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Osteopoikilosis is a Disorder Diagnosed only by Radiology

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Abstract

Osteopoekilosis is a rare hereditary sclerosing bone dysplasia, which was described simultaneously by Albers-SchÃnberg and Ledoux-Lebard in 1916, characterized by the Individualization of multiple small, homogeneous, well-defined bony lesions of rounded or oval form in the juxta-articular regions.

Keywords: Osteopoikilosis; Sclerosing bone dysplasia

Clinical Image

Osteopoekilosis is a hereditary condition, which was described simultaneously by Albers-Schonberg and Ledoux-Lebard in 1916. Osteopoekilosis is a rare hereditary sclerosing bone dysplasia [1]. Transmitted in an autosomal dominant form; the literature also describes although sporadic forms [2].

With a sex ratio of can be seen at any age. The appendicular skeleton is affected, whereas the axial skeleton is rarely affected [2]. in Osteopoikilosis, we objective the presence of numerous 2-10 mm oval or spherical densities form in the metaphysis and epiphysis of the long bones appearing in childhood and last throughout life [2]. Individualization of multiple small, homogeneous, well-defined bony lesions of rounded or oval form in the juxta-articular regions is a characteristic radiographic sign of Osteopoekilosis (Figures 1 and 2).

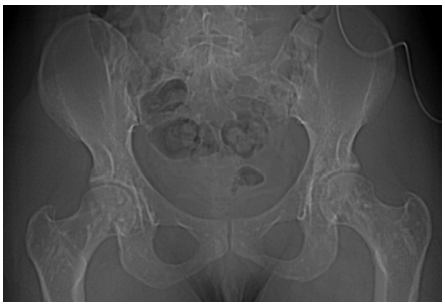

Figure 1: Frontal X-ray of the pelvis, showing multiple sclerosing bones lesions at the femoral heads, acetabulum, symphysis pubis, and ischium.

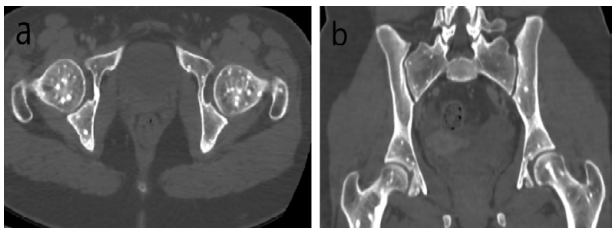

Figure 2: Abdominal-pelvic CT scan in axial section and coronal reconstruction showing dense, well-defined, and homogeneous round and oval islands at the sacrum, acetabulum, iliac wings, and femoral heads in Osteopoikilosis.

The most frequently affected sites are the epiphysis and metaphysis of the long bones, the carpus and tarsus bones, the pelvis, and the scapulae [1]. Classically, affected individuals are asymptomatic, but 15-20% describes the pain and joint effusions Osteopoekilosis may present either as an isolated anomaly or associated with other abnormalities, most commonly with skin lesions of the disseminated lenticular dermatosis type, the association of which is defined as Buschke-Ollendorff syndrome [1]. Sometimes the condition is associated with renal or cardiac malformations or endocrine disorders. Several diseases may coexist with Osteopoekilosis, such as rheumatoid arthritis, systemic lupus erythematous, reactive arthritis, ankylosing spondylitis, psoriatic arthritis, familial Mediterranean fever, dacryocystitis, scleroderma, fibromyalgia, and de Quatrain's syndrome [2].

Differential diagnoses include osteoplastic metastases, tuberous sclerosis, mastocytosis, and synovial chondromatosis [1]. Most patients are asymptomatic and do not require treatment; however, pain relief is necessary for symptomatic patients [2].

References

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