A METASTASISING ADAMANTINOMA OF THE TIBIA

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Since Fischer (1913) first described an apparently primary tumour of the tibia with the microscopic features of an ameloblastoma, a number of publications have established this interesting lesion as a rare but definite entity. Often preceded by trauma, the tumour grows slowly, has not hitherto responded to irradiation, and recurs after local removal. Amputation frequently effects a complete cure.

Earlier analyses of published reports led to its being regarded as non-metastasising, but a follow-up of these cases has revealed that regional and distant metastases, though delayed, are not uncommon (Baker et al. 1954).

The present case is considered worth recording for several reasons: 1) It is only the fourth case in the British literature; 2) a metastasis appeared in the chest wall twenty-two years after amputation; 3) this is the first instance in which a metastasis beyond the inguinal lymph nodes has been proved microscopically; and 4) the metastasis showed some response to telecobalt irradiation.

CASE REPORT

The patient, a male weigh bridge attendant, was first seen at the West London Hospital at the age of forty-two, complaining of a painful swelling in the upper third of the right leg. He had sustained a contusion at the same site a year earlier. Radiographs showed destruction of the tibia deep to the swelling, considered diagnostic of bone sarcoma, and supracondylar amputation was performed. The pathologist reported a spindle-cell sarcoma undergoing myxomatous change with cavity formation, and added: "It does not suggest a particularly rapidly growing neoplasm." Radiographs of the chest at that time showed no abnormality.

The patient remained well for nearly twenty-two years, and then developed a painful swelling over the ninth and twelfth ribs posteriorly. A biopsy at Fulham Hospital revealed an anaplastic malignant tumour, and the patient was referred to the Westminster Hospital. On examination the amputation stump was healthy and there were no palpable inguinal glands. Attached to the left lower ribs was a firm ovoid tumour measuring about eight inches by four. The axillary glands were not palpable. Radiographs revealed destruction of part of the eleventh rib, with a pathological fracture (Fig. 1). Comparison of the thoracic biopsy and the original tumour revealed similar microscopic characters, with small darkly staining cells arranged in masses with sharply defined spindles margins, and intercellular oedema, giving rise to the quasi-acinar formations that characterise the adamantinoma of the jaw (Figs. 2 to 5). Biochemical and haematological findings were normal. Telecobalt therapy was instituted using a 30 curie (20 centimetres source-skin distance) unit. Two fields, eight by six centimetres, were directed to the region of the eleventh left rib. A total tumour dose of 6,032 r. (maximum) and 4,524 r. (minimum) was given in forty-three days.

Eight months later the patient's general condition was excellent and the tumour was only a fifth of its original size. Radiographs showed that the rib lesion was calcified, but a round shadow had appeared in the lower lobe of the left lung.

DISCUSSION

The sole justification for calling these tumours adamantinomas is their histological appearance. We therefore feel free to exclude published cases unaccompanied by photomicrographs, or those that might be classed as anaplastic squamous carcinomas.
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In the most recent review Baker et al. (1954) accepted twenty-seven cases, including three of their own. In the photomicrographs of their own Cases 2 and 3 the alleged epithelial strands are almost certainly osteoid foci of fibrous dysplasia, and we therefore accept their first case only. We also regard the cases of Pérochon and Veluet (1928), of Bell (1942), and of Mangalik and Mehrotra (1952) as not satisfactorily proven. Cases included in earlier series, which we also find unacceptable, are those of Maier (1900), Simon (1923), Nové-Josserand and Tavernier (1927), Casini (1935) and Vanek (1951). We accept the other twenty-two cases of Baker et al., to which we add that of Hertz (1952) and our own case, bringing the total to twenty-four. We would, on histological grounds, include the case of Lederer and Sinclair (1954), but for the fact that these authors preferred to regard it as a malignant synovioma.

![Fig. 1](image)

Radiograph showing metastasis involving the eleventh left rib.

Of these twenty-four tumours, twenty-two have occurred in the tibia, one in the fibula (Baker et al. 1954) and one in the ulna (Anderson and Saunders 1942). In two-thirds of the cases the left tibia was involved. Fifteen of the patients were male. The age of the patient when first seen ranged from twelve to fifty-seven years.

**Pathogenesis**—Fischer (1913), who described the first case, believed the tumour to be a true adamantinoma, probably originating from a foetal cell rest during the intra-uterine period, because the potentiality of tooth-formation is limited to the stage in embryonic life during which tooth-germs are formed. The weakness of this concept is that it can neither be proved nor disproved. In recording the fourth case Ryrie (1932) noted that each tumour had been preceded by injury some months earlier, and suggested that in view of the close relationship of the bone to the overlying skin, tearing of the dermal appendages, followed by a haematoma.
Fig. 2
Original tumour showing general pattern. (Haematoxylin and eosin, ×45.)

Fig. 3
Original tumour showing cyst formation. (Haematoxylin and eosin, ×75.)
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FIG. 4
Original tumour showing intercellular oedema simulating adamantinoma. (Haematoxylin and eosin, ×160.)

FIG. 5
Metastasis in chest showing typically adamantinomatous appearance. (Haematoxylin and eosin, ×125.)

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caused a “thwarted repair” which, in the course of prolonged organisation, resulted in a tumour of basal cell type.

Neither of these theories finds much favour to-day, and it has been pointed out by Hicks (1954) that the tibia is singularly prone to minor injury. On the other hand several patients have been quite definite about the close relation between the growth and the site of injury. In some cases there was a lapse of months or years between the injury and the appearance of a lump, but in others the injury has been immediately followed by a painful swelling which persisted up to the time the tumour appeared.

Radiography of the tibia has often revealed a characteristic saucer-shaped depression of the bone below the tumour, suggesting erosion from the surface. Certainly the microscopic appearances, while not unlike the classical adamantinoma of the jaw, would do equally well for a basal cell carcinoma undergoing cystic degeneration. A number of growths have shown foci of squamous differentiation. It was suggested in another connection by King (1933) that post-traumatic epidermoid cysts resulted from a metaplasia of sweat-gland epithelium due to the presence of a subcutaneous haematoma. He was driven to this conclusion by the fact that many of his series of so-called “implantation” cysts occurred in the hands and fingers of manual workers who had sustained previous injury without breaking the skin. This concept offers some sort of answer to those who argue that the injuries preceding tibial adamantinomas have been often of a trivial nature.

In fact, the antecedent trauma has taken the following forms: contusion (9), abrasion (2), fracture (2), sprain (1), fall (1) and puncture wound (1). In four, however, a history of injury was doubtful, and in four others absent altogether. Still, the very frequency of tibial contusions and abrasions could well lead to their being completely forgotten by some patients. Perhaps the most formidable argument against the traumatic theory is the disparity between the rarity of the tumour (twenty-four cases in forty-two years) and the frequency of an injury that is the common lot of the human race.

In recent years a third theory has been advanced. In 1947 Lauche claimed that Fischer’s original case was really a synovial sarcoma, and Hicks (1954) and Lederer and Sinclair (1954) independently arrived at the same conclusion regarding the others. Bennett (1947) and King (1952) described epithelial differentiation of synovial tumours, but after a study of some thirty-one synovial sarcomas in the Westminster Hospital series we remain unconvinced of the identity of the two growths, although occasional similarities are to be found in isolated microscopic fields. In the papers of Berger (1938) and of Tillotson et al. (1951) the synovial sarcoma is represented as a highly malignant tumour, although Wright (1952) described a relatively benign variety; but in none of these articles do the photomicrographs resemble the tibial adamantinoma. Boyd (1955) found the theory of synovial origin more palatable than those involving foetal rests or thwarted repair, but we feel that the concept of a synovial tumour arising in bone is not to be accepted without question.

Metastases—For many years it was imagined that the tibial adamantinoma was only locally malignant. Baker et al. (1954) dispelled this illusion by collating the post-operative history of earlier case reports, and seventeen of the twenty-four under review are available.

Nine patients were alive and well, with no evidence of metastases, fourteen to twenty-one years after amputation (five cases) and five to eighteen years after local excision or resection and bone grafting (four cases). As the amputation cases were published between 1931 and 1939, and those treated more conservatively between 1942 and 1954, the difference in duration of survival is not significant.

The other eight patients developed metastases, and seven died—two within a year from other causes (septicaemia, pulmonary embolism) and both had microscopically proved metastases in the inguinal lymph nodes. The other five lived from four to eleven years after the initial operation, and in all of these the evidence for pulmonary or skeletal metastases was clinical or radiological, there being no necropsy. The eighth patient—our own—developed
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a metastasis in the chest twenty-two years after amputation, and this is the first case in which a distant metastasis has been proved microscopically to be of the same nature as the tibial primary.

Treatment—It is thus clear that the tibial adamantinoma is a slowly growing tumour of low grade malignancy. Recurrence after curettage or local excision is so frequent as to be virtually the rule; however, there is no guarantee that more radical measures will provide a cure. Six of the eight patients with metastases underwent amputation, in one case as the primary treatment, in four within seven to twenty-four months of the first operation, and in one after nine years. Of the nine patients alive and well, five were treated by amputation from six to forty-four months after the first operation. From this it seems probable that when the tumour is confined to bone, adequate resection, either subperiosteally (Baker and Hawksley 1931), or followed by bone grafting (Cagnoli 1944, Hertz 1952), offers as good a prognosis as amputation.

Between the years 1934–42 curettage or local excision was augmented by x-ray or radium therapy in five cases, and invariably there was local recurrence within a few months. This form of treatment was therefore abandoned, but it is worth noting that in our case an inoperable metastasis in the chest wall was considerably reduced by telecobalt irradiation.

SUMMARY

1. The so-called adamantinoma of long bones is a clinico-pathological entity, the pathogenesis of which is still in doubt. The case for its being a synovial sarcoma showing epithelial differentiation is in our view unconvincing.
2. The tumour is slowly growing, and of low grade malignancy. Apparent cure has been effected in a third of the total cases recorded by amputation or resection of the diseased bone.
3. These means, however, have not prevented metastases to the lungs and skeleton in a similar number.
4. A case is presented in which a metastasis appeared in the chest twenty-two years after amputation of the leg. This was sensitive to telecobalt irradiation, and is the first case in which a distant metastasis has been proved microscopically.

We are indebted to Sir Stanford Cade for permission to record this case. We wish to thank Dr Peter Kerley for the radiograph and the West London Hospital for their kindness in making the original notes available.

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