CLINICAL IMAGE

Spotted bone disease

Theodora Simopoulou¹



Received: 13 March 2020 / Revised: 13 April 2020 / Accepted: 21 April 2020 / Published online: 3 May 2020 © International League of Associations for Rheumatology (ILAR) 2020

Clinical case

A 34-year-old Caucasian male presented to the outpatient clinic with a 2-year history of knee and tibia pain, which was worse during bedtime resting. He reported no acute injury; however, he used to play volleyball in a non-professional team for many years. There was no morning stiffness, fever, or weight loss. Physical examination was unremarkable; the patient looked healthy with no dysmorphic features. He had no family history of musculoskeletal disease. X-ray revealed multiple, well-defined, circular, and ovoid sclerotic lesions in the femur, tibia and fibula of both legs (Fig. 1). Lesions were symmetrical, clustered around the joints, involving the epiphyses and metaphyses with a predominantly longitudinal alignment. Patient's serum calcium, phosphates, and alkaline phosphatase levels were within normal limits. The patient denied further imaging studies (bone scan/multiple X-rays). Review of a previous x-ray of his right shoulder taken 4 years ago for traumatic injury demonstrated similar lesions in the humerus. The bone lesions are typical of osteopoikilosis, or spotted bone disease. Paracetamol and nonsteroidal antiinflammatory drugs (NSAIDs) were suggested for pain control.

Discussion

Osteopoikilosis is a rare, benign skeletal disease. It is characterized by an abnormality in the endochondral bone maturation process. Osteopoikilosis is an autosomal dominant hereditary dysplasia or can be sporadic. Osteosclerotic dysplasia of bones develops during childhood and persists throughout life. The characteristic



Fig. 1 Anteroposterior X-ray view of the knees showing symmetrical well-defined, small punctuate, and ovoid sclerotic lesions affecting the femur—tibia and fibula bilaterally—typical for osteopoikilosis.

radiologic feature is multiple, sclerotic, and round or oval foci, symmetrically distributed in periarticular areas within the epiphyseal and metaphyseal regions [1]. Osteopoikilosis can be an isolated skeletal anomaly or may occur in association with cutaneous nevi called dermatofibrosis lenticularis disseminate as a component of Buscke–Ollendorff syndrome [2]. Patients are usually asymptomatic and diagnosis is an incidental finding on radiographs. However, as many as 20% may have mild articular pain and joint effusion. Long tubular bones are commonly affected [1]. The main differential diagnoses are osteoblastic metastasis, mastocytosis, tuberous sclerosis, malorheostosis, synovial chondromatosis, and osteopathia striata [2]. The symmetric distribution and lack of bone destruction as well as tendency to spare vertebral bodies and ribs are highly suggestive of osteopoikilosis. If needed, bone scintigraphy can distinguish osteopoikilosis from osteoblastic metastasis [3]. Knowledge of the pathognomonic radiographic features

Theodora Simopoulou dora.simopoulou@gmail.com

¹ Clinic of Rheumatology and Clinical Immunology, University Hospital of Larissa, Larissa, Greece

of osteopoikilosis can avoid unnecessary investigations and misdiagnosis [2].

Compliance with ethical standards

Disclosures None.

Statement of informed consent/human and animal rights and informed consent Written informed consent for publication of the clinical image was obtained from the patient.

References

- Carpintero P, Abad JA, Serrano P, Serrano JA, Rodríguez P, Castro L (2004) Clinical features of ten cases of osteopoikilosis. Clin Rheumatol 23(6):505–508
- Woyciechowsky TG, Monticielo MR, Keiserman B, Monticielo OA (2012) Osteopoikilosis: what does the rheumatologist must know about it? Clin Rheumatol 31(4):745–748
- Tuncel M, Caner B (2012) Osteopoikilosis: a major diagnostic problem solved by bone scintigraphy. Rev Esp Med Nucl Imagen Mol 31(2):93–96

Publisher's note Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.