

RAPIDLY PROGRESSIVE ATYPICAL AGGRESSIVE VERTEBRAL HEMANGIOMA: A CASE REPORT

Orthopaedics

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ABSTRACT

Vertebral hemangiomas are relatively common vascular neoplasm of endothelial cell. It is asymptomatic and often present as incidental finding. Surgery is recommended for aggressive vertebral hemangiomas if symptoms are severe or there is involvement of any neural structures. A 40-year-old female patient was admitted to our department with complain of severe back pain. Series of radiological investigation like X-ray CT Scan, MRI were suggestive of vertebral hemangioma at D11 level with involvement of posterior elements and extension into the epidural space with spinal cord compression. The patient treated surgically with intraoperative vertebroplasty with posterior spine fixation from D-9 to L-1 level. Histological examination confirmed the diagnosis of atypical hemangioma, specifically an epithelioid hemangioendothelioma.

KEYWORDS

Hemangioma; Vertebrae; Spinal cord compression, vertebroplasty

INTRODUCTION

Vertebral hemangiomas are extremely common lesion composed of blood vessels that form in the bone of spine. It is most common primary tumor of spine¹

Most vertebral hemangioma are quiescent and asymptomatic often present as incidental finding typically involve the vertebral body but rarely they can exhibit active behaviour like quickly growing and extending till posterior elements, such as the laminae, pedicles, or spinous process known as atypical VH, or involving paravertebral and epidural space with compression of spinal cord and /or nerve roots known as aggressive VH². Atypical and aggressive hemangioma are difficult to diagnose as they can mimic bony malignancies³.

Vertebral hemangioma can be observed at any age, but mostly in fifth decade of life with female predominance, most common location is thoracic and lumbar spine with most common complain of back pain⁴.

The diagnosis includes a CT scan, where the typical "polka-dot-sign" is observed due to the trabecular aspect of the involved vertebra. In addition, the diagnosis may include an MRI, where the lesion appears hyperintense on both the T1 and the T2 weighted images with contrast enhancement. Aggressive atypical hemangiomas, like epithelioid hemangiomas or hemangioendotheliomas, may demonstrate a mixed pattern, with heterogeneous signal intensity on T1 and T2 weighted MRIs that reflect the presence of inflammatory infiltration and the absence of fat. Hemangiomas are more likely to show aggressiveness when its stroma contains less fat and is more vascular. The radiologic features demonstrating the aggressiveness of the disease include the presence of a low signal on T1 weighted images and a high signal on T2 weighted images on the MRI. In addition, avid contrast enhancement may reveal the presence of cortical erosion, soft tissue stroma between the osseous trabeculae on the CT, the presence of extradural soft tissue, expansion to the posterior elements, invasion of the spinal canal, and encroachment of the spinal cord.

In most of patients hemangioma is incidental finding and does not need any treatment but when it extend to posterior element or in epidural space as in atypical or aggressive VH, require surgical treatment.

We report a rare case of a rapidly progressing aggressive hemangioma extending to posterior element and paravertebral and epidural extension at the D11 level. The case was managed by intraoperative vertebroplasty with posterior decompression and fixation from D-9 to L-1 vertebrae

presented with 10 month history of severe mid back pain with difficulty in walking and sensory disturbances in both lower limb. There is no bowel and bladder involvement. Pain is increased with activity and on standing and decreased on rest and lying down. Symptoms start as radicular pain in her legs than gradually development of unstable gait. On clinical examination, motor examination was normal (MRC Grade 5/5) with decreased sensation to light touch and painful stimulation at mid-thoracic level and bilateral lower limb. Hyperreflexia and increased tone were observed. MRI of thoracic spine shows aggressive/atypical hemangioma of D-11 segment involves vertebral body (left > right) and left-sided posterior vertebral elements. It appears hypointense on T-1W and bright on STIR images. Post contrast, it shows intense vascular-type enhancement which is slightly heterogeneous. Expansile nature with extra- osseous component occupies anterior, left lateral and left posterior epidural spaces at D11 level and causes severe compression, distortion and rightward displacement of cord. Focal abnormal intramedullary T-2W hyperintense signals at D-11 indicate compressive myelopathy or cord edema. Narrowing of left D10-11 & D11-12 foramina with compressions of exiting nerves. Involvement of D11 spinous process and right lamina. The findings were consistent with an aggressive hemangioma (Enneking Stage-3)

Operative Technique

On the basis of clinical and radiological findings posterior decompression with vertebroplasty plus pedicle screw fixation is planned. General anesthesia given to patient and placed in prone position and body was supported by pads below chest pelvis and abdomen were suspended. Incision site was marked above the affected vertebra under c arm guidance. Sufficient amount of gelfoam was scraped into small pieces and mixed with 20 mL of contrast medium and kept aside. Incisions were marked and surgical site was exposed and visualized. Pedicle screws were fixed two level above and below the affected vertebra(D-11), after that the gelfoam was injected to the site of the affected vertebral body with a 20-mL syringe with 18-G needle through both the pedicles until no more gelfoam could be injected. The leakage of the contrast medium was observed via C-arm machine. The bone cement paste was prepared for the vertebroplasty, which was performed under C arm guidance, using a thoracic puncture needle. A bipedicular approach is used approximately 4-5 mL of polymethylmethacrylate was injected slowly and carefully which was sufficiently filling the lesion without any leakage. After injecting bone cement, decompressive laminectomy was performed to decompress the spinal cord. Intraoperative biopsy was taken for histological findings. Final fixation spine done using rod and pedical screw to enhance stability.

Case Pathology

Histopathological examination revealed a soft tissue lesion composed

Case Report

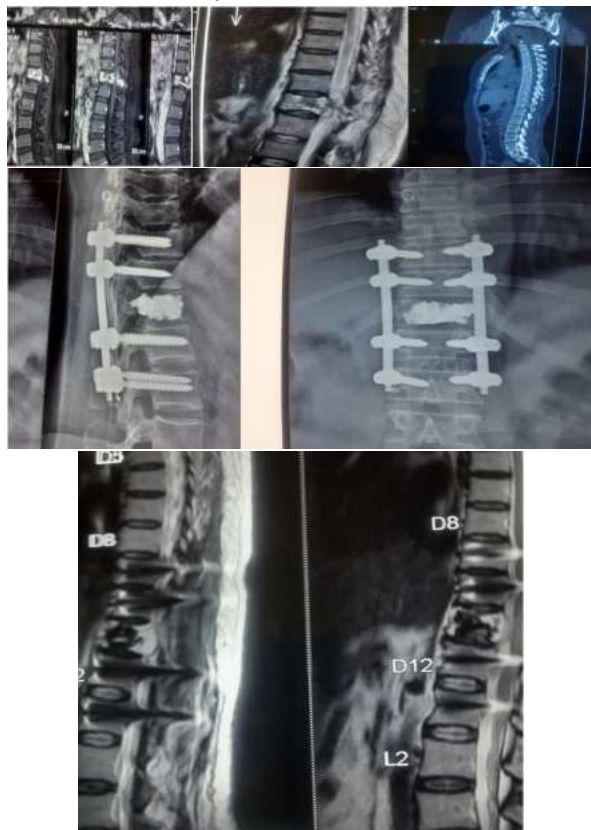
Preoperative Presentation

A 40-year-old female patient with no known chronic medical problem

of dilated and congested thin-walled capillaries with no wall thickening or muscular component as well as fatty replacement of vertebral bone marrow. The histological features were diagnostic of epithelioid hemangioendothelioma.

Postoperative Follow Up

The patient noted a slow improvement in neurological status and at a 6-month follow-up was noted to have normal gait and balance and return of normal lower extremity sensation.



DISCUSSION

Vertebral hemangioma is the most common tumor of the spine predominantly affecting lumbar and thoracic spine accounting for 10% at general autopsy⁵

They can elicit secondary reactive phenomenon such as fibrotic response showing variable histology of capillary, cavernous, arteriovenous or venous⁶

They are most commonly distributed in the spine more than that in skull. Typical hemangiomas can be diagnosed by imaging, aggressive ones pose a challenge in which case there is a need of CT guided biopsy. However, a majority of vertebral body hemangiomas are managed conservatively⁷

Rarely they cause symptoms of compression due to like epidural hemorrhage, burst fracture including radicular symptom or cord involvement.

The differential diagnosis of atypical vertebral body hemangioma includes: metastases, solitary spine plasmacytoma, multiple myeloma, lymphoma, aneurysmal bone cyst, osteosarcoma, Paget's disease and malignant vascular tumors such as epithelioid hemangioendothelioma and angiosarcoma. Their diagnosis can be challenging and requires a high level of suspicion and specialization and at the same time detailed work up to elucidate any primary or secondary malignancy

Vertebral hemangioma might also display an "atypical" appearance on MR imaging because of their histological features (amount of fat, vessels, and interstitial edema). Although the majority of VHs are asymptomatic and quiescent lesions, they can exhibit active behaviors, including growing quickly, extending beyond the vertebral body, and invading the paravertebral and/or epidural space with possible

compression of the spinal cord and/or nerve roots ("aggressive" VHs).⁸ The available literature regarding the natural history, prognosis, recurrence rates and optimal oncological and surgical treatment of AVH is very limited, although, the first description of a vertebral hemangioma with neurologic deficit was published in the 1930s⁹

Previous studies raised the recurrence rate up to 27.3 %¹⁰

The surgical approach has been widely debated, with no single approach for treatment of vertebral hemangioma, with some authors recommending decompression and radiotherapy alone, while others support a gross total resection with 360-degree reconstruction to prevent late recurrence.¹¹ Despite this, after extensive literature review, we propose that, for patients with atypical vertebral hemangiomas with mild or slowly progressive neurological symptoms, it is reasonable to attempt non-operative management with embolization, vertebroplasty, or radiotherapy. This treatment is recommended that provided symptoms are not due to spinal compression by a focal bony prominence, which is unlikely to resolve without surgery. However, for patients with severe or rapidly progressive symptoms, surgery should be considered. Aggressive vertebral hemangiomas are benign lesions that do not have metastatic potential and are not associated with mortality. In the present case, the patient complained of severe back pain with gait imbalance and sensation disturbance in lower extremity. The vertebral hemangioma was treated with intraoperative vertebroplasty and percutaneous screw fixation. Bone cement augmented percutaneous screw fixation ensures mechanical stability and prevents vertebral collapse, and bone cement itself has the exothermic reaction causing tumor necrosis. After surgery, the disabling back pain improved dramatically, and radiological studies showed no local recurrence of the tumor. Nevertheless, regular clinical and radiologic follow-up is necessary to check for tumor recurrence.

Keeping in mind, the significant destruction and expansion of posterior vertebral body, wide decompression is necessary to avoid further complications.

CONCLUSION

Although most vertebral hemangioma diagnoses are often incidental imaging findings, they can display aggressive features and rapid progression requiring surgical treatment. Diagnostic imaging awareness and an excellent understanding of the pathology is paramount to rule out other lesions that mimic aggressive hemangiomas.

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