ADAMANTINOMA OF THE TIBIA
Report of Two Cases

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Adamanatinoma of the tibia is not a common lesion, but because of its somewhat anomalous features and confusing histological picture it continues to arouse the interest of clinicians and pathologists. Much of the interest stems from the term adamantinoma which was first used by Fischer (1913) to describe a primary bone tumour involving the tibia. He used the term adamantinoma because of its resemblance to the ameloblastoma of the jaw.

Until 1940 adamantinoma of the long bones had been described only in the tibia. Since then it has been found in other long bones and also in some short bones (Anderson and Saunders 1942, Moon 1965). In 1954 Baker, Dockerty and Coventry in a review of the then published cases set out the clinical picture of the condition.

There is an equal incidence in the sexes. The age range is from twelve to fifty-seven years. The predominant site is the tibia although single cases have been reported in the femur, fibula and ulna. There is usually a history of trauma, and pain is the main symptom. The duration of the pain may range from six weeks to several years. The radiographic appearances, which are not diagnostic, show cystic and multicystic patterns with cortical destruction and some periosteal reaction.

Macroscopically the tumour is pale, grey or white. The consistency is described as firm, elastic, brain-like, sometimes soft or even gritty. There are cystic cavities which may contain straw coloured or bloodstained fluid. The tumour may involve the cortex and may penetrate the periosteum and soft tissues. Calcification may occur (Donner and Dikland 1966). Neither teeth nor enamel tissue have been found in any of the cases reported to date. The microscopic appearances vary from case to case. Three patterns are described (Baker and colleagues 1954): 1) Masses of epithelial cells are seen in islet formation, around the periphery of which columnar cells are arranged in palisade fashion. The central masses show stellate cells in a reticular formation. There are cystic areas in the centre of the reticulum. This type could be mistaken for an adenocarcinoma. One case is recorded in which secondary carcinoma was diagnosed as adamantinoma (Ackerman and Spjut 1962). 2) Islands of cells resembling basal cells are scattered throughout a fibrous stroma. Palisading of the peripheral cells occurs. Cystic areas are present with epithelial masses reminiscent of basal cell carcinoma. 3) Squamous epithelial islands are scattered throughout a fibrous stroma. There is evidence of pearl formation.

Because of the tumour's protracted course, it was believed for many years to be benign or at most only locally malignant. Local excision was advocated but this proved ineffective. Radiotherapy was also recommended, either alone or combined with local excision (Rehbock and Barber 1938, Rankin 1939, Hebbel 1940). It is now known that the incidence of local recurrence is high and that radiotherapy is ineffective. Metastases have been reported in several cases (Mangalik and Mehrotra 1952; Baker and colleagues 1954; Morgan and Mackenzie 1956; Naji, Murphy, Stasney, Neville and Chrenka 1964). Once the diagnosis is clear from the biopsy specimen amputation is recommended. Delay increases the risk of metastasis.

CASE REPORTS

Case 1—A man aged forty-two complained of a painful swelling over the front of the right tibia. There had been some irregularity of the same region since childhood. There had always been some discomfort but the acute pain was a new feature. Examination showed a small cystic swelling over the middle third of the anterior surface of the tibia, which was tender on pressure.
Radiographs were reported as consistent with a diagnosis of enchondroma (Fig. 1).
At biopsy a round tumour 5 centimetres in diameter was found. It was firm in consistency, yellow brown, and well defined from the surrounding tissues. The tumour was rather avascular. A frozen section suggested the diagnosis of adamantinoma, and histological examination showed the tumour to be composed of epithelial cells, with palisading of the peripheral areas. In many areas there was no great variation in the size and shape of the cells which had a rather coarse chromatin network and ill-defined cytoplasmic boundaries. Whorling was frequent.

No fully developed keratinisation was found. In the larger cell groups there was often some central degeneration. The appearances were those of adamantinoma (Fig. 2). 

*Treatment and progress*—The tumour was excised and bone was grafted to bridge the gap. The tumour recurred within six months. At a second operation above-knee amputation was carried out. Twenty years later the patient is still alive and well with no evidence of recurrence.

*Comment*—At the time when the histology was being discussed we were fortunate to have the opinion of Dr H. L. Jaffe, who was visiting Glasgow at that time. He confirmed the histological diagnosis of adamantinoma. It was thought that the fact that tibial deformity had been present from birth supported the theory of epithelial malformation rather than the theory of traumatic dislocation of the skin into the periosteal tissues (Ryrie 1932).

**Case 2**—A sixty-one-year-old woman had had a painful swelling of the upper shaft of the left tibia for some eighteen months. On examination there was a hot swelling one and a half inches in diameter. The skin over the tibia was free. There were no palpable lymph nodes. The radiograph was reported as consistent with an enchondroma. A second radiograph was reported as showing fibrous dysplasia (Fig. 3).
**Biopsy**—At biopsy a white, fibrous, avascular mass was noted from which a gelatinous fluid emerged. The tissue consisted of highly cellular epithelial or pseudo-epithelial masses and a less cellular dense fibrous stroma. The epithelial portion resembled a basal cell carcinoma except that the cells were more spindle shaped. There were large and small masses of cells with branching, intersecting processes. There was often distinct palisading of the boundary cells. Some cell masses had a lesser, almost myxoid, appearance and occasionally there were open clear spaces producing a reticulated appearance. The cells were small and spindle shaped with scanty cytoplasm and very hyperchromatic dense nuclei. Mitotic figures were extremely rare. Within the cell masses there were occasional zones of larger cells with paler nuclei or abundant cytoplasm resembling squamous cells and there was a single keratinised cell nest. The histological appearance was that of adamantinoma.

**Treatment and progress**—The patient had a through-knee amputation. She is now well and showing no evidence of recurrence two and a half years later.

**DISCUSSION**

Adamantinoma is of course a misleading name, signifying as it does a tumour consisting of enamel tissue. At no time has any enamel tissue been found in any of the published cases. It is around this question of the histogenesis of the tumour that most of the controversy has taken place. In 1913 Fischer stated that the origin was embryonal epithelium of the enamel germ of the teeth which are present as epithelial cell nests. Ryrie (1932) was sure that the origin of the tumour cells was a traumatic implantation of the basal cells of the skin into bone. This was said to occur even in cases where the skin was not broken. In the region of the ossifying haematoma the repair process was thwarted and tumour growth occurred. Baker and colleagues in 1954 offered no explanation for the origin of the epithelium and stated that the origin was unknown. Lederer and Sinclair in 1954 reviewed all the cases published at that time and stated that the tumours were synovial sarcomata, and Naji and colleagues (1964) supported this view. In 1957 Changus, Speed and Stewart in a comprehensive review concluded that a vascular origin had been demonstrated and that the epithelial origin had been disproved. They recommended the abandonment of the term adamantinoma in favour of malignant angioblastoma. This is supported by Elliot (1962). In 1958 Jaffe stated that the evidence pointed to an epithelial origin of the tumour. In 1969 Rosai published electron-microscopic evidence of the epithelial origin of the tumour. He stated with reasonable confidence that the tibial adamantinoma was of epithelial origin and that the evidence did not support a synovial or vascular origin. This view is supported by Saacebra, Gutierrez and Dimas (1968) who confirmed the epithelial origin of the tumour.

It is impossible to make any dogmatic statement regarding the origin of this unusual and interesting neoplasm, but our review of the histology coupled with Jaffe’s observations on the first case indicate that the growth is epithelial in origin. It is also interesting to note that the first patient (a doctor) had been aware of a swelling in the leg at the site of the tumour growth many years previously—in fact he believed that this swelling had been present since childhood. Unfortunately no investigation as to its nature was ever carried out.

The two cases presented illustrate clearly the problems posed by the tumour. The radiographic appearances are non-specific: those in both our cases were reported initially
as indicating enchondroma. This view was later revised to that of fibrous dysplasia; indeed, this confusion with fibrous dysplasia was noted by Cohen, Dahlin and Pugh (1962). Although radiographs may show no more than a small, discrete area, it may be found at the time of biopsy that the tumour has spread well beyond the area shown in the radiographs.

Morgan and Mackenzie (1956) described a similar finding in their paper. In our first case despite adequate excision and bone grafting the tumour recurred within six months. Although a successful bone grafting operation was reported by Elliot (1962) a review of the literature together with our own experience in these two cases leads us to believe that the nature of the lesion and the doubtful prognosis justify amputation.

SUMMARY

Two cases of adamantinoma of the tibia are reported. The first patient has survived twenty years after above-knee amputation and shows no sign of recurrence. The second patient shows no evidence of recurrence two and a half years after amputation through the knee.

The clinical picture of the tumour is described and the theories of histogenesis are outlined.

REFERENCES


