AN UNUSUAL BONE TUMOUR COMPLICATING PAGET'S DISEASE

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Malignant tumours complicating Paget's disease usually take the form of osteogenic sarcoma. In 1949 Russell reported three cases of malignant osteoclastoma complicating Paget's disease and gave a detailed description of the histology. The case reported here is that of a similar malignant osteoclastoma developing in the mandible of a man with long-standing and widespread Paget's disease. The case is also of interest in that skeletal changes suggestive of Paget's disease developed in two of the patient's offspring, as reported elsewhere in this Journal.*

CASE REPORT

A man of sixty-five years was admitted to Hill End Hospital under the care of Mr Rainsford Mowlem, complaining of a painful swelling of the left mandible. He gave a history of having suffered from Paget's disease for thirty years, the primary involvement being of the tibiae and left arm. Fourteen years previously an injury to the right temporal region had been followed by local overgrowth of bone. Six months before his admission a swelling had developed rapidly in the left mandibular area but had remained about the same size since a few days after its appearance. Recently it had become painful.

Examination at that time showed Paget's disease with a widely open bite and gross over-development in the symphysial region. The right temporal fossa was filled with new bone and at the left angle of the mandible there was a large swelling, confined mainly to the outer surface and not apparently cystic; within the mouth it was smooth and replaced the whole of the alveolus so that the wearing of a denture was impossible. Mandibular development in front of the swelling was excessive. Radiographs of the skull showed extensive Paget's disease with an area of destruction at the angle of the left mandible and expansion of the cortex of the bone (Fig. 1).

Treatment—Because of the length of the history and absence of progressive enlargement of the swelling during the previous six months, a clinical diagnosis of a benign tumour was made. Operation was undertaken for relief of pain (Mr Rainsford Mowlem). An incision was made below the main part of the swelling along the body of the mandible on the left side. Anteriorly this was deepened to expose normal bone, but over the swelling itself no apparent bone layer

* IRVINE, R. E. Familial Paget's Disease with Early Onset. Page 106.
remained. The facial vessels were divided and the remnants of the masseter dissected off the tumour by blunt dissection. This process was continued upwards to free the alveolar mucous membrane and, beginning anteriorly, the tumour was enucleated. It was solid but friable and left behind extensions almost in the form of loculi. Between these extensions there was a "sandy" type of osteo-fibrotic bone. This was trimmed down and the residual areas of growth were excised.

**Histological examination**—The tumour was composed of giant cells of osteoclast type in a cellular spindle-cell background (Fig. 2). The matrix cells varied in size and staining. There were moderate numbers of mitoses. The appearances were those of an osteoclastoma and suggested the possibility of malignant change.

**Progress**—Despite satisfactory healing of the wound considerable pain persisted. Within two weeks a further swelling appeared on the under and inner aspects of the left mandible in the region of the angle and increased rapidly in size. In view of the histological report of osteoclastoma, he was transferred to the Middlesex Hospital under the care of Professor B. W. Windeyer for radiotherapy. Examination (eight weeks after operation) showed extensive Paget's disease affecting most of the skeleton (Figs. 3 and 5). There was a large swelling of the left mandible, most marked at the angle of the jaw. Within the mouth there was a large ulcerated tumour protruding from the inner surface of the left mandible and extending to within two centimetres of the midline anteriorly. The swelling occupied most of the left side of the floor of the mouth and pushed the tongue over to the right. The tumour had extended up the ascending ramus of the mandible and was pushing the soft palate forwards and downwards. Radiographically, there was extensive erosion of the left mandible (Fig. 6). There was no evidence of secondary deposits in the chest. **Investigations**—Blood count was normal; acid phosphatase was 37 units, alkaline phosphatase 57.3 units, and 17-ketosteroids were 14.5 milligrams per 24 hours. **Effects of radiotherapy and further progress**—The tumour was treated by teleradium using a five-field technique, a central tumour dose of 4,100r, being delivered in twenty-three treatments over a period of thirty days. The tumour decreased in size over the next few weeks and its centre became necrotic. Large pieces of slough and necrotic bone were removed until the cavity was clean and the edges well defined. A radiograph of the mandible two weeks after the completion of treatment showed considerable ossification in the tumour (Fig. 7). This had occurred earlier than was to be expected after radiotherapy to an osteoclastoma. Nevertheless the radiographic appearances were unlike those of sarcoma and clinically the tumour appeared to be healing. Four weeks later, however, a recurrence was noticed at the anterior end of the healing lesion and this spread rapidly, involving the whole of the left mandible and infiltrating the soft tissues of the neck (Figs. 4 and 8). Despite palliative x-ray therapy his condition deteriorated rapidly and he died six weeks later (six months after local excision of the tumour and three months after teleradium therapy).
Figure 3—Photograph of patient two months after operation. The tumour of the mandible, removed by local excision, has recurred. Note extensive deformities. Figure 4—Despite early reossification after irradiation, the tumour rapidly recurred and spread to involve the whole mandible.

Fig. 5
Pelvis showing advanced changes typical of Paget's disease.
Figure 6—Radiograph of mandible immediately before irradiation. Figure 7—Two weeks after irradiation. Note reossification.

Figure 8—Eight weeks after irradiation. Pathological fracture with absorption of all new bone. Figure 9—Section of the lung metastasis found at necropsy. The tumour is a chondrosarcoma and is similar to the sections of growth in the jaw taken after death (×100).
Necropsy—All bones examined showed evidence of Paget’s disease. The skull measured 24½ inches in circumference. A large swelling was present in the mandible on the left side. The mandible was fractured and the bone destroyed, over a length of 3½ inches, by a soft white tumour which was also invading the alveolar margin and left side of the tongue. There was a small tumour in the right upper lobe of the lung which had the consistency and appearance of cartilage on section. Histological examination of the mandible showed chondrosarcoma, no osteoclastomatous areas being found. The lung metastases were similar in appearance (Fig. 9).

**COMMENT**

Since Russell (1949) drew attention to the occurrence of malignant osteoclastoma in Paget’s disease no essentially similar case can be found reported in the literature. Hilton (1950) reported two cases of osteoclastoma associated with generalised bone disease, both of which failed to respond to irradiation. One of these occurred in the skull of a man with radiographic evidence of Paget’s disease in the skull and pelvis. But both were histologically benign giant-cell tumours with no evidence of malignancy.

The case presented here has been reported in detail because the tumour followed an unusual course and appeared, in the first histological sections, to resemble those reported by Russell. Re-examination of the first sections has allowed no alternative histological diagnosis to that of malignant osteoclastoma. The six months’ history of the tumour, without change in size or symptoms, suggested at first that it was benign. Transition from benign to malignant osteoclastoma was possibly taking place at about the time of the operation. Certainly the rapid recurrence after operation suggested a malignant tumour, and the early clinical response to radiotherapy, with evidence of rapid reossification, was most unusual for a benign osteoclastoma, initial further decalcification being the typical response. Russell’s patients were not treated by irradiation and so comparison cannot be made, but the tumours reported by Hilton showed not even a temporary response to radiotherapy.

**SUMMARY**

An unusual bone tumour complicating Paget’s disease of the mandible is described. At the time of local excision the histological appearances were those of a malignant osteoclastoma, but the tumour rapidly recurred as a chondrosarcoma, which spread locally and by metastasis, and caused death within six months.

I have pleasure in recording my thanks to Professor B. W. Windeyer and Mr Rainsford Mowlem for permission to publish this case. Dr A. C. Thackray reported on the histology and produced the photomicrographs. The clinical and radiographic photographs were taken by Mr M. Turney in the Middlesex Hospital Photographic Department.

**REFERENCES**
