



Case Report

Epidural extension of dorsal vertebral D1–D7 hemangiomas with congenital cutaneous hemangiomas: Case report and literature review

Suresh S. Pillai¹, P. A. Ramsheela², Rijil Deepak³, Shinto Francis⁴, C. Jayakrishnan⁵, Ani Praveen⁶, Neena Mampally⁶

¹Department of Spine Surgery, Baby Memorial Hospital, Calicut, Kerala, India, ²Orthopedics, Baby Memorial Hospital, Calicut, Kerala, India, ³Anaesthesia, Baby Memorial Hospital, Calicut, Kerala, India, ⁴Hemato-Oncology, Baby Memorial Hospital, Calicut, Kerala, India, ⁵Neurology, Baby Memorial Hospital, Calicut, Kerala, India, ⁶Pathology, Baby Memorial Hospital, Calicut, Kerala, India.

E-mail: *Suresh S. Pillai - sureshorth@gmail.com; P. A. Ramsheela - ramshi0046@gmail.com; Rijil Deepak - rijildeepak69@gmail.com; Shinto Francis - meshinto@gmail.com; C. Jayakrishnan - jkkeecheeri@gmail.com; Ani Praveen - ani.praveenkk@gmail.com; Neena Mampally - neenamampally@gmail.com



*Corresponding author:

Suresh S. Pillai,
Department of Spine Surgery,
Arayidathu Bridge, Indira
Gandhi Road, Calicut - 673 004,
Kerala, India.

sureshorth@gmail.com

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ABSTRACT

Background: There are only rare reports of simultaneous multiple thoracic vertebral, epidural, and congenital cutaneous hemangiomas occurring at the same levels.

Case Description: A 24-year-old male presented with a progressive paraparesis attributed to multiple vertebral hemangiomas (MVH) with epidural extension (i.e. resulting in D1–D3 significant cord compression.), plus congenital cutaneous lesions at the D2–D7 levels. Following preoperative angioembolisation, a D1–D7 laminectomy was performed along with a C7–D8 pedicle screw fixation. Pathologically the bone and cutaneous lesions were spinal cavernous hemangiomas. Postoperatively, the patient regained normal function. As complete excision was not feasible, he subsequently received radiotherapy to prevent tumor recurrence.

Conclusion: MVH with multilevel epidural extension resulting in significant cord compression and congenital cutaneous lesions should undergo attempted tumor excision followed by radiation therapy where complete removal is not feasible.

Keywords: Compressive myelopathy, Congenital cutaneous hemangioma, Epidural hemangioma, Vertebral hemangioma

INTRODUCTION

Multiple vertebral hemangiomas (MVH) typically involve multiple vertebral bodies, and occasionally extend into the epidural space. They represent 2–3% of all spinal tumors, and usually follow an indolent, asymptomatic course. However, a subset may develop symptoms/signs of significant cord compression, attributed to combinations of epidural cord compression, pathological fractures, and/or hemorrhages, warranting surgery. Histologically, MVH are characterized into one of three categories; the capillary, cavernous, arteriovenous, or the venous type. Here, a 24-year-old presented with the new onset of a paraparesis attributed to MVH extending from D1 to D7 with epidural extension/cord compression and accompanying cutaneous lesions at the same levels. The spinal lesion required extensive

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thoracic cord decompression with fusion, followed by radiation therapy to address residual disease.

CASE REPORT

A 24-year-old male presented with 6 months of a progressive paraparesis that exacerbated over the past 2 months. Notably, he had a large cherry red cutaneous lesion on the left side of the thorax, (i.e. left side of the chest from sternum to mid line at the back), also involving the left arm/forearm (i.e. from the nape of neck to just below the scapula involving the lateral/medial arm/forearm) since birth [Figures 1a and b].

Neurological

On examination, he had 3–4/5 motor strength diffusely in the lower extremities and a relative sensory level below D4 level without sphincter involvement.

CT Findings

The thoracic CT showed irregular lytic lesions with coarse thickened trabeculae involving the D1 through D7 vertebral levels, also including the left 1st through 6th ribs. Axial studies showed the typical “polka-dot” pattern of vertebral involvement [Figure 2]. There was also left sided paraspinal and intracanalicular enhancing soft tissue thickening adjacent to the involved vertebrae resulting in significant cord compression.

MR findings

On the MR, tumor was hypointense on both T1W and T2W images and enhanced with contrast (i.e., encasing the cervicodorsal spinal cord from C7 to D8 with transforaminal extension). There was also a subtle increased intracord T2W hyperintensities seen at the D2–D3 vertebral levels [Figures 3–5].

Thoracic CT angiography (CTA) with embolization

The Thoracic CTA showed a highly vascular lesion extending from D1 to D7 that was mostly left-sided (i.e. extra and intracanalicular tumor), the major feeders were embolized before surgery (i.e., with gel foam and coils).

Surgery

Following a D1–D7 laminectomy, a red brown, soft lesion was encountered a leash of blood vessels. Tumor was adherent to the dorsal thecal sac and demonstrated lateral, foraminal, and left-sided paraspinal extension [Figure 6a]. The blood vessels were coagulated and removed [Figure 6b]. Finally, the



Figure 1: Patient with congenital cutaneous hemangioma of thorax, arm and forearm (left) (a) anterior and (b) posterior aspect.



Figure 2: CT of thoracic vertebrae axial view showing “polkadot” appearance.

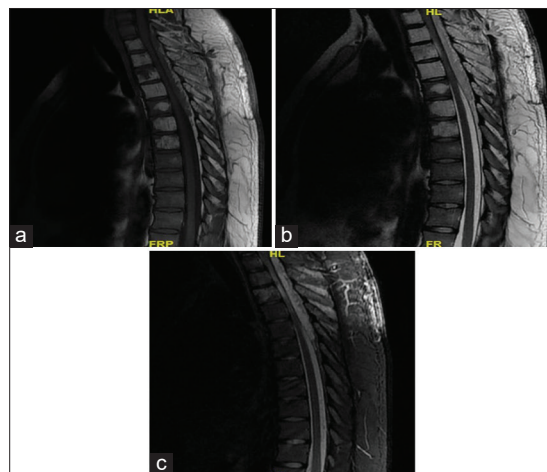


Figure 3: MRI dorsal spine Sagittal view (a) T1W image (b) T2W image (c) STIR image showing altered bone marrow signal intensity involving D1–D7 vertebrae appearing hypointense on both T1W and T2W images with contiguous involvement of posterior element. Patchy residual normal bone marrow signal noted in D1–D7 vertebrae. Extra dural/epidural lesion arising from C7 to D8 vertebrae appearing hypointense on T1W and hyperintense on T2W/STIR causing significant mass effect on cervicodorsal spinal cord. Intracord T2W/STIR hyperintense signal at D2–D3 level.

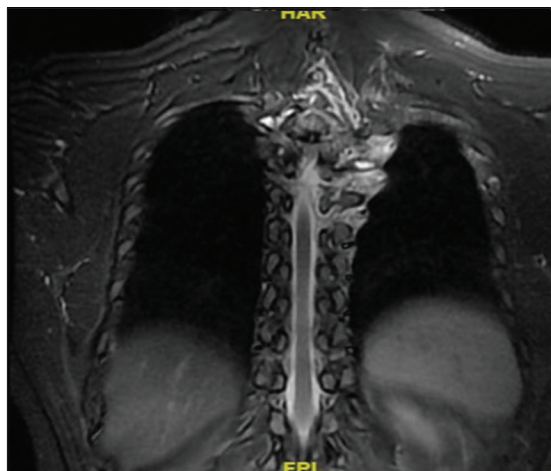


Figure 4: MRI dorsal spine coronal view/STIR image showing contiguous involvement of costovertebral joints at multiple levels noted on left side.

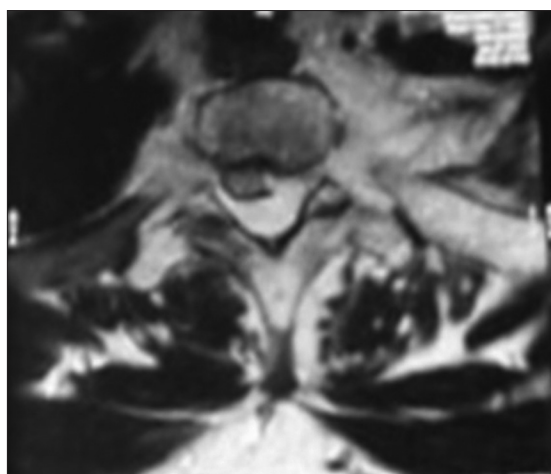


Figure 5: MRI dorsal spine axial view T2W image showing contiguous infiltration into left costovertebral joint and adjacent rib with evident altered bone marrow signal. Infiltrative extradural/epidural lesion causing significant mass effect on spinal cord and shift of cord to right side depicted.

patient underwent a pedicle screw fusion from C7 to D5–D8 [Figure 6c and Figure 7].

Pathology

The frozen sections (i.e., from the epidural, lamina, paraspinal, and skin) were all consistent with a cavernous hemangioma [Figures 8a-c and Table 1].

Postoperative recovery

The patient had an uneventful recovery; 1 week later, at discharge, he had improved and was able to ambulate without weakness. Two months later his muscle strength was 5/5 bilaterally.

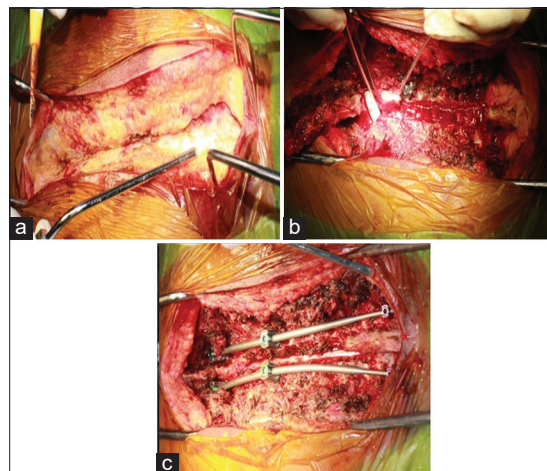


Figure 6: Intraoperative pictures (a) hemangiomatous lesion in para spinal soft tissue above dorsal vertebra (b) post laminectomy exposing epidural lesion encasing posterior aspect of cord (c) decompression of cord and instrumentation using pedicle screws and interconnecting rods.

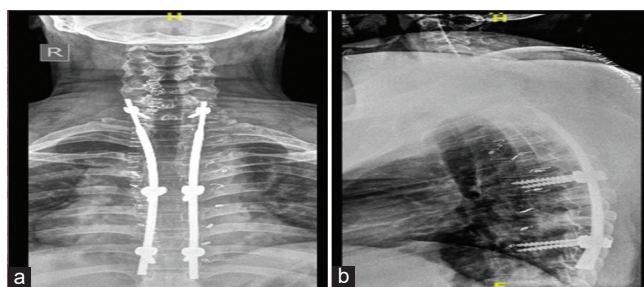


Figure 7: Post op X-ray Dorsal spine (a) AP, (b) Lateral view.

Table 1: Histopathological examination.

Specimens	HPE findings
1. Dorsal vertebrae	Closely packed, dilated and congested vascular spaces lined by endothelium with intervening loose fibrous tissue and fat. Some channels are filled with blood.
2. Epidural lesion	
3. Paraspinal lesion	
4. Vertebral lamina	Bone and normal marrow tissue with fragments of a lesion composed of closely packed dilated vascular spaces lined by endothelium with intervening loose fibrous tissue and fat.
5. Skin	The epidermis was unremarkable, papillary dermis showed dilated vascular spaces filled with blood. The adjacent dermis showed hemorrhage.

Postoperative adjunctive radiation therapy

The patient underwent postoperative radiotherapy to the tumor bed to treat surgically inaccessible tumor, and limit the chance of recurrence.

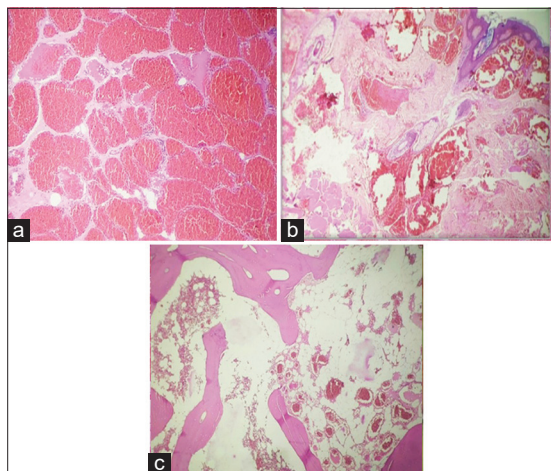


Figure 8: Histopathological examination showing Cavernous hemangioma (a) dilated and congested vascular spaces of varying size (b) dermis show dilated and congested blood vessels (arrow). Dermis show hemorrhage (c) bone with marrow showing closely packed dilated vascular spaces filled with blood.

Table 2: Diagnosis of spinal hemangiomas.

Investigations	Features
MRI	The most reliable investigation for spinal hemangioma Seen as lobulated extradural masses and show iso to hypointense signals on T1WI, hyperintense on T2WI and may show post contrast enhancement ^[8]
CT	Polka-dot' pattern of the vertebral body ^[5]
Plain radiograph	Classically has "jail house striations"
Spinal angiography	Recommended before surgery To know the vascularity of tumor
Histopathology	Lesions at all sites reveal identical features on light and electron microscopy as well as in immunohistochemistry. Cavernous angiomas are composed of sinusoidal vessels, adjacent to one another without intervening mother tissue.

DISCUSSION

According to the WHO classification, vertebral hemangiomas are considered to be a benign vasoformative neoplasms of bone.^[2] The most common site is the thoracic (approximately 60%), followed by cervical (30%) and lumbar (10%) spine.^[8] These lesions are usually confined to the vertebral bodies, but occasionally may extend into the pedicles, arches, and spinous processes.^[6] Notably, the vast majority of vertebral hemangiomas are asymptomatic, are usually diagnosed incidentally on radiological imaging or during postmortem studies.^[5] The further epidural extension of vertebral hemangioma is seen only in 1–2% of cases,

contributing to slowly progressive compressive myelopathies or radiculopathies.^[4,7]

In the case, we presented, the patient has a congenital cutaneous hemangioma at same levels as the vertebral/epidural tumor sites. Whereas, some literature describes case with concomitant occurrence of haemangiomas in other organs like liver and spleen in association with spinal haemangioma.^[3]

MR imaging is the most reliable investigation for spinal hemangioma [Table 2]. Spinal angiography is recommended prior to surgery and preoperative angioembolization of feeding vessels should be done, to avoid the risk of massive bleeding intraoperatively. Laminectomy is an effective procedure to decompress the cord without contributing to instability or deformity.^[1] If, however, vertebral body compression fractures occur, vertebroplasties and/or fusions may be warranted.^[1,5]

CONCLUSION

MVH with epidural extension plus congenital cutaneous lesions rarely occur simultaneously at the same levels. In the presence of compression, gross total excision of tumor is optimal, but subtotal removals may be followed by adjunctive radiation therapy to limit the chance of tumor recurrence.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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