Case Report | Journal of Orthopaedic Case Reports 2025 May:15(5):Page 141-145

Aggressive Thoracic Spine Hemangioma in a 13-Year-Old Boy: A Case Report with Literature Review

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Learning Point of the Article:

Understanding that vertebral hemangiomas, though typically asymptomatic, can present aggressively with neurological deficits, and recognizing the importance of early diagnosis, especially in atypical patient populations.

Abstract

Introduction: Most vertebral hemangiomas are latent and don't not require specific treatment; few cases present with symptoms, usually pain. Rarely, they manifest aggressively, causing neurological deficits from spinal cord compression.

Case Report: This case report describes a 13-year-old boy whose gait disturbance investigation revealed an aggressive T6 hemangioma.

Conclusion: Diagnosis is typically determined by imaging changes. Surgical treatment is indicated for cases with neurological deficits. Emergency decompression with laminectomy is required for rapid, progressive deficits, with radiotherapy considered based on extraosseous involvement. Level of Evidence IV. Case report.

Keywords: Hemangioma, spinal fractures, spinal cord compression.

Introduction

Hemangioma is a benign lesion of the bones, composed of newly formed capillary, cavernous, or venous blood vessels. Some of these lesions are malformations, while others, due to their growth characteristics resembling neoplasms, are considered true benign tumors [1, 2]. The term "aggressive hemangioma" refers to hemangiomas with extraosseous extension or significant bone expansion, accounting for approximately 1% of spinal hemangiomas [3].

Diagnosis is typically made through the detection of characteristic changes in imaging studies [4]. On plain radiography, hemangiomas are characterized by vertical trabeculae parallel to the vertebral bodies. Computed tomography (CT) reveals thick trabeculae and radiolucent areas, while magnetic resonance imaging (MRI) shows hyperintense signals on T1 and T2 sequences [5].

Surgical treatment is indicated for all cases with some degree of neurological deficit. Emergency decompression with laminectomy should be performed in all cases of rapid and progressive neurological deficit, with or without the need for adjuvant radiotherapy, depending on the extent of extraosseous involvement [5, 6].

We describe a rare case of aggressive hemangioma in a patient that deviates from typical epidemiology.

Access this article online

Website: www.jocr.co.in

DOI: https://doi.org/10.13107/jocr.2025.v15.i05.5588

Author's Photo Gallery


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Submitted: 11/02/2025; Review: 19/03/2025; Accepted: April 2025; Published: May 2025

DOI: https://doi.org/10.13107/jocr.2025.v15.i05.5588

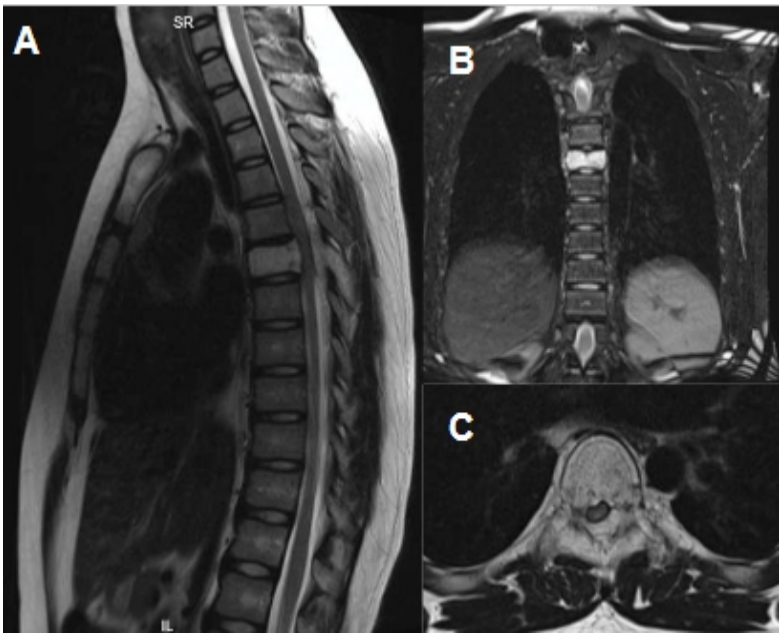

Figure 1: Magnetic resonance imaging: At the T6 level, there is an infiltrative expansive lesion affecting the bony structures, including the vertebral body and posterior elements, with a soft tissue component primarily in the posterior and lateral epidural fat, resulting in reduced dimensions of the spinal canal and compression of the dorsal spinal cord.

Case Report

Male, 13 years old, presenting with gait disturbance noted by the family without complaints of pain, evolving over 7 days. Evaluated by a neurologist, who referred him to an orthopedist. On physical examination, he presented with paraparesis in the lower limbs, classified as grade C according to the American Spinal Injury Association (ASIA) impairment scale (AIS). He denies a history of trauma or constitutional symptoms. On January 30, 2023, he underwent an MRI study (Fig. 1), revealing a lesion at the sixth thoracic vertebra (T6) with circumferential compression. Initially, a biopsy with histopathological examination was indicated (Fig. 2), revealing an aggressive hemangioma of T6.

Surgery for decompression with laminectomy without instrumentation was performed in the same internation, as there were no criteria for instability according to the Spine Instability Neoplastic Score, and the patient presented symptoms of spinal cord compression caused by the hemangioma in need of urgent therapy, without complaints of pain or changes in the height of the vertebral body. The patient was also placed in a SOMI brace and underwent physical therapy. A follow-up MRI (Fig. 3) 3 weeks later showed soft tissue and spinal cord edema. This was discussed in a multidisciplinary round, and 20 sessions of radiotherapy were recommended to prevent the progression of the extra-vertebral tumor.

Following radiotherapy sessions, a subsequent MRI (Fig. 4) 5 months after the laminectomy, revealed vertebral collapse and kyphosis (angle >20°), prompting the indication for thoracic vertebral fusion from the fourth to the eighth vertebra After a 3-

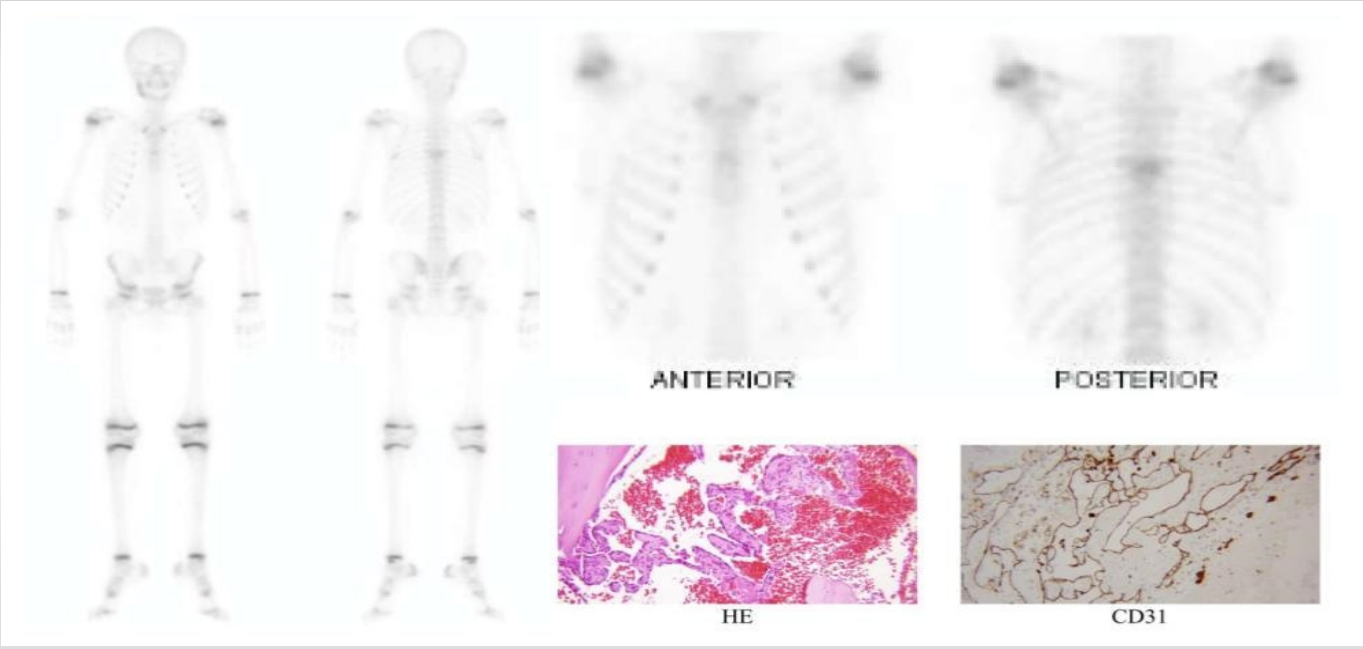

Figure 2: Scintigraphy with 99 m Technetium-MDP: Increased uptake in the right hemimandible and the sixth dorsal element suggests an infiltrative bone process. Histopathology: Bone and soft-tissue findings in the epidural region are consistent with venous proliferation, compatible with a venous hemangioma malformation without signs of malignancy. Immunohistochemistry: Compatible with hemangioma (CD31 and CD34 POSITIVE).

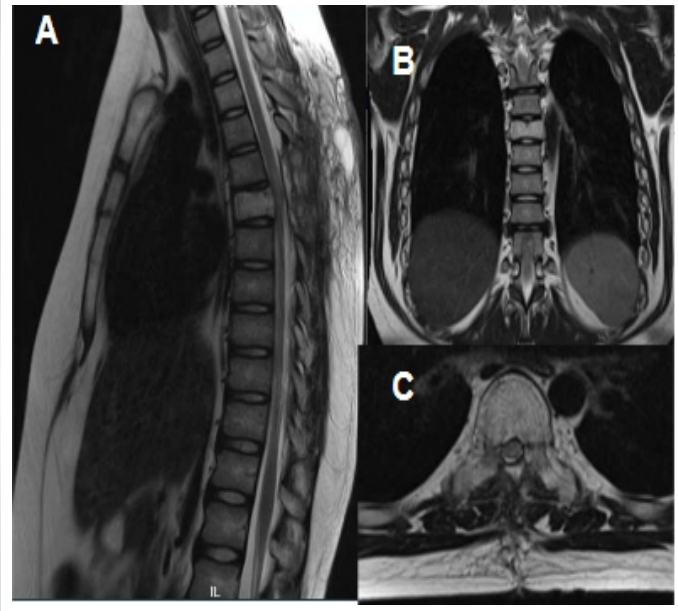

Figure 3: Magnetic resonance imaging: Signs of decompression, including laminectomies and resection of the spinous processes at the T5 and T6 levels. There is a significant reduction in the amplitude of the cerebrospinal fluid space, especially at the T6 level, and a slight increase in T2 signal intensity in the dorsal spinal cord at this level, without significant change in spinal cord thickness.

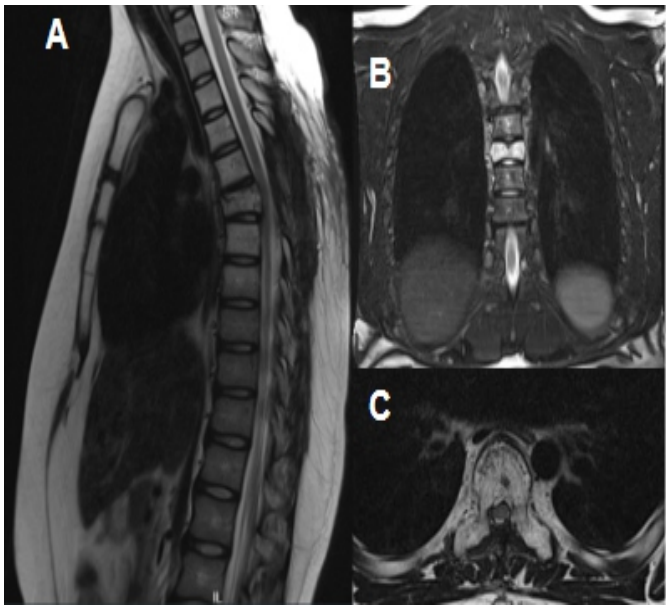

Figure 4: Magnetic resonance imaging: Signs of surgical manipulation, a slight increase in the amplitude of the cerebrospinal fluid space in the spinal canal, and a reduction in the T2 hyperintensity in the dorsal spinal cord at the T6 level. There is a significant reduction in the height of the T6 vertebral body, presenting kyphosis in image A.

month follow-up, the patient showed clinical improvement with no motor or sensory deficits, being classified as grade E according ASIA AIS. Radiological assessment (Fig. 5) demonstrated favorable outcomes, leading to the removal of the brace. The patient continued follow-up with physical therapy for muscle rehabilitation and improved mobility, with no pain complaints.

Discussion

Vertebral hemangioma is typically asymptomatic and discovered incidentally, with reported incidences of 11% in autopsy series, but symptomatic cases are much rarer at 0.9–1.2%. When symptomatic, these hemangiomas commonly affect the thoracic spine, with a noted epidemiological preference for young women around the age of 40 [1, 2]. The patient, in this case, being male and adolescent, deviates from this typical epidemiology found in the literature.

Few cases evolve with symptoms, which often manifest as pain. However, in rare instances, hemangiomas can be aggressive, showing significant vertebral expansion, extraosseous components with epidural extension, blood flow disturbances, and occasionally compression fractures, leading to spinal cord and/or nerve root compression [3, 7, 8].

Diagnosis is typically made through characteristic imaging findings. On plain radiography, lesions in the spine may appear as vertical parallel striations formed by the dissociation of bone trabeculae without widening of the vertebral body, which is considered pathognomonic, as described by Deetz in 1901. CT can show sparse bony trabeculae alternating with hypertransparent areas on axial slices. MRI demonstrates hyperintense signals in the affected vertebral bodies due to fatty substitution [4, 5].

Cases associated with back pain and without neurological deficits may be managed with periodic observation and clinical treatment, with refractory cases referred to exclusive radiotherapy or vertebroplasty [5,9]. Hemangiomas causing pain or spinal cord compression are associated with moderate-to-intense hypervascularization. If the feeding vessel also supplies the anterior spinal artery, assessed by arteriography, it can potentially be embolized as a preoperative adjunct or ligated during surgery, reducing the risk of severe intraoperative hemorrhage [6]. Surgical treatment is indicated for all cases with some degree of neurological deficit. Emergency decompression with laminectomy should be performed in all cases of rapid and progressive neurological deficit, with or without the need for adjuvant radiotherapy, depending on the extent of extraosseous involvement [5, 6].

In cases where the vertebral body is involved with extraosseous tumor extension into the spinal canal, causing spinal cord compression and neurological deficit, a more radical surgery such as corpectomy or intralesional spondylectomy with anterior reconstruction and posterior fixation has been advocated [9, 10].

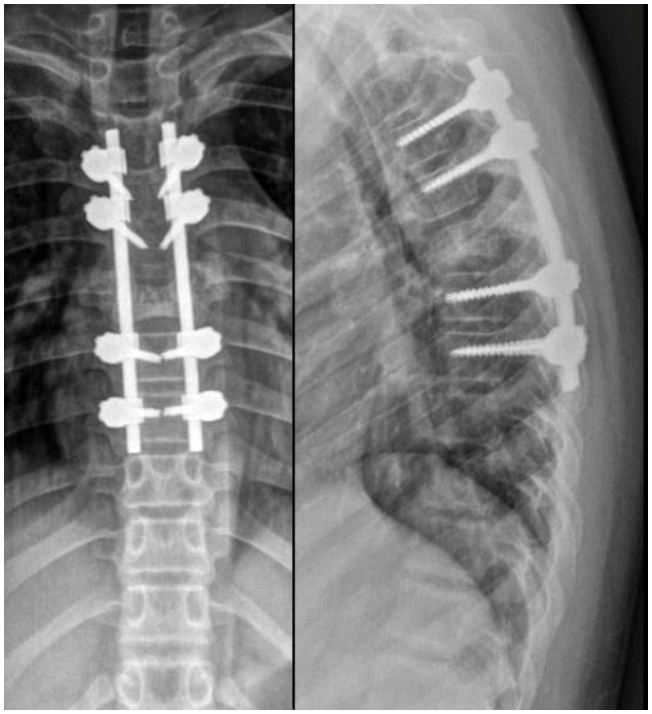

Figure 5: X-ray taken 5 months postoperatively, showing stable spinal fusion with no signs of hardware failure or recurrence.

There is no formal consensus on the ideal technique. However, the literature suggests that for asymptomatic cases without neurological deficits, minimally invasive procedures are effective. In contrast, for symptomatic cases, a combination of techniques such as preembolization, posterior decompression, subtotal resection, and vertebroplasty may offer greater benefit [11-13].

Conclusion

An accurate and early diagnosis is crucial in cases that deviate from the described epidemiology to achieve the best possible outcomes. This is particularly true for benign lesions with potential aggressiveness, impacting survival rates, complications, and overall quality of life for patients.

Clinical Message

Our case highlights the importance of early diagnosis and multimodal treatment, including laminectomy and radiotherapy, for managing aggressive vertebral hemangiomas with spinal cord compression. It outlines the management of the case and highlights the favorable outcome achieved for the patient.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given the consent for his/ her images and other clinical information to be reported in the journal. The patient understands that his/ her names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflict of interest: Nil **Source of support:** None

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Conflict of Interest: Nil
Source of Support: Nil

Consent: The authors confirm that informed consent was obtained from the patient for publication of this case report

How to Cite this Article

Gil GA, Gravina DS, Cobalchini GR, Bossardi PB, Giustina PG. Aggressive Thoracic Spine Hemangioma in a 13-Year-Old Boy: A Case Report with Literature Review. Journal of Orthopaedic Case Reports 2025 May;15(5):141-145.