

CASE REPORT

ADAMANTINOMA OF THE TIBIA IN A NINE-YEAR-OLD CHILD

B. SARISOZEN*, K. DURAK*, C. OZTURK*

Adamantinoma of the long bones is a rare, low-grade malignancy with a marked predilection for the tibia and is usually seen in patients during the second to fifth decades of life. Adamantinomas have also been reported in children, but the histological pattern in this age group is different from that seen in adults. We report a case of adamantinoma of the tibia in a 9-year-old boy. Histologically, the lesion was osteofibrous dysplasia with an epithelial component, called “differentiated adamantinoma”. An osteofibrous dysplasia-like adamantinoma (differentiated adamantinoma) may be the precursor lesion of the classic type of adamantinoma.

Keywords : adamantinoma ; osteofibrous dysplasia ; long bones.

Mots-clés : adamantinome ; dysplasie ostéo-fibreuse ; os longs.

INTRODUCTION

Adamantinoma, also called malignant angio-blastoma, primary epidermoid carcinoma of bone, or epithelial tumor of bone, is a primary intrasosseous epithelial neoplasm of low-grade malignancy with an increased tendency toward involvement of the tibia and is usually seen in patients during the second to fifth decades of life. It is a rare neoplasm, accounting for 0.33 and 0.48%, respectively, of the malignant bone tumors in the series reported by Dahlin (4) and by Schajowicz (18).

Although cellular derivation of these tumors is still in dispute, recent investigations, using immunohistochemistry and electron microscopy, support an epithelial cell origin (15, 17). However, the origin of these cells with an epithelial phenotype is still unknown (8).

Although adamantinomas have been described in children, the histological pattern in this age group is different from that seen in adults. Czerniak *et al.* (3) distinguished two types of adamantinoma : the classic form and the differentiated form. The classic form usually presents in adults, grows beyond the cortex and sometimes metastasizes. The differentiated adamantinoma occurs in the first two decades of the life, has an intracortical location and a better prognosis. Histologically, the classic adamantinoma is characterized by an abundance of epithelial cells staining strongly for cytokeratin. Differentiated adamantinomas, on the other hand, have a predominantly osteofibrous dysplasia pattern with only a small, scattered epithelial cell component.

There has been extensive speculation about a possible relationship between classic adamantinoma, osteofibrous dysplasia-like adamantinoma and osteofibrous dysplasia, all of which originate mainly in the tibial cortex and have close radiographic and histological similarities (1, 3, 10, 14, 16). Difficulties in histological classification arise when a lesion with the radiographic features of osteofibrous dysplasia is seen to have superficial characteristics of osteofibrous dysplasia on routine histological sections, but scattered individual or small aggregates of keratin-positive epithelial cells on immunohistochemical analysis are found. Some

*Uludag University Faculty of Medicine, Department of Orthopedic Surgery, Bursa, Turkey.

Correspondence and reprints : Bartu Sarisozen, Uludag University Faculty of Medicine, Department of Orthopedic Surgery, 16059 Bursa, Turkey. E-mail: bartu@uludag.edu.tr.

reports from the literature (6, 8, 13, 19) stated that osteofibrous dysplasia and adamantinoma appear to be related, and osteofibrous dysplasia may be a precursor of adamantinoma.

The different nature and indolent course of this neoplasm have led to uncertainty regarding its treatment. Most authors are of the opinion that wide local resection is an adequate local therapy for most adamantinomas of the tibia, and that amputation should be reserved for local recurrences, reconstruction failures or for the lesion with a large soft tissue component (5, 7, 9, 15, 22).

CASE

A 9-year-old boy was referred to our hospital with a three-month history of a painful mass over the proximal one-third of the anterior aspect of the right tibia. On examination, the patient was a healthy child with slight tenderness, and besides the mass over the proximal right tibia, no other findings or medical problems. Vital signs such as body temperature, blood pressure and pulse were all normal. Except for alkaline phosphatase which was 236 U (normal range : 41-117 U), all the laboratory values were within normal limits. Xrays and magnetic resonance images of the leg showed a cortical lesion with a longitudinal orientation, reactive sclerosis and loculated bubbly appearance (fig. 1 a,b). A radiographic differential diagnosis of fibrous dysplasia, eosinophilic granuloma, chondromyxoid fibroma and nonossifying fibroma was considered. Tc 99m scintigraphy showed a focal increased uptake at the lesion site. There were no other areas of abnormal increased radiotracer activity. Other imaging studies such as sonography of the abdomen and thorax CT scans were normal. An open biopsy was performed at the center of the lesion.

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. There was no pleomorphism of the spindle and epithelial

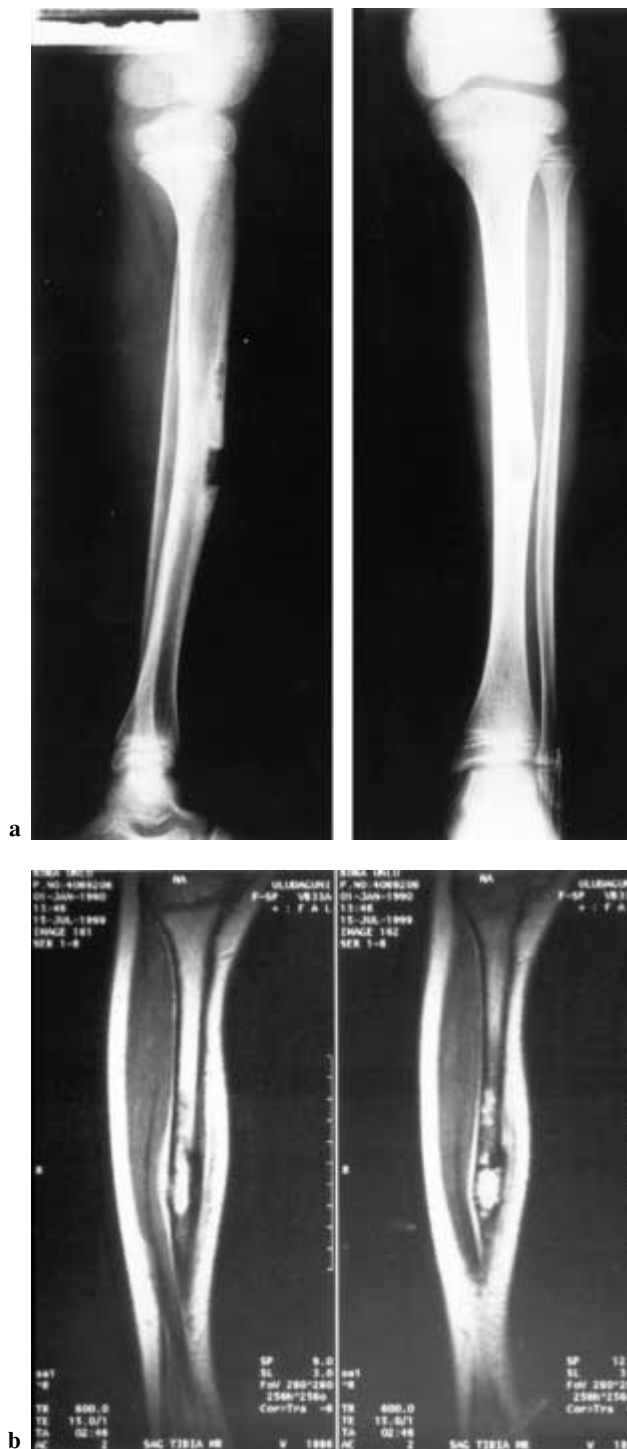


Fig. 1. — a. Direct x-rays of the leg after biopsy ; b. Magnetic resonance images showing the multiloculated tumor tissue.

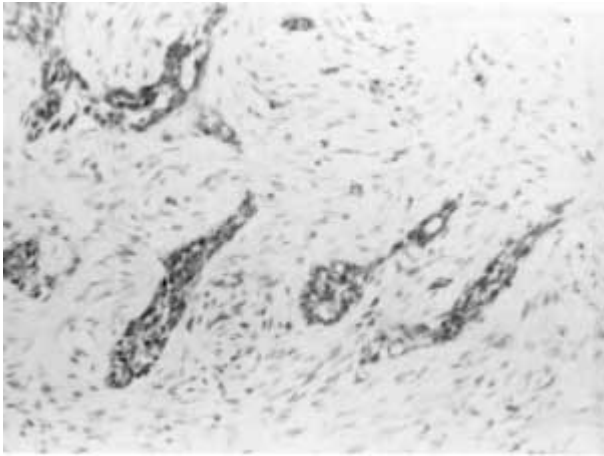


Fig. 2. — Frozen section from the tibial lesion showing scattered, small clusters of squamoid epithelial cells and a spindle-cell background. Staining for cytokeratin revealed epithelial nests.

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. Staining for cytokeratin revealed epithelial nests (fig. 2). A diagnosis of adamantinoma was made.

He was treated by wide resection (*en bloc* resection) of the lesion (10 cm long), and the involved leg was placed in an Ilizarov external fixator device. Healing of the intercalary defect was obtained by double corticotomy (carrying out trifocal osteosynthesis) with double distraction osteogenesis and compression between the transposed segments. Microscopically, the findings were similar to those made on the biopsy specimen. At the latest follow-up, 2 years after the diagnosis, the patient is free of local disease and distant spread. The length and function of the extremity are completely normal.

DISCUSSION

Primary bone tumors are unusual in children under the age of 10 years. Adamantinoma of the long bones is a rare, low-grade malignant neoplasm, which in 90% of the cases arises in the middle third of the tibia during the second to fifth

decades of life (12, 22). Radiographically, most tumors involve both the cortex and the medulla. Bone expansion is noted in the majority of the cases. In about 15% of the patients, the tumor breaks through the cortex and involves the adjacent soft tissues (11).

The neoplasm tends to recur locally and is relatively insensitive to radiotherapy (23). Wide *en bloc* resection is the radical treatment of choice. Chemotherapy may be given as an adjunct to surgery in selected cases, but is never effective by itself.

The relationship of osteofibrous dysplasia with adamantinoma is unclear. Czerniak *et al.* (3) reported that there is a continuum of lesions with classic adamantinoma at one end and osteofibrous dysplasia at the other. This hypothesis is also supported by other authors (2, 21). Mirra (14) considered this possible relationship by calling osteofibrous dysplasia “juvenile adamantinoma”. This concept, however does not rule out the possible existence of *de novo* osteofibrous dysplasia not related to adamantinoma. Sweet *et al.* (20) studied 30 cases of osteofibrous dysplasia to determine whether it is a precursor lesion to adamantinoma. They concluded that there was no conclusive evidence of a precursor role for osteofibrous dysplasia.

Since differentiated adamantinomas do not metastasize, in contrast to classic adamantinomas, the distinction between these two types of adamantinoma is important for treatment strategy.

When osteofibrous dysplasia is suspected on the basis of xrays, a biopsy should be performed at the center of the lesion, and when scattered epithelial cells are found, the possibility of progression to an osteofibrous dysplasia-like adamantinoma (differentiated adamantinoma) and ultimately to a classic adamantinoma should be seriously considered. Active follow-up with serial radiographs is advised in these patients (8).

This report describes the clinicopathologic findings in a 9-year-old boy with adamantinoma. The histology of the lesion was differentiated adamantinoma, a tumor without a potential to metastasize, with a benign course. The patient is expected to have a good prognosis, since the lesion was excised *en bloc*. Long term follow-up is needed to evaluate the final outcome.

REFERENCES

1. Alguacil-Garcia A., Alonso A., Pettigrew N. M. Osteofibrous dysplasia (ossifying fibroma) of the tibia and fibula and adamantinoma. A case report. *Am. J. Clin. Pathol.*, 1984, 82, 470-474.
2. Bridge J. A., Dembinski A., DeBoer J., Travis J., Neff J. R. Clonal chromosomal abnormalities in osteofibrous dysplasia. *Cancer*, 1994, 73, 1746-1752.
3. Czerniak B., Rojas-Corona R. R., Dorfman H. D. Morphologic diversity of long bone adamantinoma. The concept of differentiated (regressing) adamantinoma and its relationship to osteofibrous dysplasia. *Cancer*, 1989, 64, 2319-2334.
4. Dahlin D. C. Adamantinoma of long bones. In : Charles C. T., ed. *Bone Tumors. General Aspects and Data on 6221 Cases.* Lippincott-Raven, Illinois, 1978, pp. 296-306.
5. Enneking W. F. *Musculoskeletal Tumor Surgery.* Churchill Livingstone, New York, 1983, pp. 1488-1492.
6. Gaubert J., Durroux R. Adamantinoma and osteofibrous dysplasia of the tibia. *Rev. Chir. Orthop.*, 1995, 81, 333-337.
7. Gebhardt M. C., Lord F. C., Rosenberg A. E., Mankin H. J. The treatment of adamantinoma of the tibia by wide resection and allograft bone transplantation. *J. Bone Joint Surg.*, 1987, 69-A, 1177-1188.
8. Hazelbag H. M., Taminiau A. H. M., Fleuren G. J., Hogendoorn P. C. W. Adamantinoma of the long bones. A clinicopathological study of thirty-two patients with emphasis on histological subtype, precursor lesion and biological behavior. *J. Bone Joint Surg.*, 1994, 76-A, 1482-1499.
9. Huvos A., Marcove R. C. Adamantinoma of long bones. A clinicopathological study of fourteen cases with vascular origin suggested. *J. Bone Joint Surg.*, 1975, 57-A, 148-154.
10. Johnson L. C. Congenital pseudoarthrosis, adamantinoma of long bone and intracortical fibrous dysplasia of the tibia. *J. Bone Joint Surg.*, 1972, 54-A, 1355-1364.
11. Keeney G. L., Unni K. K., Beabout J. W., Pritchard D. J. Adamantinoma of long bones. A clinicopathological study of 85 cases. *Cancer*, 1989, 64, 730-737.
12. Kumar D., Mulligan M. E., Levine A. M., Dorfman H. D. Classic adamantinoma in a 3-year-old. *Skeletal Radiol.*, 1998, 27, 406-409.
13. Kuruvilla G., Steiner G. C. Osteofibrous dysplasia-like adamantinoma of bone : report of five cases with immunohistochemical and ultrastructural studies. *Hum. Pathol.*, 1998, 29, 809-814.
14. Mirra J. M. Adamantinoma and osteofibrous dysplasia. *Bone Tumors. Clinical, Radiologic, and Pathologic Correlations.* Lea and Febiger, Philadelphia, 1989, pp. 1204-1231.
15. Moon N. F., Mori H. Adamantinoma of the appendicular skeleton. *Clin. Orthop.*, 1986, 204, 215-237.
16. Park Y. K., Unni K. K., McLeod R. A., Pritchard D. J. Osteofibrous dysplasia : Clinicopathologic study of 80 cases. *Hum. Pathol.*, 1993, 24, 1339-1347.
17. Perez-Ateyde A. R., Kozakewich H. P. W., Vawter G. F. Adamantinoma of the tibia. An ultrastructural and immunohistochemical study. *Cancer*, 1985, 55, 1015-1023.
18. Schajowicz F. *Tumors and Tumor Like Lesions of Bones and Joints.* Springer, New York, 1981, pp. 384-391.
19. Springfield D. S., Rosenberg A. E., Mankin H. J., Mindell E. R. Relationship between osteofibrous dysplasia and adamantinoma. *Clin. Orthop.*, 1994, 309, 234-244.
20. Sweet D. E., Vinh T. N., 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.
21. Ueda Y., Blasius S., Edel G., Wuisman P., Bocker W., Roessner A. Osteofibrous dysplasia of long bones- A reactive process to adamantinomatous tissue. *Cancer Res. Clin. Oncol.*, 1992, 118, 152-156.
22. Unni K. K., Dahlin D. C., Beabout J. W., Ivins J. C. Adamantinoma of long bones. *Cancer*, 1974, 34, 1796-1805.
23. Weiss S. W., Dorfman H. D. Adamantinoma of long bone : An analysis of nine cases with emphasis on metastasizing lesions and fibrous dysplasia-like changes. *Hum. Pathol.*, 1977, 8, 141-153.

SAMENVATTING

B. SARISOZEN, K. DURAK, C. OZTURK. Tibia adamantinoma bij negen jarig kind : Gevalstudie.

Adamantinoma van schaftbeenderen is een zeldzaam, zwak kwaadaardig gezwel met voorkeur voor de tibia bij mensen in hun 2^o tot 5^o decennium.

Het histologisch aspect bij kinderen verschilt tegenover dit bij volwassenen.

Een geval bij een negen jarig kind wordt beschreven. Histologisch vond men een gedifferentieerd adamantinoma : dit wil zeggen, osteofibreuze dysplasie met epitheliale bekleding, wat een voorloper kan zijn van het klassieke adamantinoma.

RÉSUMÉ

B. SARISOZEN, K. DURAK, C. OZTURK. Adamantinome du tibia chez un enfant de neuf ans.

L'adamantinome des os longs est une tumeur rare, de faible malignité, qui montre une prédilection marquée

pour le tibia et se rencontre habituellement dans les tranches d'âge entre 10 et 50 ans. On a également observé des adamantinomes chez l'enfant, mais le type histologique est alors différent de celui de l'adulte. Les auteurs présentent le cas d'un garçon de 9 ans qui présentait un adamantinome du tibia. La lésion se

présentait histologiquement comme une dysplasie ostéofibreuse avec une composante épithéliale, correspondant à un «adamantinome différencié». Une lésion de ce type (adamantinome différencié) peut être le précurseur d'un adamantinome de type classique.