Clinical note
Osteopoikilosis: a major diagnostic problem solved by bone scintigraphy
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Abstract
Osteopoikilosis (OPK) is a rare disease with an unknown etiology. Although a benign condition, it may lead to diagnostic problems when the patient undergoes diagnostic imaging of the skeletal system due to various reasons like malignancy. Herein, we report 2 cases with OPK causing difficulties in the final diagnosis of the cases which was resolved with the contribution of bone scintigraphy and clinical follow-up.

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Osteopoiquilia: un importante problema diagnóstico solucionado con una gammagrafía ósea

Resumen
La osteopoiquilia es una enfermedad rara aunque benigna de etiología desconocida que puede causar problemas diagnósticos cuando se hace diagnóstico por imágenes del sistema esquelético por diferentes motivos como la malignidad. Presentamos dos casos clínicos con osteopoiquilia en las cuales las dificultades para el diagnóstico final de los casos se solucionaron con la contribución de la gammagrafía ósea y el seguimiento clínico.

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Introduction
Osteopoikilosis (OPK) is a rare disease with an estimated incidence of 1/50,000.1–3 It is transmitted as an autosomal dominant pattern with an unknown etiology, although recent studies have identified some genetic defects as the cause of OPK.4,5 Several clinical abnormalities associated with OPK have been reported, including rheumatoid arthritis, synovial chondromatosis, and more frequently, dermatofibrosarcoma (such as dermatofibrosis lenticularis disseminata, keloid formations and scleroderma-like lesions).3,6 Most common roentgenographic findings are numerous, well defined circular or ovoid sclerotic lesions usually in symmetric distribution.1 Radiologically, the differential diagnosis of OPK includes osteopathia striata, melorheostosis, tuberous sclerosis, sclerotic bone metastases (especially from prostate cancer) and osteoma.

Although OPK is a benign condition, it may lead to diagnostic problems when the patient had diagnostic imaging of skeletal system due to various reasons like malignancy. This paper presents 2 cases with OPK causing difficulties in the final diagnosis of the cases which was resolved with the contribution of bone scintigraphy and clinical follow-up. Both cases have been found incidentally on CT taken for other purposes than OPK.

Case report
Case 1
A 16 years old patient with a history of back pain underwent whole body bone scintigraphy, followed by pelvic SPECT-CT imaging to increase the accuracy. Bone scintigraphy and pelvic bone SPECT images were normal. However the CT portion of the pelvic SPECT-CT and X-ray survey revealed numerous sclerotic foci in carpal, tarsal and pelvic bones without any corresponding uptake on bone scan (figs. 1 and 2). Regarding the patient’s age, absence of known malignancy, normal physical examination as well as normal bone scintigraphy and the distribution of the lesions on CT, OPK was considered as the most likely etiologic of sclerotic foci and the patient was followed accordingly.

Case 2
A 77 years old patient with complicated gastric ulcer underwent CT before surgery. Abdominal CT revealed gastric wall thickening and an enlarged (7.8 × 8.5 cm) and heterogenous prostate gland and presence of multiple sclerotic lesions in pelvic bones and vertebras which were suspicious for metastases (figs. 3 and 4).

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Figure 1. A) Normal whole body bone scintigraphy and B) Multiple, symmetric sclerotic lesions in iliac bones at pelvic SPECT-CT.

Figure 2. A: Pelvic SPECT-CT and B: X-ray images showed sclerotic foci in bilateral femurs and carpal bones.

Figure 3. Abdominal CT images showed multiple sclerotic lesions in vertebras (A) and pelvic bones (B).
His PSA level was close to the upper limit: 3.79 ng/ml (0–4) and he had no associated skin conditions and bone deformities. He had no prior diagnosis of OPK. In order to find out the etiology of the sclerotic lesions, patient underwent whole body bone scintigraphy which was interpreted as normal except bladder diverticula (fig. 5). Later, he had transurethral biopsy of prostate which was interpreted as benign prostate hyperplasia (BPH). The patient was accepted as OPK and followed accordingly.

**Discussion**

OPK is a rare sclerosing bone dysplasia of unknown etiology. It is characterized by the presence of multiple and often symmetrical foci of dense spots in the spongy bone tissue or in the inner bone cortex. The major differential diagnostic considerations in cases of widespread focal round or oval radiodense lesions are osteoblastic metastases.\(^1\)\(^2\) In these situations bone scintigraphy plays a crucial role in distinguishing OPK from primary bone tumors or osteoblastic bone metastases. Scan findings are usually normal in patients with OPK but may reveal slightly increased activity, as reported by An YS et al reflecting active osseous remodeling.\(^7\)\(^8\) In both cases presented in this paper, the lesions of OPK have been found incidentally. In case 1, X-rays and SPECT-CT images showed the numerous, mostly symmetric, sclerotic foci in tarsal and carpal and pelvic bones without any corresponding osteoblastic activity on bone scan (figs. 1 and 2). Metastases generally have a predilection for vertebrae, ribs and the diaphysis of long bones with rare involvement of the carpal or tarsal bones.\(^9\) The distribution of the lesions as determined by CT and X-ray helped the final diagnosis of OPK. Although the case 1 demonstrated the typical radiographic features of osteopoikilosis (figs. 1 and 2) Case 2 was rather complicated. There were two features of Case 2 making it difficult to reach a final diagnosis: First was the lumbar spine involvement of OPK. It has been reported that the involvement of the ribs, clavicles, spine and skull is rare and less marked.\(^1\)\(^3\) Moreover, these lesions may increase or decrease in size and number or even disappear on follow-up. Due to such changes over time, the presentation may be more atypical in elderly as happened in our patient. The second difficulty of Case 2 was the presence of enlarged heterogeneous prostate on CT which further increased the suspicion of metastases from prostate cancer. Considering above findings, sclerotic bone metastasis was thought among the possible explanations for the sclerotic lesions on CT. However normal bone scintigraphy helped to exclude the metastatic origin. Normal bone scintigraphy with sclerotic lesions at CT can also be explained by hormonotherapy in prostate cancer patient, however normal PSA levels without hormono or chemotherapy and biopsy reported as BPH further supported the diagnosis of OPK. The patients were followed up 10 months with no signs of malignancy. The OPK lesions may create another problem as indicated by Kennedy JG et al.\(^10\) Other than leading to false positive interpretation for bone metastases, these lesions may mask osteoblastic metastasis in patients with breast cancer and prostate cancer. The clinical and scintigraphic correlation is crucial in patients with OPK to avoid such misinterpretations.

**Figure 5.** A) Whole body bone scintigraphy were normal except activity protruding from bladder (black arrow) which is compatible with bladder diverticula as seen on CT scan (B: white arrows).
Conclusion

OPK is a rare bone dysplasia. Although the disease is benign in nature it may create problems to reach an accurate diagnosis in some cases. The characteristic imaging findings especially bone scintigraphy may help to reach the final clinical decision.

Conflict of interests

The authors declare not to have any conflict of interests.

References