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Mysterious but not serious?

Gayle Hann,¹ Stefanie Catherine Thust,² Emma Devereux¹¹Paediatrics Department, North Middlesex University Hospital NHS Trust, London, UK;²Radiology Department, Guy's and St Thomas' NHS Foundation Trust, London, UK

Correspondence to Dr Gayle Hann, gaylehann@yahoo.com

DESCRIPTION

A previously well 13-year-old boy presented with a 3-month history of right-sided hip pain responding to

Figure 1 Pelvic x-ray demonstrating multiple sclerotic lesions in both femora and pelvis.

Figure 2 X-ray of the asymptomatic hip featuring multiple sclerotic lesions in the proximal left femur and adjacent pelvis.

simple analgesia. Physical examination and blood tests were unremarkable. A pelvic x-ray (figure 1) revealed multiple sclerotic lesions in both femora, worse on the asymptomatic left side (figure 2) with further lesions in the pelvis. Review of a previous x-ray taken for minor trauma 2 years earlier demonstrated similar lesions in the right humerus. Following discussion with radiology, a diagnosis of osteopoikilosis was made. Osteopoikilosis (osteopathia condensans or osteosclerosis fragilis congenita) is an osteosclerotic dysplasia, seen in males and females of all ages but rarely before 3 years of age.¹ Inherited and sporadic cases have been identified as well as a possible link to dwarfism and dystocia. Genetic studies suggest autosomal dominant inheritance with possible increasing penetrance.² The radiographic findings of osteopoikilosis consist of multiple 3–10 mm, well-defined sclerotic foci² often symmetrically distributed and usually clustered in epiphyses and metaphyses of tubular long bones. Cases of osteosarcoma, chondrosarcoma and giant cell tumours in patients with osteopoikilosis have been reported, although no definite association has been established.¹ While the imaging appearances of osteopoikilosis are pathognomonic, other conditions may give rise to similar appearances. These include melorheostosis, osteopathia striata, enostoses, mastocytosis, tuberous sclerosis but also primary bone malignancy and metastases. Importantly, 25% of osteopoikilosis patients also manifest cutaneous lesions (dermatofibrosis lenticularis disseminate).¹ Knowledge of the pathognomonic radiographic features of osteopoikilosis can avoid multiple investigations as well as unnecessary anxiety in patient and physician alike.³

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Competing interests None.

Patient consent Obtained.

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