



Case report

Adamantinoma : The first report from Syria

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ABSTRACT

Adamantinoma is rare, representing less than 1 % of all primary bone tumors. It is a slow growing low grade tumor which is often clinically, radiologically and histologically mistaken for many other tumors. In this report, we present a case of Adamantinoma in a 17-year-old Syrian adolescent. The patient presented with pain and swelling anterior aspect of his left leg for last six months. We analyzed the clinical, radiographical and pathological characteristics of our patient, which helped us to reach the final diagnosis. The aim of this report is to document the presence of Adamantinoma in Syria to help other Syrian physicians considering this disease in the differential diagnosis if they face similar presentations.

1. Introduction

Adamantinoma of long bone is a malignant miscellaneous tumor, according to WHO Classification of Bone Tumors in 2020. It is a biphasic locally aggressive or malignant tumor characterized by a variety of morphological patterns, with a variable epithelial component within a bland osteofibrous component. The first case report is attributed to Maier in the year 1900 [1]. In 1913, Fisher reported the first tibia adamantinoma [2]. To our knowledge this tumor has not previously been reported in Syria. This case report has been reported in line with the SCARE Criteria [6].

2. Case presentation

A 17-year-old Syrian adolescent patient presented to our orthopedic department in Tishreen University Hospital with a history of pain and swelling anterior aspect of his left leg for last six months. Pain was continuous in nature, aggravated by activities and relieved by rest. The patient denied any history of trauma or injury. Medical and social histories were unremarkable. On physical examination there was a bony hard immobile swelling of about 3 cm * 2 cm over the anterior aspect of left leg. The overlying skin was warmer but not adherent to underlying structures.

A radiograph of the left tibia showed intracortical lesion of the diaphysis of the tibia with oval shape radiolucency (Fig. 1A, B). On computed tomography (CT), an intracortical isodense lesion involving the anterior cortex of the midshaft of the tibia was detected (Fig. 1C).

T1 and T2-weighted MRI revealed increased signal lesion involving the anterior cortex of the midshaft of the tibia (Fig. 1D-F).

Intraoperatively, the incision was made at the anterior side of the leg layer by layer until the periosteum. Then the periosteum was incised at one side and performed curettage until the distal and proximal edge. Then, the defect was filled with bone graft and periosteum was sutured back. The specimen was sent to the department of pathology for histopathological study.

Grossly the fragmented specimen was tan gray, soft to firm, measured in aggregate 3x1x1 cm. Microscopic inspection of the H&E stained sections revealed biphasic tumor composed of intermingled epithelial and osteofibrous components. The epithelial sheets with irregular lumens were consisted of basaloid cells showing peripheral palisading. No mitotic activity or necrosis was detected. Adamantinoma, classic type, was our diagnosis. To confirm our diagnosis immunohistochemical stains were performed. The epithelial sheets of the tumor were positive for Pan CK, but the mesenchymal component was negative (Fig. 2, c). Moreover, both components were positive for vimentin and negative for CD99, S100 (Fig. 2, D). Finally, adamantinoma was diagnosed based on the immunohistochemical results and routine microscopic findings. The constellation of the morphology on H&E stained section and the results of the IHC aid to confirm our diagnosis and exclude fibrous dysplasia, adamantinoma-like Ewing sarcoma, and metastatic carcinoma.

The patient was put in a posterior splint for 4 weeks and instructed non-weight bearing for 6 weeks, after that he was allowed to start partial weight bearing. Full weight bearing was allowed at 8 weeks. The

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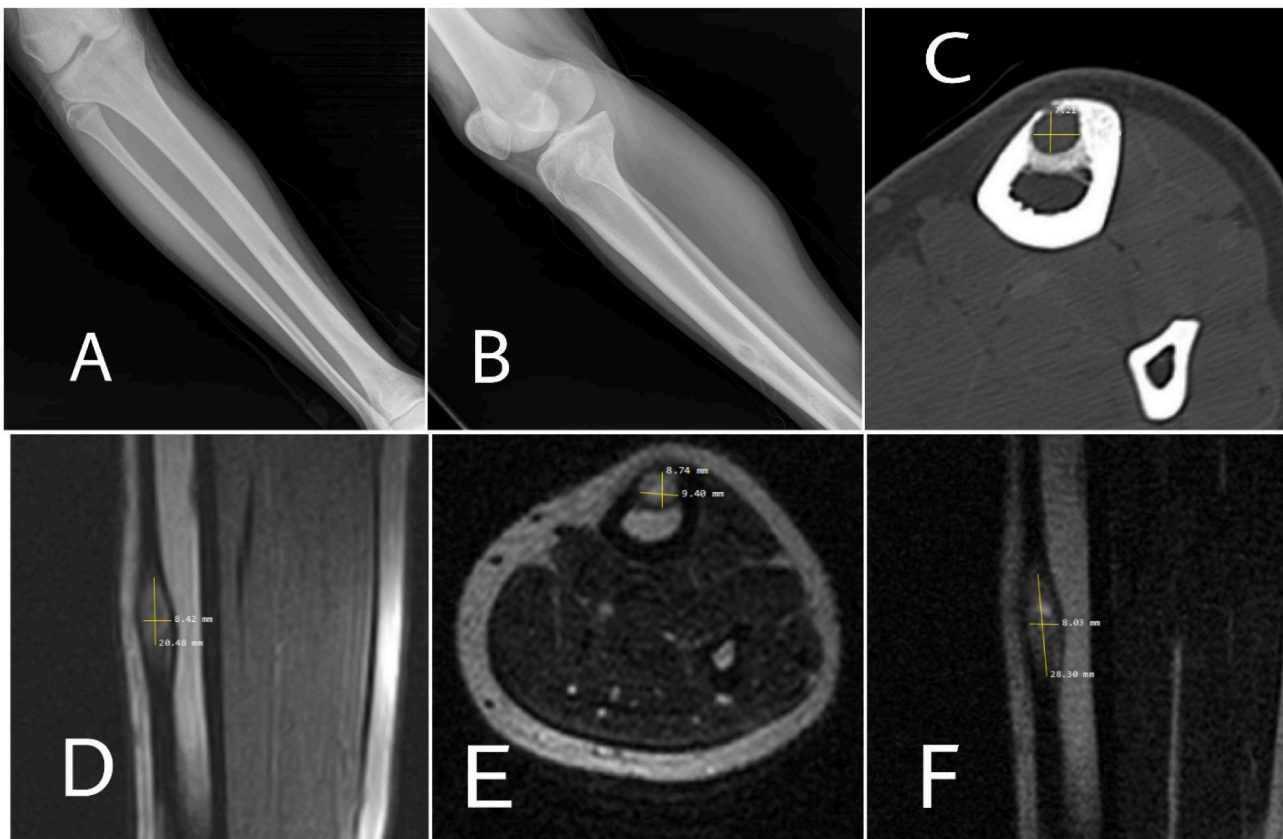


Fig. 1. (A) X-ray AP view, (B) X-ray lateral view: intracortical lesion of the diaphysis of the tibia with oval shape radiolucency. (C) CT- Bone window: intracortical isodense lesion involving the anterior cortex of the midshaft of the tibia. (D) MRI T1, (E-F) MRI T2: increased signal lesion involving the anterior cortex of the midshaft of the tibia.

patient had local recurrence one year after the treatment without any metastasis and underwent to another surgery. We hope that we could make a long-term clinical and radiological monitoring to the case and document the data related to it.

3. Discussion

Adamantinomas represent 0.1 to 0.5 % of all primary bone tumors. In the huge majority of cases, they involve the anterior cortex of the tibial diaphysis [1]. In their initial stage they are located at the affected bone's cortex and they can be asymptomatic or have a very slight symptomatology, while during the advanced stage they affect the perilesional soft tissue and the bone marrow, and symptoms such as pain and limb deformity increase [5]. In our case the pain and swelling were the primary presentations. In histopathological terms, an adamantinoma is a tumor characterized by the presence of a variable proportion of epithelial cells within osteofibrous tissue. There are two histological sub-types: osteofibrous dysplasia-like adamantinoma (predominance of the osteofibrous component) and classic adamantinoma (predominance of the epithelial component) [1]. Four histological patterns have been described for classic adamantinoma: basaloid, spindle, tubular, and squamous, of which the basaloid and tubular patterns are the most common [2]. In our case classic type, was the diagnosis.

Adamantinoma is typically a slowly-proliferating, locally-invasive tumor. There is a slight male predominance. The ages of patients with adamantinoma were older and less equally distributed over the second to fourth decades; nevertheless, the tumor can occur in patients from 3 to 86 years [2]. In X-ray images, an early adamantinoma appears as a small, unifocal, or more rarely bifocal, eccentric cortical lesion (tibia and ipsilateral fibula), with no associated periosteal reaction. At a more

advanced stage, the tumor has a 'soap bubbles' appearance. Peripheral osteosclerosis is common, and cortical destruction and aperiosteal reaction may also be associated [1]. In our case the lesion was involved to the anterior cortex of the tibia but without periosteal reaction. CT is better for showing the edge and internal structure of the lesion including calcification or ossification [2]. MRI is the firstline examination to perform to determine the extension of the tumor into the bone marrow and neighboring structures. Two types of presentation have been described: a solitary or bifocal lobulated lesion or many small nodules grouped into one or more foci [1]. Adamantinoma metastasizes in about 15 to 30 % of cases by both hematogenous and lymphatic routes to other parts of the body, usually to the lungs or lymph nodes; bone and abdominal viscera make up a minority [3]. Block resection of the tumor with healthy margins, then reconstructive surgery are considered as the most appropriate treatment for an adamantinoma [1]. Since adamantinomas are low-grade lesions with an extremely low mitotic index, they are radioresistant lesions. The use of chemotherapy for this disease has not proven to be effective [5]. With suitable surgical treatment, the prognosis for adamantinoma is excellent (10-year survival rate of 87.2 %). However, long-term clinical and radiological monitoring is necessary because of the risk of local recurrence or metastasis, which can occur many years after the initial treatment [1]. In our case, the patient had local recurrence one year after the treatment without any metastasis and underwent to another surgery. We hope that we could make a long-term clinical and radiological monitoring to the case and document the data related to it.

4. Conclusion

Diagnosing of adamantinoma can be very challenging due to the rarity

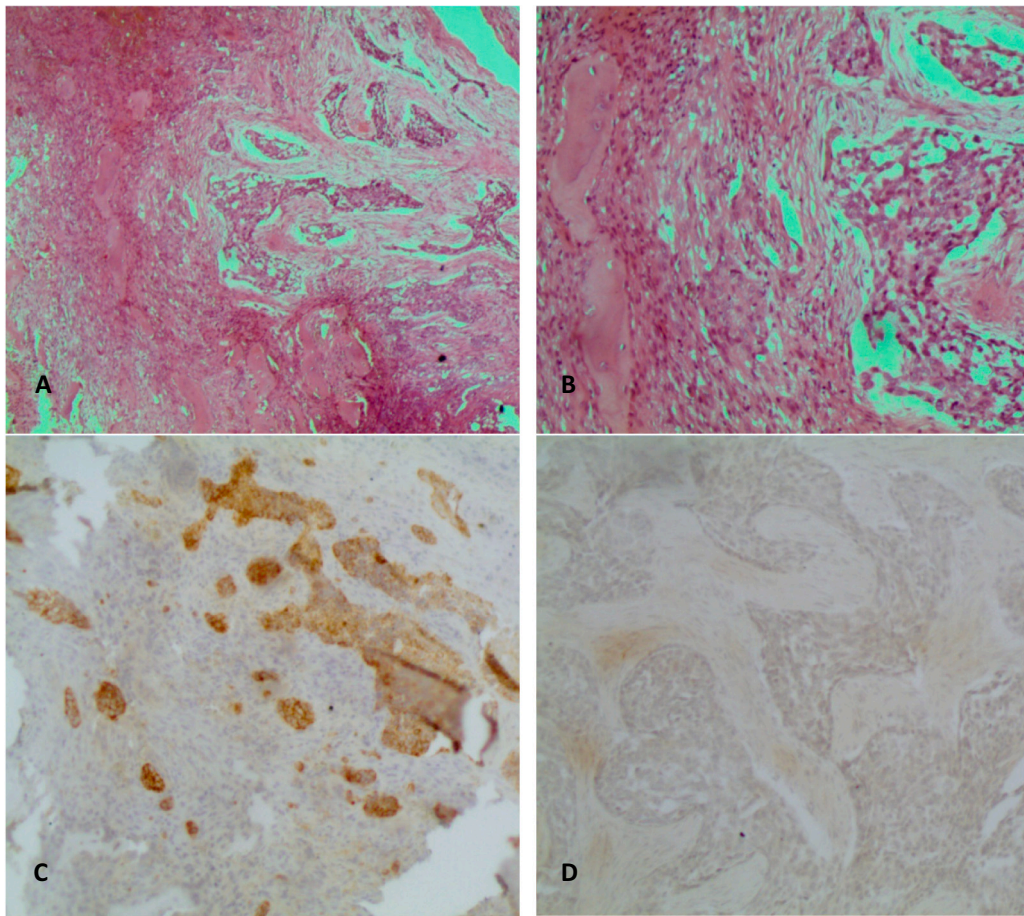


Fig. 2. H&E (A and B) and IHC (B and C) microscopy of adamantinoma (A&B) Biphasic tumor composed of epithelial and fibrous mesenchymal components ($\times 40$ and $\times 200$), (C) epithelial component is positive for panCK ($\times 100$), and (D) negative for CD99 ($\times 200$).

of the tumor. It should be one of the differential diagnoses in every osteolytic bone lesion in adolescents. Professional assessment of the radiological findings is necessary for diagnosis, but the pathological examination remains the essential tool for accurate diagnosis of adamantinoma in all cases.

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Ethical approval

The study was exempted from ethical approval in our institution, Tishreen University Hospital.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Mohammad Haj Ali: Conceptualization, Methodology, Validation,

Writing – original draft, Investigation, Project administration.

Zein Alabdin Alhrysheh: Conceptualization, Methodology, Validation, Investigation, Supervision.

Ali youssef: Methodology, Investigation, Resources, Writing – review & editing, Supervision.

Rana Issa: Investigation, Writing – review & editing, Visualization.

Registration of research studies

Not applicable.

Guarantor

Mohammad Haj Ali.

Declaration of competing interest

The authors declare no conflicts of interest.

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