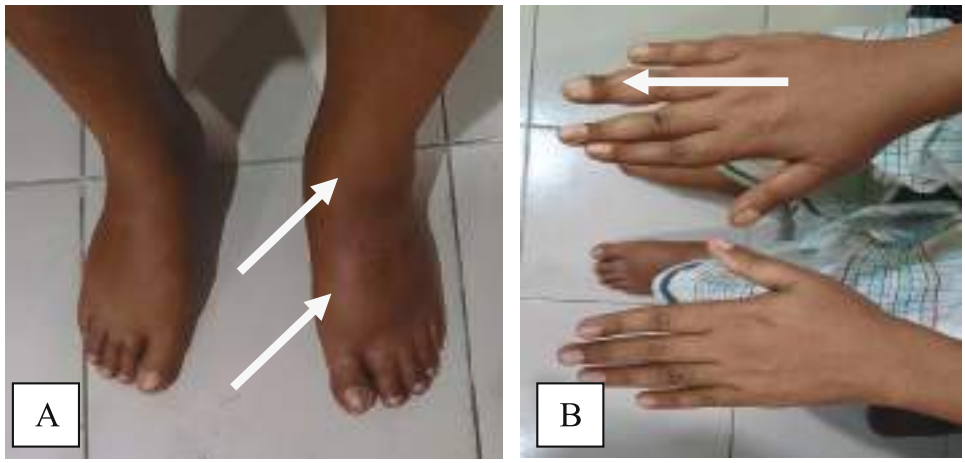

Figure 1: Swelling involving left foot & ankle (A) and small joints of right hand (B).

A plain radiograph of both upper and lower limbs, including both hip bones, revealed irregular cortical thickening of the external aspect of the tibia, femur, and tubular bones of the hand and foot, appearing as dense sclerosis and giving a molten wax appearance (Figure 2). Multiple rounded sclerotic lesions were also noted around the hip and knee joints. Based on the findings, Melorheostosis with Osteopoikilosis was proposed as a diagnosis.

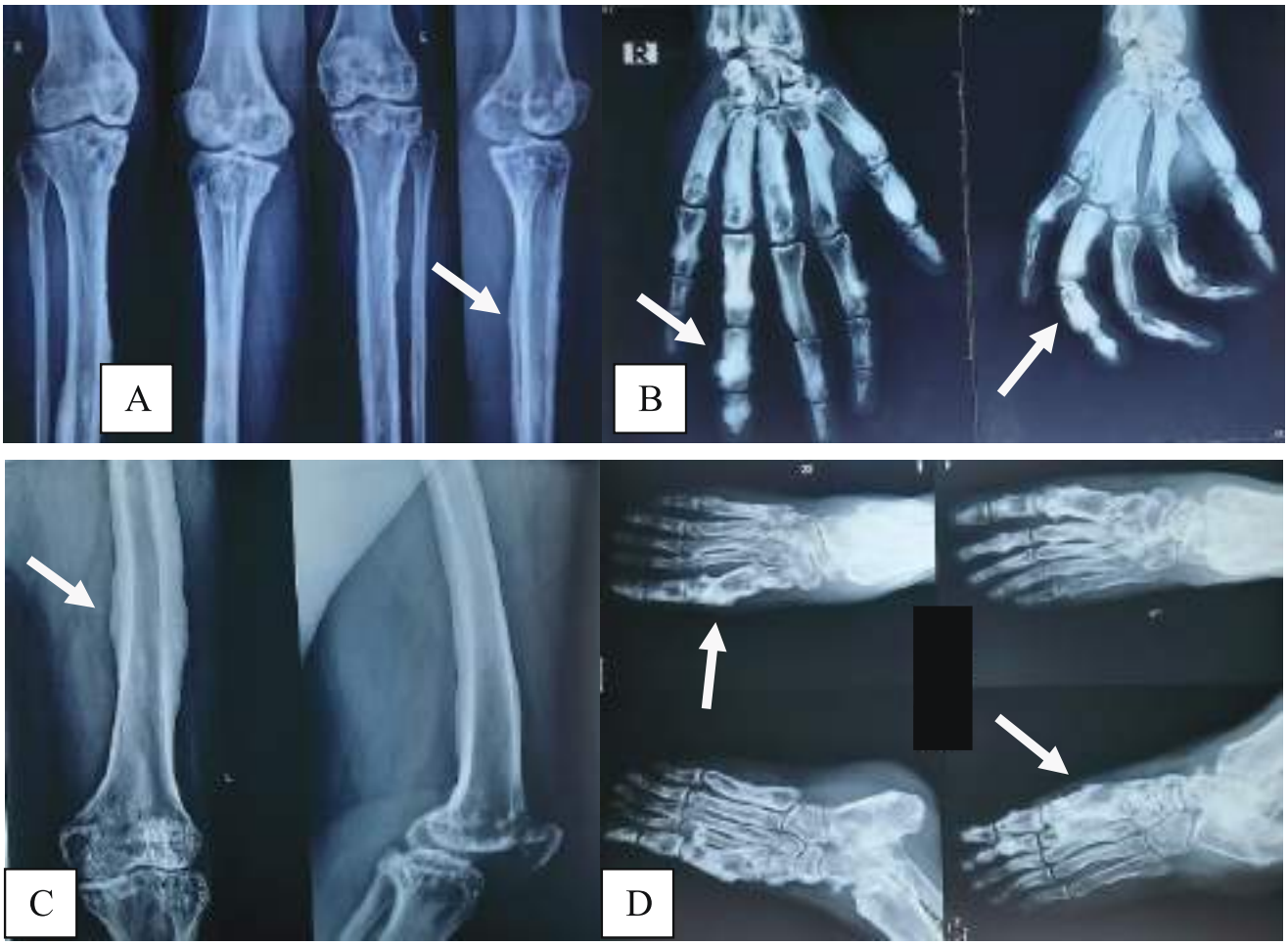

Figure 2: Cortical thickening showing typical dripping wax appearance in plain X-ray of long bones (A, C). Dense cortical and extra-cortical bone formation is also seen in right hand and left foot (B, D).

The patient was referred to our center for a full-body skeletal survey, which included a ^{99m}Tc-Technetium MDP bone scan. The scan was performed in both the anterior and posterior views three hours after the administration of 20 mCi of ^{99m}Tc-MDP. Skeletal scintigraphy confirmed the radiological findings of involvement in long bones (both femoral shafts, right tibia) as well as tubular bones of the hand and foot (metacarpal bones and phalanges of the right hand, small joints of both feet). Besides the appendicular skeleton, mildly increased radiotracer concentrations were also seen at the right clavicle, right scapula, left 6th–8th ribs, right SI joint, part of the left hip bone, and multiple vertebrae denoting axial skeletal involvement (Figure 3).

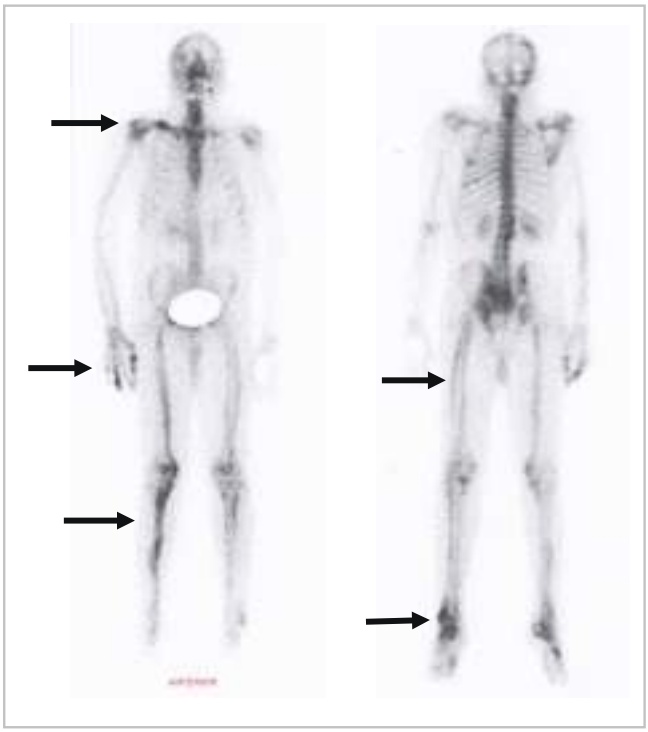

Figure 3: Bone scan showing radiotracer concentration in upper shaft & lower aspect of right tibia, metacarpal bones & phalanges of right hand and small joints of both feet. Linear tracer activity is noted at cortical regions of both femoral shafts with absence of medullary activity, likely due to cortical thickening. Besides, lateral aspect of right clavicle, right scapula, left 6th-8th ribs (posteriorly), right SI joint, acetabular region of left hip bone, and multiple dorso-lumbar vertebrae are also affected.

DISCUSSION

Melorheostosis, or candle bone disease, is infrequently mentioned in nuclear medicine literature because conventional radiography is usually considered a fairly informative imaging modality for making the diagnosis. However, the complementary role of bone scintigraphy in this pathology has been documented by several authors.

Chang et al. described a case where they observed excellent correlation between the scintigraphic and radiographic distribution of lesions (5). Our findings also supported this statement and, moreover, revealed previously undetected lesions. Melorheostosis can be monostotic, polyostotic, or monomelic in presentation, with the monomelic form being the commonest variant (6). Kumar R et al. reported a monostotic monomelic case in 2014, and C. C. Teoh et al. found another patient with the polyostotic monomelic variety (two long bones in the ipsilateral leg) in 2019 (3, 4). Both had hyperpigmentation of the overlying skin, which is found in 17% of the cases (3). Our patient not only had extensive polyostotic involvement of one upper and two lower limbs but also the axial skeleton, which is extremely rare in this disease (7). Plain radiography was inconclusive about the axial skeletal involvement (Figure 4).

Figure 4: Plain X-ray pelvis and spine A/P & lateral view. There were apparent sclerotic changes in right 12th rib, L1-2 vertebrae and right SI joint, but interpretation is inconclusive due to overlying extensive bowel shadow and inability to explore the full length of spine and ribs.

According to previous studies, melorheostosis results in mild to moderately avid ^{99m}Tc MDP accumulation in areas of sclerosis, possibly due to increased bone turnover or increased bone mass in the affected region, while other benign sclerotic conditions like osteopoikilosis and osteopathia striata do not generally produce a positive

bone scan. Therefore, skeletal scintigraphy might be useful to distinguish between overlapping osseous lesions, to make an accurate diagnosis, and also to avoid inappropriate treatment (5, 8). The patient showed localized sclerotic lesions along with typical "dripping wax"-like cortical thickening in a plain radiograph, which suggested co-existence of Melorheostosis with Osteopoikilosis. A bone scan could not rule out an osteopoikilosis lesion because there was already widespread involvement, resulting in superimposition of pathologies.

MRI is beneficial to delineate soft tissue involvement more accurately or explore the possibility of malignant degeneration (5). But whole-body screening with MRI is not cost-effective for an osseous pathology. This is the advantage bone scintigraphy holds over other imaging options: being able to provide a whole-body skeletal survey in a single setting with relatively less expense.

CONCLUSION

Melorheostosis is a rare entity among various causes of osteosclerosis, which can be distinguished by its clinical manifestation, radiographic findings, and scintigraphic correlation. This disease can result in remarkable morbidity despite its benign nature, and symptomatic management is necessary to alleviate patient suffering. Our study described a rare case of polyostotic melorheostosis, demonstrating once again that the bone scan is a reliable tool for whole-body screening which can supplement other investigations and improve diagnostic accuracy in the majority of skeletal disorders.

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