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Melorheostosis with an associated para-articular enhancing soft tissue mass

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ABSTRACT

Melorheostosis is a rare non-hereditary sclerosing bone dysplasia which predominantly affects the appendicular skeleton. Although melorheostosis is typically recognized as an osseous lesion, associated soft-tissue components have been reported. Advanced imaging with MRI may allow for more complete evaluation of these soft tissue components; however, there is little information regarding their MRI characteristics which may lead to confusion with malignant processes. We present a case of melorheostosis in a 32-year-old woman with an associated paraarticular enhancing soft tissue mass and emphasize discriminating this from soft tissue sarcoma.

1. Introduction

Melorheostosis is a rare non-hereditary sclerosing bone dysplasia first described by Léri and Joanny in 1922 [1]. Classically the radiographic appearance demonstrates flowing cortical hyperostosis, likened to dripping candle wax [2]. The appendicular skeleton is predominantly affected, usually in a monomelic (one limb) distribution with either mono- or polyostotic involvement [3]; rarely, more than one limb or the axial skeleton may be involved [2,4].

Although melorheostosis is typically recognized as an osseous lesion, there can be an associated soft tissue component [5–7]. These masses are often para-articular and may be contiguous with the nearby cortex [8]. Histologically, the soft tissue component can be variegated, including osseous, chondroid, vascular, fibrocartilaginous, or fibrolipomatous tissue [6,9–11] which is often incompletely characterized by radiography or scintigraphy. MRI may provide more complete evaluation; however, there remains a paucity of information regarding the MRI characteristics of the soft tissue component [4,5,9,12]. Accurate characterization of the soft tissue component is important because these lesions can be misinterpreted as aggressive soft tissue sarcomas rather than the benign manifestations of melorheostosis that they represent [4], potentially leading to unnecessary biopsy or surgical intervention. Herein, we present a case of melorheostosis with an associated para-articular enhancing soft tissue mass characterized by radiographic and MR imaging and provide a brief literature review of this entity.

2. Case report

A 32-year-old Hispanic woman presented with progressive right knee and leg pain for the past year; this was accompanied by occasional stiffness, limited knee flexion, and paresthesia. Physical examination revealed palpable prominences and tenderness at the lateral femoral condyle and proximolateral fibular shaft. No overlying skin findings were seen and laboratory evaluation was normal.

Radiographs of the right tibia-fibula and knee demonstrated undulating cortical hyperostosis at the lateral femoral condyle and proximolateral fibula (Fig. 1). An amorphous region of soft tissue mineralization was seen adjacent to the lateral femoral condyle.

The soft tissue component was incompletely characterized radiographically and there was clinical concern for malignancy; therefore, an MRI was performed. By MRI, the mineralized portion of the soft tissue mass showed low-signal on T1- and T2-weighted sequences without enhancement on post-contrast images (Fig. 2). In the non-mineralized portion of the mass, T1- and T2-weighted images showed heterogeneous but predominantly intermediate signal intensity. On post-contrast images, the soft tissue mass demonstrated both peripheral and heterogeneous internal enhancement.

3. Discussion

Melorheostosis is a rare benign skeletal dysplasia of uncertain etiology, but there are two major theoretical explanations. Murray and McCredie first described the segmental distribution seen in melorheostosis as sclerotomal, i.e. corresponding to a sensory spinal nerve of the skeleton [8]; they hypothesized that insult/injury to the neural crest during embryogenesis could lead to this sclerotomal distribution. Fryns et al. suggested that melorheostosis may be related to somatic mosaicism, similar to Proteus syndrome (a rare congenital asymmetric overgrowth syndrome involving any germ layer), and may be due to an

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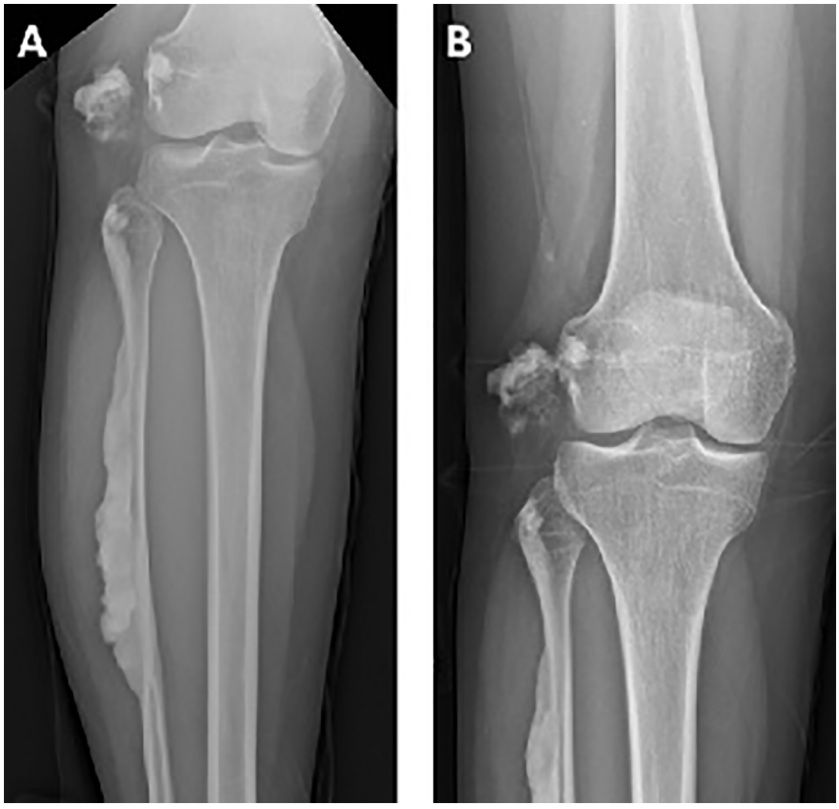

Fig. 1. A and B Frontal radiographs of the right tibia and fibula (A) and knee (B) demonstrate the classic "candle wax" cortical hyperostotic lesions along the right fibula and lateral femoral condyle. Myositis ossificans-like soft tissue ossifications are seen lateral to the lateral femoral condyle.

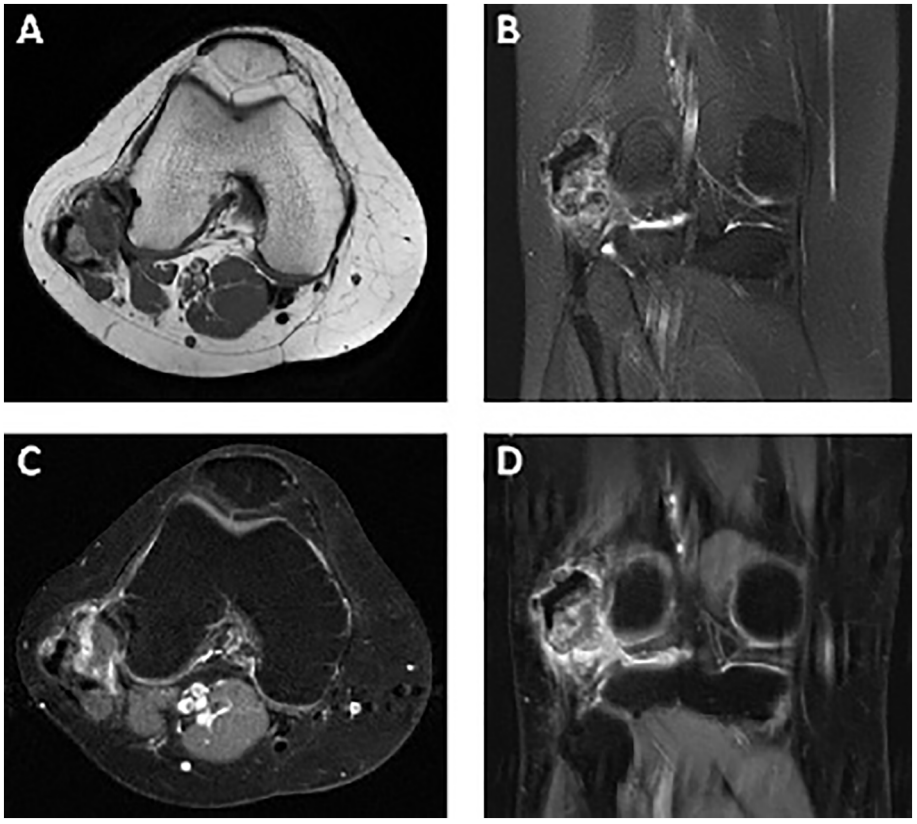

Fig. 2. A and B Axial T1-weighted (A) and coronal T2-weighted fat suppressed (B) MR images demonstrate decreased signal intensity within the soft tissue mass corresponding to regions of mineralization on radiographs. Heterogeneous but predominantly intermediate signal is seen within the non-mineralized portion of the soft tissue mass on both T1- and T2-weighted images. C and D Axial (C) and coronal (D) T1-weighted fat suppressed post-contrast MR images demonstrate enhancement along the periphery of the soft tissue mass as well as heterogeneous enhancement within the mass. No enhancement is seen in the mineralized portion of the mass.

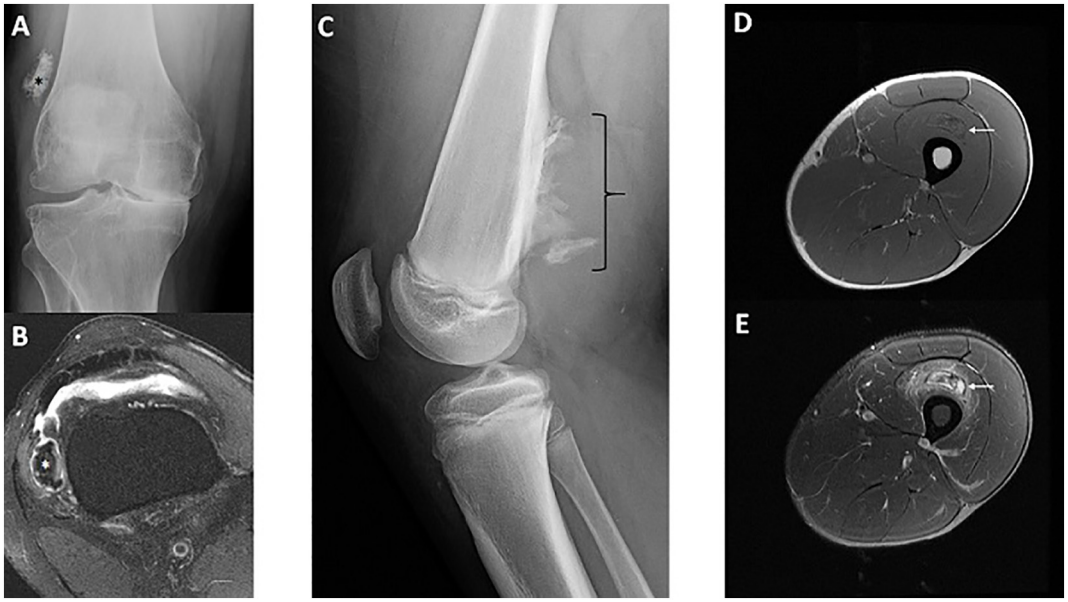

Fig. 3. A and B Frontal radiograph (A) and axial T2-weighted fat suppressed MR image (B) demonstrate a large osteochondral body (asterisks) in the lateral aspect of the suprapatellar recess consistent with secondary synovial osteochondromatosis; note, the severe joint space loss of the medial femorotibial compartment. C Lateral radiograph demonstrates a large exophytic mass with dense central ossification at the posterior aspect of the distal femoral metadiaphysis (bracket), consistent with a parosteal osteosarcoma. D and E Axial T1 (D) and T2-weighted fat suppressed (E) MR images demonstrate a somewhat ovoid lesion with central heterogenous signal, a peripheral rim of hypointense signal (arrows) indicating the zonal organization of mature bone, and surrounding edema within the vastus intermedius muscle—findings consistent with myositis ossificans after a soccer-related injury.

early post-zygotic mesenchymal mutation causing asymmetric involvement of skeletal structures, often with accompanying soft tissue changes [13].

The incidence of melorheostosis is 0.9 per million people affected. Men and women are equally affected and may present at any age, but 40–50% of cases manifest by age 20 [14]. Although benign, melorheostosis can cause significant morbidity: patients may present with chronic or progressive pain, limb deformity, or restricted movement due to contracture and fibrosis [15]. Occasionally, patients may develop muscle atrophy secondary to nerve impingement, with at least two cases reported [4].

The radiographic appearance of melorheostosis has been extensively reported. Our case, like others, demonstrates the classic flowing cortical hyperostosis, as depicted along the lateral femoral condyle and fibula. The soft tissue component—understandably—has variable radiographic appearance depending on the degree of mineralization. The para-articular soft tissue mineralization seen in our case has been previously characterized as a myositis ossificans-like pattern [3].

The MRI appearance of melorheostosis, including any soft tissue component, is less well described. Although many soft tissue abnormalities have been reported in association with melorheostosis, including linear scleroderma; fibrosis with associated joint contracture(s); and masses of fibrous, adipose, lymphatic, vascular, or osseocartilaginous tissue, many of these lesions were not evaluated by MRI. Despite this limitation, the MR appearance may be apprehensible with a knowledge of the histologic constituents. In our case, as expected, the mineralized portions of the soft tissue mass corresponded to low-signal intensity and lack of enhancement. The non-mineralized portions showed a more variable appearance with heterogeneous but predominantly intermediate signal intensity. Based on prior histologic evaluation of similar soft-tissue components, this is speculated to represent fibrovascular tissue with variable collagen content [4]. The peripheral enhancement seen surrounding the mass is presumably secondary to corresponding inflammatory change [9], but the heterogeneous internal enhancement is noteworthy. Only one other case has been shown to have this marked internal enhancement pattern and that case was initially erroneously interpreted as sarcoma [4]; however, subsequent biopsy revealed non-malignant fibrovascular tissue. This highlights the variability of the MR imaging appearance of the soft tissue component of melorheostosis and emphasizes the importance of recognizing the soft tissue component as a manifestation of melorheostosis to prevent misdiagnosis and unnecessary workup or treatment.

The differential diagnosis of melorheostosis with an associated para-articular soft tissue mass may include synovial osteochondromatosis (primary or secondary), surface sarcoma (e.g. parosteal osteosarcoma), and myositis ossificans (Fig. 3). Secondary synovial osteochondromatosis, more common than the primary type and most commonly due to osteoarthrosis, shows nonuniform osteochondral nodules within a joint with associated joint space loss; parosteal osteosarcoma presents as an exophytic lesion arising from the adjacent bone with or without an associated soft tissue mass; and myositis ossificans demonstrates a mass with zonal organization (i.e. mature bone peripherally surrounding myxomatous tissue composed of vascularized sheets of spindle cells with subsequent development of giant cells and differentiation into fibroareolar connective tissue [16]) typically involving the large muscles of the lower extremities after a traumatic injury.

In conclusion, we demonstrate a case of melorheostosis with an associated para-articular soft tissue mass with heterogenous internal enhancement on MR imaging that should not be misinterpreted as a sarcoma.

Declarations of interest

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