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CASE REPORT

Recurrent Hemangioendothelioma of the Spine

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ABSTRACT

Introduction: Hemangioendotheliomas are vascular neoplasms that rarely involve the neuraxis.

Case report: A 50-year-old male patient complaining of severe pain and weakness on both his legs. Patient had history of spinal tumor operation 13 years ago. On examination, patient showed tenderness on his back and no localizing signs.

Investigations: X-ray and 3D reconstructive CT scan of the dorsal spine showed involvement of D11 and L1 with collapsed D12. MRI showed kyphosis deformity and significant cord compression.

Treatment: Preoperative embolization was performed followed by transthoracic removal of the tumor and D12 vertebral body which was followed interbody vertebral cage. A second surgery was undertaken to further excise the tumor and stabilize the spine.

Results: Patient was relieved of his symptoms and neurological deficit postoperatively.

Conclusion: A rare case of spinal hemangioendothelioma was treated with embolization and two step surgery requiring stabilization.

Keywords: Hemangioendothelioma, Dorsal spine, Stabilization.

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INTRODUCTION

Hemangioendothelioma (HE) is a vascular neoplasm composed of endothelial cells and is considered to be of an intermediary pathology between hemangioma and frankly malignant angiosarcoma.[1] Hemangioendothelioma is not uncommon in the soft tissues and has also occured in the oral cavity, mediastinum, bone and various superficial and deep body areas.[2] However, neuraxis involvement is infrequent.[3,4]

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We describe a rare case in the literature of a recurring HE case causing gross instability of the dorsal spine.

CASE REPORT

A 50-year-old male patient presented with history of imbalance and numbness on both lower legs since 3 years ago. He was operated on his dorsal spine in 1996 in a place several hundred kilometers from Mumbai due to spinal tumor causing instability of the dorsal spine with hystopathology report unknown. Tumor was excised and the spine was stabilized with Harrington rods and onlay auto bone grafts from the iliac crest.

He remained symptom free for almost 13 years when he presented to us the first time in 2009, with severe pain in the back due to dislocated Harrington rods (Figs 1A and B). In view of severe pain, the Harrington rods were removed but it was not an easy job as the rods were fully submerged into the new bone formation.

Being relived of pain patient promised us that he would return soon for evaluation and management of tumor recurrence but it took him more than 3 years to report to us again. Clinically, he had some weakness in the lower limbs. He could walk but he was limping and now had developed severe pain locally which made it impossible for him to turn in bed easily.

He was thinly built and did not suffer from any other ailment. He did not smoke or chew tobacco.

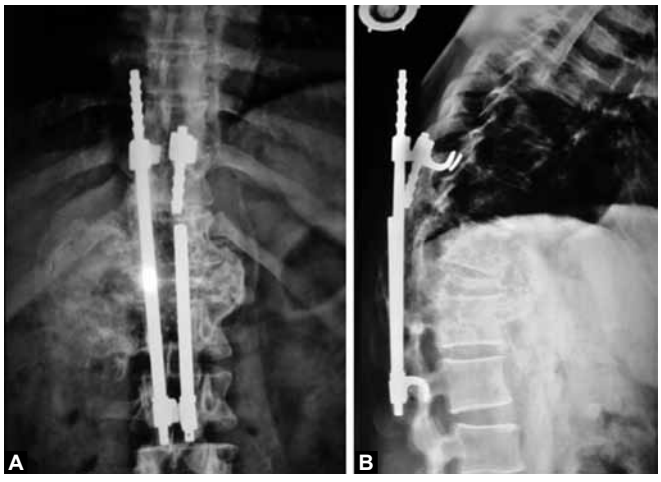
On examination, patient was afebrile and showed tenderness on his low back. Both SLR were restricted to 20º and both extrimities weak with power 4/5. Peripheral pulses in both limbs were normally felt. Bladder and bowel were normal.

INVESTIGATIONS

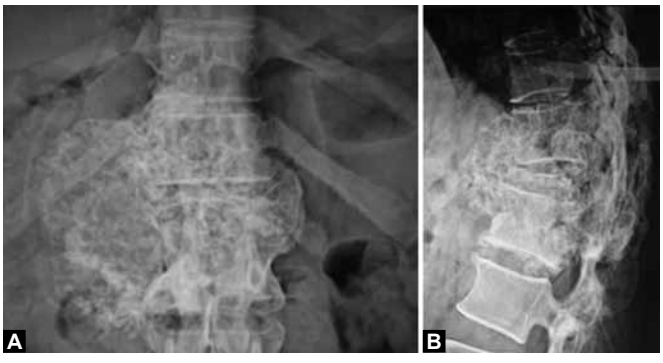
Laboratory findings were unremarkable and digital X-ray of the dorsal spine showed recurrence of the tumor causing total collapsed of the D12 (Figs 2A and B). A 3D reconstructive CT scan demonstrated a 360º recurrence with involvement of D11 and L1 (Figs 3A to C). MRI showed kyphosis deformity and significant compression of the spinal cord (Figs 4A and B).

MANAGEMENT

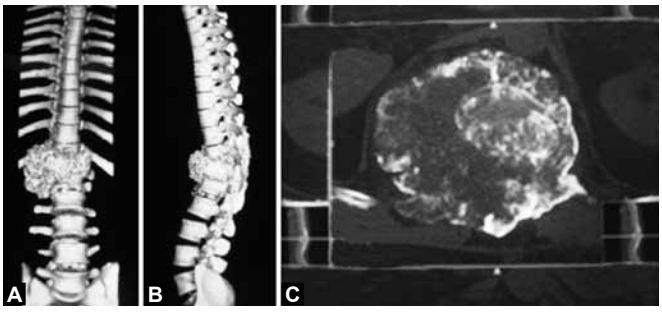
Digital subatraction angiography (DSA) was performed and showed tumor of moderate vascularity. It was decided



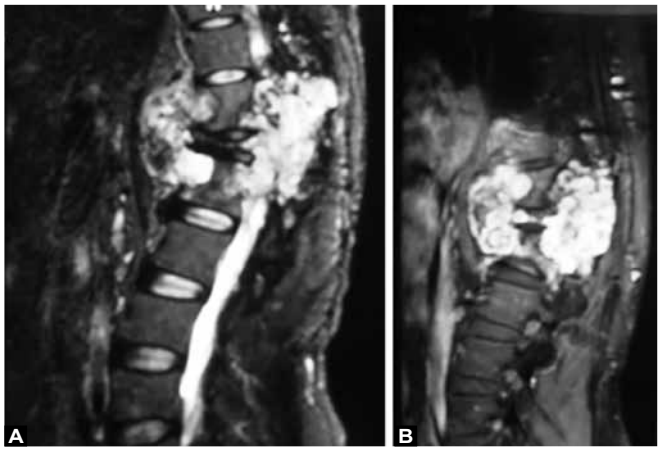
Figs 1A and B: X-rays showed broken and dislocated Harrington rod and recurrence of tumor



Figs 2A and B: 360° recurrence more marked on the right side. D12 showing totally collapsed vertebra with tumor involvement at D11 and L1



Figs 3A to C: CT images and 3D reconstruction showing gross tumor involvement of the dorsal vertebra



Figs 4A and B: MRI DL spine showing kyphosis deformity and significant compression of the spinal cord

to embolize this tumor which revealed significant decrease in vascularity (Figs 5A and B).

After embolization patient underwent tumor removal and stabilization surgery which was done in two phases.

OPERATIVE FINDINGS

We first performed transthoracic removal of the tumor along with removal of D12 vertebral body which was followed by stabilization using interbody vertebral cage and fixation with screw and rods (Figs 6A and B). The main bulk of the tumor was infiltrating the vertebral body and the paravertebral tissue; it was purple and vascular with a soft consistency. Another surgery after 3 months was undertaken to further excise the tumor and stabilize the spine with pedicle screws (Figs 7A and B).

PATHOLOGY REPORT

The pathology specimen consisted of multiple fragments of brownish bony tissue with similar appearance from both surgeries. The tissue was decalcified and processed further according to standard techniques. Histologic sections were stained with hematoxylin and eosin. The lesion consisted of a spindle cell neoplasm admixed with bony trabeculae, pieces of cartilage and fibrous tissue. Tumor cells were mildly pleomorphic and show intracytoplasmic vacuoles containing red blood cells. Bone destruction was seen. The backgound was myxohyaline and few giant cells were present. The immunohistochemical profile showed tumor to be positive for vimentin, negative for CK, and patchily positive for CD31 (Figs 8A to C). The cells were also negative for S-100 and NSE with MIB-I proliferative index less than 1%. The histological features confirmed a HE.

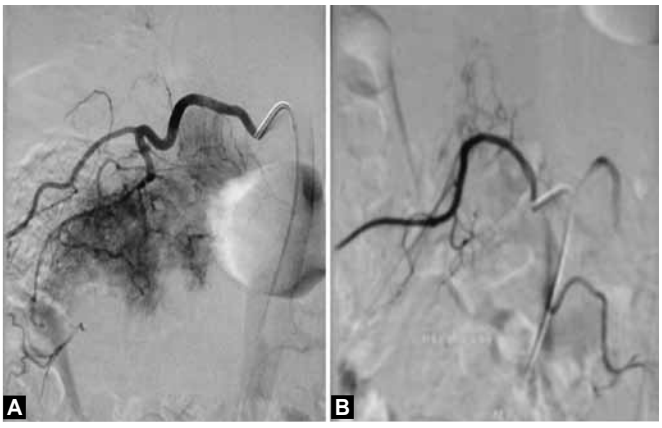
POSTOPERATIVE COURSE

Patient improved symptomatically and was given radiotherapy. He was discharged when his condition was satisfactory. He has been followed up for 6 months.

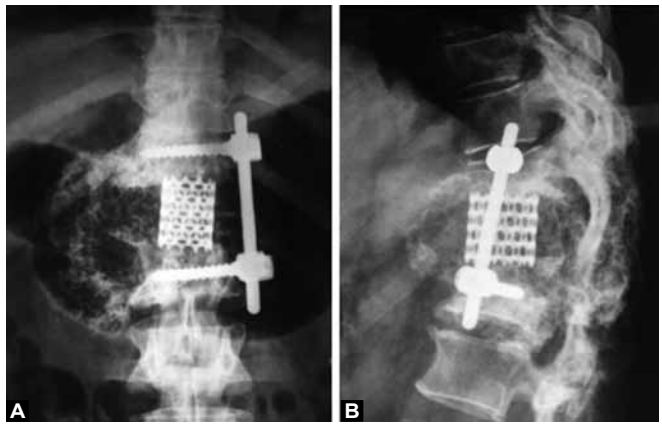
DISCUSSION

It was in the early 1980s, with the first edition of the Enzinger and Weiss textbook, that a refined and thoughtful approach to the use of the term 'Hemangioendothelioma' was introduced. Specifically, Enzinger and Weiss utilized the term to describe the small subset of vascular tumors which did not fit neatly into either the benign or malignant categories at that time.[5]

Hemangioendothelioma is a vascular neoplasm composed of endothelial cells and is considered to be of an intermediary pathology between hemangioma and frankly



Figs 5A and B: (A) Moderately vascular tumor, (B) substantial reduction in vascularity following embolization

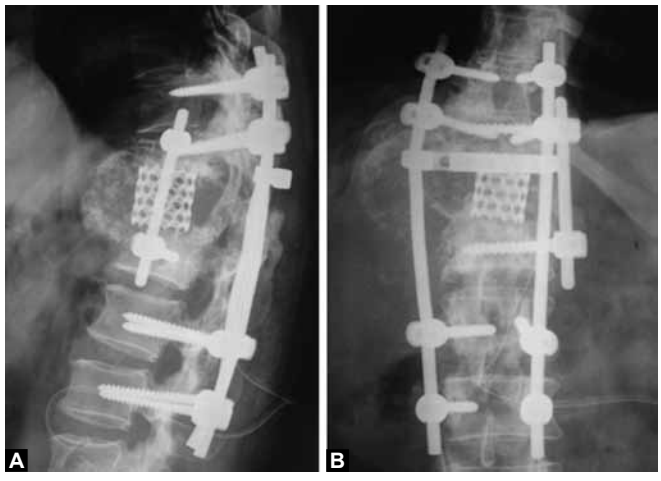


Figs 6A and B: Excision of Left and anterior tumor with reconstruction with interbody cage and stabilization with screws and rod

malignant angiosarcoma. Tumors with significant cellular atypia and mitotic activity are associated with more aggressive clinical behavior. A large percentage of HE may recur at the original site, or in close proximity. These tumors can occur in almost all locations but rarely involve the neuraxis.[2,5]

It can occur in at every age but has its maximum distribution in patients in the fifth decade. Its etiology and risk factors are not known. Clinically, HE is most frequently diagnosed with primary lesions located in the liver, lung, gastrointestinal tract, head and neck, heart, central nervous system or bone.[6]

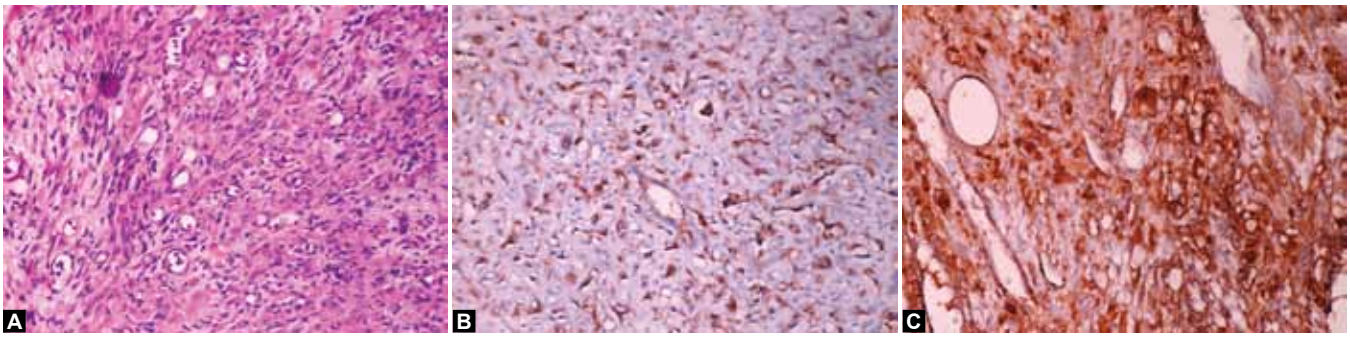
Most commonly affected bones include the skull, vertebrae and long bones. The appearance of this extremely rare tumor can be solitary, multifocal in the same bone, or polyostotic. A purely osteolytic appearance have been found to either expand or remain stationary, can occur and cortical erosion is not uncommon. Therefore, one of the multiple sequelae of HE is a pathologic fracture due to erosion and destabilization of the cortical bone. The coarse trabecular pattern, or honeycomb pattern, is suggestive of a vascular tumor.[6,7] But the radiologic findings are rather nonspecific and therefore, the diagnostic finding has to be done in a multidisciplinary board. Besides radiographic studies such as plain X-ray studies, CT scanning and MR imaging, tumor biopsies are a cornerstone in the diagnosis of HE.[6]



Figs 7A and B: Excision of laterally placed tumor on the right side and posteriorly placed tumor mixed with bone grafts followed by stabilization

The most frequent complaints of patients with spinal HE are pain in the neck and the back accompanied with numbness in the extremities. Some times, they may give rise to severe symptom of cord compression. In her report, Ma et al four out of five of her patients only had backache and sensory disturbance, whereas one presented with paralysis.[8]

During surgical treatment for spinal HE, how to control the intraoperative blood loss is a critical problem that must be faced. Sybert et al reported a case of spinal HEs who did



Figs 8A to C: (A) Cellular areas alternating with cavernous vascular spaces, H & E stain (B) positive immunohistochemical staining for CD 31 in the endothelial cells lining the vascular spaces, some spindle cells, and a few large cells, (C) positive immunohistochemical staining for vimentin

not have embolization, and the surgery had to be terminated due to profuse bleeding.[9] Because, as a vascular tumor, HEs have a substantial potential for intraoperative blood loss. To solve this problem, angiographic evaluation and selective embolization of the involved vertebrae should be applied if possible as was done in our case.[8]

Treatment options and prognosis of spinal HE are sources of controversy, not least because of the small number of patients and the short follow-up periods. We present management guided by the basic oncological principles of maximal safe resection and subsequent radiotherapy for residual disease is the usual course of action. Diffuse disease is probably best treated with chemotherapy.[4,7]

The case presented in this report is unique in many aspects. Firstly, besides being a rare disease involving a rare site (spinal cord), the lesion involved a whole motion segment of the dorsal spine causing instability. Secondly, this tumor had recurred slowly causing minor symptoms associated with the extent of the tumor. And last, we approached this tumor with two step surgery, after preoperative embolization in an attempt to decrease vascularity, this tumor was resected and dorsal spine was stabilized.

CONCLUSION

This case report shows the intricacies and pitfalls in diagnosis and treatment of HE and further adds on to the existing database on the course of disease, therapy and outcome of this rare disease.

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