Skeletal Radiol (1998) 27:406–409

CASE REPORT

Classic adamantinoma in a 3-year-old

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Abstract Classic adamantinoma of the long bones is a rare, low-grade malignant neoplasm arising most often in the tibia and usually in patients during the second to fifth decades. Although adamantinomas have been described in children, the histologic pattern in this age group is different from that seen in adults and resembles osteofibrous dysplasia. The usual pattern of adamantinoma in children has been termed "differentiated adamantinoma" and follows a benign course. We report a case of adamantinoma in the proximal tibia of a 3-year-old patient. The lesion had abundant epithelial component with formation of keratin pearls, a pattern that has been described only in classic adamantinoma occurring in adults. Since differentiated adamantinomas are essentialy benign and classic adamantinomas are low-grade malignancies, the finding of a classic variant at this young age raised important therapeutic and prognostic issues.

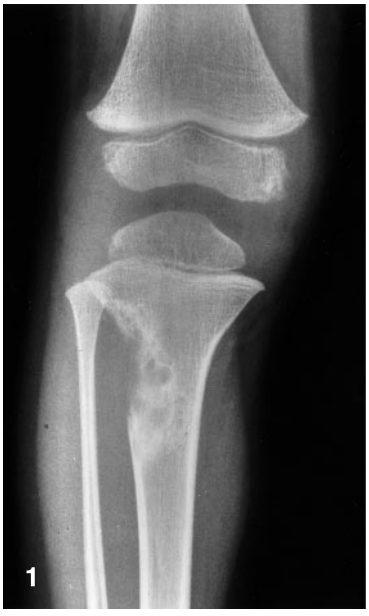
Key words Adamantinoma · Classic · Tibia · Child

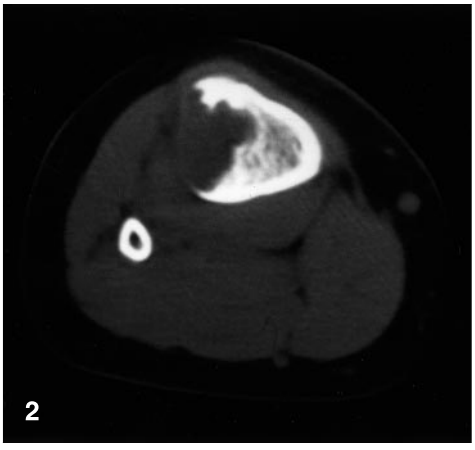
Introduction

Extragnathic adamantinoma is a primary intraosseous epithelial neoplasm of low-grade malignancy with a marked predilection for the tibia. Czerniak et al. [1] distinguished two types of extragnathic adamantinoma: the classic form and the differentiated form. The classic form usually presents in adults, grows beyond the cortex, and sometimes metastasizes. Differentiated adamantinoma occurs in the first two decades of life, has an intracortical location, and a favorable prognosis. Histologically, classic adamantinoma is characterized by an abundance of epithelial cells which stain strongly for cytokeratin. Differentiated adamantinomas, on the other hand, have a predominance of osteofibrous dysplasia pattern with only a small scattered inconspicuous epithelial cell component.

Case report

A 3-year-old boy presented with complaints of intermittent pain in the right calf, which started after a fall. On examination, the patient was a healthy child with slight tenderness over the proximal right tibia and without other findings or medical problems. Radiographs showed a destructive lesion of the lateral diametaphysis of the proximal tibia with cortical loss and a matrix of sclerosis and osteolysis (Fig. 1). CT confirmed the presence of a destructive lesion that was predominantly osteolytic, involving the cancellous bone and destroying the cortex (Fig. 2). A differential diagnosis of non-ossifying fibroma, eosinophilic granuloma, or a chondromyxoid fibroma, was entertained. A bone scan demonstrated





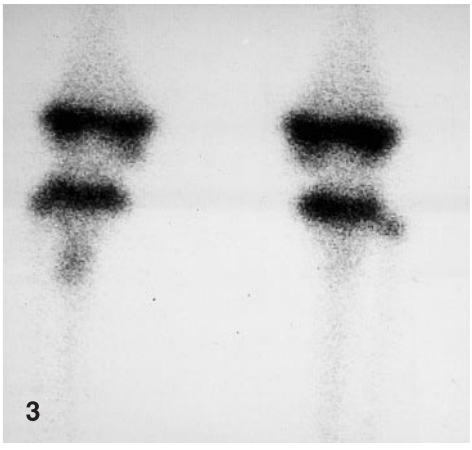


Fig. 1 Anteroposterior radiograph shows an eccentric, lytic, metaphyseal lesion with cortical destruction but no periosteal reaction. The lesion has a lobulated margin with a thin rim of sclerosis

Fig. 2 Axial CT section near the midportion of the lesion shows no mineralized matrix or periosteal reaction. The lobulated lesion involves the lateral cortex, but a thin rim of intact periosteal tissue is evident. There is no soft tissue mass

Fig. 3 Bone scan shows minimal activity in the area of the lesion

minimal increased uptake in the lesion, with intensity less than that observed in the epiphysis (Fig. 3). There were no other areas of abnormal increased radiotracer activity.

Curettage of the lesion with bone graft was planned. An intraoperative frozen section was obtained, which revealed a pauci-cellular fibro-osseous lesion with two keratin pearls (Fig. 4). A diagnosis of adamantinoma was suggested. Surgery was aborted, further biopsies were obtained, and the patient was scheduled for follow-up.

Histologic examination of the biopsy specimens showed a spindle-cell lesion with variable cellularity. The lesion was quite cellular in some areas, with a vague storiform pattern. In other areas the lesion was more collagenized and hypocellular. On careful examination, scattered small clusters of squamoid epithelial cells could be seen (Fig. 5). These plump cells were seen to line slit-like spaces in some areas, appearing to be plump endothelial cells. No keratinization of these cells was observed. No keratin pearls were seen. There was no pleomorphism of the spindle or epithelial cells. No mitotic figures were seen. Some biopsy fragments had tiny spicules of woven bone lined by plump osteoblasts in a fibrous background, resembling osteofibrous dysplasia (Fig. 6). Staining for cytokeratin revealed an abundant epithelial component in the neoplasm (Fig. 7). A diagnosis of adamantinoma was made.

The patient underwent resection of the proximal tibia just distal to the physis, with preservation of the growth plate. The defect was filled by use of a bone transport technique. The specimen consisted of a 5×3×3-cm piece of proximal tibia without the articular surface. Externally the bone had some soft tissue attached, but was otherwise unremarkable. On cut section in the coronal plane, a lesion, 3.5×1.0 cm, tan and partly red, was seen involving the lateral medullary cavity and adjacent cortex. The lesion had eroded the cortex, but did not extend beyond the periosteum. Microscopically, it was similar to the biopsy specimen, without evidence of keratinization. Post-operative course was complicated by a fracture. At the 12-month follow-up, the patient is disease free with, a healed stable extremity and a functioning growth plate.

Discussion

Primary bone tumors are unusual in children under the age of 5 years. Radiologic differential diagnosis, in our patient, included non-ossifying fibroma or fibroxanthoma, Langerhans cell histiocytosis, and chondromyxoid fibroma. Adamantinoma of the long bones is a rare, low-grade malignant neoplasm, that in 90% of the cases arises in the middle third of the tibia during the second to fifth decades of life [2]. Radiographically, most tumors involve both the cortex and the medulla and are frequently seen as multiple well-circumscribed lucent defects of variable size, with areas of sclerosis within the tumor and at the periphery. Bone expansion is noted in the majority of cases. In approximately 15% of cases, the tumor breaks through the cortex and involves the adjacent soft tissues [3].

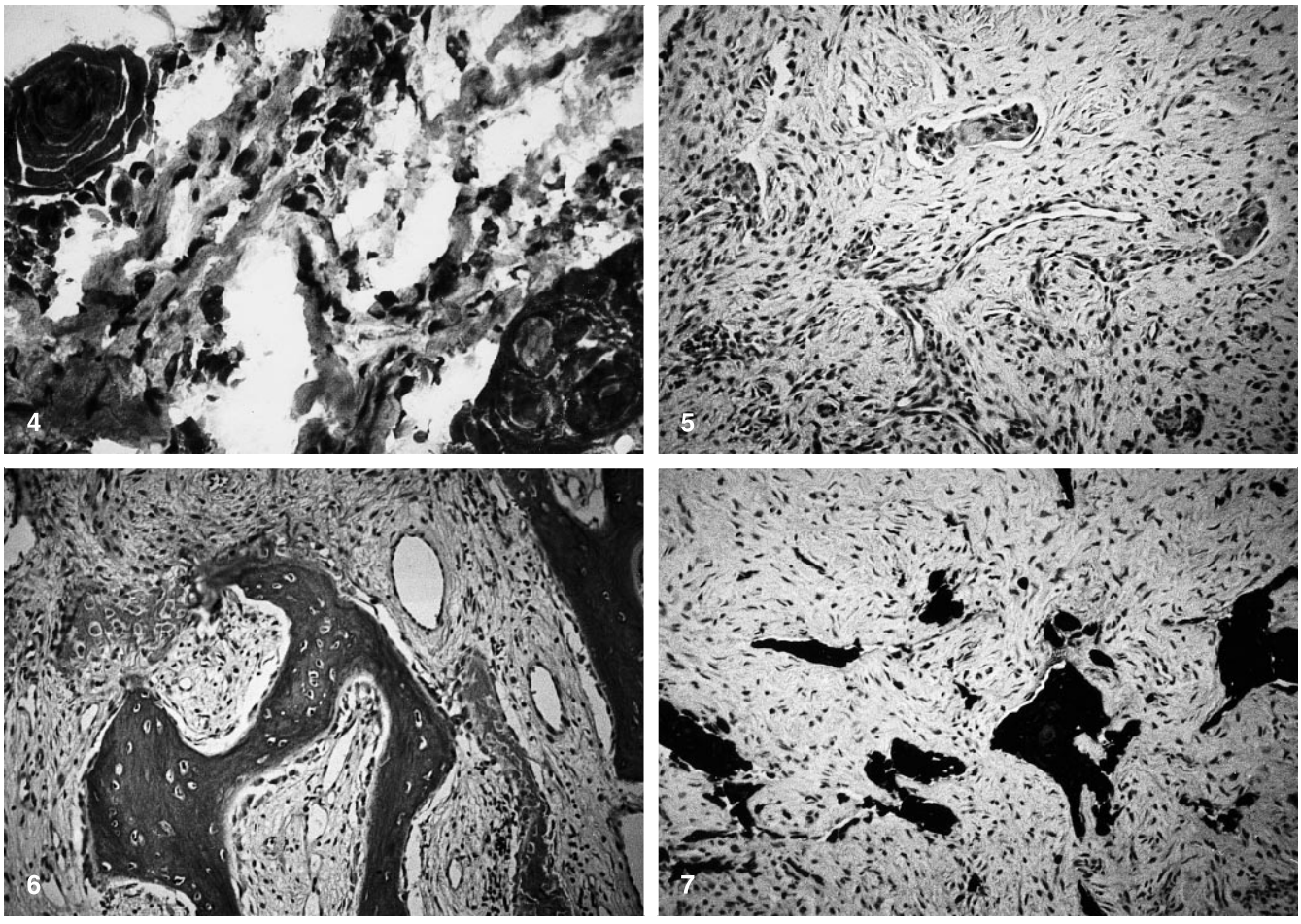


Fig. 4 Frozen section from the tibial lesion showing two keratin pearls and a spindle cell stromal background

Fig. 5 Adamantinoma demonstrating areas of spindle cell proliferation with nests of squamous cells. Some of the plump cells seem to line slit-like spaces

Fig. 6 Osteofibrous dysplasia-like areas in adamantinoma. There is spindle cell proliferation with production of osteoid that is lined by plump osteoblasts

Fig. 7 Adamantinoma stained with antibodies to cytokeratin. The squamous cell nests are strongly positive

The neoplasm tends to recur locally and is relatively insensitive to radiotherapy [4]; wide en bloc resection is the procedure of choice for initial treatment.

Czerniak et al. in 1989 [1] first proposed the concept of "differentiated adamantinoma" and established the radiologic, histologic, and immunohistologic features that distinguish this subtype from the more common "classic adamantinoma". The classic form usually presents in older patients, grows beyond the cortex, and sometimes metastasizes. Histologically, classic adamantinoma is characterized by an abundance of epithelial cells which stain strongly for cytokeratin. The differentiated adamantinoma, on the other hand, occurs at a young age (first two decades), has an intracortical location, and histologically shows a uniform predominance of an osteofibrous dysplasia-like pattern, with only small scattered inconspicuous epithelial cell component.

The relationship of osteofibrous dysplasia (OFD) to differentiated adamantinoma is unclear. Czerniak et al. [1] believe that there is a continuum of lesions, with classic adamantinoma at one end and OFD at the other. They postulate that in patients aged less than 20 years, an intracortically situated adamantinoma becomes dominated by a secondary reparative process having the histologic features of OFD. Continuation of this reparative process leads to elimination of recognizable tumor cells from the lesion and spontaneous regression – a hypothesis also supported by other investigators [5, 6]. This concept, however, does not rule out the possible existence of de novo osteofibrous dysplasia not related to adamantinoma. Many other investigators have suggested that OFD may be a precursor of adamantinoma [7–9]. Sweet et al. [10] studied 30 cases of OFD to determine whether it is a precursor lesion to adamantinoma. They concluded that there was no conclusive evidence of a precursor role for OFD.

Since differentiated adamantinomas do not metastasize, in contrast to classic adamantinomas, which are low-grade malignant neoplasms, distinction between these two types of adamantinoma has important therapeutic implications. The patient described in this report showed abundant epithelial component with islands of squamous cells, showing intercellular bridges and foci of keratin pearl formation. Keratin pearl formation is rare even in classic adamantinoma. In a study of 85 cases, well-formed squamous cells with keratin pearls were seen in only 9 cases [3]. Interestingly, none of these nine patients had recurrence of the tumor, but one patient developed metastasis. The authors concluded that of all the histologic features that were evaluated, the presence of squamous differentiation protected against recurrence, but not against metastasis.

This report describes the clinicopathologic findings of a 3-year-old patient with adamantinoma. The histology of the lesion would argue in favor of classifying it as a classic adamantinoma, a tumor with a potential to metastasize. The age of the patient, however, would favor a benign course. The patient is expected to have a good prognosis, since the lesion was excised en bloc. A lengthy follow-up will be needed to determine the final outcome.

References

1. Czerniak B, Rojas-Corona RR, Dorfman HD. Morphologic diversity of long bone adamantinoma. The concept of differentiated (regressing) adamantinoma and its relationship to osteofibrous dysplasia. Cancer 1989; 64:2319–2334.
2. Dahlin DC, Unni KK. Bone tumors: general aspects and data on 8,542 cases, 4th edn. Springfield, Illinois: Charles C. Thomas 1986:346–355.
3. Keeney GL, Unni KK, Beabout JW, Pritchard DJ. Adamantinoma of long bones. A clinicopathologic study of 85 cases. Cancer 1989; 64:730–737.
4. Weiss SW, Dorfman HD. Adamantinoma of long bone: an analysis of nine new cases with emphasis on metastasizing lesions and fibrous dysplasia-like changes. Hum Pathol 1977; 8:141–153.
5. Ueda Y, Blasius S, Edel G, Wuisman P, Bocker W, Roessner A. Osteofibrous dysplasia of long bones – a reactive process to adamantinomatous tissue. J Cancer Res Clin Oncol 1992; 118:152–156.
6. Bridge JA, Dembinski A, DeBoer J, Travis J, Neff JR. Clonal chromosomal abnormalities in osteofibrous dysplasia. Cancer 1994; 73:1746–1752.
7. Hazelbag HM, Taminiau AHM, Fleuren GJ, et al. Adamantinoma of long bones: a clinicopathological study of thirty-two cases with emphasis on histologic subtype, precursor lesion and biological behaviour. J Bone Joint Surg Am 1994; 76:1482–1499.
8. Springfield DS, Rosenberg AE, Mankin HJ, Mindell ER. Relationship between osteofibrous dysplasia and adamantinoma. Clin Orthop 1994; 309:234–244.
9. Hazelbag HM, Van den Broek LJCM, Fleuren GJ, Taminiau AHM, Hogendoorn PCW. Distribution of extracellular matrix components in adamantinoma of long bones suggests fibrous-to-epithelial transformation. Hum Pathol 1997; 28:183–188.
10. Sweet DE, Vinh TN, Devaney K. Cortical osteofibrous dysplasia of long bone and its relationship to adamantinoma. A clinicopathologic study of 30 cases. Am J Surg Pathol 1992; 16:282–290.