

Primary telangiectatic osteosarcoma of the cervical spine

Case report

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Telangiectatic osteosarcoma (TOS) is one of the 8 subtypes of osteosarcoma that infrequently affects the spine. The radiopathological features of TOS overlap with those of more benign entities, most commonly the aneurysmal bone cyst), and therefore is a significant diagnostic challenge. It is a rare but well-described entity in the thoracolumbar and sacral spine, and to the authors' knowledge has not been previously reported in the cervical spine.

The authors report the case of a 15-year-old boy who presented with a 6-month history of neck pain and torticollis. He underwent preoperative glue embolization followed by a staged subtotal C-5 spondylectomy and posterior fusion for a C-5 vertebral body lytic expansile lesion. Histopathological examination showed the lesion to be TOS. The surgery was followed by adjuvant radiation and chemotherapy with a favorable outcome at the 1-year follow-up. This report reiterates that TOS is an important differential diagnosis for aneurysmal bone cyst and giant-cell tumor of the spine, as its biological behavior and clinical outcome differ from those of these more benign lesions, which it mimics. (<http://thejns.org/doi/abs/10.3171/2011.12.SPINE111037>)

KEY WORDS • telangiectatic osteosarcoma • cervical spine • aneurysmal bone cyst

TELANGIECTATIC osteosarcoma is one of 8 subtypes of osteosarcoma.²² It is more common in children and adolescents,^{8,20} and TOS of the spine accounts for 2% of all cases of primary vertebral osteosarcoma.⁹ Although isolated case reports have described TOS in the thoracolumbar^{1,9,12,14} and sacral^{18,20} spine, its occurrence in the cervical spine has not been reported before in the English literature. Despite its obvious differences from the other osteosarcoma subtypes, TOS has been confused with benign entities such as GCT, hemangioendothelioma, and ABC.⁸ Misdiagnosis can delay treatment and thereby possibly affect outcome.^{7,17}

The aim of this report is to make the reader cognizant of TOS when confronted with lytic expansile vertebral lesions of the spine. We report the case of a 15-year-old boy, discuss the radiological and pathological features of TOS, and address the management of this condition and outcomes.

Abbreviations used in this paper: ABC = aneurysmal bone cyst; GCT = giant-cell tumor; TOS = telangiectatic osteosarcoma; VB = vertebral body.

Case Report

History and Examination. This 15-year-old boy presented with a 6-month history of neck pain and head tilt to the right. Examination revealed torticollis with paraspinal muscle spasm and no neurological deficit. There was no cervical spine tenderness.

Imaging Findings. A cervical spine radiograph showed an expansile bony lesion involving the anterior and posterior elements of the C-5 vertebra with anterolisthesis of C-5 over C-6 (Fig. 1A). A CT scan showed a collapsed C-5 VB with multiloculated lytic areas replacing the body and the posterior elements (Fig. 1B and C). Magnetic resonance imaging showed multiple fluid levels within the collapsed body and its posterior elements (Fig. 2). The posterior superior endplate of C-6 was compressing the thecal sac. There was no paraspinal soft-tissue involvement. The preoperative differential diagnosis included ABC and GCT.

This article contains some figures that are displayed in color online but in black and white in the print edition.



FIG. 1. **A and B:** Preoperative lateral cervical spine radiograph and sagittal CT reconstruction images showing collapse of the C-5 VB and anterolisthesis of C-5 over C-6. **C:** Axial CT images of the cervical vertebrae at corresponding levels (arrows) showing expansile lytic lesions predominantly involving posterior arch elements.

Angiography showed significant hypervascularity in the region of the collapsed C-5 vertebra, with arterial supply from multiple tiny tortuous cervical branches of both vertebral arteries and thyrocervical trunks and venous shunting into the cervical venous plexuses (Fig. 3A and B).

Preoperative Embolization. The day before surgery, the patient underwent direct percutaneous embolization (Fig. 3C and D) of the tumor with sodium tetradecyl sulfate foam (Setrol; Samarth Life Sciences) and 17% *N*-butyl-2 cyanoacrylate glue (Histoacryl, Braun-Aesculap AG), with about 80% reduction in tumor vascularity. A selective right thyrocervical angiogram performed after percutaneous embolization showed some residual tumoral supply (Fig. 3C); this vessel was however not embolized as it was communicating with the right posterior inferior cerebellar artery.

Operative Procedure. A 2-staged surgery was per-

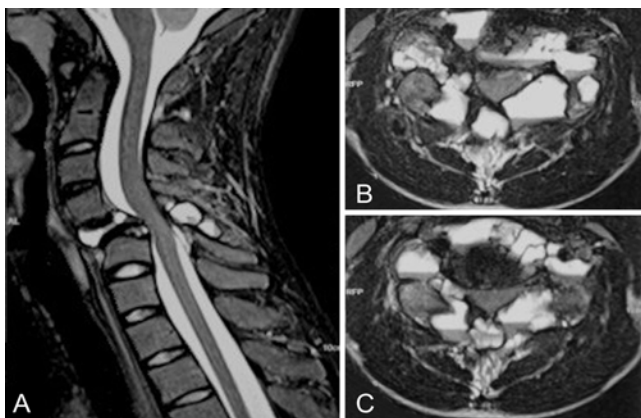


FIG. 2. **A:** Preoperative sagittal T2-weighted MR image showing a collapsed C-5 VB with anterolisthesis resulting in compression of the thecal sac. **B and C:** Corresponding axial images showing expansile and lytic cystic lesions with fluid levels, predominantly involving the posterior arch elements of C-5, encroaching into the canal and causing further cord compromise.

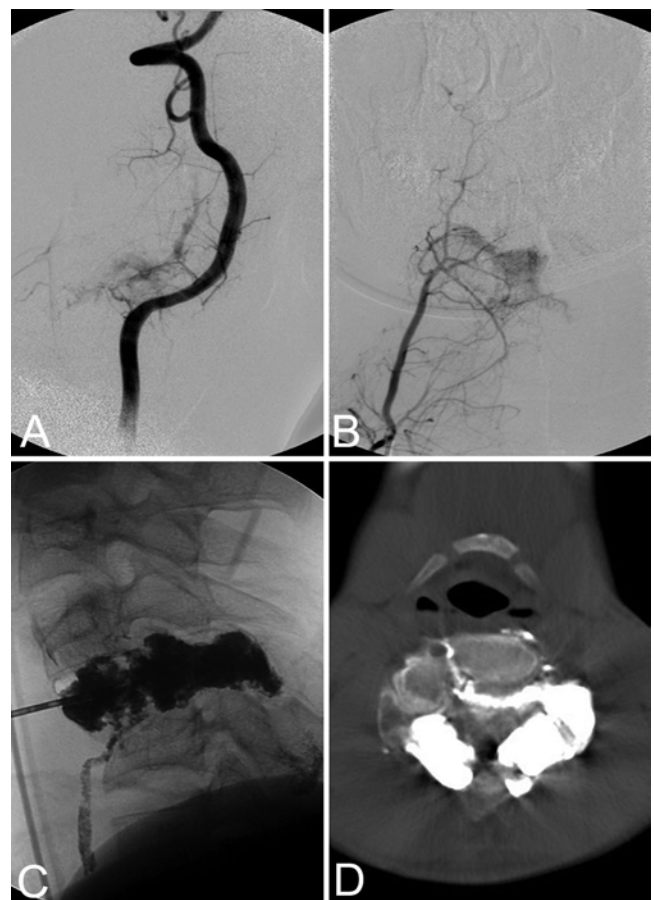


FIG. 3. **A and B:** Selective angiograms of the left vertebral artery (A) and right thyrocervical trunk (B) showing abnormal tumor blush in the C-5 vertebra supplied by these vessels. **C and D:** Lateral fluoroscopic image obtained during percutaneous injection of sodium tetradecyl sulfate foam and 17% *N*-butyl-2 cyanoacrylate glue into the collapsed C-5 VB and posterior arch elements (C) and corresponding axial CT section obtained after the embolization procedure (D).

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formed. First, a C5–6 central corpectomy with an iliac bone autograft was done. Intraoperatively, the C-5 VB was found to be collapsed with lateral ballooning of the body, without involvement of the adjacent disc spaces, which were normal. The C-5 VB was bluish in color and was replaced by tissue that appeared to be soft, destroyed bone. The tumor was superficially infiltrating the posterior longitudinal ligament and could be easily dissected off it. The C-6 VB was also removed to decompress the thecal sac and improve cervical alignment (Fig. 2). Ten days later the patient underwent a C-5 laminectomy and removal of the tumorous lateral mass. A C3–7 fusion was performed with lateral mass screws, rods, rib autograft, and femoral head allograft. Intraoperatively, the C-5 spinous process, lamina, and lateral mass were found to be expanded and replaced with soft tumor. Because of the preoperative percutaneous embolization, there was only limited bleeding. The patient's postoperative course was uneventful with no worsening of his neurological condition and resolution of the pain and torticollis. He was advised to wear a Philadelphia collar for 6 months.

Histopathology. The histopathological sample obtained at the first surgery (corpectomy) showed sclerotic cancellous bone with foci of hemorrhage, osteonecrosis, and spindle cell proliferation typical of an ABC. No malignant cells were seen. The specimen obtained from the posterior elements at the second surgery, viewed under low power (Fig. 4A), was composed of variably sized vascular spaces containing blood, lined by benign-looking multinucleated giant cells and separated by fibrocellular septae. The solid areas showed sheets of spindle cells with pleomorphic hyperchromatic nuclei, several bizarre cells, and tumor giant cells with a few atypical mitotic figures. There were foci of malignant osteoid formation. These features were consistent with TOS (Fig. 4B and C).

Adjuvant Therapy. A CT scan of the thorax and an ultrasound of the abdomen did not show any metastasis. The patient underwent 3 cycles of chemotherapy with cisplatin, doxorubicin, isophosphomide, and etoposide. This treatment was followed by conventional radiation therapy (60 Gy in 30 fractions) and 3 more cycles of the same chemotherapy.

Follow-Up. At the 1-year follow-up, the patient had resumed school and had no neurological deficits. A CT scan showed minimal residual disease in the right C-5 lateral mass (Fig. 5).

Discussion

Primary bone tumors involving the spine are rare in children. The histological subtypes include osteoid osteoma, osteoblastoma, osteochondroma, osteosarcoma, eosinophilic granuloma, Ewing sarcoma, ABC, chordoma, mesenchymal chondrosarcoma, GCT, fibrous dysplasia, fibroma, angiosarcoma, and hemangioma.⁵ The current World Health Organization classification of osteosarcoma of bone includes 8 categories: conventional, telangiectatic, small cell, low-grade central, secondary, parosteal,

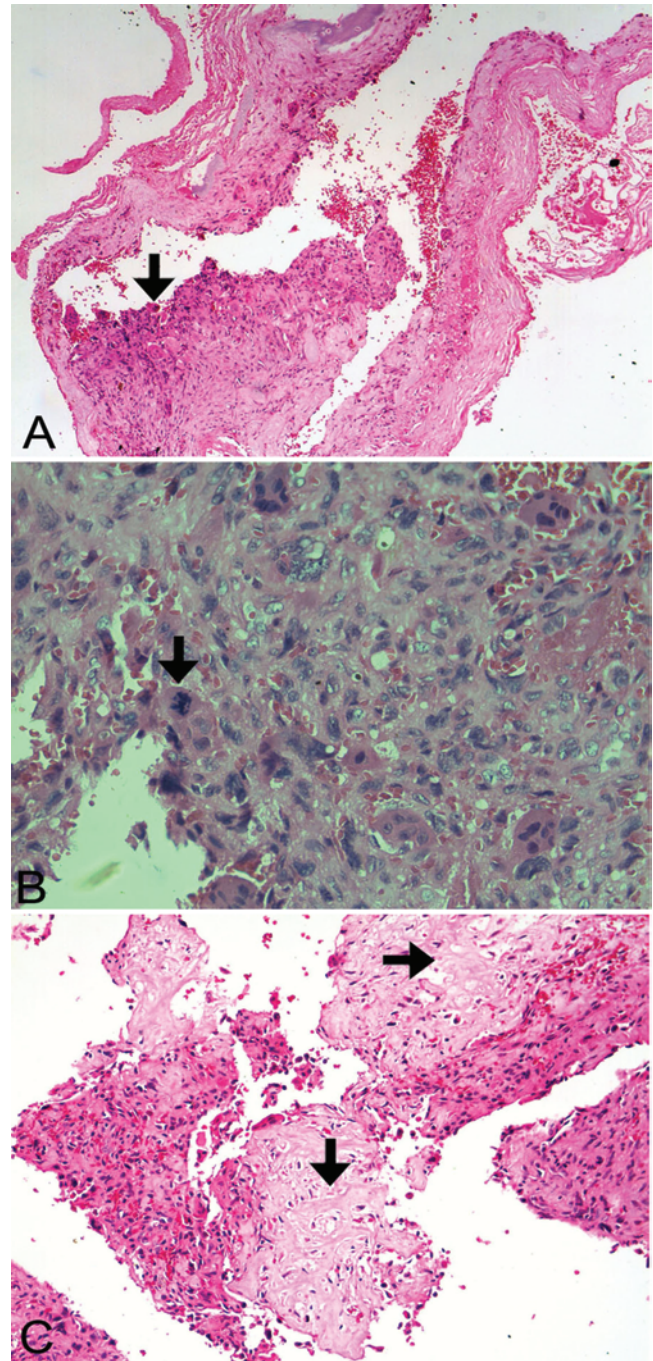


FIG. 4. **A:** Low-power photomicrograph showing the wall of the cavitated lesion lined by tumor tissue containing multinucleated giant cells (arrow) and surrounded by reactive new bone (in the upper left corner), mimicking an ABC. **B:** High-power photomicrograph showing the solid areas within the septa containing highly pleomorphic malignant cells with atypical mitoses (arrow) and multinucleation. **C:** Photomicrograph showing foci of osteoid formation within the tumor. H & E, original magnification $\times 50$ (A), $\times 400$ (B), and $\times 200$ (C).

periosteal, and high-grade surface.⁴ Telangiectatic osteosarcoma is an unconventional subtype with well-defined radiological and pathological features.^{3,13,20} It accounts for 2%–12% of tumors in the appendicular skeleton,^{8,22} especially long bones—most often the distal femur or

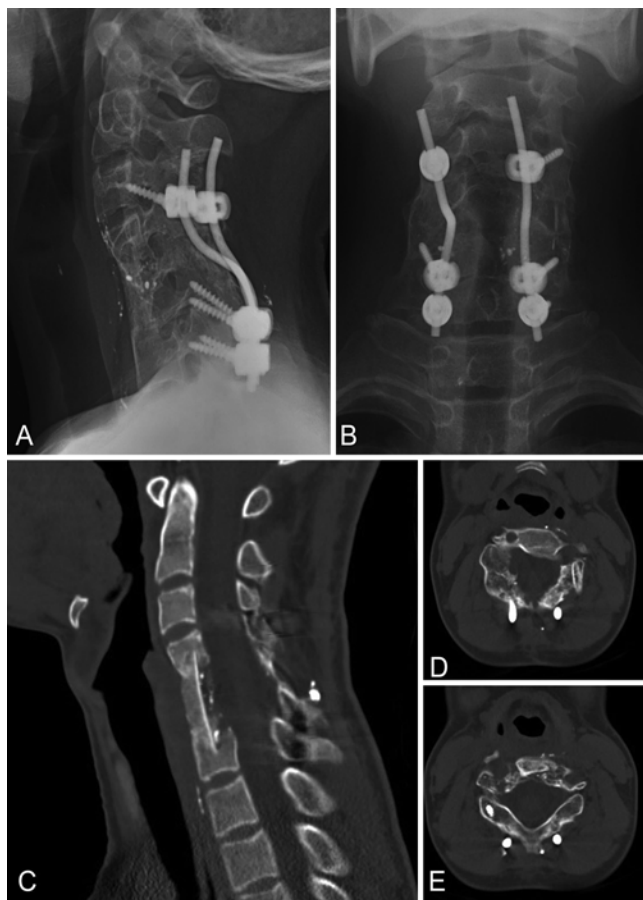


Fig. 5. One-year postoperative follow-up. Cervical spine anteroposterior and lateral radiographs (**A and B**) and sagittal (**C**) and axial (**D and E**) CT images showing improvement in the spine alignment and widening of the bony canal. There is minimal residual disease in the right C-5 lateral mass. The graft has fused anteriorly and the posterior positioned implant and bone graft can be seen.

proximal tibia. Telangiectatic osteosarcoma of the spine accounts for 2% of all primary vertebral osteosarcomas.⁹

Clinical Features

In a large series of 124 patients with TOS, 60% were male and the most frequently affected age group was 11–16 years.⁸ In another series of 24 patients with TOS, 67% were male and the median age was 15.7 years.²⁰ Our patient was male and was 15 years of age. Aneurysmal bone cysts affecting the spine also occur in the same age group, with a slight female preponderance.⁶

The most common symptom in a child harboring a bony lesion of the cervical spine is neck pain. Additional symptoms include impaired range of motion in the neck, head tilt, tightness and weakness of the limbs, paresthesias, numbness, and urinary difficulty, depending on the extent of cord involvement and duration of symptoms.⁵ Quite often the symptom gets noticed or is given importance after trivial trauma. Our patient presented with neck pain and head tilt without any clinical features of spinal cord compression.

Radiology and Histopathology

The differential diagnosis for a lytic expansile lesion (as seen in our case) on a cervical spine radiograph and CT in this age group includes ABC, GCT, eosinophilic granuloma, osteoblastoma, fibrous dysplasia, and osteosarcoma.¹⁰ The conclusive diagnosis of TOS is based on radiological and pathological criteria, as defined in the World Health Organization classification.²⁰ These criteria are summarized by Weiss et al.²⁰ as follows: “1) predominantly lytic bone mass with minimal sclerosis on radiographs, 2) grossly cystic medullary mass with no or minimal solid or sclerotic component, and 3) histological features consisting of bone-forming tumor with notable blood-filled spaces separated by septae lined by and/or containing malignant tumor cells with prominent nuclear atypia and limited osteoid deposition.”

In the largest published series,⁸ the diagnosis of TOS required the presence of a predominantly lytic (more than 90%) destructive lesion of bone with only minimal lesional sclerosis on radiographs. In the Mayo Clinic series, the presence of sclerosis was grounds for elimination of this diagnosis.¹¹ The presence of nodular septal thickening, osteoid matrix mineralization, and cortical destruction and infiltrative margins on CT imaging can aid in distinguishing TOS from ABC.²² Giant-cell tumor is another hemorrhagic tumor with an expansile lytic radiographic appearance that can be confused with TOS. It most often affects the sacrum.¹⁶ Giant-cell tumor is known to be a more solid mass with intermediate signal intensity similar to that of muscle on T1-weighted MR images and low to intermediate signal intensity on T2-weighted MR images.²² Hemorrhage and fluid levels are frequently identified in MR imaging studies.¹³ The finding of fluid levels is a nonspecific one, reported in other tumors, including TOS and ABC.¹⁹ Our patient had predominantly expansile and lytic lesions affecting the C-5 vertebral anterior and posterior elements with minimal sclerosis on CT and multiple fluid levels on MR imaging (Figs. 1 and 2). Preoperatively, ABC or GCT was suspected, rather than TOS.

When viewed under low-power magnification, TOS can be mistaken for an ABC (Fig. 4A).¹ These 2 types of lesions contain strikingly similar histological features, as described above, and the key to distinguishing TOS from an ABC is in demonstrating malignant cells with osteoid deposition. The histopathological specimen obtained from our patient in the first surgery (corpectomy) did not reveal any malignant cells and hence a diagnosis of ABC was provided. This was revised after tumor from the posterior elements revealed bizarre, giant tumor cells with osteoid formation. Review of the previous (corpectomy specimen) slides showed no evidence of malignancy.

The addition of immunohistochemical analysis does not aid in the diagnosis of TOS or ABC, nor does it help differentiate between these 2 entities. Molecular tests for ABC and TOS are not routinely done and neither of these 2 entities is associated with any disease-specific chromosomal abnormality.⁴

The transformation of an ABC into TOS as well as other subtypes of osteosarcoma has been previously reported.² Whether the two entities can coexist is debatable. This emphasizes the need for sending an adequate tissue

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sample for histopathological examination as well as the need for careful review of slides in these cases. If we had based our treatment on the first biopsy report of ABC from the corpectomy sample, our patient would not have been treated aggressively with adjuvant therapy and may have had a less favorable outcome.

Treatment

Treatment for TOS includes a combination of aggressive surgery with good margins, radiotherapy, and multimodality chemotherapy.^{15,21} For TOS in the appendicular skeleton, estimates of 5-year event-free survival ($58.3\% \pm 11.9\%$) and overall survival ($66.8\% \pm 11.6\%$) were similar to those for patients with other osteosarcoma subtypes.²⁰ Weiss et al.²⁰ recommend that patients with TOS should be treated with conventional osteosarcoma protocols, as the outcomes are similar if not better than with other osteosarcoma subtypes. They found the absence of local progression on chemotherapy, and the use of 3 or more chemotherapeutic agents to be important factors that predicted a better outcome.²⁰

There have been very few case reports discussing the management of spinal TOS, and the outcome has generally been dismal.^{1,12} The anatomical peculiarity of the spine makes it difficult to achieve an oncological resection. Murakami et al.¹² described 2 patients with TOS of the thoracic spine and total paraplegia who underwent a total spondylectomy with complete resection of the spinal cord (and preoperative chemotherapy in 1 case). Both patients died within 6 months. Amritanand's group¹ reported on a patient who underwent a laminectomy and curettage for an extensive thoracic TOS. The patient did not undergo adjuvant radiation and chemotherapy as advised and died within 1 year of surgery. Nishida et al.¹⁴ and Simon and Irwin¹⁸ have reported 4- and 5 year-survival of patients with thoracic and sacral TOSs, respectively, after partial excision followed by adjuvant chemotherapy and radiotherapy.

Preoperative embolization of vertebral tumors is known to be a useful adjunct to surgery, facilitating resection by minimizing blood loss, improving visualization, and reducing surgical time.⁵ Our patient underwent glue embolization followed by subtotal resection with cervical stabilization. Postoperatively he was treated with adjuvant radiation and multimodality chemotherapy with 4 drugs. At 1-year follow-up, there was significant reduction in the residual disease (Fig. 5); the patient had no neurological deficits and had resumed a normal lifestyle. Long-term follow-up will indicate if this treatment protocol was appropriate.

Conclusions

Telangiectatic osteosarcoma is a rare yet distinct subtype of osteosarcoma with few reports of cases involving the spine. To our knowledge, this is the first mention of this entity in the cervical spine. Misdiagnosis of the tumor as an ABC is quite likely due to the radiological and pathological similarities of the 2 lesion types. Early diagnosis with adequate surgical excision and adjuvant radiation and chemotherapy provides good short-term outcome. There is a need for long-term follow-up data.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Joseph. Acquisition of data: Joseph, Turel, Singh, Moses. Analysis and interpretation of data: Joseph, Turel, Singh, Moses. Drafting the article: Turel, Singh, Moses. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Joseph. Study supervision: Joseph.

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