Atypical Presentation of Aggressive Vertebral Hemangioma with Spinal Cord Compression

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Abstract
Symptomatic aggressive vertebral hemangiomas are rare sub variety of much common benign asymptomatic vertebral hemangioma presenting with bone expansion, extra osseous extension, epidural soft tissue component, compression fracture leading to neural and or cord compression. It can present with diagnostic dilemma especially in elderly patient mimicking serious pathology like metastasis. We present a case report of an elderly female presenting with progressive paraparesis, diagnosed for aggressive vertebral hemangioma and managed by preoperative embolisation and decompressive surgery. This article highlights the imaging spectrum including spinal angiography in such conditions.

Keywords: aggressive hemangioma; angiography; cord compression; extradural component

1. Introduction
Hemangiomas are congenital benign vascular malformation of dysembryogenetic or hamartomatous origin.[1] Majority of the vertebral hemangiomas are asymptomatic and are incidental findings on imaging with only less than 1% presenting with features of radiculopathy or cord compression known as aggressive vertebral hemangiomas.[2] Here we present a case of aggressive vertebral hemangioma involving D4 vertebra presenting with paraparesis secondary to extradural component with aim of highlighting the various imaging spectrum of this unusual sub type along with features on digital substraction angiography.

2. Case Report
68 years old female presented with gradual progressive weakness in bilateral lower limbs. Examination showed a spastic paraparesis with grade IV weakness in both legs with patchy sensory loss.

Radiograph dorsal spine AP-Lat view showed vertical linear streaks over D4 vertebral body (Figure 1).

Figure 1: Radiograph dorsal spine lateral view showing coarse, vertical, trabecular pattern over D4 vertebra with osseous reinforcement (trabecular thickening)

Axial computed tomography (CT) scan at the level of the lesion showed bony expansion with classic “polka dot” appearance over D4 body and posterior element. There was an illdefined anterior epidural mass causing cord compression (Figure 2).
On MRI study the lesion showed low signal intensity with scattered areas of increased signal on T1 weighted images with marked hyper intensities on T2 weighted images. Axial T1,T2 weighted images showed irregular lobulated vertebral margin of D4 body and multiple small hypo intense vascular flow voids within and bilobed configuration epidural component causing cord and neural compression (Figure 3).

Post contrast MR images showed intense enhancement over D4 body, posterior element with clear delineation of bilobed configuration enhancing epidural component causing neural and cord compression.

Above imaging features suggested a vascular lesion with aggressive hemangioma being the most likely diagnosis and recommendation was made for angiography.

Digital subtraction spinal angiography through right femoral artery route using 5F cobra catheter revealed abnormal vascular blush and vessels over D4 vertebra feeded from bilateral segmental dorsal arteries. (Figure 4)
Looking at the vascular nature of the lesion, preoperative endovascular embolisation was performed using polyvinyl alcohol particles on next day followed by definitive decompressive laminectomy and debulking of epidural mass.

At 15 days follow up she had made a satisfactory sensory and motor function recovery.

3. Discussion

Hemangiomas are common benign vasoformative neoplasm or developmental condition of endothelial origin that closely resembles normal vessels. These are benign osseous lesion of dysembryogenetic origin or a hamartomatous lesion.[1]

First described by Perman in 1926, followed by Bailey and Bucy in 1930, vertebral hemangiomas are commonest benign lesions of the spinal column with an estimated incidence of 10-12% based on large autopsy series and reviews of spinal radiographs. Typically these are incidental findings and are symptomatic in only 0.9 to 1.2% [2] of adults and are known as aggressive vertebral hemangiomas presenting with radiculopathy or spinal cord compression, commonly seen in thoracic spine.

These aggressive hemangiomas presenting with signs and symptoms of myelopathy can mimic serious pathology like metastasis, lymphoma, and tuberculosis.

Aggressive vertebral hemangiomas are characterized by bone expansion, extra osseous tumor extension, disturbance of local blood flow, epidural hemATOMA, epidural soft tissue component, compression fracture of involved vertebra leading to compressive symptoms.[3]

Microscopically hemangioma can be capillary, cavernous, arteriovenous and venous types. In bone, the capillary and cavernous types are common. The histological pattern is characterized by the proliferation of anomalous thin walled blood vessels and sinuses lined by endothelium between the thickened, vertically oriented trabeculae of bone. The dilated vascular channels are set in a stroma of fat. The ratio of fat to vascular tissue wrapped between the pillars of bone determines the signal intensity on MRI.

Aggressive vertebral hemangioma in elderly usually presents with varied differential diagnosis due to alarming symptomatology requiring a dedicated radiological imaging. These lesions are characterized by thickening of vertically striated trabeculae which preserve the functional axial load bearing capacity. This appearance as seen on radiograph and CT are described as “polka-dot”, “honeycomb”, “corderoy cloth”, “jail bar”, “salt and pepper” with decrease in the density of vertebra due to fatty marrow content.[4] In addition CT scan can also detect posterior element involvement, bony expansion, epidural hematoma and soft tissue component and cord compression.

MRI plays a pivotal role in evaluating the patient with backache, radiculopathy or myelopathy by diagnosing benign pathology like aggressive hemangioma and differentiating it from more serious malignant aetiologies.

In asymptomatic vertebral hemangioma, MRI showed increase signal intensity on both T1 and T2 weighted images with alternate hypointense areas. Chemical shift images and histological studies demonstrated that adipose tissue causes the increased signal intensity, while the hypointense areas represent vascular flow voids.

Aggressive vertebral hemangiomas contain less fat and more vascular stroma, thereby producing a low signal on T1 weighted images. Presence of higher T2 weighted signal, coarsened trabeculae, multiplicity helps in differentiating it from the metastatic lesion. Also MRI helps in detecting the involvement of posterior element and epidural component and status of the spinal cord.

In radiologic evaluation of vertebral hemangiomas, Laredo et al[5] described six features which were seen significantly more often in symptomatic vertebral hemangioma: location between D3 and D9, involvement of the entire vertebral body, extension to the neural arch, an expanded cortex with indistinct margins, an irregular honeycomb pattern, and soft tissue mass. They also suggested that fatty vertebral hemangioma may represent inactive form while soft tissue content at CT and low signal at MRI may indicate a more active vascular lesion with potential to compress the spinal cord.

Workup for aggressive vertebral hemangioma also includes spinal angiography to determine vascularity, locating the feeding artery and draining veins. It also plays an important role in intra arterial embolisation as a preoperative adjuvant procedure.

Our patient was confidently diagnosed for aggressive vertebral hemangioma with epidural component causing cord compression using CT and MRI followed with digital subtraction spinal angiography to delineate the vascular blush over D4 vertebral body.

The management of symptomatic vertebral hemangioma requires a combined multi modality approach including surgical decompression, preoperative endovascular embolisation, radiotherapy, absolute alcohol ablation, percutaneous
Surgery is mainstay treatment option including bony decompression by laminectomy or vertebrectomy with excision of soft tissue component. [6]

Due to its high vascular nature, surgery alone is associated with increased mortality due to blood loss, illdefined surgical planes, formation of epidural hematoma and its related complication. [7]

Endovascular embolisation using polyvinyl alcohol, cyanoacrylate is now recommended preoperative therapy to reduce the bleeding and its related complication.[8]

There has been considerable increased use of percutaneous injection of methyl methacrylate for stabilizing vertebra at risk of collapse or for alleviating severe backache. [9]

Percutaneous sclerotherapy is another adjuvant therapy using sclerosants like absolute alcohol and recently sodium tetradecyl sulphate. [10]

Our patient was managed by preoperative transarterial embolisation using polyvinyl alcohol particles followed by definitive laminectomy and debulking of the epidural mass with surgeon reporting less intraoperative bleeding with good surgical planes.

4. Conclusion

Most often the vertebral hemangiomas are incidental benign imaging findings; however a rare subset of aggressive hemangioma may present with devastating neurologic sequel. It is important to keep aggressive vertebral hemangioma as a differential diagnosis in any patient presenting with myelopathic symptoms and obtain early imaging, especially MRI to guide proper treatment. Additional imaging modality such as angiography is now recommended adjuvant diagnostic and therapeutic option for definitive management.

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