Acta Scientific Orthopaedics (ISSN: 2581-8635)

Volume 7 Issue 4 April 2024

Case Report

Adamantinoma of the Tibia; Case Report

Carlos Humberto Franco Moran[1], Alicia Tamayo Figueroa[2], Ragnar Calzado Calderón[2]*, Ingrid Quesada Villa[3], Jessael Eliecer Ramírez Bateca[1] and Vladimir Andrés Vásquez Encalada[1]

[1]*First Degree Specialist in Orthopaedics and Traumatology, Cuba*

[2]*Specialist in Orthopedics and Traumatology Report of a Case, Cuba*

[3]*Specialist in First Degree in Comprehensive General Medicine and First Degree in Pathological Anatomy, Cuba*

***Corresponding Author:** Ragnar Calzado Calderón, Specialist in Orthopedics and Traumatology Report of a Case, Cuba.

DOI: 10.31080/ASOR.2024.07.0938

Received: March 13, 2024

Published: March 26, 2024

Abstract

Adamantinoma is a malignant tumor of epithelial origin with a low incidence. This article aims to describe the clinical, radiology and anatomopatology diagnosis of a patient with adamantinoma of the tibia. A 60 years old female patient is presented with the diagnosis of adamantinoma located in the upper middle third of the right tibia. The most relevant clinical findings are pain and swelling in the anterior aspect of the middle and upper thirds of the right leg, imaging shows extensive intracortical osteolytic lesion with bone destruction at the cortical level of the anterior tibia with periosteal reaction and increased density of soft parts. The treatment carried out was excision and biopsy of the tumor, the histopathological results report a predominance of epithelial cells with squamous differentiation and non-extensive keratin production, in addition to dermosomes and monofilaments basement membrane, which allows it to be framed within the Adamantinomas of the appendicular skeleton.

Keywords: Adamantinoma; Bone Neoplasms

Introduction

Adamantinoma is a primary bone tumor of rare onset, low degree of malignancy and epithelial origin. Its nomenclature comes from the Greek and means very hard. It was first observed in 1900 in the shaft of the ulna by Maier C [1,2] who identified it as a carcinoma. It was coined by Fisher. B. in 1913 because of its morphological similarity to the Adamantinoma of the mandible [3], but histologically they are not related. Campanacci M in 1976 named it osteofibrodysplasia of the tibia and fibula because of its anatomical location, origin, development and histological resemblance to fibrous dysplasia [4]. It accounts for between 0.1% and 0.3% of primary bone tumors [1,5,6], men are most often affected and in men, the tumor is more aggressive. The highest incidence occurs between 10 and 35 years of age [7-9], but cases have been reported from 2 to 86 years of age and individuals who have black skin color are predisposed. It usually appears mainly in the tibia (80%-90% of cases) [1-3,5,9].

The clinical manifestations are insidious and slow-onset. The signs and symptoms will depend on the extent and location of the tumor, the most common being the increase in volume with or without pain of the affected area, deformities and pathological fractures may also appear [1,2,5].

Adamantinoma appears as a lytic, elongated, eccentric, circumscribed lesion on plain radiographs.1, 9 With tomography and MRI, non-specific findings are observed, although intraosseous extension, lesions at distances, and the presence of metastatic lesions can be appreciated [1,10,11].

Macroscopically it is lobed, well defined, gray or white, elastic. You may have pockets of hemorrhage and necrosis [1,3,4,10,12].

Microscopically, it is composed of epithelial cells surrounded by a fibrous stroma formed by spindle-shaped collagenous fiber-pro-

ducing cells. Malignant epithelial cell nests are columnar in nature with palisade-like periphery. In some cases, the fibrous stroma contains bony spicules with osteoblast margins reminiscent of osteofibrous dysplasia [1,3,7].

The aim of this study was to show the clinical, radiological and pathological diagnosis made to a patient with Adamantinoma of the tibia treated at the Tumor Service of the Frank País Orthopedic Scientific Complex.

Case Report

A 60-year-old female patient, black skin color, with a history of controlled arterial hypertension, glaucoma and right breast nodule, with a history of three months ago having begun to present pain at the level of the upper middle third of the anterior aspect of the right leg accompanied by an increase in volume in that area, for this reason she went to the doctor on several occasions and performed treatments with analgesics. anti-inflammatories, and physiotherapy without improving the painful symptoms.

She was seen in another hospital where a radiographic study of her right leg was performed, where osteolytic lesions were observed at the level of the middle and upper thirds of the right tibia, so she was admitted to the Tumor Service of the Frank País International Orthopedic Scientific Complex for study and treatment.

On physical examination, the patient presented claudication at the expense of the right lower limb, where there was a diffuse increase in volume on the anterior aspect of the upper third of the right leg, seven centimeters in diameter, with smooth and shiny skin without changes in color or collateral circulation. This tumor is rounded, with a smooth surface, irregular edges, painful on superficial and deep palpation, adhering to deep planes, with a slight increase in local temperature. Mobile, soft and painless lymphadenopathy is palpable in the right inguinal region. The rest of the physical exam is normal.

Complementary exams

HB 11.4 G/L.	When: 115.7 mmol/l
HTO: 0.35%.	Glucose: 7.7 mmol/l
Erythrocyte sedimentation rate: 4.7 mm	Alkaline phosphatase: 262 U/l
Uric Acid: 303.0 mmol/l	C-reactive protein: negative
Calcium: 2.4 mmol/l	Rheumatoid factor: negative
Fosforo: 0.7 mmol/l	HIV-negative
Serology: non-reactive	

Table 1

Imaging studies

- **Plain radiograph:** On the anterior aspect of the proximal third of the shaft of the right tibia, an osteolytic image of six centimeters with poorly defined contours is observed, which destroys the anterior cortical and extends intracortically and distally with little trabeculation inside. (Anteroposterior and lateral projection).
- **CT scan:** In the sections and reconstructions carried out, extensive intracortical osteolytic lesion and bone destruction at the level of the anterior cortical of the diaphysis of the right tibia with an extension of plus or minus 19 cm in sagittal section associated with periosteal reaction, increase in volume and density of the adjacent soft tissues, in addition to towards the external aspect of the distal metaphysis of said tibia, an osteolytic lesion measuring 1.5 cm with irregularity and rupture of the cortical in several segments and another of the same characteristics on the posterior aspect (Figure 1 and Figure 2).

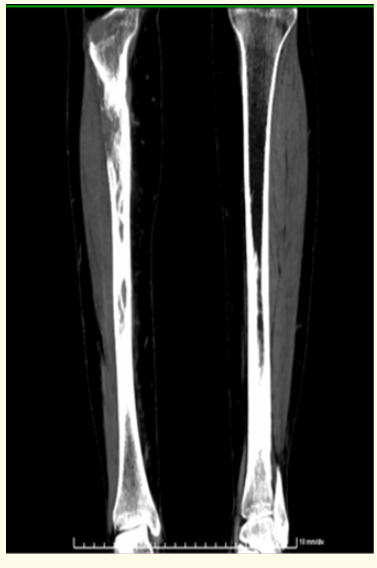

Figure 1

Chest X-rays and CT scans

In the sections and reconstructions performed, small hyperdense micronodular images located in the lower lobe of both lungs are observed, of possible secondary origin.

Scintigraphic bone survey

Intense fixation of the radiopharmaceutical 99mTc-MDP in the entire upper and middle third of the right tibia and in its distal me-

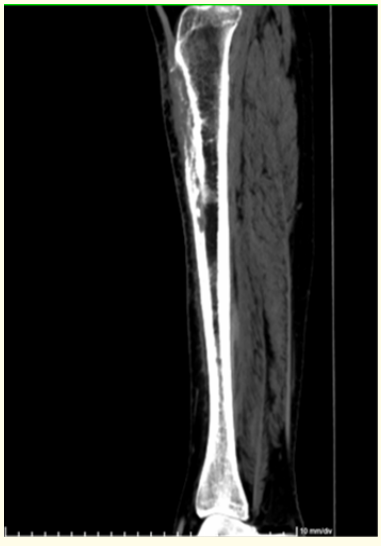

Figure 2

taphysis (Figure 3), and at the level of the vertebral body of the 5th dorsal vertebra (Figure 4). Rest of the study within normal limits.

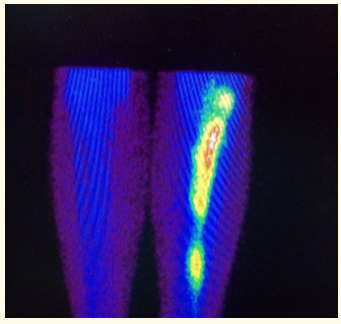

Figure 3

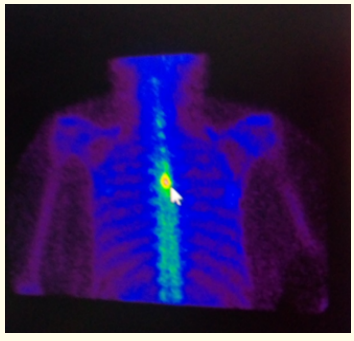

Figure 4

The patient is taken to the operating room for biopsy sampling. An anterior approach is performed in the proximal third of the right tibia, the skin and subcutaneous cellular tissue are detailed, the periosteum is opened and through a window carved into the external cortical of the tibia, abundant tumor tissue with a dense fibrous appearance of a whitish pearly color is curetted, the cavity is washed with saline solution, hydrogen peroxide and povidone iodine and the surgical plans are closed. A posterior antalgic splint was placed for a period of two weeks, followed by the placement of an orthopedic device such as a posterior silicone splint until definitive treatment was decided to avoid pathological fracture.

- **Histology:** Macroscopic description: Pathological anatomy received reddish-brown bone scraping material with a volume of 1 cubic cm.
- **Microscopic description:** Several fragments of bone tissue are observed under the microscope, almost entirely replaced by nests or islets of epithelial cells resting in a fibrous stroma without nuclear atypia or mitotic activity. The epithelial cells are arranged in these nests that have palisade on the periphery, they also form cell cords that leave gland-like spaces, empty or with fluid. Epithelial cells are eosinophilic, have sparse well-defined cytoplasm, dense nuclear chromatin, and low degree of nuclear atypia. Bone trabeculae, small foci of necrosis, and hemorrhages are observed. Presence of low mitotic activity.

Immunohistochemistry

- Vimentina (V): Intensely positive.
- Cytokeratin (CK): focally positive
- S 100- Negative
- CD34 Negative

Discussion

Adamantinoma is an uncommon primary bone tumor with a low degree of malignancy and debated histogenesis. Its diagnosis is the result of the variable clinical picture and imaging and pathological studies [1-3].

The most widely accepted theory about its origin mentions the displacement of the basal epithelium from the skin during embryonic development and is supported by its predominant presentation towards the anterior cortical of the tibia, where encondral ossification occurs very close to the skin [2,3].

It occurs mainly in male, Negroid patients, in mature skeletons, usually between the second and fifth decade of life, with an average between 25 and 35 years of age in 75% of patients [1-3], although it has been reported in children, with two years of age [9,13].

He has a predilection for lake bones. Its most common location is in the tibia (80% - 90%) although it can be seen in other long bones such as the forearm and humerus as well as in the hands, feet, ribs and spine [1-3,7,14-18].

On the anterior aspect of the tibia. Localized pain, pathological fracture, and bone deformity are other features that may appear [19]. Jain D and Anoumou MN are of the opinion that adamantinoma is characterized by a very slow clinical course, local recurrences, and pulmonary metastases [1,19]. The duration of symptoms, depending on the location and extent, can vary from weeks to years, and usually only non-specific symptoms are present: pain and swelling [1,9,12,20].

Adamantinoma appears as a lytic, elongated, eccentric, circumscribed lesion on plain radiographs. The anterior cortex of the diaphysis of the distal tibia is the most affected site (85% - 90%), in 10% there is a synchronous location in the ipsolateral fibula [1,17,18,20].

Usually, the lesion has several lytic defects separated by sclerotic bone, which gives it a bubble-like appearance [1,17]. It can spread to the entire bone with "satellite lesions" [17]. There is cortical rupture but little periosteal reaction. According to Greenspan A, areas of cortical destruction in the tibia present tooth imaging that helps distinguish the tumor [17]. The lesion may go through the cortex and spread to the soft tissues.

There may be multiple lesions next to normal tissue. The existence of a soft tissue mass is a sign of tumor aggressiveness and is not accompanied by calcification. According to Desai SS., *et al.* location in the tibia and intracortical development are two arguments indicative of adamantinoma [7,9,14,17].

Computed tomography (CT) shows cortical damage and extension to soft tissues, and intraosseous extension of the tumor is visualized. It plays an important role in the detection of lung metastases [1,9,14].

Magnetic resonance imaging (MRI) helps determine the degree of intra- and extraosseous involvement [11,18,20]. They are especially useful for detecting distant cortical foci, intramedullary extension, and soft tissues; determine tumor-free margins, and plan reconstructive surgery. MRI shows hypointensity on T1 imaging and hyperintensity on T2 imaging, typical of most tumors [1,11]. Occasionally, a liquid-liquid level may be seen. Van der Woude HJ., *et al*. describe two morphological patterns: solitary lobed focus and multiple small nodules in one or more foci [1,11,20].

Bone scintigraphy reveals an increase in 99mTc-Methylphosphonate uptake in all three phases of the study and may show the coexistence of lesion in the ipsilateral fibula [21].

Macroscopically it is lobed, well defined, gray or white, elastic. You may have pockets of hemorrhage and necrosis.

Microscopically, it is composed of epithelial cells surrounded by a fibrous stroma formed by spindle-shaped collagenous fiber-producing cells. Malignant epithelial cell nests are columnar in nature with palisade-like periphery. In some cases, the fibrous stroma contains bony spicules with osteoblast margins reminiscent of osteofibrous dysplasia [2,7-9,20].

Adamantinoma is a low-grade, biphasic malignant tumor. It is classified into two types: differentiated Adamantinoma or osteofibrous dysplasia type and classic Adamantinoma [1] which, due to the different growth patterns, have been described in some histological varieties, such as:

- A tubular variety in which epithelial cells are arranged in narrow cords and glandular spaces that are the result of loss of cell cohesion, rather than glandular formation per se.
- **Basaloid variety**: Reminiscent of basal cell carcinoma with the presence of peripheral palisade in the nests of epithelial cells.
- **Squamous variety:** Reminiscent of squamous cell carcinoma well differentiated by the formation of keratin pearls and globes.
- **Variety of spindle cells:** It is given by the spindle appearance of neoplastic epithelial cells, producing patterns reminiscent of mesenchymal neoplasms with storiform and fascicular patterns. The key to its identification is the tendency to form fissures or tubular spaces.
- **Adamantinoma type Ewing Sarcoma or Adamantinoma type Ewing:** This is an uncommon variety. It is composed of anastomosed cords of small, round, uniform cells arranged in a myxoid stroma. However, the demonstration of a translocation on the chromosome (11:22) of this tumor favors a close

pathogenetic relationship with Ewing's sarcoma, rather than with a classic Adamantinoma [8,16].

- **Differentiated adamannoma or osteofibrous dysplasia type:** Which presents a pattern similar to osteofibrous dysplasia with few nests of epithelial cells.

A few varieties can often be seen intermingled in the same tumor.

IHC (immunohistochemistry)

Despite the histological varieties of Adamantinoma, all of these are positive for pancytokeratin, suggesting a common histogenesis for all Adamantinoma subtypes. In addition, they are vimentin positive. These immunohistochemical patterns are preserved in local recurrences and metastases.

Adamantinoma differs from other bone and soft tissue neoplasms with recognized epithelial characteristics such as synovial sarcoma, chordoma and epithelioid sarcoma, as they lack immunoreactivity for keratins [8,18], express CD99, BCL2 and CD34, a myoepithelial marker [3].

The positivity of epithelial cells for the proliferation marker Ki-67, antibodies to epidermal growth factor and fibroblast growth factor type 2 demonstrates that this component constitutes the primary proliferation of neoplastic cells capable of stimulating reactive fibroblast growth [11,22].

The current treatment of Adamantinoma is aimed at salvage of the limb by en bloc resection of the tumor with effective oncological margins, plus reconstruction [23,24], which offers fewer recurrences and increases survival to 10 years. Amputation has not been shown to improve survival compared to limb-sparing surgery, and should be reserved for cases where en bloc resection cannot be performed with oncological margins, there is soft tissue extension, reconstruction fails, or local recurrences occur [1,23,24]. Unfortunately, neither radiotherapy nor chemotherapy has demonstrated efficacy in treating this lesion. Adamantinoma is highly radio-resistant and has low chemosensitivity [9].

Adamantinoma presents a wide variety of histological and radiological appearances, but there is no discernible predictive sign of clinical course [9,21]. Factors associated with an unfavorable clinical outcome include male sex, younger than 20 years of age, initial intralesional treatment, short duration of symptoms, pain at presentation, and poor squamous differentiation of the tumor [23], late diagnosis, and the type of treatment used [20]. The main prognostic factor for relapses is compromised surgical margins.

Adamantinomas are slow-growing, sometimes lasting more than 15 years from symptom onset to surgical treatment without metastases. Between 15% and 20% of cases metastasis occurs in advanced stages of the disease [20,24,25], being more frequent in the lungs (15%), skeleton, lymph nodes (7%), pericardium and liver [6]. The median survival with metastatic disease is 12 years.

Adamantinoma should be differentiated by its clinical and radiological similarities from metastases, epithelial tumor to vascular tumor, fibrous dysplasia, and osteofibrous dysplasia.

Therefore, we conclude that our patient underwent curettage of the lesion, to take a sample for biopsy, being evidenced from the clinical, radiological and anatomopathological point of view that it is Adameninoma of the long bones. She presented a good post-surgical clinical evolution and is being treated oncologically while waiting for future procedures for the treatment of the lesion.

Conflict of Interest

There is no conflict of interest among the authors of this paper.

Summary

Adamantinoma is a malignant tumor of epithelial origin with a low incidence. The aim of this article is to describe the clinical, radiological and pathological diagnosis of a patient with Adamantinoma of the tibia. We present a 60-year-old female patient with the diagnosis of Adamantinoma located in the middle-upper third of the right tibia. The most relevant clinical findings are pain and inflammation in the anterior aspect of the middle and upper thirds of the right leg, imaginologically extensive intracortical osteolytic lesion with bone destruction at the level of the anterior cortical of the tibia with periosteal reaction and increased density of soft tissues. A biopsy of the tumor was performed, and the histopathological results reported a predominance of epithelial cells with squamous differentiation and non-extensive keratin production, as well as a basement membrane, dermosomes and monofilaments, which allowed it to be included within the Adamantinomas of the appendicular skeleton.

Bibliography

1. Alicia Tamayo Figueroa., *et al*. "Adamantinoma of the tibia". *Cuban Journal of Orthopedics and Traumatology* 31.2 (2017): 9-12.

2. Jain D., *et al*. "Adamantinoma: A clinicopathological review and update". *Diagnostic Pathology* 3 (2008): 8.

3. Fischer B. "About a primary adamantinoma of the tibia". *Virchow's Arch [B]* 12 (1913): 422-441.

4. Most MJ., *et al*. "Osteofibrous dysplasia and adamantinoma". *Journal of the American Academy of Orthopaedic Surgeons* 18.6 (2010): 358-366.

5. Landeras RM., *et al*. "Adamantinoma of tibia. Review of a case". *Radiology* 41.1 (1999): 62-64.

6. McWilliams S and Weerakkody Y. "Adamantinoma". *Radiopaedia* (2017).

7. Hogendoorn P and Kanamori M. "Adamantinoma". En: Fletcher C, Bridge JA, Hogendoorn P, Mertens F (eds). WHO: Classification of tumors of soft tissue and bone. 4th ed. Lyon: IARC (2013): 343-345.

8. Gaubert J and Durroux R. "Adamantinoma of the tibia and osteofibrodysplasia". *Revue de Chirurgie Orthopédique et Traumatologique* 81.4 (1995): 333-337.

9. Qureshi AA., *et al*. "Current Trends in the Management of Adamantinoma of Long Bones: An International Study". *The Journal of Bone and Joint Surgery* 82A.8 (2000): 1122-1131.

10. Torrents C., *et al*. "Adamantinomade long bones. Radiological diagnosis". *Radiology* 32 (1990): 173-176.

11. Van der Woude HJ., *et al*. "MRI of adamantinoma of long bones in correlation with histopathology". AJR *American Journal of Roentgenology* 183.6 (2004): 1737-1744.

12. Ulmar B., *et al*. "Classical and atypical location of Adamantinomas - presentation of two cases". *Onkologie* 29.6 (2006): 276-278.

13. Van Rijn R., *et al*. "Adamantinoma in childhood: report of six cases and review of the literature". *Pediatric Radiology* 36 (2006): 1068-1074.

14. Keeney GL., *et al*. "Adamantinoma of long bones. A clinicopathological study of 85 cases". *Cancer* 64.3 (1989): 734-737.

15. Dini LI., *et al*. "Adamantinoma of the spine: case report". *Neurosurgery* 59.2 (2006): E426.

16. Anoumou MN., *et al*. "Adamantinoma of the Tibia: A Case Report". *Journal of Orthopaedic Case Reports* 4.1 (2014): 15-17.

17. Greenspan A. "Other bone tumors, malignant bone tumors II: miscelaneous tumors". En: Greenspan A (ed). Orthopaedic imaging: Apractical approach. 5th ed. Philadelphia: Wolters Kluwer, Lippincott Williams and Wilkins (2011): 754-755.

18. Desai SS., *et al*. "Adamantinoma of Tibia: A Study of 12 Cases". *Journal of Surgical Oncology* 93.5 (2006): 429-433.

19. Khan ., *et al*. "Leg pain and swelling in a 22-yearold man". *Clinical Orthopaedics and Related Research* 448 (2006): 259-266.

20. Czerniak B. "Dorfman and Czerniak´s Bone Tumors". 2nd edition. Philadelphia: Elseiver Saunders (2016): 1495.

21. Benevenia J., *et al*. "Imaging rounds. Multifocal adamantinoma of the tibia and fibula". *Orthopaedic Review* 21.8 (1992): 996-1000.

22. Castañedo-Rodríguez., *et al*. "Adamantinoma: An uncommon pathology in children". *Advances in Biomedicine* 7.1 (2018).

23. Criollo F., *et al*. "Adamantinoma of the tibia: surgical management with intercalar allograft. About a case". *Revista Colombiana de Cancerología* 19.2 (2015): 111-118.

24. Hoshi M., *et al*. "Surgical treatment for adamantinoma arising from the tibia". *Journal of Orthopaedic Science* 10.6 (2005): 665-670.

25. Kanakaraddi SV., *et al*. "Adamantinoma of the tibia with late skeletal metastasis: an unusual presentation". *The Journal of Bone and Joint Surgery* 89B.3 (2007): 388-389.