

OSTEOPOIKILOSIS: AN INCIDENTAL FINDING IN A PATIENT WITH OLECRANON FRACTURE

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Introduction

Osteopoikilosis (osteopathia condensans disseminata, spotted bone; OPK) is a rare bone dysplasia. The skeletal changes of osteopoikilosis were first described by Stieda in 1905 followed by Albers-Schoenberg (1915). It has an autosomal dominant inheritance, and can be observed equally in both sexes and in all age groups. It is an uncommon disorder with an estimated incidence of 1/50,000.¹ The etiology and pathogenesis remain unclear. It is usually asymptomatic and is a coincidental finding at radiographic examination. However, mild joint pain and swelling may be seen in 15–20% of cases. The most common localization of these lesions is in the phalanges of the hand, carpal bones, metacarpals, metatarsals, foot phalanges, pelvis, femur and humerus. There is no involvement of the clavicles, ribs or skull, typical of osteopoikilosis. In the affected bone these lesions are seen in the epiphysis and metaphysis. These multiple, sclerotic, roundish lesions vary in size from a few millimeters to several centimeters. They are scattered within the cancellous bone and concentrated near the joint surface.² In 25% of the cases osteopoikilosis is associated with skin manifestations (dermatofibrosis lenticularis disseminata, keloid formation, plantar and palmar keratomas), rheumatic and skeletal disorders (arthritis, exostoses, osteitis condensans ilii, melorheostosis, spinal stenosis, chondrosarcoma, osteosarcoma), organ anomalies (aorta coarctation, double ureter, growth abnormalities, hare lip, dental abnormalities), and endocrine dysfunction (diabetes mellitus).³ Histologically, osteopoikilosis is defined as focal condensations of compact lamellar bone within the spongiosa and is related to other osteosclerotic disorders like osteopathia striata, fibrous dysplasia

and melorheostosis. This disorder may resemble osteoblastic metastasis, mastocytosis and tuberous sclerosis.⁴

Case Report

A 33 years old man was presented to us in the emergency room with the history of road traffic accident. He slept from motor bicycle and sustained injury in his right elbow. He has no associated other injury. After usual pain management, we offered radiograph of injured elbow.

On reviewing his radiographs, in addition to olecranon fracture; diffusely distributed round sclerotic foci were also seen (Fig.1). Patient was asymptomatic before RTA. He was initially managed with back splint. In order to confirm the diagnosis, additional radiographs were performed that included, shoulder (Fig. 2) and pelvis (Fig. 3). These entire radiographs revealed diffusely distributed round sclerotic foci. Bone scintigraphy for whole body with Tc-^{99m} was normal. In order to exclude any other pathology laboratory work up was also done that included complete blood count, sedimentation, C-reactive protein, rheumatoid factor, thyroid, renal, liver function tests, calcium, phosphorus, alkaline phosphatase, parathyroid hormone, urinalysis that were all normal. Bence Jones protein and tumor markers were negative. In order to confirm the autosomal dominant inheritance of these lesions, the medical histories of the closest family members were taken and radiographs of the wrist were also done. We found no signs of similar lesions. We thus concluded it was a sporadic case. Finally the diagnosis of osteopoikilosis was made. Lastly the fracture was fixed with tension band wiring (Fig. 4).

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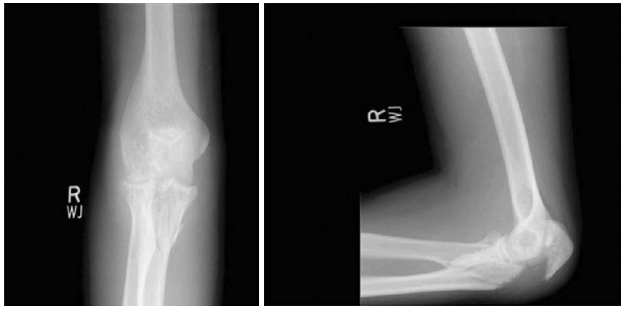


Figure 1: AP and lateral radiographs of elbow showing fracture of olecranon process and associated multiple small, round sclerotic lesions of variable sizes.



Figure 2: Radiograph of the shoulder showing multiple ovoid radiodense sclerotic foci of the proximal humerus and scapula adjacent to the glenohumeral joint.



Figure 3: AP pelvic radiograph showing numerous round or oval-shaped sclerotic foci of varying size in both proximal femora and in iliac bone, predominantly in regions adjacent to sacroiliac and hip joints.



Figure 4: AP and lateral radiographs of elbow after open reduction and internal fixation with tension band wiring.

Discussion

Osteopoikilosis, osteopathia condensans disseminata, is an osteosclerotic dysplasia with an unclear etiology and pathogenesis which takes place during childhood and persists for life. The pattern of inheritance is autosomal dominant, although sporadic cases do exist, as in our case (since no family member had the disease).^{1,2,3} The diagnosis is usually made incidentally after radiographic review is done for another reason as in our case for elbow injury, assessing numerous small, well-defined, homo-geneous, circular-to-ovoid foci of increased radio density clustered in periarticular osseous regions. Sites of predilection include phalanges (100%), carpal bones (97.4%), metacarpals (92.5%), and foot phalanges (87.2%), metatarsals (84.4%), tarsal bones (84.6%), pelvis (74.4%), femur (74.4%), radius (66.7%), ulna (66.7%), sacrum (58.9%), humerus (28.2%), tibia (20.5%), and fibula (2.8%).⁵ The ribs, skull and vertebrae are typically spared as in our case. Typically patients are asymptomatic as is the case of our patient but 15–20% may have slight articular pain and joint effusion. The number of sclerotic lesions in a single bone range from 1 to 1,000 and they can both increase and decrease in number, size and radiodensity or even disappear eventually. Histologically, osteopoikilosis is defined as focal condensations of compact lamellar bone within the spongiosa.⁶

The major differential diagnostic considerations in cases of widespread focal round or oval radiodense lesions are osteopoikilosis, osteoblastic metastases, mastocytosis, tuberous sclerosis and synovial chondromatosis. The symmetric distribution, the predisposition for epiphyseal and metaphyseal involvement, and the uniform size of the lesion are features that suggest osteopoikilosis.⁷ A bone scan is essential in distinguishing osteopoikilosis from primary bone tumors or osteoblastic bone metastases. Scan is usually normal in patients with osteopoikilosis, but may show slightly increased tracer uptake.⁸ Bone scan for whole body with Tc-^{99m} was normal in our patient. Several other rheumatologic diseases can coexist with OPK, such as rheumatoid arthritis, lupus erythematosus, reactive arthritis, ankylosing spondylitis and familial mediterranean fever^{3,4,6}, but these were absent in our patient. Various developmental abnormalities have been reported to be associated with osteopoikilosis, including: coarctation of the aorta, double ureter, pubertas praecox, urogenital defects, growth

abnormalities, peptic ulcer, diabetes mellitus at the endodermal strata level, arthritis, exostoses, osteitis condensans ilii, Klippel-Feil syndrome, melorheostosis, spinal stenosis, cervical myelopathy, dacryocystitis, giant cell tumor, fibrous dysplasia, chondrosarcoma, osteosarcoma, synovial chondromatosis at the mesodermal level; facial abnormalities, hare lip, dental abnormalities, dermatofibrosis lenticularis disseminata, keloid formation, and plantar and palmar keratomas at the ectodermal level.^{9,10} None of these developmental abnormalities were seen in our patient.

Since our patient had trochanteric fracture, Good fracture was observed in our case despite presence of these lesions. Multiple cases have been reported in literature which describe no adverse effect of osteopoikilosis on fracture healing. In the five cases of osteopoikilosis observed by Sim E, no delay of bony union was observed. Sim concluded that there was no negative influence of osteopoikilosis on fracture healing.¹¹

In another case report by Buyukbebeci O et al, radiological features of osteopoikilosis were found in a patient with subtrochanteric femoral fracture. Healing with callus formation occurred in this patient within normal time duration.¹²

Similarly femoral neck fracture in another patient with osteopoikilosis was reported to be successfully treated with partial femoral prosthesis by Zahar A et al.¹³

Moreover healing within normal time frame in cases of fractures of the hand has also been reported in literature, in patients with concomitant osteopoikilosis.¹⁴ Osteopoikilosis thus can be termed to have no deleterious influence on fracture healing.

Conclusion

In conclusion, whenever uniform multiple radio opaque lesions are found on radiographic examination, osteopoikilosis must be in the differential diagnosis before invasive diagnostic procedures and dangerous and unnecessary treatments are planned.

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