

Melorheostosis mimicking synovial osteochondromatosis

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Melorheostosis is an uncommon, sporadic, sclerosing bone lesion that may affect the adjacent soft tissues. It has been associated with many entities such as osteopoikilosis, soft tissue vascular malformations, bone and soft tissue tumors, nephrotic syndrome, segmental limb contractures, osteosarcoma, desmoid tumor, and mesenteric fibromatosis. Synovial osteochondromatosis is a benign neoplasia of the hyaline cartilage presenting as nodules in the subsynovial tissue of a joint or tendon sheath. The intra-articular extension of melorheostosis mimicking synovial osteochondromatosis has not been reported before. In this article, the authors describe an unusual case mimicking synovial chondromatosis arising as a result of melorheostosis and their characteristic imaging findings.

Melorheostosis is an uncommon benign sclerosing bone disease that may affect the adjacent soft tissues.¹ Although it is a non-hereditary (sporadic) disease, it has been observed in families of osteopoikilosis (an autosomal dominant trait).² It has thus been suggested that melorheostosis probably represents a “second hit” mutation of the gene causing osteopoikilosis.³ Melorheostosis has been reported to be associated with multiple other entities such as vascular malformations, nephrotic syndrome, segmental limb contractures, osteosarcoma, desmoid tumor, and mesenteric fibromatosis;⁴⁻¹² however, its intra-articular extension mimicking synovial osteochondromatosis has not been described before. The authors describe the classic imaging features of melorheostosis with the complicating feature of intra-articular extension resulting in multiple loose bodies mimicking synovial osteochondromatosis.

CASE

A 46-year-old woman presented with a 10-year history of chronic right knee pain and stiffness. She had undergone 2 surgical procedures in the past for the same, 1 open and 1 arthroscopic at an outside institution, the nature of which were not known. However, she con-

tinued to suffer from persistent pain and stiffness. Her history was significant for hyperthyroidism, for which she underwent partial thyroidectomy. On examination, the patient had a flexion contracture of approximately 0/30/80 degrees. Diffuse tenderness was present medially and laterally about the joint. There was no obvious ligamentous instability and no signs of active inflammation. There was no loss of muscle tone or sensations in the limb. An MRI showed hypointense lesions of the femur, tibia, and patella on all imaging sequences. Additionally, there was synovial thickening of intra-condylar notch and multiple loose bodies mimicking synovial osteochondromatosis (**Figures 1 and 2**). A correlative radiography series of the right knee (**Figure 3**) characteristically showed dense sclerotic lesions of the femur, tibia, and patella in a classic “candle dripping” appearance in keeping with melorheostosis. The intra-articular bodies were identified as an extension of the melorheostosis process mimicking synovial osteochondromatosis.

DISCUSSION

Melorheostosis is an uncommon periosteal hyperostosis of the cortex of long bones. It is a benign disease that may be asymptomatic for many years. The abnormal os-

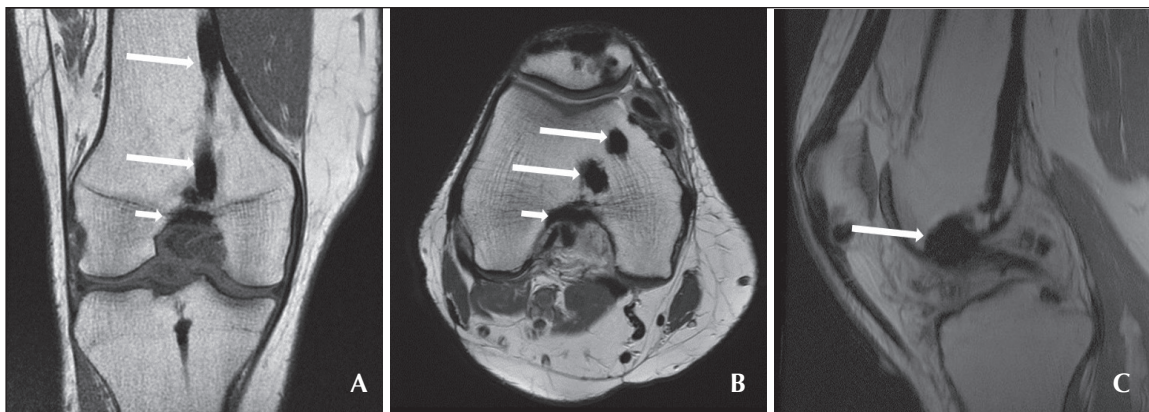


Figure 1. T1W MRI of the right knee. A (coronal view), B (axial view), and C (sagittal view) show hypointense lesions of the femur, tibia, and patella (large arrows). Notice synovial thickening of intra-condylar notch and multiple loose bodies mimicking synovial osteochondromatosis (small arrows).

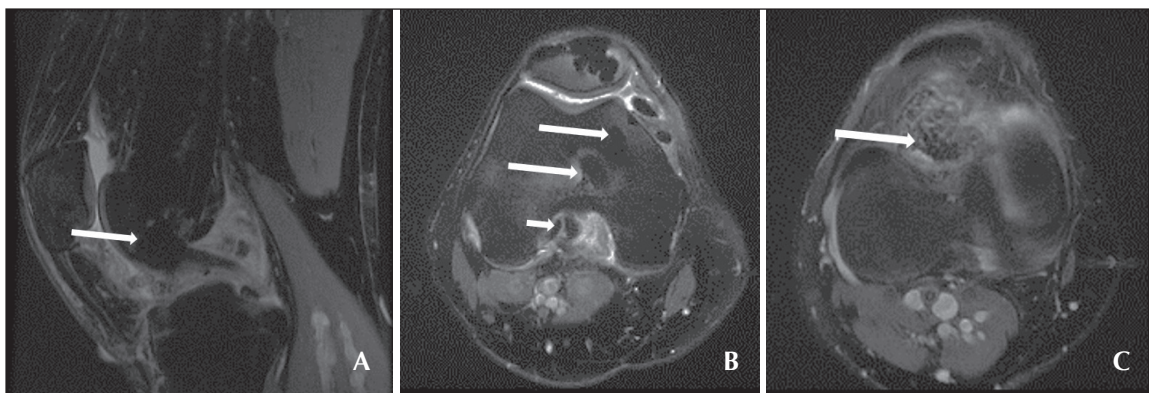


Figure 2. Fat suppressed T2W MRI of the right knee. A (sagittal view) and B and C (axial views) show hypointense lesions of the femur, tibia, and patella (large arrows). Notice synovial thickening of intra-condylar notch and multiple loose bodies mimicking synovial osteochondromatosis (small arrows).

sification in the cortex of the long bone may involve the soft tissues leading to joint contractures and fibrosis. Clinical presentation includes deformity, chronic pain, stiffness, and restricted joint movement in the involved limb, causing significant morbidity associated with the entity.¹ It tends to be segmental and unilateral, and may affect one or multiple bones of the limb, with the lower extremity more commonly involved than the upper.¹³ There is a predilection for the diaphyseal and the epiphyseal regions of the long bones in a typical sclerotomal distribution akin to our case.¹⁴ Though long bones of the limbs are more commonly affected, it may rarely involve the spine, skull, and facial bones.⁴

Plain radiographs of melorheostosis show undulating cortical thickening of the involved bone. It is also appreciated on computed tomography (CT) scans with high attenuation. These areas of thickening are seen encroaching on the medullary canal due to the involve-

ment of the endosteum. Marked increase in radionuclide uptake is seen on bone scan.¹ Soft tissue involvements on CT images show a variable degree of mineralization ranging from predominant mineralization with small soft tissue components to nonmineralization with small foci of calcification. MRI reveals hypointensity on all pulse sequences. MRI of these soft tissue masses shows heterogenous signal intensity due to the presence of various types of tissues. The mineralized areas show hypointensity on all sequences, while the non-mineralized areas with fibrovascular tissue and collagen show isointense signals on T1W images and iso- to hyperintense signals on T2W images. Fat-containing areas may show hyperintensity on T1W images and isointensity on T2W images.^{14,15} Extension into the joint can mimic synovial osteochondromatosis on MRI, and plain film correlation may become essential for the correct diagnosis as observed in this case.

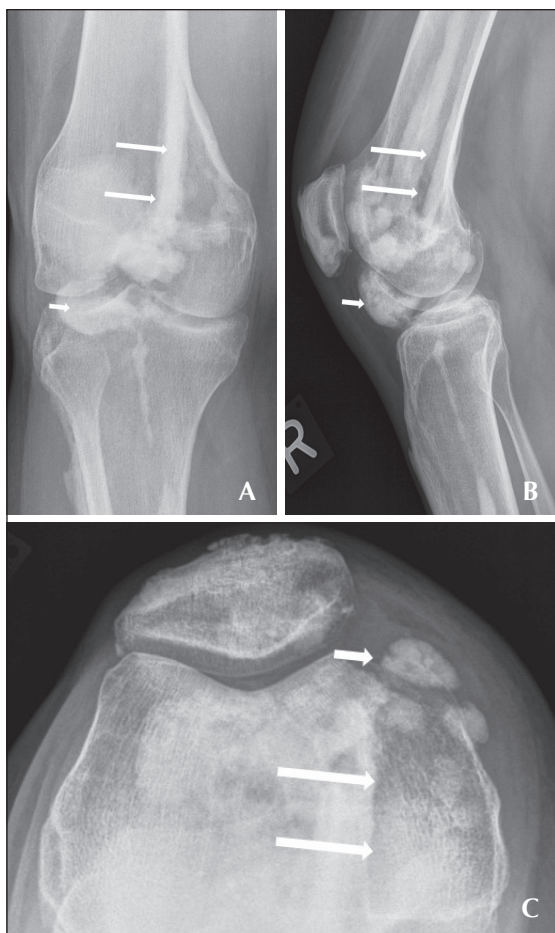


Figure 3. Corresponding plain radiographs of the right knee. A (AP view), B (lateral view), and C (skyline view) show dense sclerotic lesions of the femur, tibia, and patella in a classic “candle dripping” appearance in keeping with melorheostosis (large arrows). Notice extension into the intra-articular space mimicking synovial osteochondromatosis (small arrows).

Melorheostosis is has been widely reported to be associated with a multitude of clinical entities, such as osteopoikilosis and osteopathia striata,⁴ anomalies of blood vessels or lymphatics,^{5,6} osteosarcoma,⁷ fibrolipomas,⁸ desmoid tumors,⁹ multicentric fibromatosis,¹⁰ minimal change nephrotic syndrome,¹¹ and hypophosphatemic rickets.¹² We did not find any other association in this patient.

Synovial osteochondromatosis is a benign neoplasia of the hyaline cartilage presenting as nodules

in the subsynovial tissue of a joint or tendon sheath. The knee is the most commonly involved joint.¹⁶ The usual clinical presentation is pain, swelling, and restriction of joint movement with articular tenderness and crepitus. It is primarily a monoarticular disease with rare cases of polyarticular involvement being reported.¹⁷ A long-standing disease may lead to secondary osteoarthritis and asymmetric joint space reduction with bone erosion. Imaging findings of synovial osteochondromatosis are usually pathognomic. Plain radiographs show multiple intra-articular calcifications distributed throughout the affected joint. This typical punctate mineralization appearance is called “ring-and-arc pattern.” A central focus with single peripheral rim of calcification may give rise to a “target appearance.”¹⁸ Several patterns have been described on MRI of this lesion. Corresponding to regions showing calcification on radiographs or CT scans, there is a homogeneous, intermediate, intra-articular isointensity to muscle on T1W images and hyperintensity on T2W images, with focal areas of low signal intensity on all pulse sequences. Another pattern corresponding to foci of endochondral ossification on CT and radiographs has similar features as discussed in this paragraph; however, it also includes hyperintense foci (isointense to fat) with a peripheral rim of hypointensity.^{16,19}

The management of melorheostosis is difficult due to the formation of joint contractures and fibrosis of joint space. It includes splinting, tendon lengthening, excision of fibrous and osseous tissue, sympathectomy, implant arthroplasty, corrective osteotomy, and amputation, if severe.^{4,14} Surgical release has also been tried with varying degrees of success.²⁰ The treatment of choice for synovial osteochondromatosis (intra-articular or extra-articular) is surgical resection.²¹

To summarize, melorheostosis can extend into the joint in a contiguous sclerotome distribution and produce intra-articular bodies mimicking synovial osteochondromatosis.

Conflicts of interest

Dr. Chhabra has research grants from GE-AUR (GERRAF), Siemens Medical Solutions, and Integra Life Sciences. He also serves as a research consultant with Siemens CAD group.

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