Disappearing hemipelvis: Low grade osteosarcoma, an unusual and poorly described variant of Paget's Sarcoma

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ABSTRACT

Paget's sarcomatous transformation is a rare and potentially fatal complication of Paget's disease. Histologically, it is typically described as a high-grade and extremely aggressive malignancy. We present an unusual radiographic series from a patient diagnosed with a low-grade Paget's osteosarcoma, a very rare and poorly described variant of the disease.

1. Introduction

Paget's sarcomatous transformation is a rare and potentially fatal complication of Paget's disease. It accounts for approximately 5% of all osteosarcomas.¹ Histologically, it has been described as a high-grade and extremely aggressive malignancy. We present the unusual radiographs of a patient diagnosed with a low-grade Paget's osteosarcoma, a very rare and poorly described variant of the disease.

In this radiographic review, we illustrate the unusual features of localised low-grade Paget's osteosarcoma of the pelvis and proximal femur, with lytic resorption and complete loss of the hemi-pelvis. We also identify the histological features that are consistent with low-grade Paget's Osteosarcoma.

2. Case presentation

A previously healthy 73-year-old man presented to the orthopaedic department for management of an ankle fracture. During an admission to hospital for this, he raised concern regarding ongoing left hip pain. Our patient detailed a prolonged history of gait disturbance and discomfort on weight-bearing, but described no pain at rest or at night. Subsequent radiographs demonstrated a lytic lesion on the left side of his pelvis, involving the ischium (Fig. 1). A bone scan showed evidence of markedly increased uptake at this site.

With the exception of gastroesophageal reflux disease and hypertension, no significant past medical history was noted. The patient did not report a family history of Paget's disease, however as he was adopted an accurate assessment was difficult to obtain.

Given the concerning areas of lysis and activity on bone scan, open biopsy was organised. The histological appearance was suggestive of active Paget's disease (Photo 1), but with no obvious evidence of malignancy. In spite of this, additional biopsies were organised, owing to a strong clinical suspicion of underlying malignancy.

A second core biopsy included new woven bone and showed irregular trabeculae of pre-existing lamellar bone undergoing aggressive osteoclastic resorption. The intervening marrow was almost completely replaced by fibro-connective tissue and areas of haemopoiesis. Again there were no obvious features of malignancy.

The patient was placed under radiographic surveillance. The radiographs taken over the five years since his initial presentation (Figs. 2–6) record the unusual progression of the disease. Subcapital fracture was followed by subtrochanteric femoral fracture and complete resorption of the hemipelvis and proximal femur. A third biopsy, taken three years after the initial presentation, was unlike the previous two biopsies. The morphological features were consistent with low to intermediate grade spindle cell sarcoma, with no convincing lineage of differentiation (Photo 2). Subsequently, the case was presented to the pathologists at the multidisciplinary sarcoma meeting and discussion

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Fig. 1. Radiograph AP Pelvis at presentation.

Photo 1. Histology shows bone within which there is florid osteoclastic resorption coupled with marked osteoblastic activity. There is mild peritrabecular fibrosis. The appearances are consistent with with Paget's Disease of bone. H/E stain, original magnification $\times 20$.

was held at the Scottish National Sarcoma meeting. A diagnosis of an exceptionally rare low-grade Paget's osteosarcoma was made. This is an unusual and poorly described variant of Paget's osteosarcoma, which normally presents as high-grade and extremely aggressive. Review by

the Professor of pathology at the Manchester Royal Infirmary provided consensus on the diagnosis.

Zoledronic Acid was first prescribed following diagnosis of Paget's disease. This had an good initial result with our patient improving in

Fig. 2. Radiograph AP Pelvis at 1 year post presentation.

both pain and ambulation. Further progression of the disease process lead to two further treatments with Zoledronic Acid. After still further progression the patient was commenced on three monthly injections of Denosumab however this also proved ineffective and injections were stopped. Following diagnosis of osteosarcoma, treatment options including resection, chemotherapy and radiotherapy were discussed, however the patient and the team decided non-operative symptomatic treatment was the best option.

3. Discussion

In 1877, Sir James Paget first described a case of *Osteitis Deformans*: an inflammation of the bone.² His name would become eponymous with the condition. Today the prevalence of Paget's disease is approximately 4–5%; Barker et al report a 5.4% prevalence in British adults older than 55 years.^{3–5}

Paget's disease is characterised by local disruption of normal bone

remodelling. Sarcomatous transformation is a rare event in Paget's disease.⁶ The reported incidence ranges from 0.15% to 1%.^{7,8} The conventional high-grade presentation of Paget's osteosarcoma is extensive, both locally and systemically.⁹ As such, the prognosis of high-grade Paget osteosarcoma is poor; few patients survive beyond 5 years.^{1,6,10,11}

Low-grade, also known as intraosseous well-differentiated osteosarcoma, is an intramedullary bone-producing tumour that accounts for approximately 1–2% of all osteosarcomas.¹² Low-grade osteosarcomas have been reported to have a better prognosis than high-grade variants, with slower growth and increased survival after treatment.¹³ However, the low incidence of low-grade osteosarcomas and the overlap in presentation with benign conditions often leads to a delay in diagnosis and treatment.¹⁴

Given the rarity of this presentation, it may be reasonable to question whether low-grade osteosarcoma is a form of Paget's sarcomatous transformation, or whether it is a separate disease process. A further

Fig. 3. Radiograph AP Pelvis at 2 years post presentation.

example in the literature is that of Franchi et al.¹³ Two patients presented with characteristics of Paget's disease and subsequently developed low-grade osteosarcoma. As Paget's osteosarcomas are assumed to be high-grade, the authors concluded that although the low-grade osteosarcomas had histological features suggestive of Paget's disease, it could not be Paget's sarcomatous transformation.¹³ No other cases of low-grade Paget's osteosarcoma in the literature could be found.

4. Conclusion

This is a very rare presentation of a poorly described variant of Paget's osteosarcoma. Whilst it is typically high grade and extremely aggressive, this low-grade tumour has been exceptionally aggressive in

its resorption of bone, leading to complete loss of the hemipelvis and proximal femur. We present this unique radiographic series for review by surgeons with an interest in this disorder.

Disclaimer

None.

Conflict of interest

No conflicts.

Fig. 4. Radiograph AP Pelvis at 3 years post presentation.

Fig. 5. Radiograph AP Pelvis at 4 years post presentation.

Fig. 6. Radiograph AP Pelvis at 5 years post presentation.

Photo 2. The normal bone architecture is replaced by fibroblastic tissue of low cellularity with minimal cellular atypia. Small islands of woven bone are being produced within the fibroblastic tissue. H/E stain, original magnification $\times 20$ (inset $\times 2$).

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