Osteopoikilosis of the knees, incidental MRI findings

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Section: Musculoskeletal system
Area of Interest: Musculoskeletal system
Procedure: Imaging sequences
Imaging Technique: MR
Special Focus: Pathology Case Type: Clinical Cases
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Patient: 24 years, female

Clinical History:

A 24-year-old woman presented to the radiology service with long term bilateral pain in the knees worsened by strenuous physical activity. The range of motion of the knees was not affected. Other joints were unremarkable. No associated skin lesions. Chondromalacia patellae was suspected.

Imaging Findings:

Magnetic Resonance Imaging (MRI) was considered to explore possible intra-articular pathologies. In Proton Density (PD) and T1-weighted sequences were seen multiple, well-defined, millimeter-sized low signal foci of sclerotic areas located in the metaphysis and epiphysis of the femur, tibia and fibula. There were few foci seen also on the patella. No sign of intra-articular injury was shown.

Discussion:

Osteopoikilosis is a benign condition first documented by the German surgeon Alfred Stieda in 1905 and attributed to Heinrich Ernst Albers-Schönberg, a German radiologist and surgeon who published it in 1915 [1,2,3,4]. In 1916, a French radiologist named René Ledoux-Lebard introduced the term osteópoecilie meaning ‘spotted bones’[2,5].

The disorder is a rare form of osteosclerotic dysplasia, with an incidence estimated in 1:50.000, with no gender preference, although new evidence suggests slightly more males affected[4,6].

Also referred to as spotted bones or osteopathia condensans disseminate, osteopoikilosis has an autosomal dominant inheritance, associated with a heterozygous mutation in the LEMD3 gene, or a sporadic form, characterized by defective endochondral bone formation[6,7].

Typically, an incidental imaging finding, osteopoikilosis is usually clinically asymptomatic, although there are reported cases of articular pain or joint effusion, despite established clear evidence is not known[4,6].

The lesions typically have and uniform size and symmetrical periarticular distribution in the axial and appendicular skeleton, with metaphyseal and epiphyseal involvement, presented as small round areas of low signal intensity on MRI, measuring 2–10 mm in diameter[2,4,6,7], better seen on T1 weighted sequences.

The main differential diagnosis involves sclerotic metastases and other sclerosing dysplasias, as enostosis, ostepathia striata, pycnodysostosis and osteopetrosis. All these conditions can be diagnosed and differentiated by
radiologic features[7].

Despite both osteopoikilosis and osteopathia striata have periarticular distribution, the first one presents with multiple sclerotic, rounded or oval foci whereas the second by linear areas of sclerosis, whereas enostosis presents with scattered sclerotic foci and no specific distribution. All the conditions above are asymptomatic. Osteopetrosis is characterized by diffuse sclerosis or bone-within-bone pattern and may present with anemia and increased risk of fractures. On pycnodysostosis is seen a diffuse pattern of sclerosis, instead of spotted or rounded foci. In contrast, osteoblastic metastases show lack of internal architecture and irregular margins, although the possible periarticular distribution[7].

Accordingly, differential from other pathologies is based on the symmetric distribution, location and absence of bone destruction[4].

Regardless of the radiological findings of osteopoikilosis, in some cases, differentiation with primary or secondary sclerotic bone lesions can be complicated, especially in patients with known cancer history. As osteopoikilosis lesions are not affected by bone scintigraphy, this imaging method may aid in differentiating malignancy[2, 8].

Osteopoikilosis is considered a ‘don’t touch’ injury and there is no need for routine follow-up or further studies[4,9].

‘Written informed patient consent for publication has been obtained.’

Differential Diagnosis List: Osteopoikilosis, Enostosis, Ostopathia striata, Osteopetrosis, Pycnodysostosis, Osteoblastic metastases

Final Diagnosis: Osteopoikilosis

References:

Description: Sagittal section of T1 image showing rounded sclerotic areas in the patella and in the metaphysis and epiphysis of the femur and tibia. Origin: Department of Radiology, Nossa Senhora do Perpétuo Socorro Hospital, Alfenas, Brazil.
**Description:** Sagittal T1 image showing low signal sclerotic foci in the femoral, tibial and fibular epiphysis. **Origin:** Department of Radiology, Nossa Senhora do Perpétuo Socorro Hospital, Alfenas, Brazil.
Description: Sagittal section of PD image showing rounded sclerotic areas in the epiphysis of the femur and tibia. Origin: Department of Radiology, Nossa Senhora do Perpétuo Socorro Hospital, Alfenas, Brazil.
**Description:** Axial section of PD image showing periarticular sclerotic areas in the tibia and fibula.

**Origin:** Department of Radiology, Nossa Senhora do Perpétuo Socorro Hospital, Alfenas, Brazil.
Description: Axial PD image showing periarticular sclerotic areas on femur and patella. Origin: Department of Radiology, Nossa Senhora do Perpétuo Socorro Hospital, Alfenas, Brazil.
Description: Coronal section of PD image showing periarticular sclerotic foci on femur and tibia.
Origin: Department of Radiology, Nossa Senhora do Perpétuo Socorro Hospital, Alfenas, Brazil.
Description: Sagittal section of T1 image presenting millimeter-sized sclerotic areas in the femoral and tibial metaphysis and epiphysis. Low signal foci noted in the patella. Origin: Department of Radiology, Nossa Senhora do Perpétuo Socorro Hospital, Alfenas, Brazil.
Description: Sagittal section of PD image presenting millimeter-sized sclerotic areas in the metaphysis and epiphysis of the femur and tibia. Origin: Department of Radiology, Nossa Senhora do Perpétuo Socorro Hospital, Alfenas, Brazil.
**Description:** Axial section of PD image showing sclerotic areas in the femoral epiphysis. There are no signs of intra-articular injury. **Origin:** Department of Radiology, Nossa Senhora do Perpétuo Socorro Hospital, Alfenas, Brazil.
**Description:** Coronal PD image showing sclerotic foci in the femoral and tibial epiphysis. **Origin:** Department of Radiology, Nossa Senhora do Perpétuo Socorro Hospital, Alfenas, Brazil.