Hip pain

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Diagnosis: Bisphosphonate-related atypical subtrochanteric femoral fracture in patient with Paget disease.

Paget disease of bone is a progressive disorder characterized by an increased rate of bone turnover and remodeling leading to excessive bone resorption and disorganized bone formation [1]. Bowing deformities occur in the long bones of the extremities and pathologic fracture is a common complication, manifesting as a transverse fatigue fracture. The lateral cortex of the femur is most often affected, as the bowing deformity creates greater tensile force along the lateral aspect [2]. Three major phases of Paget disease have been described. The lytic (incipient) phase is characterized by predominant osteoclastic resorption. The mixed (active) phase features decreasing osteoclastic and increasing osteoblastic activity. The blastic (late inactive) phase demonstrates declining osteoblastic activity, and the process transitions to a quiescent state [3]. Radiographic appearance of these stages is classic. In the incipient phase of the disease, osteoclastic activity is demonstrated by a wedge-shaped radiolucency in the metaphysis and diaphysis, often described as a “flame” or “blade of grass” appearance. In the later stages, radiographs demonstrate increasing cortical and trabecular thickening in the involved bone, correlating with increasing osteoblastic activity [3]. MRI shows thickened cortical bone as a rim of low signal

intensity, with the medullary cavity containing normal fat signal [4].

Although more often asymptomatic, Paget disease may be associated with focal bone pain, for which bisphosphonate therapy is considered among the first-line treatment options [5]. Bisphosphonates suppress disease progression by down-regulating osteoclastic activity and decreasing the rate of bone turnover, as clinically evident by a decline in alkaline phosphatase and improvement in bone pain [6]. During and after treatment with bisphosphonates, Pagetic bone, like non-diseased bone, is at risk of atypical fracture. Atypical proximal femoral insufficiency fractures involve the lateral aspect of the subtrochanteric femoral neck, and are associated with long-term bisphosphonate therapy [7–9]. There is some controversy whether such fractures are a result of the disease process itself, rather than bisphosphonates [9]. Radiographic features of atypical femoral fracture include subtle transverse linear lucency with a mound-like thickening of the overlying lateral cortex, referred to as “cortical beaking”. This cortical beaking is unique to bisphosphonate-related fractures, and has not been described in insufficiency fractures occurring in Pagetic bone in the absence of bisphosphonate therapy [7, 8]. Atypical femoral fractures may be treated conservatively with limited weight-bearing, rest, and pain medication, but outcomes are poor [10]. Thus, patients will often undergo surgical intervention either with intramedullary nailing or arthroplasty [10].

The patient in this case had a history of Paget disease of the right proximal femur treated with bisphosphonate therapy. Her images demonstrate disorganized bony overgrowth involving the proximal right femur, typical of Paget disease. The subtle cortical beaking apparent along the lateral aspect of the right subtrochanteric femoral neck is suggestive of bisphosphonate-related atypical femoral fracture. On the confirmatory PD FS MRI image, there is hypointense signal at the fracture line, with surrounding hyperintense linear signal along the lateral

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femoral cortex representing focal periosteal reaction. Due to this fracture, the patient underwent right total hip arthroplasty. Follow-up radiograph shows the hardware, along with increased lucency of the fracture line.

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