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Case Report

Aggressive vertebral hemangioma masquerading as neurological disease in a pediatric patient

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ABSTRACT

Aggressive hemangioma is a rare vertebral lesion in pediatric patients which can present with deteriorating neurological function. It can mimic malignancy on imaging, particularly as it regularly has an extrasosseous soft tissue component. We present a case of a 13-year-old male who presented with a three month history of lower extremity weakness that was found to have an infiltrative mass at T10 with associated cord compression from epidural extension of the lesion. In this report we review the characteristic imaging findings associated with aggressive hemangioma, including its appearance on read-out segmented diffusion-weighted images. It is imperative that radiologists who interpret studies of children be aware that this lesion exists and what it looks like, as it can be associated with massive hemorrhage if encountered unexpectedly during surgery.

Introduction

Vertebral hemangiomas are the most common benign angiomatous lesions of the spine. The prevalence of hemangiomas increases with age, being most common after middle age [1]. They have an estimated incidence of ten to twelve percent in the general population, and most commonly affect the thoracic spine. Although the specific prevalence of vertebral hemangiomas in the pediatric population has not been reported, they have been described as "extremely rare," with only a limited number of cases reported in the literature [2]. The majority of vertebral hemangiomas are incidentally identified and are asymptomatic, although they may rarely become symptomatic due to osseous expansion and/or extraosseous extension. These "aggressive" hemangiomas constitute approximately 1% or less of all spinal hemangiomas [3]. Aggressive hemangiomas can easily be misdiagnosed as malignancy if not correctly identified on preoperative imaging, with potentially catastrophic consequences due to intraoperative blood loss [4].

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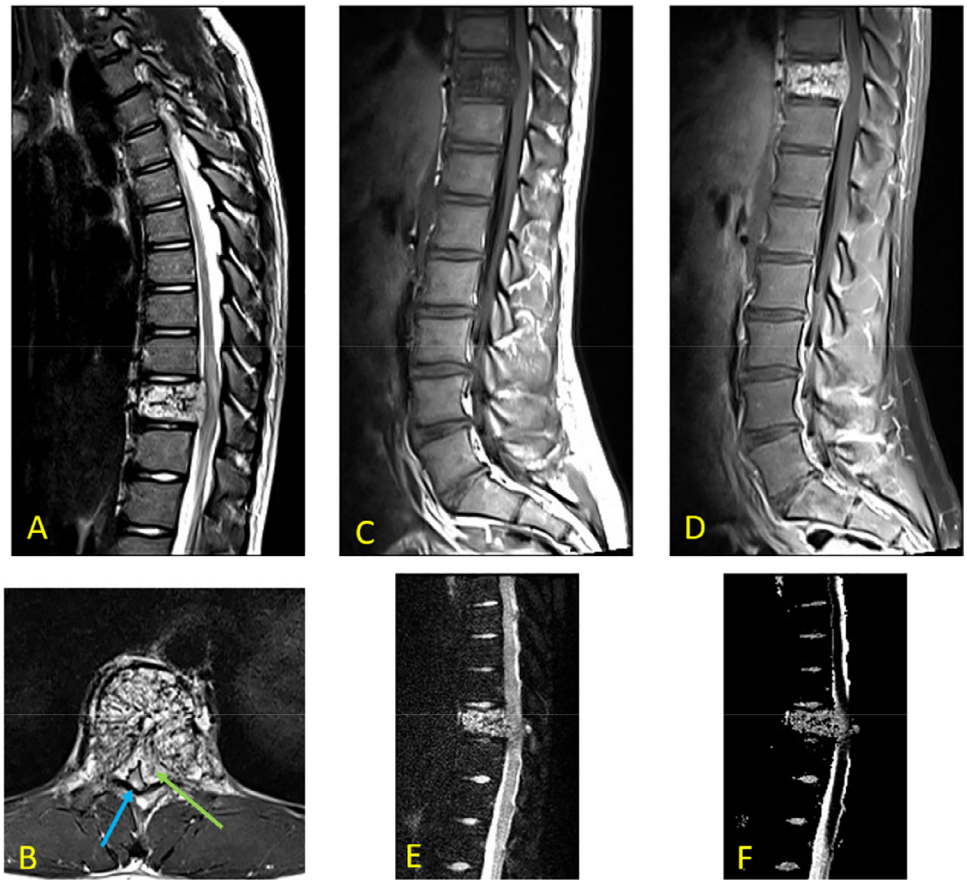

Fig. 1 – MRI of the thoracic spine without and with contrast. There is an infiltrative mass throughout the T10 vertebrae that is T2 hyperintense (A), with multiple internal T2 hypointensities likely reflecting a combination of trabecula and flow voids. There is a significant epidural component (green arrow) with resulting narrowing of the vertebral canal and cord compression, causing abnormal edema-like-signal within the cord (blue arrow) (B). The mass results in diminished T1 marrow signal (C) and enhance avidly (D). The mass demo^{nst}rates facilitated diffusion on the readout-segmented DWI sequence (DWI, E and ADC, F). (Colored version of figure is available online.)

Case summary

Presentation and physical examination

A 13-year-old boy with a history of asthma presented with a 3-month history of lower extremity weakness and balance problems resulting in worsening gait. Onset of his symptoms was not associated with trauma, illness, or new medications. At presentation, he could only walk short distances without holding onto something for support. He reported multiple falls since the onset of his symptoms with inability to catch himself.

He was initially seen by an outside orthopedist, who referred him to Child Neurology at our institution. His initial neurological exam was significant for a positive Rhomberg and weakness in hip flexion and ankle dorsiflexion. Reflexes were intact and there was no dysmetria or tremor.

Imaging findings

Initial outpatient magnetic resonance imaging (MRI) of the spine demonstrated a mass throughout the T10 vertebral body which extended into the posterior elements, as well as into the epidural and paraspinal regions (Fig. 1). The mass demonstrated heterogeneous T2 signal hyperintensity, with multiple internal serpentine areas T2 signal hypointensity felt to be most indicative of flow voids. The mass enhanced avidly, and demonstrated facilitated diffusion. There was resulting severe narrowing of the vertebral canal at the level of the mass due to its epidural extension, with associated effacement of the intrathecal CSF space and deformity of the cord at this level. T2 signal hyperintensity within the cord was compatible with cord edema. There was also associated narrowing of the bilateral T10/T11 neuroforamen due to the extension of the mass into these areas. No additional vertebral or spinal lesion was identified.

Subsequent computed tomographic (CT) imaging of the thoracic spine redemonstrated the lesion centered within the T10 vertebrae (Fig. 2). The vertebral body demonstrated thickened trabecula with a somewhat striated appearance on sagittal and coronal images. There were scattered areas of apparent osteolysis throughout the vertebrae including within the inferior aspect and along its left lateral margin. There was asymmetric enlargement of the segmental arteries at the T10

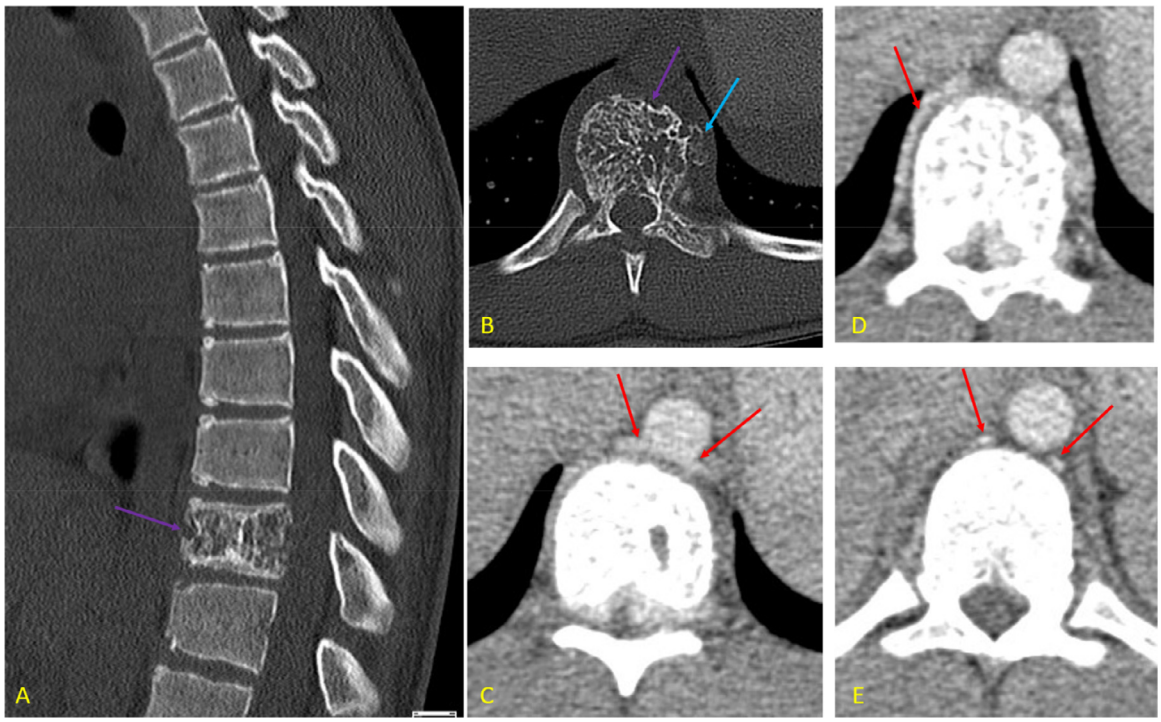

Fig. 2 – CT of the thoracic spine without and with contrast. Unenhanced sagittal (A) and axial (B) reconstructions through the T10 vertebrae demonstrate accentuation of the trabecular markings, which appear striated on the sagittal images. There are scattered areas of osteolysis (purple arrows), as well as a small anomalous ossific protuberance (blue arrow). Enhanced thin axial reconstructions demonstrate asymmetric enlargement of the segmental arteries at this level (red arrows in C and D), compared with the T11 level (red arrows in E). (Colored version of figure is available online.)

level, and asymmetric enlargement of the paravertebral veins draining into the azygous/hemiazygos system at the level of the lesion. CT of the neck, chest, abdomen, and pelvis did not identify any additional lesions.

Surgical findings

Upon identification of the mass, the patient and his family were immediately directed to the pediatric emergency department (ED), and the patient was started on IV dexamethasone. Both pediatric neurosurgery and pediatric hematology-oncology were consulted.

Due to the suspected hypervascularity of the lesion, and as the leading preoperative differential consideration based off the imaging characteristics was an aggressive hemangioma, prior to surgery the patient underwent an angiogram with the intent to preoperatively embolize the lesion (Fig. 3). The endovascular neurosurgeon initially localized the artery of Adamkiewicz to left T12 radicular artery, and subsequently catheterized the left T10 radicular artery in anticipation of embolization. Prior to initiating the embolization, a selective injection through the vessel demonstrated what appeared to be an accessory/duplicated artery of Adamkiewicz. As such, the right T10 radicular artery was then selectively catheterized and 4 mL of Onyx-18 was injected into the lesion. The embolization was terminated when a small amount of reflux of contrast was identified in the contralateral (left) radicular artery.

After the angiogram, the patient underwent surgical resection of the lesion. This was accomplished by performing a T10 corpectomy/laminectomy/medial facetectomy with placement of an expandable cage at T10 (Fig. 4). Even with adequate pre-operative embolization there was still significant intraoperative blood loss (2.5 L), required four units of intraoperative packed red blood cells and two units of fresh frozen plasma. The patient also underwent arthrodesis of T8-T12 using both surgical instrumentation and bone allograft.

Pathology

The surgical pathology specimen was described as “hemangioma.” The lesion was located between bony trabeculae and was composed of vascular channels of varying sizes and foci of tightly packed capillary vasculature with an anastomosing growth pattern (Fig. 5). The nuclei were benign with no significant atypia or pleomorphism. Additionally, no significant mitotic activity was identified. Evidence of embolization prior to surgery was present in the examined sections. Immunohistochemically stained sections using antibodies to CD34 highlighted the numerous small vascular channels within the areas of solid anastomosing growth.

Outcome

The patient was discharged to an inpatient rehabilitation center following his hospitalization. At the time of this report (ap-

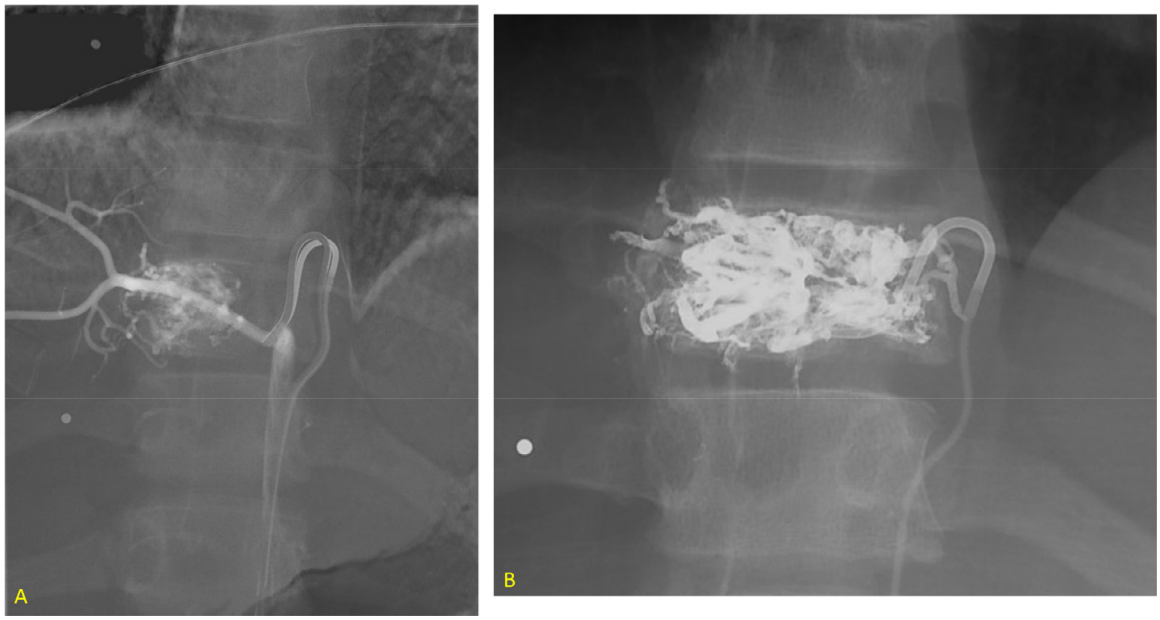

Fig. 3 – Angiographic images obtained during the embolization procedure. A representative image (A) after injection into the right T10 radicular artery illustrates the hypervascularity of the lesion prior to embolization (B).

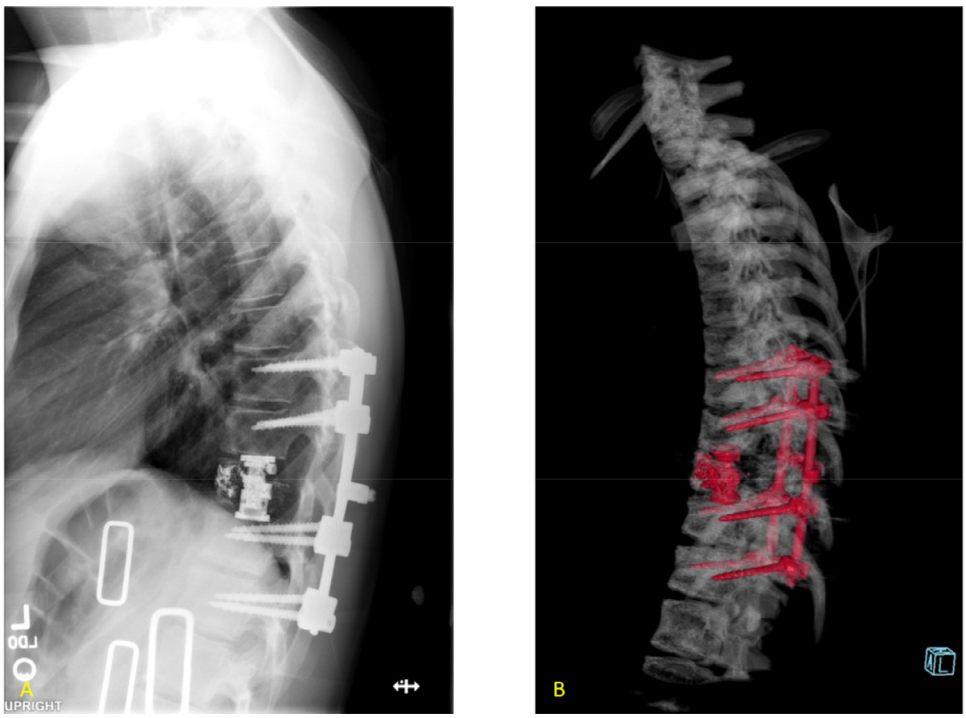

Fig. 4 – Postsurgical changes including T10 corpectomy, placement of an expandable cage at T10, and posterior instrumented fusion of T8 through T12. Lateral radiograph (A) and volume-rendered 3D recon from CT (B).

proximately 2 months after his surgery) the patient is able to ambulate independently without orthotic or assist device, and has had no falls. His gait is slow and sometimes off-balance, which has been improving. He continues to undergo physical therapy.

Informed consent was obtained from the patient's family to submit the case for publication as a case report.

Discussion

Vertebral hemangiomas are relatively common, benign vascular lesions that affect the vertebral column. They account for approximately 2%-3% of all spinal tumors, and occur with an estimated incidence of 10%-12% in the adult population.

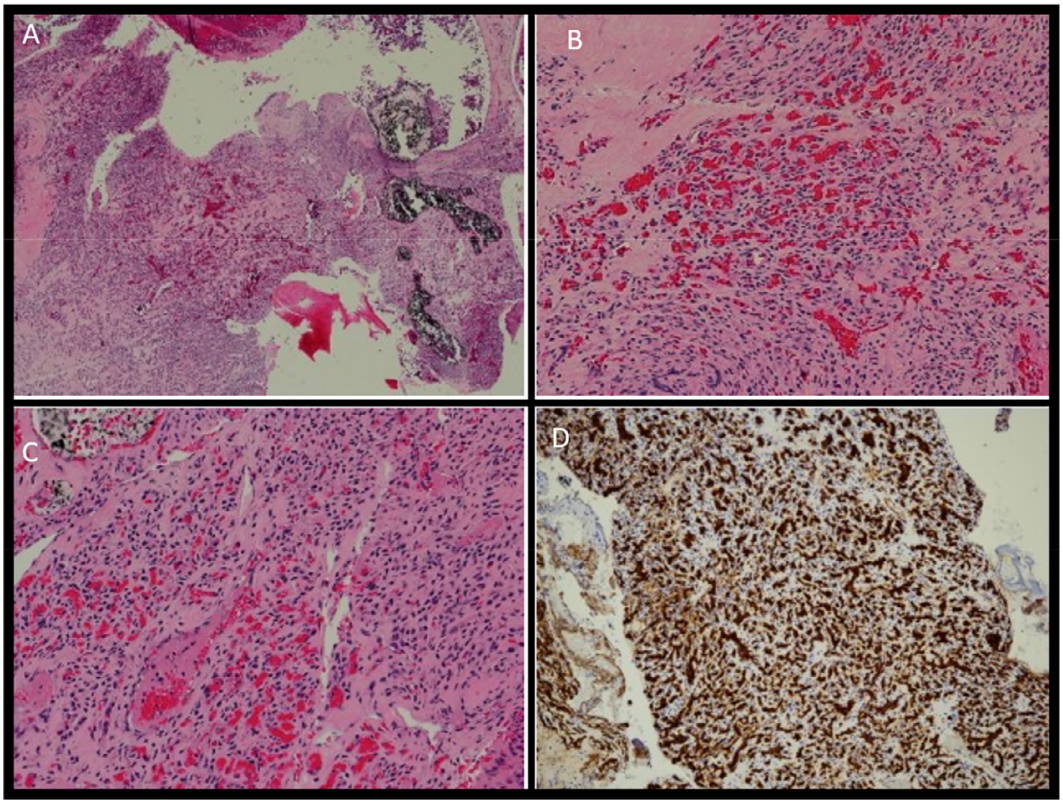

Fig. 5 – Histologic images of the lesion. (A) 4× image showing bony trabeculae with a vascular lesion with tightly compressed capillary vascular channels and evidence of embolization prior to surgery (B) 10× image showing a benign vascular lesion with tightly packed capillary vascular channels (C) 20× with no significant atypia or mitotic activity. (D) The vascular channels are highlighted with an immunohistochemical stain to CD34.

These lesions may rarely enlarge and cause pain or neurological deficit due to spinal cord compression, expansion of the vertebral body and/or posterior elements, or pathologic fracture [5]. These lesions are referred to as "aggressive hemangioma" when they are associated with extraosseous extension. There is a known association between aggressive hemangiomas and pregnancy, which is perhaps attributable to hormonal changes associated with pregnancy [6]. Rare reports of vertebral hemangiomas in the pediatric age group have suggested that they tend to behave more aggressively compared to adults, noting that these patients have generally been in their adolescence suggesting that a hormonal influence may also be involved [4].

When a vertebral lesion causing cord compression in a pediatric patient is encountered by imaging, it is imperative to identify imaging characteristics that could indicate an aggressive hemangioma. Surgery of aggressive hemangioma is often associated with massive hemorrhage from these highly vascular tumors, and preoperative transarterial embolization of the vessels feeding the hemangioma has been shown to decrease intraoperative blood loss and associated morbidity [5]. Aggressive hemangiomas within the vertebrae have been described in the literature as being misdiagnosed preoperatively by imaging as lymphoma, myeloma, metastasis, schwannoma, chordoma, and aneurysmal bone cyst [7]. Particularly in the pediatric population, it is important to note that Ewing sarcoma can have a similar imaging appearance [1].

Characteristic imaging findings of a vertebral hemangioma can help to distinguish it from other spinal lesions. The majority of hemangiomas are seen in the thoracic and lumbar spine, and typically confined to the vertebral body. Hemangiomas in the vertebrae characteristically have thickened trabeculae and a coarse "honeycomb" appearance on imaging [1,5]. CT often shows a polka-dot appearance that represents a cross-section of reinforced trabecula [1,4].The appearance of a hemangioma on MRI is dictated by its histopathologic features – high vascularity, interstitial edema, and interspersed fat. T1 and T2 signal hyperintensity is related to extent of adipocytes or vessels and interstitial edema, respectively. The lesions typically enhance [1].

Aggressive vertebral hemangiomas are identified by having an associated epidural mass and/or paravertebral extension. Additional imaging characteristics that have been reported in the literature that can be associated with aggressive hemangiomas include osteolytic bone destruction, vertebral compression fractures, lesions involving multiple segments, and epidural compression of the spinal cord by either the lesion

itself or by associated osseous compression [7]. The identification of flow voids can be an additional useful imaging characteristic [1].

In our case, diffusion-weighted imaging (DWI) of the lesion provided additional useful information in helping to delineate the lesion preoperatively from other more common pediatric vertebral lesions, such as Ewing Sarcoma or neuroblastoma metastasis. It has been demonstrated that, in general, malignant vertebral lesions usually demonstrate a lower apparent diffusion coefficient values compared with benign vertebral lesions [8]. The high apparent diffusion coefficient value in our patient's lesion helped us to differentiate it preoperatively from one of these malignant lesions. Although DWI of the vertebrae and spine has historically been challenging due primarily to susceptibility artifact from the bone, the recent development by some MRI vendors of readout-segmented DWI sequences has helped to mitigate these artifacts and to make DWI imaging of the spine and vertebrae more practical [9]. At our institution, we routinely incorporate a readout-segmented DWI sequence when imaging vertebral, spinal, and paraspinal masses.

At the time of embolization, our patient was found to have duplication of the Artery of Adamkiewicz. This congenital variant has been reported to occur in approximately five percent of patients [10]. Had this not been identified by the endovascular neurosurgeon prior to the planned embolization of the left T10 radicular artery, the procedure could have resulted in partial paralysis of the patient.

Multiple treatment strategies have been reported in the literature for aggressive hemangioma including a combination of percutaneous vertebroplasty, transarterial embolization, radiation therapy, decompressive laminectomy, and vertebrectomy [5,11]. Recurrent aggressive hemangioma after surgical decompression that required further therapy have been reported [11].

Consent statement

Informed consent was obtained from the patient's family to submit the case for publication as a case report.

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