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Youssef Mahdi, Lamiaa Rouas, Abdelouahed Amrani, Abderrahmane Malihy, Najat Lamalmi, Zaitouna Alhamany

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Introduction: Telangiectatic osteosarcoma is a rare and aggressive subtype of intramedullary osteosarcoma. The distal femur is the most common site.

Case Report: We present a case of telangiectatic osteosarcoma of the femur in a 15-year-old male. The tumor was neglected for six months. The patient had severe anemia with hemoglobin of 5 g/dL. Radiography showed an expansive lytic lesion of the distal end of the femur with cortical destruction, focal periosteal reaction and a pathologic fracture. It showed massive extension to adjacent soft tissue. Given the advanced stage of the disease, an amputation of the limb was performed. Gross examination of the resected distal femur revealed a 22x5 cm intramedullary multicystic hemorrhagic and destructive tumor with cortical destruction and extension into the adjacent soft tissues. Histologically, the tumor showed blood-filled spaces lined by giant cells, separated by septa with malignant tumor cells. An immature bone osteoid kind was observed. Numerous mitotic figures were noted. The resection margins were negative. The staging showed no metastasis. The decision was to complete by 6 cycles of chemotherapy. The postoperative course was unremarkable.

Conclusion: In terms of telangiectatic osteosarcoma, pathological analysis is critical in positive and differential diagnosis.

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Keywords: Telangiectatic, Osteosarcoma, Cystic, Malignant

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INTRODUCTION

Telangiectatic osteosarcoma is a rare, aggressive morphologic variant comprising 2–4% of osteosarcomas [1–3]. Pathologic fracture is a clinical feature of this malignant bone-forming tumor [1]. This tumor resembles aneurysmal bone cyst radiologically and microscopically with large cavity containing blood filled cystic spaces separated by septa [4, 5]. There is usually invasion of soft tissues and formation of a large palpable mass resembling a soft tissue tumor at clinical examination. Historically, the preferred treatment for telangiectatic osteosarcoma was amputation. More recently, chemotherapy has changed the prognosis of this tumor, which is considered worse than conventional osteosarcoma in the first observations [1, 4–6].

We present a rare case of telangiectatic osteosarcoma in the femur of a 15-year-old boy with a brief review

of literature and emphasize on diagnostic difficulties, differential diagnosis, and treatment.

CASE REPORT

A 15-year-old boy was presented with a six month history of a growing mass in left thigh. The lump had been neglected. It grew until it reached the subcutaneous zone and ulcerated. The remainder of his medical history was non-contributory. He presented to the Children's Hospital. He was very pale and asthenic.

He had severe anemia with hemoglobin of 5 g/dL. The rest of routine laboratory tests were within normal levels. Radiography showed an expansive lytic lesion of the distal end of the femur with cortical destruction, focal periosteal reaction and a pathologic fracture (Figure 1). It showed massive extension to adjacent soft tissue. Given the advanced stage of the disease, an amputation of the limb was performed.

At the pathology laboratory, we received a piece of the left lower limb amputation (Figure 2). We opened it. Gross examination of the resected distal femur revealed a 22x5 cm intramedullary multicystic hemorrhagic destructive tumor with cortical destruction and extension into the adjacent anterior and posterior soft tissues. The latter component of the mass was larger than the intraosseous tumor (Figure 3). The articular surface was not involved. We cut the femur in its longest axis with electric saw (Figure 4). Fixation in 10% neutral buffered formalin for 48 hours and decalcification of bone with nitric acid were performed. Slice of bone section was included in full. Many specimens from soft tissues and the surgical margins were also taken. Microscopic view of histological specimens of tumor stained with hematoxylin and eosin showed prominent blood filled cysts with malignant stroma in septa separating cysts (Figures 5 and 6). It contained atypical tumor cells oval or round of variable size with osteoblast-like multinucleated giant cells and a variable amount of immature osteoid (Figures 7–12). Numerous mitotic figures were noted (Figure 13). The tumor was located at 6 cm from the bone limit and 2 cm at the edge of the soft tissues. The resection margins were negative. The marrow, scooped and submitted separately, was negative.

The staging (bone scintigraphy, chest X-ray and abdominal ultrasound) showed no metastasis. The decision was to complete by six cycles of chemotherapy (Adriamycin (60 mg/d) and cisplatin (120 mg/d)). The postoperative course was unremarkable.

DISCUSSION

Telangiectatic osteosarcoma is a very rare tumor that account for less than 4% of all cases of osteosarcoma [1–3]. Paget was the first to describe it in 1854 [4, 5, 7].

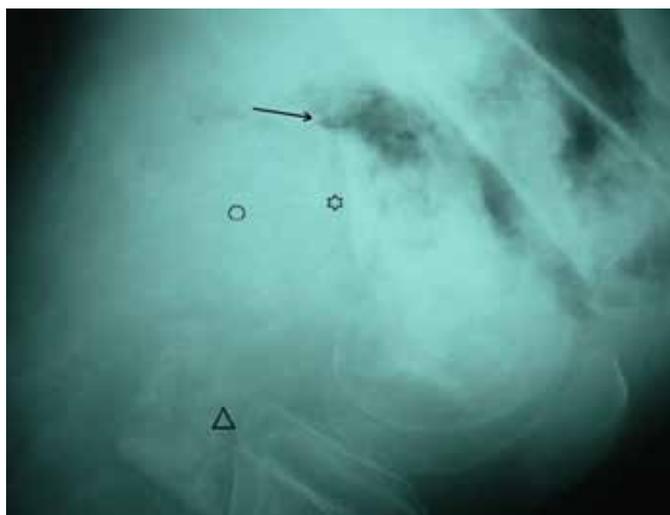


Figure 1: Plain radiograph showing an expansive and ill-defined destructive tumor of distal femur. There was focal periosteal reaction (star), cortical destruction and pathologic fracture (arrow). Massive extension to adjacent soft tissue was observed (circle). Articular surface and tibia were not involved (triangle).

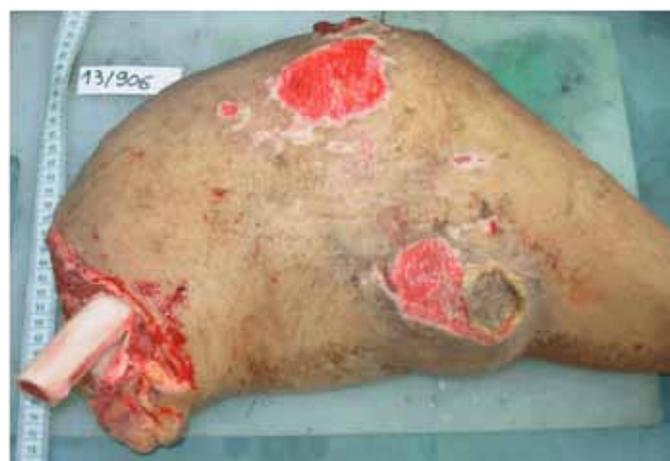


Figure 2: A large mass bulging from the soft tissues of the distal left thigh. The overlying skin is ulcerated.

Originally, it was thought to be a variant of conventional osteosarcoma, but it has several distinctive features.

It most frequently occurs in the second decade of life and has a male predominance (1.5:1 male/female ratio) [1–4]. The peak incidence and the anatomic distribution are similar to that of conventional osteosarcoma [1–5]. Most tumors occur in the metaphyseal region of long tubular bones [1, 2]. The distal femoral metaphysis is the single most common anatomic site, followed by the upper tibia and proximal humerus or proximal femur [1–4].

Telangiectatic osteosarcoma is rapidly expansile and behave aggressively. Clinical manifestations resemble those of conventional osteosarcoma, including pain and palpable mass [1, 2, 4, 5]. One characteristic clinical finding of this tumor is pathological fracture, being present in one-fourth of the cases, due to massive bone destruction [1].

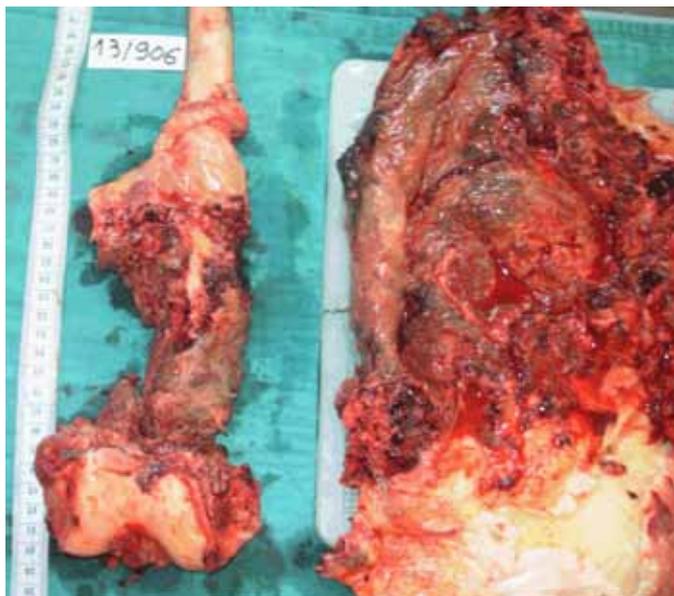


Figure 3: Expansile hemorrhagic tumor replacing proximal end of femur. Massive extension into soft tissue from intramedullary osteosarcoma of femur forms large encircling mass with hemorrhagic locules in fleshy background.



Figure 4: Four sections of the specimen using a band saw showing the tumor.

Delayed diagnosis reported in previous publications varies from 3–4 months [2, 5, 7]. In our case, the causes of the delay in the diagnosis were fear of the diagnosis and the treatment and a low socio-economic status. Indeed, the mother was a housewife and the father did not have fixed income. This made difficult to access to the hospital and the diagnostic tests necessary at first times.

The diagnosis of telangiectatic osteosarcoma requires presence of a predominantly lytic destructive lesion of bone with no or minimal lesional sclerosis on radiographic imaging [1, 4]. Periosteal bone formation is

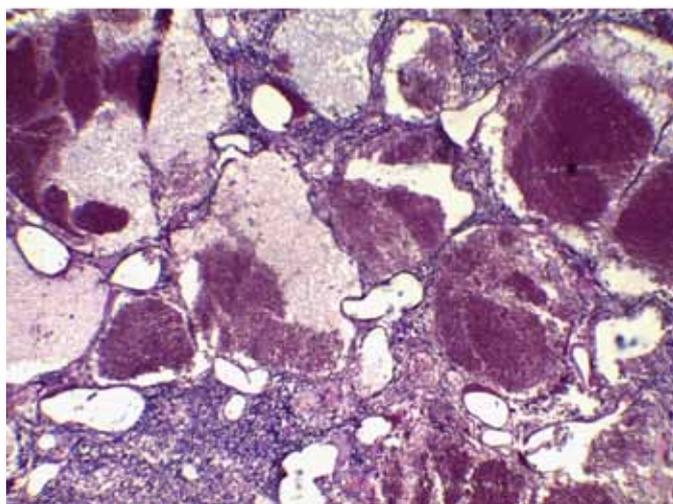


Figure 5: Low magnification showing large blood-filled cystic spaces with septa composed of loose hemorrhagic tissue with sarcomatous cells (H&E stain, x400).

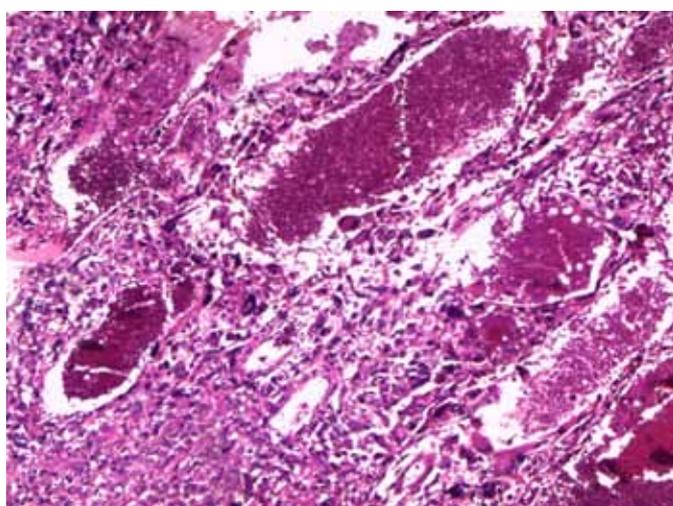


Figure 6: Medium-power magnification shows cystic spaces without lining surrounded by septa containing highly atypical malignant cells (H&E stain, x100).

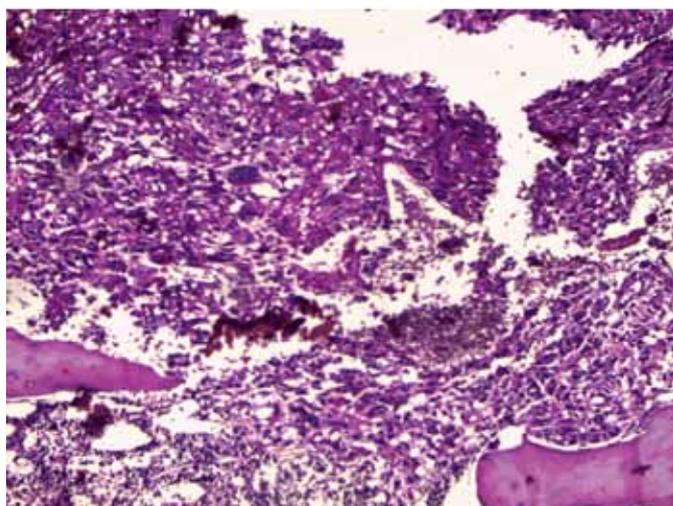


Figure 7: Medium-power view of hemorrhagic area with floating, noncohesive tumor cells, infiltrating trabecular bone (H&E, stain, x100).

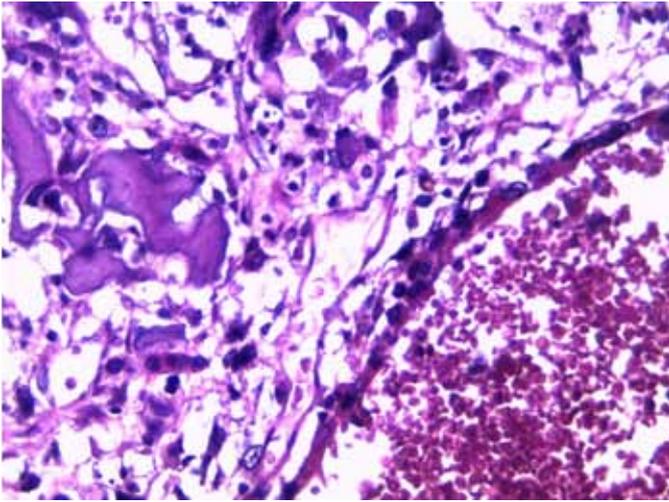


Figure 8: Septa shows highly pleomorphic stromal cells and tumor giant cells bordering spaces without endothelial linings. Note minimal osteoid.

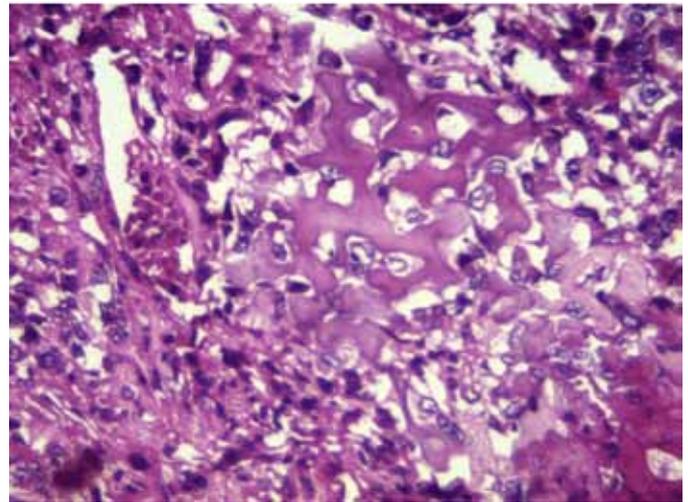
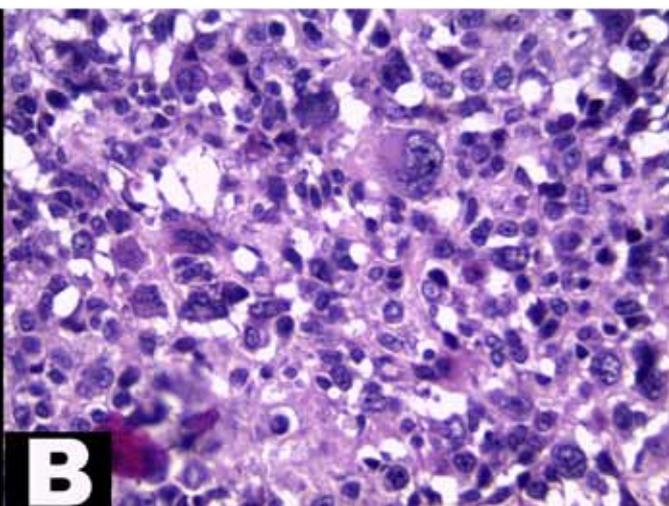
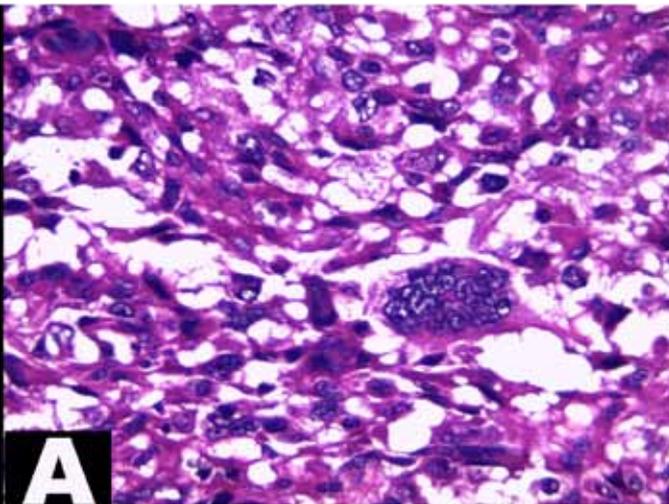


Figure 10: Tumor osteoid in septum bordering blood-filled space. Higher magnification showing irregular, finely divided (lace-like) strands of osteoid (H&E stain, x400).



Figures 9: (A, B) High magnification showing nuclear pleomorphism of sarcomatous septal cells and tumor giant cells (H&E stain, x400).

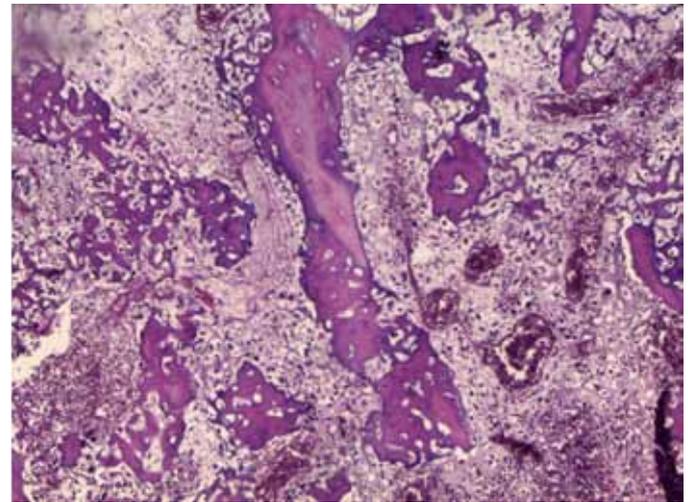


Figure 11: Telangiectatic osteosarcoma with blue spiculated bone.

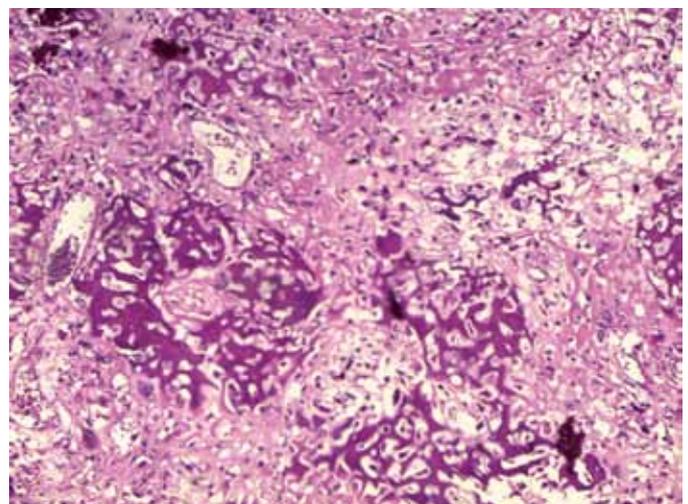


Figure 12: Telangiectatic osteosarcoma. Malignant osteoid is deposited in a lace-like pattern.

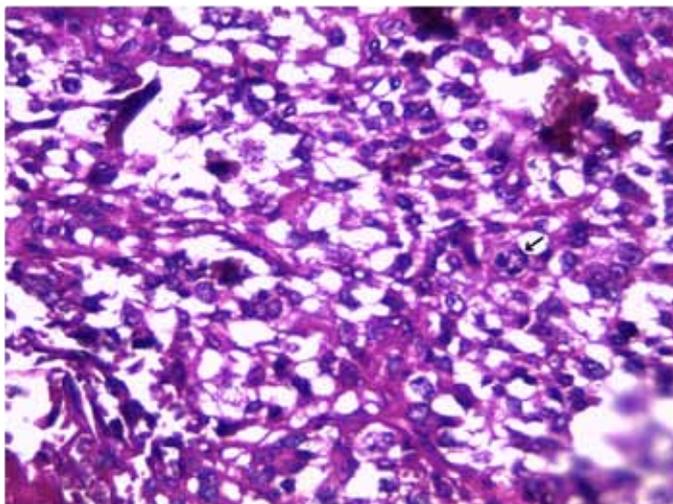


Figure 13: High magnification showing anaplastic cellular features and mitotic activity (arrow).

absent or minimal and is referred to as onion skin [1, 2]. The tumor commonly shows extension into soft tissues [1, 7]. Imaging features alone would also be compatible with an aneurysmal bone cyst, Ewing sarcoma, Langerhans cell histiocytosis, fibrosarcoma and malignant fibrous histiocytoma [8]. The pathologic findings enable confident exclusion of these radiologic differential diagnosis [4].

In our case, the gross findings were very suggestive of telangiectatic osteosarcoma. We had the gross appearance of an expansile hemorrhagic tumor with predominantly cystic spaces filled with blood, described as “a bag of blood” [1, 2, 4]. The tumor can have a honeycombed appearance with spongy areas containing smaller cysts. There is no fleshy or sclerotic bone formation. Features of invasive growth are often observed with extensive irregular cortical erosion and/or complete disruption of cortical continuity and massive invasion of soft tissue [1, 2, 4].

Microscopic findings revealed highly pleomorphic cells and foci of bone matrix formation between blood filled cyst like spaces, confirming the diagnosis of telangiectatic osteosarcoma. In some cases, where septa have broken down, atypical stromal cells may be identified, free-floating within the blood clot [4]. The amount of osteoid varies, but usually fine and lacelike osteoid is observed in minimal amount [1, 4, 6]. However, this feature is not essential to establish the diagnosis of telangiectatic osteosarcoma [4].

At this advanced local stage, telangiectatic osteosarcoma should be differentiated from an osteosarcoma of soft tissue. In the later, the tumor must arise in the soft tissues without any attachment to periosteum or bone [9].

The most important morphologic differential diagnosis is the benign ABC due to radiographic and

gross appearance’s similarities. Microscopic examination is the only definitive diagnostic aid [4]. The presence of atypical or malignant stromal cells identified in telangiectatic osteosarcoma is never seen in aneurysmal bone cyst (ABC) [4]. The presence of atypical and/or overtly malignant cells is enough to rule out the diagnosis of ABC [4]. Furthermore, osteosarcoma is associated with aggressive growth features as indicated by cortical destruction and extension into the surrounding soft tissues [5]. In contrast, aneurysmal bone cysts cause marked expansile remodeling of bone and cortical thinning but lack true soft-tissue involvement [5].

Prognosis in the modern era is similar to conventional osteosarcoma [1, 4–6]. Indeed, Farr et al and Mervak et al found no prognostic differences between the two entities [10, 11].

Surgical resection of the tumor, limb-salvage as possible, is the treatment of choice, preceded by neoadjuvant chemotherapy [4–6]. Following resection, the histological response must be evaluated according to Huvos score based on the percentage of necrosis [12]. Depending on the grade, chemotherapy regimens are adapted after surgery [12]. In our case, given the emergency status, the patient was treated by primary amputation.

CONCLUSION

The case is reported because of the rarity of telangiectatic osteosarcoma along with the important role of pathological examination in making the positive and differential diagnosis. In our case, we had the typical histological appearance. Despite the locally advanced stage, the patient showed no metastasis.

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Author Contributions

Youssef Mahdi – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revising it critically for important intellectual content, Final approval of the version to be published

Lamiaa Rouas – Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Abderahmane Malihy – Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Najat Lamalmi – Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Zaitouna Alhamany – Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Abdelouahed Amrani – Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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