Paget Disease of the Bone Presenting as Leontiasis Ossea

Carvalho Ruben*, Pereira Inês and Sapeira Isabel
Hospital Distrital de Santarém, Imaging Department, Santarém, Portugal

*Corresponding author: Carvalho Ruben, Hospital Distrital de Santarém, Imaging Department, Santarém, Portugal, Tel No.: +351-243400200; E-mail: rubenpcarvalho@gmail.com


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Clinical-Medical Image

Paget disease of the bone is a chronic disorder of unknown etiology that typically results in enlarged, deformed bones due to abnormal and excessive bone remodeling. The disease which can be monostotic or polyostotic and is characterized initially by an increase in bone resorption, followed by a disorganized and excessive formation of bone, leading to pain, fractures, and deformities. The classically described radiological appearances are of expanded bone with a coarsened trabecular pattern,

Figure 1: Incidental finding on CT of the skull (axial) shows thickening of the calvarium and widening of the diploic space, features of Paget disease of the bone

Figure 2: The radiographs of the skull (frontal and lateral views) show marked diffuse hyperostosis, cortical thickening, and accentuation of the trabecular pattern of the facial and skull bones, an exuberant presentation of Paget’s osteitis deformans of the skull, historically described as Leontiasis Ossea.
but it can present differently according to the disease stage: in the active or osteolytic phase, aggressive bone resorption and lytic appearance are the hallmarks while on the quiescent and inactive stage there is a predominance of cortical thickening and sclerosis, and finally, it can have a mixed pattern presentation. The pelvis, spine, skull, and proximal long bones are most frequently affected. We depict an exuberant case of this disease with craniofacial involvement resulting in the historical eponym of Leontiasis Ossea (lion mask face) (Figures 1 and 2).