Acta orthop. scand. *50*, 97–101, 1979

ADAMANTINOMA TIBIAE

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A case with characteristic roentgenographic and microscopic findings of adamantinoma tibiae is reported. Knee disarticulation with early ambulation is the treatment of choice and was performed on this patient. Limb ablation followed by Unna Paste Bandage to prevent oedema and permit early prosthetic fitting facilitates rehabilitation. The 1 year follow-up showed excellent prosthetic function and no evidence of residual tumour.

Key words: adamantinoma tibiae; knee disarticulation; early prosthetic fitting; Unna Paste Bandage

Accepted 8.viii.78

Adamantinoma is a rare primary tumour of long bones. It may occur in all regions but the tibia is the most frequent site. According to Dahlin (1967) it represents only 0.33 per cent of primary malignant bone tumours in the files of the Mayo Clinic. The present tumour is the first case of adamantinoma tibiae registered in the Swedish Cancer Register since it was started in 1958. There are at present about 1000 malignant skeletal tumours registered, which means an incidence of about 0.1 per cent for adamantinoma tibiae (J. Ericsson 1978, personal communication). There are only about 100 cases reported in the literature (Huvos & Marcove 1975). The term adamantinoma tibiae was first used by Fischer (1913) because of the alveolar and tubular arrangement of the cells resembled that of adamantinoma of the jaw bone (ameloblastoma). This pattern of islands and strands of cells forming alveolar or tubular structures is characteristic and pathognomonic (Jaffe 1958, Dahlin 1967, Lichtenstein 1972).

There is considerable controversy regarding the *pathogenesis*. Some authors consider this tumour to be a form of synovial sarcoma (Lederer & Sinclair 1954, Hicks 1954) while others believe that the cell clusters are of angioblastic origin (Changus et al. 1957). Based on electronmicroscopic studies, Rosai (1969), and also Unni et al. (1974), concluded that the palisading cells in the fibrous stroma were of epithelial origin.

Adamantinoma tibiae is a slow growing and potentially malignant tumour with a marked tendency to local recurrence and with the possibility of metastasizing through the lymphatics and blood vessels even after very long periods of time (Jaffe 1958, Moon 1965). A multicentric origin of the tumour has been suggested as an explanation for the apparent frequency of recurrence and delayed metastases (Unni et al. 1974).

Roentgenograms demonstrate the lesion in its usual tibial location as a well marginated trabeculated rarefaction of the cortex (Jaffe 1958, Radiological atlas of bone tumours). The roentgenologic picture is frequently misinterpreted as fibrous dysplasia.

Most authors consider radiotherapy in-

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Figure 1. The lesion is located anteriorly in the diaphysis of the tibia and is fairly well-defined with sclerosis at the margin with the normal bone. In some parts the tumour has a coarse trabecular structure. The cortex is partly destroyed and in places it is only paper thin.

effective (Jaffe 1958, Lichtenstein 1972, Unni et al. 1974). In 1972, however, Zand et al. reported a patient who at the age of seven had been treated with cobalt irradiation and who was free from recurrence or metastasis 9 years later.

The following conclusions can be drawn from the reported cases regarding surgical treatment:

Local curettage. This treatment is often followed by recurrence and is inadequate.

"En bloc" resection. This has been done in selected cases with good long-term results.

Amputation. Amputation is the treatment of choice when radical "en bloc" resection

would produce technically unacceptable results. An active regime of early prosthetic fitting may contribute to the patient's acceptance of limb ablation and leads to optimal rehabilitation.

CASE REPORT

A 32-year-old married woman was first seen in March, 1977, with a 6-month history of a painless swelling below the left knee. There was no history of preceding trauma. Since the age of 17 she had been on Difhydan (Fenytoin) medication because of epilepsy and had been free from attacks for at least 10 years.

On physical examination there was a slightly tender firm swelling on the anterior aspect of the proximal half of the left tibia. Roentgenograms of the left tibia showed an oval multiloculated cystic lesion that was 12 cm long (Figure 1). The lesion had destroyed a portion of the anterior cortex 4 cm below the tibial tubercle. Arteriograms were normal. Cytologic findings after thin-needle biopsy were inconclusive. Tissue from an open biopsy was interpreted as probable adamantinoma tibiae. Radical "en bloc" resection and preservation of knee function was not possible due to the size and site of this neoplasm. After some hesitation the patient agreed to a through-knee amputation to be followed by early prosthetic fitting.

The knee disarticulation, *ad modum* Dederich (1970), was performed on May 3, 1977. To prevent swelling of the stump and facilitate early ambulation an Unna Paste Bandage was applied. Casting for a temporary prosthesis was made 1 week postoperatively after change of the dressing. The first sockets were given quadrilateral shape for tuberoischial support and relief of the stump end. Six weeks postoperatively (June 16, 1977) she was equipped with a total contact thigh socket and a Lyquist prosthetic knee mechanism. This improved her function and it was fully accepted from a cosmetic point of view. The postoperative procedures are summarized in Table 1. They were performed in a stimulating but non-irritating atmosphere and were well tolerated (Table 1).

Dissection of the *amputation specimen* demonstrated the anterior portion of the tibia from a level 3 cm below the tibial plateau to 12 cm distally to be involved by a firm grey tumour (Figure 2). The tumour was confined to bone. The anterio-medial cortex was infiltrated and very thin, but it was not perforated. As in the pre-operative biopsy specimen, the microscopic picture was dominated by a loose fibrous stroma

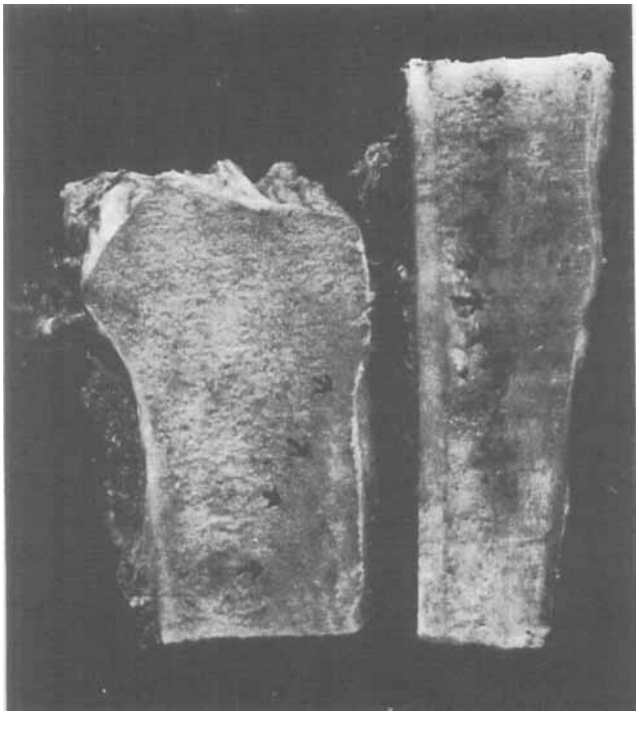

Figure 2. Cut surface of the surgical specimen shows a greyish-white, rather sclerotic appearing tumour in the anterior part of the left tibia extending from 3 cm below the tibial plateau distally for 12 cm.

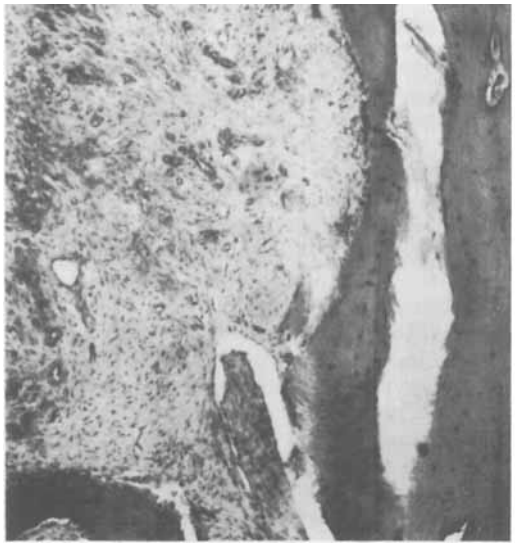

Figure 3. The tumour is confined to the bone and infiltrates the anterior cortical bone. As in the preoperative biopsy, the microscopic picture is dominated by loose, vascular connective tissue without cellular atypia resembling that of fibrous dysplasia.

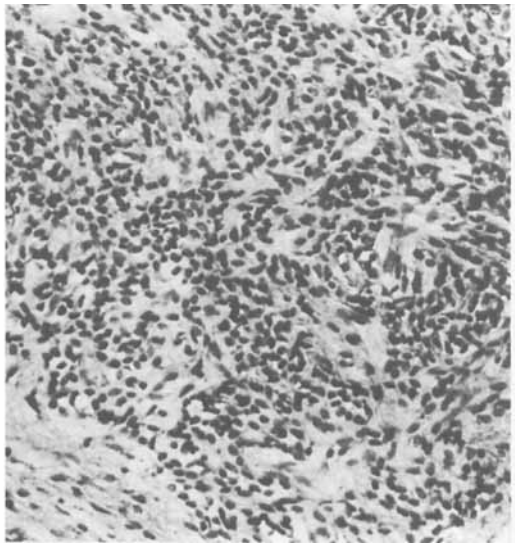

Figure 4. Scattered cellular areas are seen. In some areas nests of elongated or cuboid cells give the impression of a vascular tumour.

Table 1. Summary of the postoperative management

Date	Management	Comment
May 3, 1977	Through knee amputation (*ad modum* Dederich) Unna Paste Bandage	
May 9, 1977	Change of dressing Casting for temporary prosthesis	No swelling
May 11, 1977	Walking exercises begun	Quadrilateral socket with tuberoischial support
May 24, 1977	Extraction of AO-pins used to fix the patella	
May 26, 1977	New high socket	
June 16, 1977	Socket shortened	Partial end bearing
June 23, 1977	Definitive prosthesis (Total contact thigh socket and Lyquist knee mechanism)	
November, 1978	Follow-up. No complaints	Ambulating well with prosthesis and re-employed

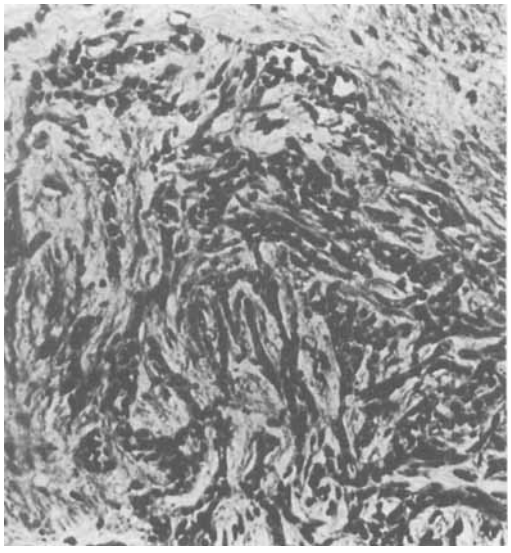

Figure 5. Other areas consist of elongated or cuboid cells in tubular arrangements that are characteristic of adamantinoma. Palisading of cells can also be seen.

without cellular atypia (Figure 3) in which were islands and strands of cells. These cells were elongated or cuboidal and occurred in nests that gave the impression of a vascular tumour (Figure 4). In other areas strands and islands of cuboid epithelial cells were present which had a tubular arrangement and demonstrated palisading (Figure 5). These were characteristic of adamantinoma tibiae.

The 1.5 year follow-up has shown the patient to be well and to have good prosthetic function. There is no evidence of recurrence of the tumour.

ACKNOWLEDGEMENT

The authors express their gratitude to G. Moberger, M.D., and G. Söderberg, M.D., Tumörbiologiska Institutionen, Karolinska sjukhuset, Stockholm, for help in the histopathological analysis and to B. Stener, M.D., Gothenburg, and U. Nilsonne, M.D., Stockholm, for advice.

REFERENCES

Changus, G. W., Speed, J. S. & Stewart, F. W. (1957) Malignant angioblastoma of bone. *Cancer* **10,** 540–559.

Dahlin, D. (1967) *Bone tumors*, pp. 204–211. Charles C Thomas, Ill.

Dederich, R. (1970) *Amputationen der unteren Extremität.* pp. 85–88. Georg Thieme Verlag, Stuttgart.

Fischer, B. (1913) Uber ein primäres Adamantinom der Tibia. *Frankfurt. Z. Path.* **12,** 422–441.

Hicks, J. D. (1954) Synovial sarcoma of the tibia. *J. Path. Bact.* **LXVII,** 151–161.

Huvos, A. G. & Marcove, R. C. (1975) Adamantinoma of long bones. *J. Bone Jt Surg.* **57**-A, 148–154.

Jaffe, H. L. (1958) Adamantinoma of limb bones. *Tumors and tumorous conditions of the bones and joints*, pp. 213–223. Lea & Febiger, Philadelphia.

Lederer, H. & Sinclair, A. J. (1954) Malignant synovioma simulating "adamantinoma of the tibia". *J. Path. Bact.* **LXVII,** 163–168.

Lichtenstein, L. (1972) Dermal inclusion tumors in bone (so-called adamantinoma of limb bones). *Bone tumors,* 4th ed., pp. 343–348. The C. V. Mosby Co., St Louis.

Moon, N. (1965) Adamantinoma of the appendicular skeleton. *Clin. Orthop.* **43,** 189–213.

Radiological atlas of bone tumours (1966). *Adamantinoma of the limb bones.* Vol. 1, pp. 213–214. The Williams & Wilkins Co.

Rosai, J. (1969) Adamantinoma of the tibia. *Amer. J. clin. Path.* **51,** 786–792.

Unni, K. K., Dahlin, D. C., Beabout, J. W. & Ivins, J. C. (1974) Adamantinomas of long bones. *Cancer* **34,** 1796–1805.

Zand, A., Chambers, G. H. & Street, D. M. (1972) So-called "adamantinoma of long bones". *Clin. Orthop.* **86,** 178–182.

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