Chondrosarcoma as inaugural manifestation of monostotic Paget’s disease of bone

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LETTERS TO THE EDITOR

To the Editor,

Sarcomatous degeneration is one of the serious and rare complications of Paget’s disease of bone. Osteosarcoma is the most common secondary tumour, while other variants such as chondrosarcoma are extremely uncommon. We describe a unique case of Paget’s chondrosarcoma of the pelvis in an elderly female patient, with no previous established diagnosis of Paget’s disease of bone. We emphasize clinical and radiologic aspects that should raise suspicion of malignant transformation, revealing good correlation with the final histopathological diagnosis.

INTRODUCTION

The malignant transformation of Paget’s bone disease is a rare complication, with an estimated incidence of 0.95%.1 It typically occurs in long-term disease with polyostotic distribution, presenting mostly as osteosarcoma. In contrast, chondrosarcoma is a neoplastic variant uncommonly reported in clinical cases, whose most frequent sites are the humerus, femur and tibia2–3.

CLINICAL CASE

An autonomous 84-year-old woman, with arterial hypertension, was observed in the emergency department due to disabling left hip pain for 3 months, with no history of previous trauma. Physical examination revealed oedema of the limb up to the groin and painful hip in all planes of motion. Radiographs revealed areas of sclerosis and lytic lesions in the left iliac bone, suggestive of Paget’s disease of bone (Figure 1A). Cortical disruption in the inferior ischial ramus was noted. At follow-up, a complementary study was performed. Laboratory tests revealed elevation of alkaline phosphatase 308U/L (normal<120), lactate dehydrogenase 274U/L (normal<225) and c-reactive protein 101mg/L (normal<3). Bone scintigraphy showed intense hyperfixation corresponding to the suspected area, denouncing a monostotic involvement. Pelvis computed tomography scan (CT) revealed alteration of the bone trabeculation in the left iliac and sacrum and a massive mass centered in the left ischium with thin calcifications. The lesion extended to the hip joint and to the adductor and obturator thigh muscles, causing bulging of the pelvic organs (Figure 1B, C and D). These findings were suggestive of chondrosarcoma. Magnetic resonance imaging (MRI) showed the extensive mass (120x145x227mm) with central necrotic region and heterogeneous contrast uptake (Figure 1E). Due to suspicion of sarcomatous transformation of Paget’s disease of bone, the patient was admitted to perform a biopsy. Histological examination confirmed the presence of chondrosarcoma. No osteogenic differentiation was present. Thoraco-abdomino-pelvic CT scan revealed pulmonary metastases, bilateral hydronephrosis and thrombosis of the left common, internal and external iliac veins and right external iliac vein, in a compressive relation with the neoplastic mass. Due to rapid clinical deterioration, the complexity of a potential surgical approach and lack of conditions for chemotherapy or radiotherapy, the patient was integrated into palliative care. She died 7 months after the onset of symptoms.

DISCUSSION

Our case report highlights that the axial location of chondrosarcoma may be associated to a delay in diagnosis, since tumour growth in the pelvic space can be quite extensive until it becomes symptomatic. Also pagetic bone hypervascularity promotes early and frequent distant metastasis, which contributes to its aggressive course and unfavourable vital progno-
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Furthermore, although the pelvis is the preferred location of Paget’s disease of bone and osteosarcoma, there are no documented cases of Paget’s chondrosarcoma in that region, either in the isolated or polyostotic form.

No laboratory parameter can be seen as a consistent indicator of malignant transformation due to the pronounced variation of alkaline phosphatase levels, however some characteristic imaging features can raise that hypothesis. Cortical breakthrough in the affected bone, as observed in the described case, and a soft tissue mass are cardinal radiologic findings suggestive of malignancy. CT and MRI scans are helpful to better delineate a suspected pagetic site and to differentiate a subtype according to the lesion matrix. Nevertheless, biopsy is critical to establish a definitive diagnosis and to characterize the histological subtype, since not all tumors associated with Paget’s disease of bone are malignant, such as giant cell tumor which, although extremely rare, is associated with a longer survival.

CONCLUSION

Paget’s disease of bone can often remain silent until secondary complications arise. In addition to the rarity of the manifestation and the histological variant, this seems to be, to our knowledge, the first case of Paget’s chondrosarcoma located in the pelvis described in the literature.

REFERENCES


FIGURE 1. A) Anteroposterior view of pelvic radiograph showing lytic and sclerotic lesions involving the left iliac bone, consistent with Paget’s disease and cortical disruption in the inferior ischial ramus (arrows). Pelvis CT (B, C, D) reveals a coarsened trabecular pattern and cortical thickening in the left iliac and sacrum suggesting Paget’s disease (black arrows) and a massive destructive mass, centered in the left ischium with thin calcifications and a large soft tissue component suggestive of chondrosarcoma (white arrows). T1-weighted MRI image (E) shows the extensive mass (120x145x227mm) (white arrows) which extends to the hip joint and to the adductor and obturator thigh muscles, causing bulging of the pelvic organs.