# SARCOMA SUPERIMPOSED ON PAGET'S DISEASE OF THE FEMUR

## A CASE REPORT

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The tendency toward sarcomatous change demarcates Paget's disease from all other metabolic bone diseases. This complication may develop in an affected bone (or even several affected bones), whether the total skeletal involvement is limited or extensive. Many writers claim that its incidence is between 5 and 10%, though Jaffe[1] stated that it is difficult to be precise about the overall incidence. Where the skeleton is heavily involved by the disease, the incidence seems to approach 10%, but if the numerous cases of non-clinical Paget's disease of limited extent are taken into account, the overall incidence is probably lower than 2 or 3%. Jaffe's estimate was confirmed by Poretta *et al.*,[2] who found only 16 cases of sarcoma among 1,753 examples of Paget's disease of bone seen at the Mayo Clinic, an incidence of 0·9%. These writers reviewed the English-language literature and collected a total of 128 cases of sarcomatous transformation in Paget's disease. Barry[3] reported that, although there is a high incidence of Paget's disease in Australians of Anglo-Saxon origin, the development of sarcoma in Paget's disease is rare in Australia; he estimated it at less than 1%.

Although Paget's disease of bone is fairly commonly encountered by the practising clinician, the occurrence of sarcomatous changes in a bone is so unusual that it excites comment when it is met. It is for this reason, and also because the malignant change apparently developed while the patient was under treatment for the far more common complication of a pathological fracture, that this case was thought to be worthy of record.

### CASE REPORT

M.M., a Coloured male, aged 58 years, was admitted to hospital complaining that he had injured his left thigh at the time of a fall. On X-ray examination (Fig. 1) it was seen that he had a pathological fracture at the junction of the middle and lower thirds of the shaft of the femur, which showed evidence of Paget's disease. This disease was also present in the skull and pelvis. No radiographs were taken of the rest of the skeleton. Four days after the injury on 10 May 1961, an open reduction was performed and a Küntscher nail was

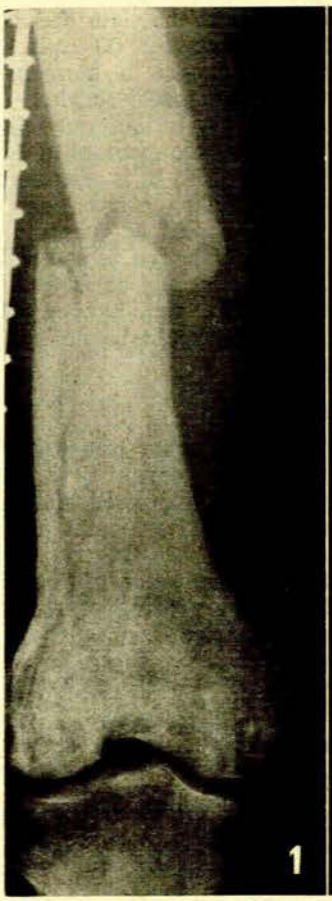


*Fig. 1.* A pathological fracture at the junction of the middle and lower thirds of a femur which is affected by Paget's disease.

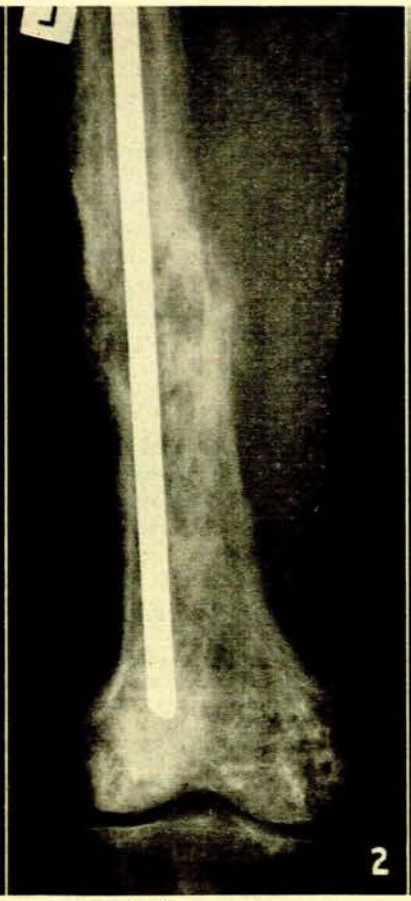


*Fig. 2.* Three months after internal fixation the fracture is uniting.

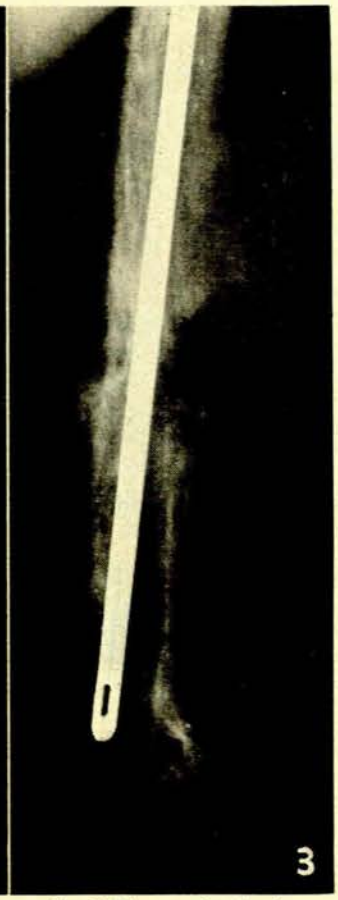


*Fig. 3.* Five months after internal fixation there is a destructive lesion of the bone at the site of the previous fracture.

inserted to maintain the reduction. The postoperative course was uneventful and the patient was discharged on 19 June, being allowed partial weight-bearing on crutches.

He was seen at the follow-up clinic on 9 July, and radiographically and clinically nothing untoward was found. By 28 August he was walking well, using one stick, and he had 90° of knee movement. The X-ray plate (Fig. 2) showed the fracture to be uniting. On 15 September he had 110° of knee movement.

He was seen again on 17 November and he now complained of severe pain of a few weeks' duration in his left thigh. Clinical examination showed a hot, fluctuant swelling enveloping the thigh at the level of the middle of the operation scar. The swelling was aspirated and 80 ml. of serosanguinous fluid were obtained. On X-ray examination a destructive lesion involving mostly the posterior aspect of the femur was seen (Fig. 3). On 22 November a biopsy was performed and microscopic examination of the sections (Fig. 4) showed a spindle-cell sarcoma with quite marked anaplasia and frequent mitotic figures. A few osteoclast-like giant cells were also present. Some new bone was found enmeshed in fibrous tissue, and this was no doubt related to the callus formation.

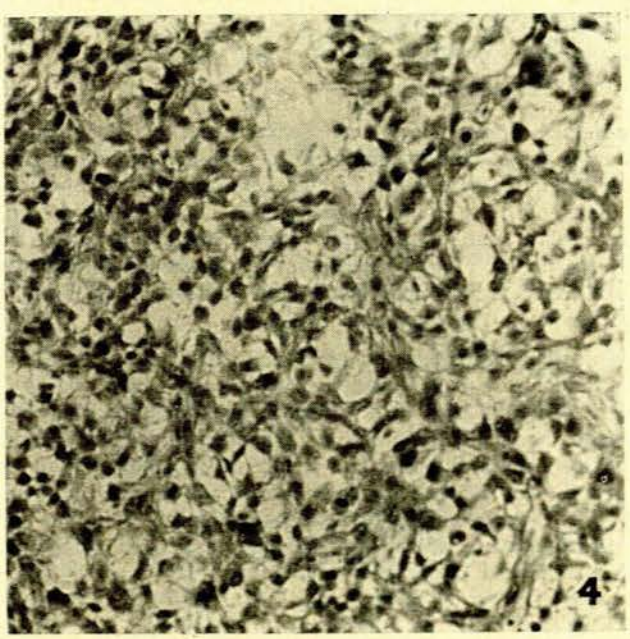


*Fig. 4.* Photomicrograph (× 150) shows a spindle-cell sarcoma with quite marked anaplasia and frequent mitotic figures. A few osteoclast-like giant cells are also present.

The patient was referred for palliative radiotherapy, but the radiotherapists considered the lesion unsuitable. Preparations were then made to perform a disarticulation through the hip-joint, but unfortunately the patient's condition deteriorated rapidly and he died on 30 December. Examination of the autopsy specimen showed marked bone destruction and a large haemorrhagic tumour-mass at the site of the previous fracture (Figs. 5 and 6).

## DISCUSSION

This patient presented with a pathological fracture of a femur affected by Paget's disease. The disease was not monostotic, but also affected the skull and pelvis; a full skeletal survey was not undertaken. Five months later, after the fracture appeared to be healed, the patient rapidly developed severe pain and considerable swelling at the previous fracture site. Radiographs suggested, and a biopsy confirmed, the presence of a sarcoma. Poretta *et al.*[2] found that, in a series of 78 patients with long-bone sarcomatous involvement, 18% had had recent fractures in the area where the malignant growth later developed. In some cases the fracture had healed before the sarcoma

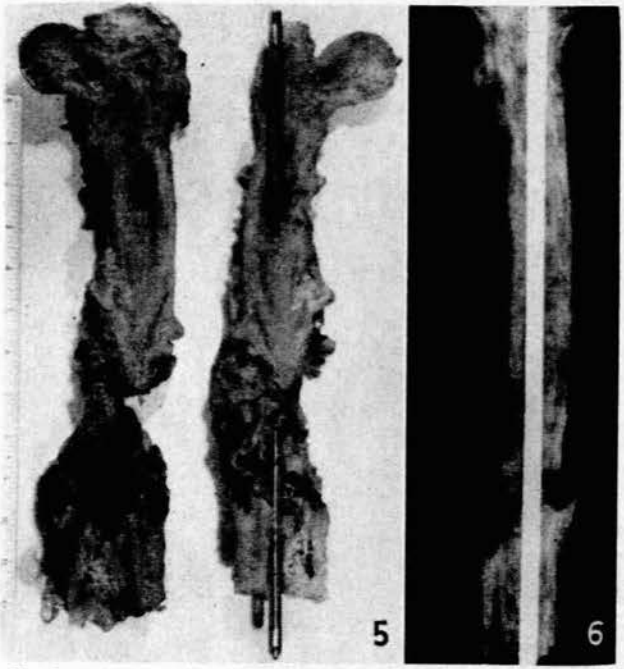


*Fig. 5.* Autopsy specimen showing a marked lytic lesion of bone and a bulky, fleshy haemorrhagic neoplasm.

*Fig. 6.* Radiograph of the autopsy specimen showing involvement by Paget's disease and the bone destruction caused by the superimposed sarcoma.

developed, and in others it had not healed. The average time interval between the fracture and the appearance of the first symptoms of the neoplastic change was 3·2 months. Lake[4] mentioned that the two peaks of incidence of bone sarcoma, viz. adolescence and the sixth decade, have two common factors of importance: (1) actively growing vascular bone; and (2) changes in hormonal balance occurring at the extremes of gonadal activity.

The prognosis of this complication is uniformly depressing; only five two-year survivals were recorded in the 128 cases reviewed by Poretta *et al.*[2] It is therefore important that severe pain and/or swelling of recent onset in any patient with Paget's disease should be considered an extremely sinister development, although sarcoma is such a rare complication. The patient should be subjected to the closest clinical, radiographic and biochemical scrutiny. When doubt persists there should be no hesitation in performing a biopsy.

SUMMARY

1. A case of Paget's disease of the femur complicated by the development of a sarcoma is presented.

2. The sarcoma was apparently superimposed after a pathological fracture had healed, and five months after the fracture occurred.

3. Although sarcoma is a rare complication of Paget's disease of bone, severe pain and/or swelling of recent onset should be considered of sinister significance.

I am indebted to my brother, Mr. Alec Singer, for his helpful criticism and for his assistance in obtaining the autopsy specimen. I am grateful to my Registrar, Dr. W. Johnson, for keeping the notes and radiographs intact. My thanks are also due to Mr. G. McManus and Mr. B. Todt for the photographs.

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3. Barry, H. C. (1961): *Ibid.*, **43A**, 1122.
4. Lake, M. (1951): *Ibid.*, **33B**, 323.