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(CASE REPORT)

Telangiectatic osteosarcoma of the lumbar spine in a 15-Year-Old Girl: A Diagnostic Pitfall Mimicking Benign Lesions

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Abstract

Background: Telangiectatic osteosarcoma (TOS) is a rare, aggressive osteosarcoma variant that can mimic benign lesions such as aneurysmal bone cyst (ABC) or giant cell tumor (GCT). Its occurrence in the spine is exceptional and may lead to diagnostic delays. Case: We report the case of a 14-year-old girl presenting with progressive paraparesis and low-back pain. MRI revealed an expansile lytic lesion at L3. Initial histopathology after decompressive laminectomy suggested a giant cell–rich lesion. The patient improved transiently, but symptoms recurred within six weeks. A second surgery with anterior tumor excision was performed; histology then favored an aneurysmal bone cyst. Owing to the rapid recurrence and aggressive local behavior, a third expert histopathological review confirmed telangiectatic osteosarcoma. Conclusion: Telangiectatic osteosarcoma can closely mimic benign cystic bone lesions. Awareness of its aggressive evolution and careful histopathological review are essential for accurate diagnosis and timely oncologic management.

Keywords: Telangiectatic Osteosarcoma; Spine Tumor; Aneurysmal Bone Cyst; Diagnostic Pitfall; Adolescent; Case Report

1. Introduction

Telangiectatic osteosarcoma (TOS) is a rare subtype of osteosarcoma, accounting for approximately 2–4% of all osteosarcoma cases. It is characterized by large blood-filled cystic spaces resembling an aneurysmal bone cyst (ABC), often leading to misdiagnosis. Involvement of the spine is particularly uncommon and poses major diagnostic and therapeutic challenges. We report a spinal case of TOS initially misdiagnosed twice as a benign lesion before final confirmation, to highlight diagnostic and management lessons.

2. Case presentation

We present the case of a 14-year-old girl with no significant medical history who was admitted in our department after a 6-month history of low back pain, initially managed with analgesic treatment resulting in mild symptomatic improvement. One month prior to her admission she began to develop neurologic symptoms including: lumbar radiculopathy, bilateral lower limb progressive paralysis, urinary urgency and intermittent incontinence associated with worsening of her back pain.

General exam revealed a stable patient with no hemodynamic or respiratory problem, but the patient had a painful face and was unable to stay in a strict dorsal decubitus for a prolonged period, local tenderness with a paravertebral mass on the left lumbar region.

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Neurological assessment found a cauda equina syndrome associating sever proximal lower limb weakness (1/5 on the left and 2/5 on the right) with moderate impairment of knee flexion bilaterally with hypoesthesia in the left L3 territory, no saddle anesthesia, and patellar reflexes were abolished bilaterally, the ASIA scale grade C.

CT-scan revealed a heterogeneous paravertebral mass on the left side with an osteolytic lesion of the L3 vertebra resulting in vertebra plana (complete vertebral body collapse). The lesion showed extension into the spinal canal with compression of the dural sac **(see Figure 1)**.

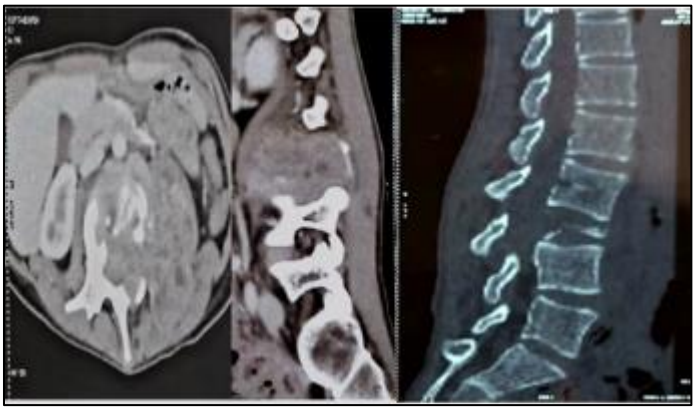

Figure 1 CT scan showing osteolytic lesion of L3 vertebra with complete destruction of the vertebral body

MRI demonstrated a process involving L3with hypointense signal on T1-weighted images and hyperintense signal on T2-weighted sequences. The lesion showed heterogeneous enhancement after gadolinium administration, with multiple fluid-filled cavities creating a multilocular appearance. There was involvement of the vertebral body and left posterior arch with invasion of the left psoas muscle and significant spinal canal compromise **(see Figure 2)**.

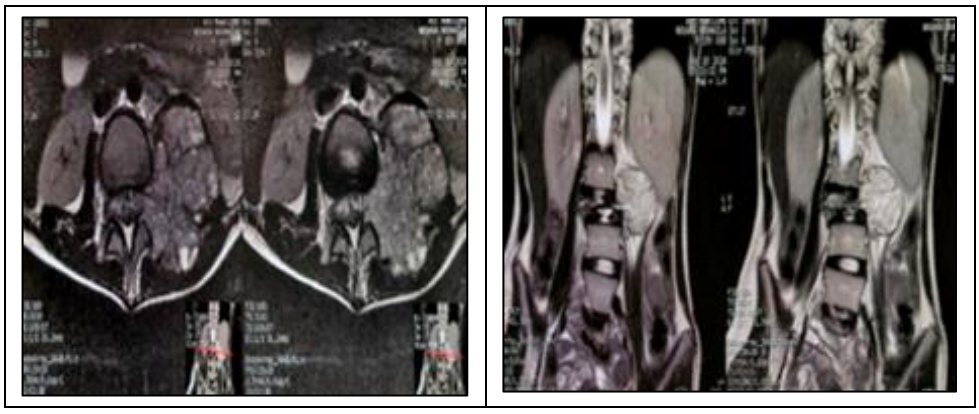

Figure 2 MRI demonstrating heterogeneous lesion with fluid-filled cavities

For our first intervention we decided to do three level laminectomy L2-L3-L4 with posterior instrumentation. Due to the highly hemorrhagic tumor, we could only achieve a partial resection. After surgery, the patient showed significant clinical improvement of pain with almost a normalization of her lower limb weakness (patient started walking again). The pathological initial diagnosis was giant cell tumor.

Six weeks postoperatively, the patient experienced clinical deterioration with recurrence of paraparesis and lumbosacral radiculopathy. Follow-up imaging revealed dramatic tumor progression with doubling of the lesion size and increased number and volume of fluid-filled cavities compared to preoperative studies **(see Figure 3)**.

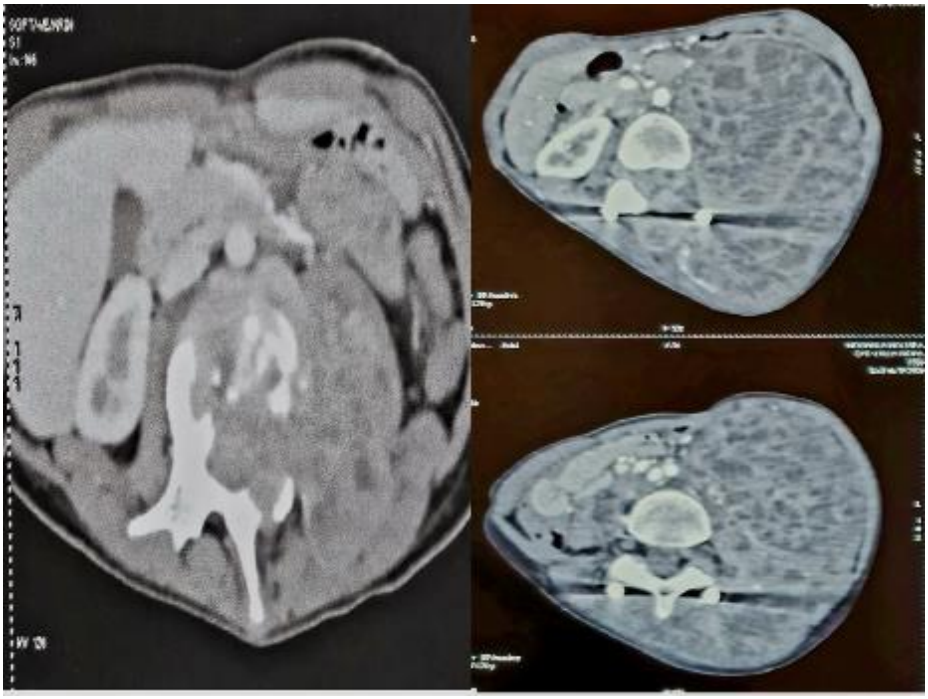

Figure 3 Comparative CT images before and after tumor progression

Following multidisciplinary team discussion involving neurosurgery, oncology, and urology departments, a decision was made for direct tumor resection via retroperitoneal approach in collaboration with the urology team to achieve better tumor exposure and more complete resection.

The tumor demonstrated a highly hemorrhagic appearance with multiple blood-filled septations and cystic spaces. The lesion was extremely vascular with significant bleeding during manipulation. This time, we could achieve a subtotal resection and the patient could regain he strength **(see Figure 4)**.

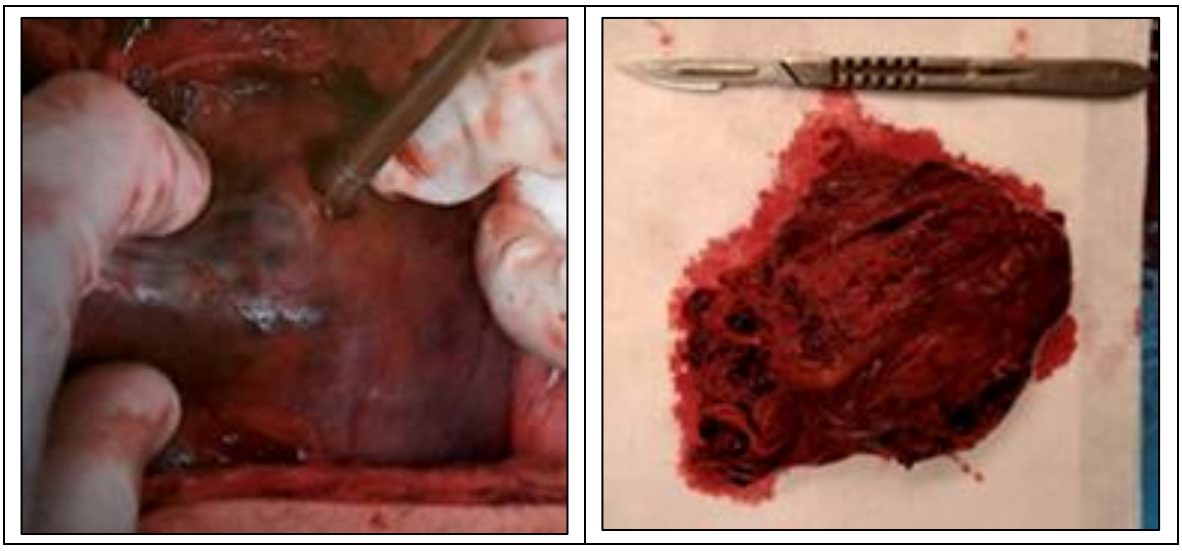

Figure 4 Intraoperative photograph showing hemorrhagic tumor with blood-filled septations

The second histopathological diagnosis was aneurysmal bone cyst (ABC). The discordance between the aggressive clinical behavior and the benign histopathological diagnoses prompted a new pathological review and expert consultation. The definitive diagnosis was **telangiectatic osteosarcoma**. Patient was sent to oncology for emergent chemotherapy.

The lesion had been misdiagnosed twice before the correct malignant diagnosis was established, highlighting the diagnostic challenges posed by this rare osteosarcoma variant.

3. Discussion

Osteosarcoma can be distinguished according to WHO 2020 5th edition (**1**) into:

3.1. Central (medullary) osteosarcomas

Conventional (osteoblastic, chondroblastic, fibroblastic), telangiectatic, low-grade central, and small-cell

3.2. Surface (peripheral) osteosarcomas

Parosteal, periosteal, and high-grade surface

3.3. Telangiectatic osteosarcoma characteristics

- Represents 2-12% of all high-grade osteosarcoma
- Must have >90% telangiectatic component for classification
- Characterized by blood-filled cystic spaces separated by septa containing highly malignant cells
- Histologically mimics aneurysmal bone cyst but with critical malignant features
- Main differential diagnosis challenge is with ABC

3.4. Epidemiology (spinal location)

Telangiectatic osteosarcoma (TOS) accounts for about 2–4% of osteosarcomas overall (**2**). It's occurrence in the spine is extremely uncommon (0.08% of all osteosarcomas is the percentage given by Amitranand et al, in their case report (**3**)). Though it is difficult to say as reports are mostly isolated case studies. The disease shows a bimodal age distribution, with a first peak in the adolescence (10-14 y.o) and a second peak in adults older than 65 year of age.

Wang el al (**4**), reported a total of eight case reports published so far in the English literature of spinal TOS, our patient would be the ninth one.

3.5. Histopathologic diagnostic pitfalls

Throughout the literature we find a high misdiagnosis rates, mainly due to samples inadequacy, it can be either misdiagnosed as benign lesions like ABC or CGT or sometimes as other type of osteosarcomas (**5, 6**). Core-needle biopsy has been shown to misdiagnose TOS as ABC in 18-35% of cases due to the absence of viable high-grade sarcomatous cells in small tissue samples (**5, 7**) Because of the lack of immunohistochemical markers, the diagnosis relies entirely on morphological and clinical-radiological correlation.

Macroscopic examination of telangiectatic osteosarcoma reveals a hemorrhagic multicystic lesion filled with blood clots (which mimic that of ABC). Histologically, TOS shows blood-filled spaces lined by septa containing malignant stromal cells producing osteoid (**8**). Small biopsies may capture only cystic areas and miss malignant osteoid, leading to misdiagnosis as ABC or GCT (**5, 7**). Thus, repeat or open biopsy and expert pathology review are critical.

Sometimes, laboratory investigations may provide additional diagnostic clues, platelet, lactate dehydrogenase (LDH), alkaline phosphatase (ALP), and white blood cell levels were significant predictive factors for distinguishing telangiectatic osteosarcoma from aneurysmal bone cyst. These parameters demonstrated high values in the case of TOS. In our case, the platelets and white blood cells levels were normal and we did not evaluate ALP or LDH (**5, 9**).

3.6. Clinical and radiological features

Spinal osteosarcoma presents in almost all patients with a stereotypical presentation: local pain and rapidly progressing (in a few weeks) to paraparesis or sphincter disturbances. More than 70% of patients exhibit neurologic deficit, indicating locally advanced disease at presentation (**10**).

TOS appears as an expansile osteolytic lesion with cortical destruction and multiloculated cavities showing fluid-fluid levels on MRI/CT often with an associated soft-tissue mass, which allows distinction from aneurysmal bone cyst. These features mimic ABC or GCT.

Signs suggesting malignancy that should alert the surgeon include: cortical destruction with epidural expansion, soft-tissue extension, enhancing septa, and rapid growth (or in our case regrowth, 6 weeks after the first surgery).

3.7. Management

Current treatment options for spinal TOS lack sufficient evidence-based support given the small number of reported cases.

But the management of TOS in general, follows that of every high-grade osteosarcoma which combines chemotherapy both before and after surgery. Indeed, the advent of chemotherapy in the 80s and 90s increased the survival rate from 20% in the 70s to 60% - 70% in the 90s (**11, 12**).

The main surgical aim is the removal of the entirety of the tumor "en bloc" rather than intralesional debulking. While this is relatively easy to do in extremity lesions, for the spinal ones the "en bloc" spondylectomy as described by Tomita and Kawahara (**13**) is a complex surgery, requiring often a combined anterior-posterior approach with many intraoperative challenges (prolonged operative time, massive bleeding, risk of vascular/ neural injury).

For our case, we could achieve a subtotal resection after two stage surgery, but we were misled two times by the pathologists which resulted in a delay in diagnosis. And for a high-grade osteosarcoma a delay will most likely reduce the overall survival which is already low in the case of spinal TOS.

3.8. Outcome and prognosis

When managed adequately, TOS has survival similar to conventional osteosarcoma (5-year OS 50–70%) (**14, 15**). However, spinal TOS - same as any high-grade spinal osteosarcoma - prognostic is poorer compared to extremity osteosarcoma due to incomplete resections and local recurrence (**16,17**). Prognosis will depend on margin status, metastases, and chemotherapy response. Early diagnosis improves outcomes.

4. Conclusion

Telangiectatic osteosarcoma is a highly aggressive tumor that may mimic benign cystic bone lesions. Radiologic-pathologic correlation, early multidisciplinary management, and expert histopathologic review are vital to prevent diagnostic delay and improve outcomes.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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