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Case Report

Osteopoikilosis - Case report

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Key words: osteopoikilosis; osteopecilia; bone density; spotted bone; cancellous.

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Abstract

Osteopoikilosis is a rare bone anomaly usually benign which may be confused with other severe primary diseases or dangerous metastatic bone lesions. We present one new case describing typical radiological findings with no clinical signs or other associations. No malignant transformation or other complications happened in the last ten years.

Osteopoikilosis is found in most cases incidentally, and in most cases is not associated with other serious medical complications. Screening of family members may be considered.

Introduction

Osteopoikilosis is a typical rare bone disorder related to bone architecture characterized by the presence of small, multiple round or ovoid areas of increased bone density. Albers-Schönberg in 1915 made the first known description in literature [1]. The disorder is a hereditary anomaly transmitted in autosomal dominant trait [2, 3, 10, 11]. The degree of genetic penetration is relatively high. The origin is unknown. Biochemical studies are generally normal. There are no clinical signs, and the disorder is discovered casually from X-rays taken for other purposes. Bone scintigraphy is normal in patients with osteopoikilosis The spongiosa is widely affected in cancellous or even in tubular bones.

Case Report

The patient presented here is a 42-years old male complaining of acute ankle pain after daily long walking. He was a village teacher walking about 10 km daily. The routine X-ray has taken showed unusual findings which were interpreted as some kind of malignancy from the alarmed family practitioner. The local radiologist specialist was not enough experienced with such rare findings so send him to our consulting office (Fig. 1).

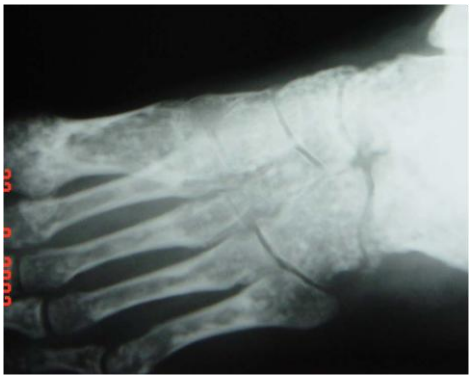

Figure 1: X-ray of metatarsal and tarsal ones of left foot showing small sclerotic areas in all five metatarsal bones and tarsal bones also. Note that sclerotic areas are more intense in spongisa in epi-metaphysis.

Pelvic (Fig. 2) and skull x-rays were taken and foot X-ray were repeated (Fig. 3). After a serious evaluation the diagnosis of osteopoikilosis was established. Biochemical analyses including erythrocyte sedimentation rate, alkaline and acid phosphatase, leukocytes were quite normal.

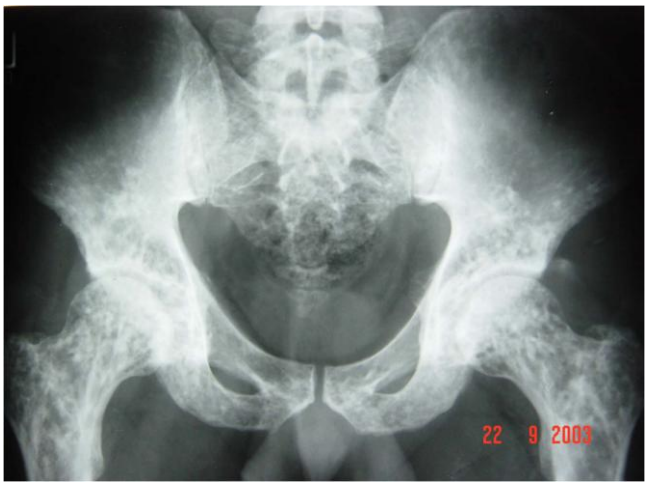

Figure 2: Pelvic X-ray with multiple sclerotic round shape areas especially around the acetabular dome and femoral neck and head. Trochanteric region in widely affected. The iliac bones have little involvement above the dome. The sacrum also shows dense sclerotic zones.

In reality the ankle pain was tendinitis of anterior tibial tendon. No joint swelling was found. Metatarsal and tarsal bones were free of pain. No local edema or tenderness around the foot was seen objectively. Anti-inflammatory drugs, rest, ice packages were prescribed which resulted in complete relief from pain within a week after the visit. No other metabolic disorders were found. The screening of other family members did not result in other cases of osteopoikilosis. The patient is under periodic annual follow-up for about ten years with no signs of malignant transformation. No more clinical complaints are found.

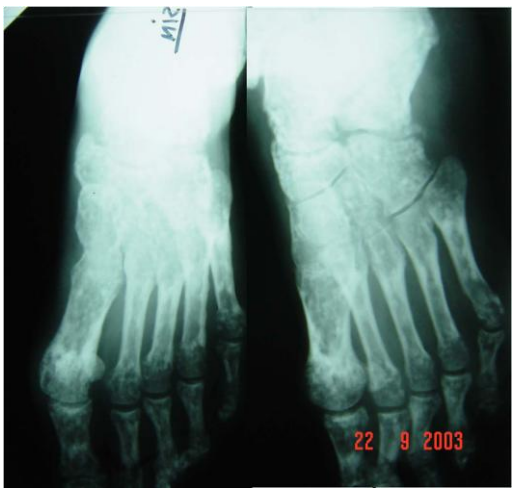

Figure 3: Both feet showing the same radiological view (repeated by us). One can note the similarity of “spotted bone” image with Madura Foot disease, with the differences that are signs of bone erosions and destruction.

Discussion

Radiological there are small areas of sclerosis ranging in size from some millimetres to 1 centimetre [1-5, 10]. These are round or ovoid in shape in proximity with normal cancellous zones. It is believed not to advance, but change in shape may happen. The lesions appear early and persist throughout the life [2, 4-10]. The age distribution is between 15-60 years and only rarely are described geriatric cases. There are no reports of complete resolutions of sclerotic foci. Late remodelling may happen in some foci. The major sites of involvement include long tubular bones, carpal bones, tarsal bones, pelvis, sacrum, and scapulae [2]. The ribs, clavicle, spine, and skull are typically spared. The typical radiologic view is that of “spotted bone”. The long bones are affected in the epi-metaphyseal region especially hands and feet [6]. The disorder is known also as: *Osteopathia Condensas Disseminata, Osteopecilia, Osteodermatopoikilosis, Osteosclerosis Generalisata, Spotted Bones.*

This anomaly is a radiologic curiosity more than real disease because rarely this disorder is accompanied with anomalies needed medical treatment. The anomaly has genetic nature. Scleroderma, palatoschisis, syndactyly, dwarfism and melorheostosis may be assoccaited with osteopoiklosis. The disorder is not symptomatic, but incorrectly diagnosed patients may have expensive studies for other important and dangerous conditions including metastatic lesions of the skeleton [8].

Patients with joint pains especially in upper limbs are described by Paraskevas et al [9]. Some times is associated with connective tissue nevus called *dermatofibrosis lenticularis disseminata* or the Buschke – Ollendorf syndrome [10]. In such cases the dermatosis shows small asymptomatic papules. Rarely are white or yellow discs or plaques.

Malignant complication or association with osteopoikilosis is rarely reported. Osteosarcoma [11], giant-cell tumor [12] chondrosacoma [13] are described.

Differential diagnosis is made with Paget disease, tuberous sclerosis, multiple myeloma, metastatic bone disease, Hodgkin disease, sickle cell disease, Madura foot, leukemia etc. Osteopoikilosis is considered as one of the skeletal “Don’t touch” lesions.

Family members should be screened with a radiograph of the hand and knee and when possible with pelvic X-ray. We believe that CT-Scanning and MRI is rarely needed when there are no other clinical suspicious findings.

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