PAGET'S DISEASE AND METASTATIC CARCINOMA

A CASE REPORT

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In patients over 60, both Paget's disease and metastatic carcinoma are not uncommon, but it is rare for both to occur in the same patient and in the same bone. One such patient is reported; she had primary carcinoma of the vulva which metastasised to a femur already affected by Paget's disease.

In February 1983 a 67-year-old woman presented with a skin lesion in the posterior triangle of her neck 11 months after a radical vulvectomy for vulval carcinoma (Fig. 1). While she was in hospital awaiting excision of this lesion, her leg gave way unexpectedly and she was found to have a pathological fracture through an area of Paget's disease in the mid-shaft of the femur (Fig. 2).

Radiographs of her chest showed no abnormality. Except for the femoral lesion, a bone scan was normal, as was a liver scan. Blood tests showed an ESR of 39 mm in the first hour, an alkaline phosphatase of 375 iu/l and a serum calcium level of 2.54 mmol/l.

At an operation for open reduction and intramedullary nail fixation a firm tumour was found at the fracture site. This involved the medullary canal, had spread 8 cm along the cortex and extended a little through it. Histological examination of biopsies showed Paget's disease with a coexistent well-differentiated squamous cell carcinoma (Figs 3 and 4) which had metastasised from the previously removed vulval lesion; the lesion in the patient's neck was also excised and found to be another metastasis from the same primary lesion. The patient had radiotherapy but died two months later.

DISCUSSION

Sarcomatous change is a well recognised complication in about 1% of cases of Paget's disease, but in advanced disease it is estimated to affect about 10% (Jaffe 1958). The tumour is usually an osteosarcoma but fibrosarcoma, chondrosarcoma, malignant giant-cell tumour and poorly differentiated sarcoma have all been described (Barry 1969; Spjut et al. 1979), as have myeloma and lymphoma (Copelan 1964). Although patients

Fig. 1 Microphotograph of the carcinomatous vulval lesion (× 8).

Fig. 2 A radiograph of the whole femur shows extensive Paget's disease and a pathological fracture.
over 60 years old not infrequently have Paget's disease and metastatic carcinoma, their coexistence at the same site is rare (Wilner and Sherman 1966; Agha et al. 1976); it was first reported by Castleman and McNeill in 1956. Metastatic carcinoma in Paget's disease has been recorded from primary sites in the breast, lung, kidney, prostate and colon (Wilner and Sherman 1966); tumours at the first four sites are often recognised as metastasising to bone. In the present case, however, the primary site was one which does not normally metastasise to bone, yet it had implanted preferentially in the femur affected by Paget's disease.

Cardiac output values as high as 13.3 l/min have been recorded in cases of Paget's disease (Edholm, Howarth and McMichael 1945). Such increased perfusion could reasonably be expected to cause haematogenous metastases to implant preferentially in bone affected by Paget's disease. According to Howarth (1953), cardiac output is unlikely to increase considerably if less than 35% of the skeleton is affected by Paget's disease, or if the alkaline phosphatase level is below 300iu/l. There was no ostensible evidence of increased cardiac output in the reported patient, but she did have an alkaline phosphatase of 375iu/l. This finding suggests the presence of active Paget's disease with a consequent increase in blood flow, and any site affected by the disease might therefore have been seeded preferentially by blood-borne metastases.

In cases of pathological fracture of bone affected by Paget's disease it thus seems advisable to consider biopsy of the fracture site, especially if the patient has a history of malignancy. The histopathologist must be given the patient's complete history so that a diagnosis of sarcomatous change due to Paget's disease is not arrived at without considering alternatives; a secondary deposit from another site may be more amenable to treatment and have a better prognosis than that associated with Paget's sarcoma.

REFERENCES


