J Cancer Res Clin Oncol (1982) 104: 315–320

Journal of
Cancer Research
and
Clinical Oncology

Short Communications

An Unusual Case of Adamantinoma of Long Bones

M. Altmannsberger [1], H. Poppe [2], and A. Schauer [1]

[1] Pathologisches Institut der Universität Göttingen, Robert-Koch-Str. 40, D-3400 Göttingen, Federal Republic of Germany
[2] Radiologische Klinik der Universität Göttingen, D-3400 Göttingen, Federal Republic of Germany

Summary. Adamantinoma of the tibia is a rare bone lesion, in which it is impossible to predict the biological behaviour. A case of extremely late local recurrence and lung metastasis is presented. Additionally, the patient developed pneumothorax on the right side months before intrapulmonary metastasis could be detected by X ray. Finally 1 week before her death the young woman suffered from paraneoplastic severe hypercalcemia leading to hypercalcemic coma and pancreatitis.

Key words: Adamantinoma – Pneumothorax – Paraneoplasia – Hypercalcemia

Introduction

Adamantinoma of long bones is an uncommon tumor with a histological pattern reminiscent of adamantinoma of the jaw. The term is a misnomer because the tumor possesses no potential to produce enamel. Normally the tumor follows a benign course, but in 10–15% of all cases adamantinoma metastasizes (Weiss and Dorfmann 1977). We present a case showing uncommon behaviour and extremely late metastasis, as up until now only a small number of cases have been published.

Clinical Data, Radiographic and Microscopy Findings

In 1964, a 14-year-old girl attended the clinic complaining of a swelling in the anterior middle part of the tibia. X-ray examination revealed an osteolytic, multicystic lesion; the cortex had been made to bulge out by the tumor (Fig. 1). Histologically, the pre-existing bone was destroyed by the tumor, which consisted of two different parts. The first part was composed of epithelial-like cells that were arranged in nests and strands. The second part of the tumor consisted of a fibrous stroma, resembling the stroma seen in fibrous dysplasia, and so the diagnosis of adamantinoma of long bones was made. As the tumor was sharply delineated, the patient was only treated by excochleation with tumor-free margins.

Offprint requests to: Dr. M. Altmannsberger (address see above)

0171-5216/82/0104/0315/$1.20

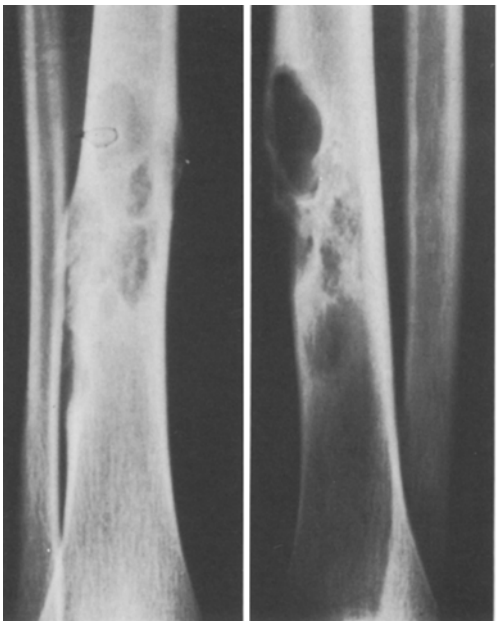


Fig. 1.

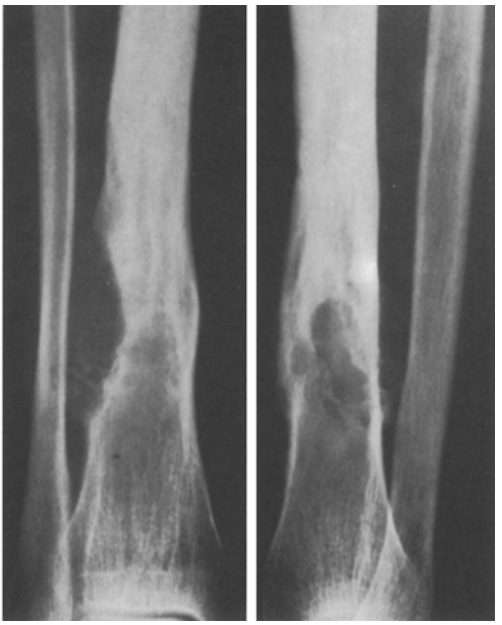


Fig. 2.

Fig. 1. Osteolytic destruction of tibia diaphysary medulla in combination with subcorticol, vesicular compacta osteolysis. Note the watch-glass-like elevation of corticalis lamella

Fig. 2. Local recurrence in the distal tibia diaphysis with large laterodorsal extraosseous component 10 years after excochleation and spongiosa filling

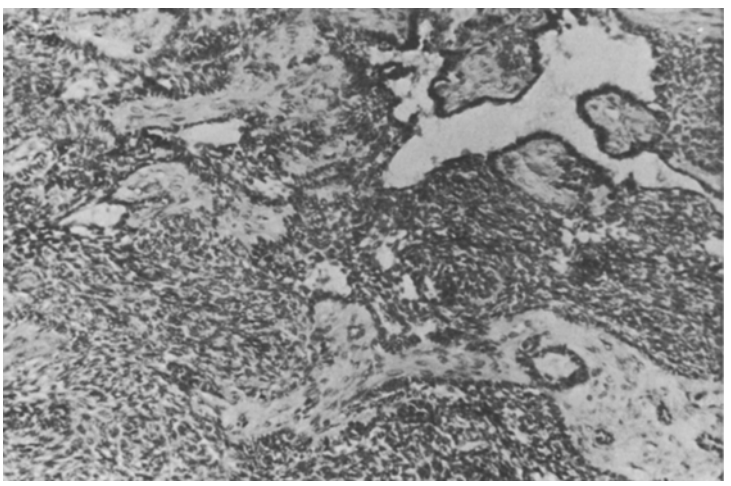
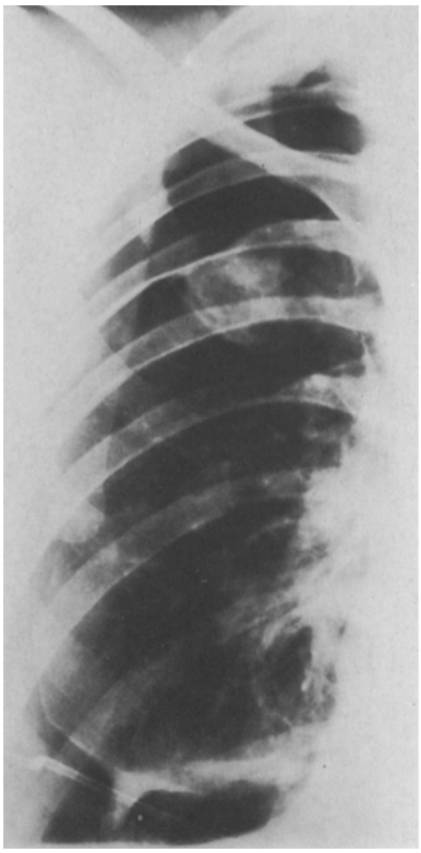


Fig. 3. Typical adamantinoma with epithelial-like tumor-cell formations arranged in a pseudoglandular pattern with intervening fibrous stroma. Note also tumor-cell formation in strands and nests (HE, $\times 200$)



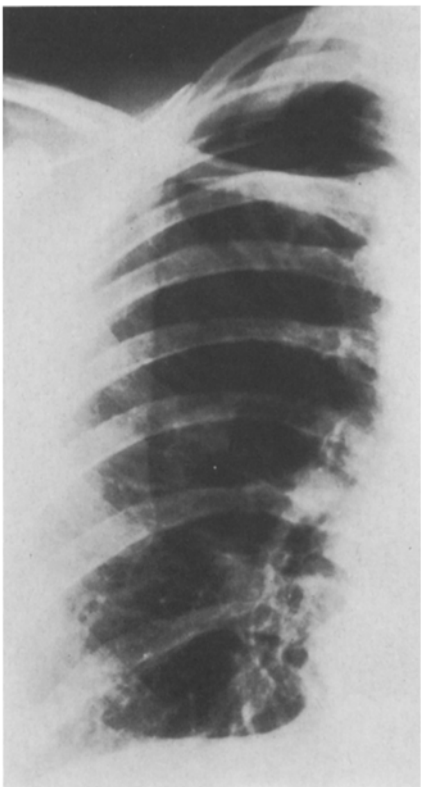


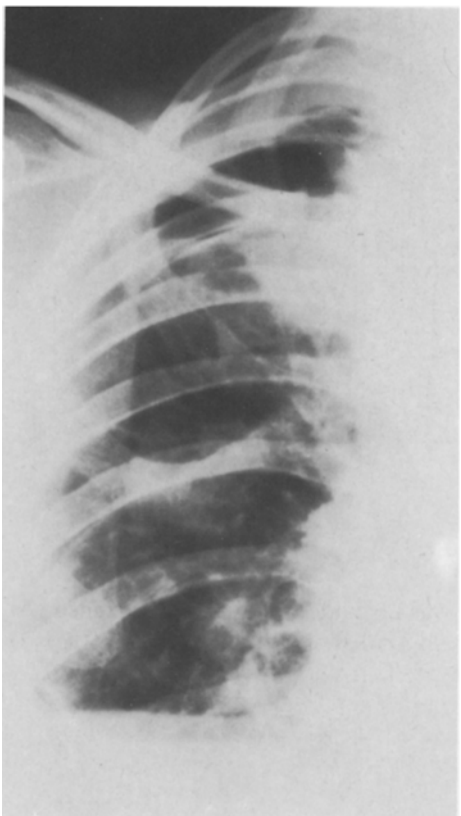
Fig. 4. **Fig. 5.**

Fig. 4. Right lung; pneumothorax and lung collapse without evidence of intrapulmonary metastasis

Fif. 5. Right lung; intrapulmonary metastasis near the hilus. Note rest of pneumothorax after drainage

For 10 years little happened, but in 1974 the tumor recurred at the previous site. Radiologically, the recurrent tumor demonstrated a multiple osteolytic lesion surrounded by multifocal areas of sclerosis. Contrary to X rays made in 1964, a large extraosseous tumor component was now evident (Fig. 2). Histological examination showed the same tumor as in 1964, but in contrast to results of the first histological test the cellularity and mitotic activity were highly increased (Fig. 3).

As the recurrent tumor had a large extraosseous component, a monobloc excision could not be performed and amputation was the treatment of choice. After another 5 years without any signs of recurrence or metastasis, a routine X ray of the chest revealed a pneumothorax on the right side. The patient had no complaints and despite a careful radiographic examination, the etiology of the pneumothorax could not be clarified, especially as no signs of lung metastasis were found (Fig. 4). In October 1979, a widening of the mediastinum to the right could be demonstrated. Between October 1979 and the middle of January 1980, the patient suffered from pneumothorax on the right and the left side. In the right pleura, an effusion could be demonstrated and at the same time intrapulmonary metastasis were evident (Fig. 5–7) At the end of January, the young woman developed a severe hypercalcemia (24 mg), which produced severe pancreatitis and later renal failure. The syndrome could not be improved and led to hypercalcemic coma. A week later the patient died from irreversible circulatory failure.



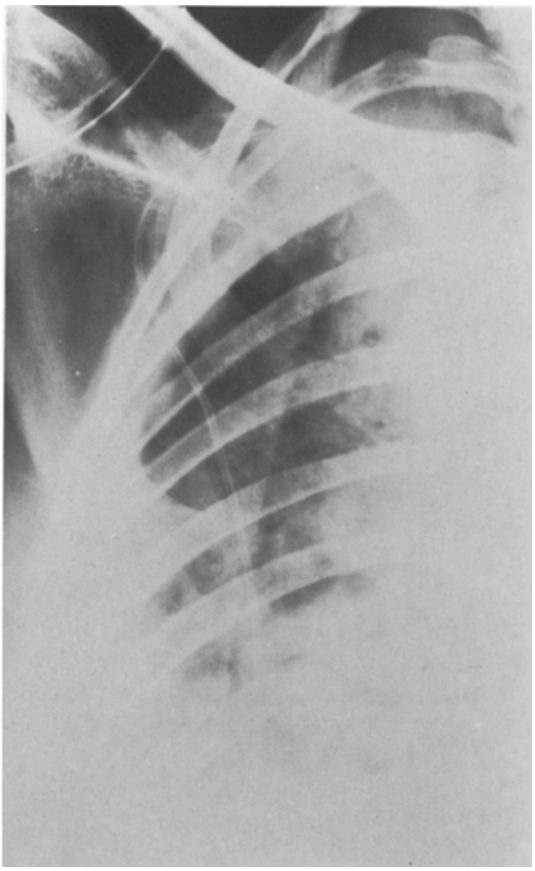


Fig. 6. **Fig. 7.**

Fig. 6. Right lung; intrapulmonary metastasis in the right middle and lower lobe and seropneumothorax with collapse of lung

Fig. 7. Advanced metastasis in the apical segment of the upper, middle and lower lobe. Note constant sero-pneumothorax and partial collapse of right lung

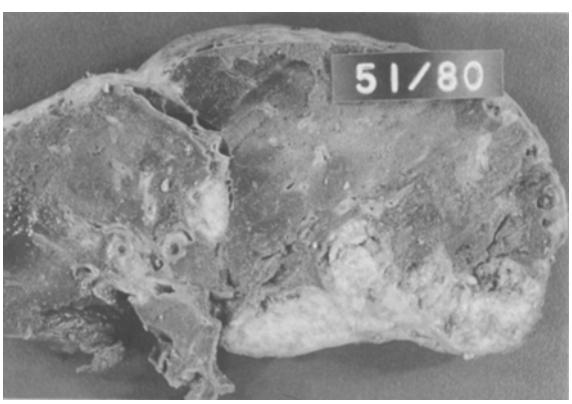


Fig. 8. Cross section of the right lung. The right pleura is covered by a tumor layer with a diameter of 2 cm. Note also intrapulmonary metastasis

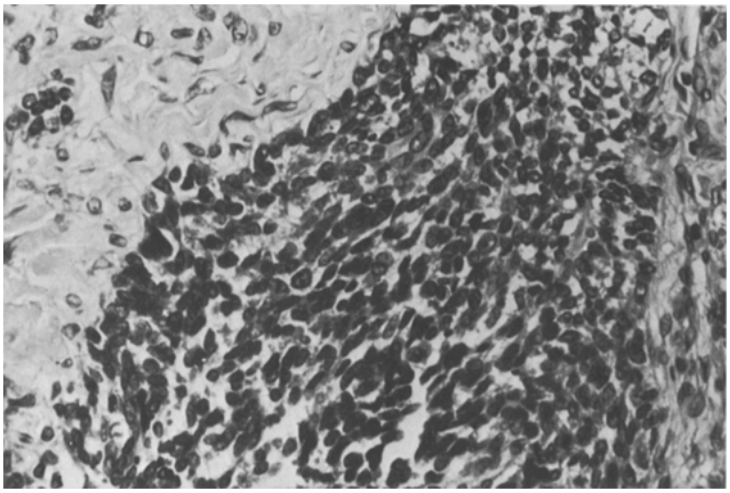


Fig. 9. High magnification of lung metastasis reveals the cellularity of the tumor (HE, 680 ×)

Autopsy Findings

In the amputation stump of the right leg there was no evidence of local recurrence and in the regional lymph nodes no tumor could be demonstrated either. On the contrary, the lungs showed diffuse nodular metastasis in all lobes and massive outbreak of the tumor in the right pleural cavity. The right pleura was covered by a tumor layer with a diameter of 2 cm, causing a total atelectasis of the right lung (Fig. 8). Contrary to these findings, the chest X ray 8 days before death showed in part ventilation of the right upper lobe. Histologically, the metastasis reavealed the same picture as the primary tumor and there was no evidence of a carcinoma of the bronchi or pleura (Fig. 9). A solitary bone metastasis in the third thoracic vertebra body could only be detected microscopically. The hypercalcemia caused severe haemorrhagic pancreatitis with multiple fat necrosis and acute peritonitis. The cause of death was an irreversible circulatory failure produced by the pancreatitis.

Discussion

In this case report, we want to emphasize the unusual course of disease, the recurrent formation of pneumothorax and the hypercalcemia. Adamantinoma of long bones is a rare bone tumor, mostly found in the tibia of adults in their twenties and thirties (Unni et al. 1979; Huvos 1979). In most cases, the tumor follows a benign course, but in 15–20% metastasis appear (Weiss and Dorfman 1977). Histologically, it is impossible to predict the biological behaviour of the tumor. Normally metastasis occur in the first 2 years after diagnosis but in this case the tumor was discovered in a 14-year-old patient. In the first histological examination the tumor had no features that allowed one to predict early metastasis and the first recurrence with the large extraosseous component did not appear until 10 years after diagnosis. Moreover, it is very surprising that in

spite of the high mitotic activity in the recurrent tumor, the lung metastasis appeared 6 years after the tumor had recurred. In this case, the development of pneumothorax could only be explained by a bronchopleural fistula close to the pleura and originating in intrapulmonary metastasis, although lung metastasis could not be deteced by X rays. It is very interesting that, in the literature, two other cases of metastasizing adamantinoma are reported, in which pneumothorax precedes the lung intrapulmonary metastasis by several months (Winter 1976). The hypercalcemia occurring a few days before death must be interpreted as a paraneoplastic syndrome, because during autopsy extensive bone metastasis or primary hyperparathyreodism could be ruled out; the four parathyroid glands were of normal size. For the pathogenesis of paraneoplastic hypercalcemia two possibilities are considered:

(1) the production of PTH or PTH-like substances in the tumor (Roof et al. 1971; Powell et al. 1973) and

(2) the production of ectopic prostaglandins (Seyberth et al. 1975; Klein and Raisz 1970).

In patients with myeloma an osteoclast-activating factor is also discussed (Mundy et al. 1974). In our case the PTH was several times in the normal range, so that prostaglandins seem to be the most likely factor for hypercalcemia. The histiogenesis of adamantinoma of long bones is so far not clear and is not discussed in this report.

References

Huvos AG (1979) Bone tumors – Diagnosis, treatment and prognosis. WB Saunders, Phildelphia Toronto London

Klein DC, Raisz GL (1970) Prostaglandins: Stimulation of bone resorption in tissue culture. Endocrinology 86:1436

Mundy GR, Raisz GL, Cooper RA (1974) Evidence for the secretion of an osteoclast stimulating factor in myeloma. N Engl J Med 291:1041

Powell D, Singer FR, Murray TM (1973) Nonparathyroid humoral hypercalcemia in patients with neoplastic diseases. N Engl J Med 289:176

Roof BS, Carpenter B, Fink DJ (1971) Some thought on the nature of ectopic parathyroid hormones. Am J Med 50:686

Seyberth HW, Serge GV, Morgan JN (1975) Prostaglandins as mediators of hypercalcemia associated with certain types of cancer. N Engl J Med 293:1278

Unni KK, Dahlin DC, Beabout JW, Ivins JC (1974) Adamantinomas of long bones. Cancer 34:1796–1805

Weiss SW, Dorfman MB (1977) Adamantinoma of long bone. An analysis of nine new cases with emphasis on metastasing lesions and fibrous dysplasia like changes Human Pathol 8:141–153

Winter WG jr (1976) Spontaneous pneumothorax heralding metastasis of adamantinoma of the tibia. Report of two cases. J Bone Joint Surg (Am) 58:416–417

Received November 15, 1981/Accepted March 12, 1982