Monostotic paget’s bone disease of the pelvis

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Description

A 63-year-old woman, recently diagnosed with right breast cancer, was referred for a CT scan for extension workup. The CT scan revealed an enlarged right coxal bone with cortical thickening, thick trabeculae and multiple areas of sclerosis, responsible for cortico-medullary dedifferentiation, respecting the joint space confirming the diagnosis of Paget’s disease in the right hemi-pelvis (Figure 1). She was asymptomatic, and her serum alkaline phosphatase was slightly elevated at 182 U/L (normal values: 30-120 U/L).

Discussion

Paget’s disease is a chronic, non-inflammatory metabolic disease of unknown origin characterised by focal lesions of bone resorption and increased, disorganised bone formation associated with marrow fibrosis and increased vascularisation [1].

Usually asymptomatic, but occasionally symptomatic, diagnosed at the bone remodeling stage where bone distortion and expansion lead to musculoskeletal events, bone fragility, neuromuscular or cardiovascular complications, or a sarcomatous transformation [2].

PDB has a predilection for the axial skeleton, affecting the pelvis in 21-75% of cases. However, its monostotic form is the least frequent (10-35%) [3].

Paget’s disease usually begins in one site and progresses throughout the bone, without extending to adjacent bones [4].

Conventional radiology and CT can show distinctive radiological signs of each phase.

The initial phase is characterized by geographic osteolysis with an increased resorption border.

The intermediate phase of osteosclerosis where Paget’s disease is most often diagnosed is marked by cortical thickening, bone expansion, cortico-medullary dedifferentiation due to the creation of a new endosteal bone impinging on the medullary cavity with accentuated trabecular marks (Figure 1).

In the late osteosclerotic form, excessive bone neoformation is apparent, manifesting as a diffuse increased bone density and size [5].

Bone turnover markers, especially alkaline phosphatase, reflect osteoblastic activity and can be used as a marker of therapeutic response [5].
Figure 1: Axial (a) and Coronal (b) CT of the pelvis showing right coxal cortical thickening, intra trabeculation with coarse thick trabeculae, bone expansion, and multiple areas of sclerosis, (arrow) consistent with Paget's disease.

Declarations

Author’s contributions: All authors contributed to this work. All authors have read and approved the final version of the manuscript.

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References


