BONE SARCOMA IN PAGET'S DISEASE

Report of a Case with Long Survival

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The prognosis in sarcomata secondary to Paget's disease is distinctly bad, and we have found only one five-year survival (Sherman and Soong 1954) and six two-year survivals reported in the English literature (Coley and Sharp 1931; Hansen 1942; Mercer and Duthie 1955; Porretta, Dahlin and Janes 1957; Platt 1947). Professor Dorothy S. Russell tells us that one of H. Osmond-Clarke's patients (Case 5, 1949), in whom a seemingly highly malignant tibial tumour complicated Paget's disease, was still alive without sign of recurrence ten years after amputation.

The following is a case report of a man who lived for almost eight years after local excision of a sarcoma of a humerus which was the site of Paget's disease.

CASE REPORT

In 1918, when the patient was twenty-four years old, he noticed that his right leg was becoming bowed and was occasionally painful. The deformity slowly increased and when he was thirty-eight years old his left leg began to bow and he also noticed that his head was becoming larger. In June 1940, at the age of forty-six, his right leg "gave way" and he was unable to stand on it. He was admitted next day to the Massachusetts General Hospital and

![Fig. 1](image)

Humerus showing Paget's disease with an osteolytic lesion in the mid-shaft.
was found to have a transverse fracture of the tibia and fibula, the bones being affected by Paget's disease. Radiographs of the skull, pelvis and left femur also showed evidence of Paget's disease. The fracture was treated by open reduction and internal fixation with a metal plate and screws. Sepsis, which at first appeared to be superficial, later developed into chronic osteomyelitis, requiring an above-knee amputation in February 1948. No evidence of tumour was found in the amputated limb. After the operation partial necrosis of the skin flaps developed.

While in the hospital in June 1948 he called attention to a painless swelling of his right arm which he had discovered two weeks before. On examination, a hard, immovable, irregular, non-tender mass measuring three inches by three inches was found arising from the antero-medial aspect of the humerus. Radiographs (Fig. 1) showed evidence of Paget's disease in the humerus, and there was an osteolytic defect in its middle third involving about half of the diameter of the shaft. The chest film was negative.

*Operation*—The lesion was diagnosed clinically as an osteogenic sarcoma and it was decided to attempt local resection without preliminary biopsy. At operation the tumour was found to have invaded only the brachialis muscle. The tumour, brachialis muscle and six inches of the humerus were excised, allowing approximately a two-inch margin of almost normal bone on either end of the tumour mass. The gap was bridged by a bone graft consisting of eight inches of the left fibula, fixed to the humerus with one screw proximally and two screws distally. Histological examination of the specimen confirmed the diagnosis of osteogenic sarcoma.

*Progress*—Difficulty was encountered in controlling the position of the arm in a spica: bone contact between the graft and upper humerus was lost (Fig. 2), and a frank pseudarthrosis developed later (Fig. 3). Although great care was taken in protecting the nerve trunks a radial
nerve palsy was noted after operation. Severe arthritic changes in the remaining leg and the impaired function of the arm confined him to a wheel-chair. In spite of the pseudarthrosis

![Image](https://example.com/figure4.jpg)

**Fig. 4**
Photograph of specimen showing thickening of cortex and sarcoma invading brachialis muscle. (Scale in centimetres.)

![Image](https://example.com/figure5.jpg)

**Fig. 5**
Section of specimen taken from resected margin of humerus showing active Paget’s disease of bone. (Haematoxylin and eosin, ×125.)

and the radial nerve palsy, function of the hand and of the extremity as a whole was so good that further surgery was deemed inadvisable. His general health remained good until February 1956 when he was readmitted to the Massachusetts General Hospital with acute urinary retention. After decompression of the bladder he remained very ill for several weeks with a
labile electrolyte balance, but recovered and was discharged in March 1956. He died in April 1956 of cardiac failure seven years and ten months after resection of the sarcoma.

![Fig. 6](image)

**Fig. 6**

Section of tumour showing plump tumour cells with abundant mitotic figures separated by amorphous intercellular substance. (Haematoxylin and eosin, ×450.)

![Fig. 7](image)

**Fig. 7**

Section of tumour showing pleomorphic cells with more scanty intercellular substance than in Figure 6. The intercellular substance contains abundant collagen. (Haematoxylin and eosin, ×450.)

While he was in the hospital in 1956 clinical and radiological examination revealed no evidence of local recurrence or pulmonary metastases. No necropsy was secured.

_**Pathology of the specimen**—_The specimen consisted of part of the humerus fourteen centimetres long (Fig. 4). A firm fibrous mass had destroyed part of the cortex, producing an elliptical
defect six centimetres long and four centimetres wide. It had penetrated to the opposite cortex but there was no extension along the medullary cavity. The tumour was ovoid, with its greatest dimension where it had invaded the brachialis muscle and, though well circumscribed, did not have a capsule. The cortex of the humerus was irregularly thickened at the part adjacent to the tumour.

Microscopically, the cortex from the resected margin of the specimen showed active Paget's disease with numerous osteoblasts and osteoclasts and areolar fibrosis of the intertrabecular spaces (Fig. 5). Early conversion of the cortex into a mosaic of angular lamellar systems was present. The tumour had invaded brachialis muscle bundles, but no invasion of blood vessels was seen. Multiple sections of the tumour showed a sarcoma with fairly numerous and often atypical mitotic figures. Many areas were composed of plump tumour cells separated by a pale pink intercellular substance resembling osteoid tissue (Fig. 6). Elsewhere, whorled masses of elongated tumour cells were intermixed with abundant collagen (Fig. 7). Transitions between these fields were frequent, and collagen bundles could be followed into the more amorphous pink substance. No new bone trabeculae or calcification were seen.

DISCUSSION

This tumour was originally diagnosed on histological examination as an osteogenic sarcoma, but a review of the sections reveals no true osteoid or bone. As we believe that osteogenic sarcoma cannot be diagnosed in the absence of either osteoid or bone, we consider that this tumour is a fibrosarcoma. Lichtenstein (1952) states that fibrosarcoma occurring in Paget's disease "is not unusual" and it seems likely that some of the sarcomata of Paget's disease that have been reported as osteogenic sarcomata are, in fact, fibrosarcomata. This finding does not detract from the rarity of the long survival after local excision.

The early age of onset of Paget's disease in this man and the lack of pain in his tumour are both of interest and may have a bearing on his survival.

REFERENCES