

CASE REPORT

EPIDURAL EPITHELOID HEMANGIOMA OF CERVICAL SPINE PRESENTING WITH COMPRESSIVE MYELOPATHY: A CASE REPORT

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ABSTRACT: Cervical spinal epidural epitheloid hemangioma is a rare entity. All epidural spinal cord tumors should be considered in differential diagnosis. Surgery is indicated when there is compression over the cord or spinal instability. We report a case of 16 year old boy presented with neck pain and spastic quadriparesis. On imaging, the cervical spine showed posteriorly placed extradural spinal tumour extending from C2 to C7 causing compression over the cal sac and cord. Cervical laminectomy was done and tumor excised completely from the extradural compartment. Spasticity subsided after surgery. High index of suspicion and accurate histological diagnosis is a major key for appropriate management.

KEYWORDS: Cervical spine, Epitheloid hemangioma, Epitheloid hemangioendothelioma, Extradural, Spastic quadriparesis

INTRODUCTION: Hemangiomas in epidural space are rare. They constitute around 4% of all epidural tumours.¹ These are not to be considered as true vascular neoplasms but rather malformations of the microcirculation.¹ All the epidural spinal cord tumours should be considered in differential diagnosis. We report a case of cervical epidural epitheloid hemangioma in 16 year old boy who presented with neck pain and spastic quadriparesis.

CASE REPORT: A 16 year old boy presented with neck pain, paresthesias and weakness of all four limbs for a period of 4 months. Clinically he had decreased vibratory and position sense below C3 and spastic quadriparesis. MRI Cervical spine (Fig. 1) showed intra-spinal, extradural, lobulated mass extending from C2 to C5, anterolaterally causing compression over the cord, maximum compression being at C3 level. The lesion is isointense on T1W and isointense with hyperintense Centre on T2W images. The lesion is not enhancing after administration of contrast.

Cervical laminectomy and excision of the tumour was planned. C1 to C6 laminectomy was done. Tumor was completely extra-dural, moderately vascular, greyish pink in colour and firm in consistency. There was no infiltration into the dura or extension into neural foramina. Tumor was excised completely after incising denticulate ligaments on one side. Post op MRI cervical spine (Fig. 2) showed complete excision of the tumor.

Histopathology demonstrated (Fig. 3) multiple vascular spaces lined by plump endothelial cells with moderate eosinophilic cytoplasm with occasional hobnail projections into luminal space. The stroma is oedematous with foci of inflammatory cell infiltrates. Histological features are consistent with epitheloid hemangioma. Immuno-histochemistry was not done.

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Patient improved immediate post-operative period and spasticity subsided but paresthesias persisted for a few months. No radiotherapy was given and at 1 year follow up there was no recurrence of the tumour.

DISCUSSION: In 2000 WHO classification, epithelioid hemangioma is placed in benign tumours (Grade 0) and separated from epithelioid hemangioendothelioma which was considered a borderline or malignant lesion (Grade I).²

The differential diagnosis for spinal epidural hemangiomas before surgical resection include neurofibroma, meningioma, epithelioid hemangio-endothelioma, angioliipoma, disc herniation, synovial cysts, granulomatous infection, pure epidural hematoma, and extramedullary hematopoiesis.^{1,2,3} In our case histopathology confirmed the diagnosis of epithelioid hemangioma.

Based on MR signal intensity characteristics, spinal epidural hemangiomas can be categorized into 4 types: type A for a cyst like mass with T1 hyperintensity, type B for a cyst like mass with T1 isointensity, type C for a solid hyper-vascular mass and type D for an epidural hematoma.¹ Anterior epidural space of the lumbar spine was a common location of types A and B, whereas the posterior epidural space of the thoracic or cervical spine were common in types C and D.¹ Present case fits into type C with cord compression and no vertebral body involvement. As the lesion here is circumscribed, hence complete excision is possible.

Epithelioid hemangiomas have thin walled sinusoidal vascular spaces of varying sizes, which were surrounded by loose connective and adipose tissue, with no necrosis, haemorrhage, or degenerative changes. They have a circumscribed lobular lesion made up of a different sized gland-like or tubular structure lined with plump cuboidal cells and occasional hobnail projections into luminal space. In addition, tall and enlarged endothelial cells imparting so-called 'tombstone-like features' may be seen projecting into the lumen. Transition from solid cords to tubules forming lumens containing blood recapitulating vessel formations may be present. The immunostains of the lining cells show strong immuno-reactivity for cytokeratin (AE1/AE3) and vascular markers CD31, CD 34, and Factor VIII-related antigen².

Epithelioid hemangiomas have to be differentiated from hemangioendothelioma which often show infiltrated borders, cells arranged in clusters or nests or cords, arise from single vessel, without attempted lumen formation, areas of calcification with extensive areas of myxoid and hyalinized stroma.²

Conservative management can be considered in patients without cord compression or instability. Complete excision is the treatment for epidural hemangiomas. Incomplete surgical removal or gross residue prompts post-operative radiotherapy. Pre-operative embolisation is another modality for highly vascular tumors.⁴ In complete surgical removal of a spinal epidural hemangioma because of diffuse bleeding or minimal exposure during surgery might result in the persistence of clinical symptoms or recurrence.¹ Therefore, proper preoperative planning and complete resection in the first operation is essential.

CONCLUSION: Cervical spinal epidural epithelioid hemangioma is very rare. It should clearly be differentiated from hemangioendothelioma as the treatment strategies of the two are completely different. The general principles of achieving cord decompression and tumor control are

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important⁵. Good prognosis can be assured if the tumour is resected completely. Incomplete excision requires adjuvant radiotherapy. Epithelioid hemangioendothelioma requires more aggressive therapy.

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Figure 1: Preoperative MRI of the cervical spine with CVJ showing extradural, lobulated mass extending from C2 to C5, anterolaterally causing compression over the cord, maximum compression being at C3 level. The lesion is isointense on T1W (a) and isointense with hyperintense centre on T2W images (b+c). The lesion is not enhancing on contrast administration (d+e).

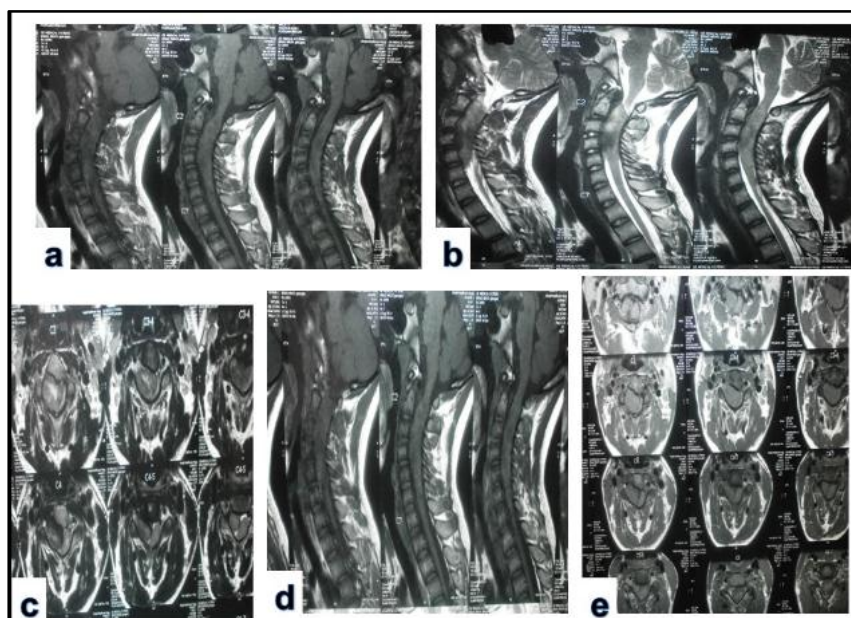


Figure 1a, 1b, 1c, 1d & 1e

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Figure 2: Post-operative MRI of cervical spine T1W (a) and T2W (b) sagittal images showing complete excision of tumor with restoration of anterior and posterior CSF spaces.



Figure 2a & 2b

Figure 3: Histopathology: Low (a+b) and high (c) magnification slides of the specimen demonstrate moderate cellularity, multiple arborising vascular channels (vs) lined by plump endothelial cells (pc), moderate eosinophilic cytoplasm (ec) with occasional hobnail (hn) projections into luminal space. There are no giant or spindle shaped cells. There is no necrosis or haemorrhage. The stroma is oedematous with foci of inflammatory cell infiltrates without any myxoid or hyaline background. Histological features are consistent with epithelioid hemangioma.

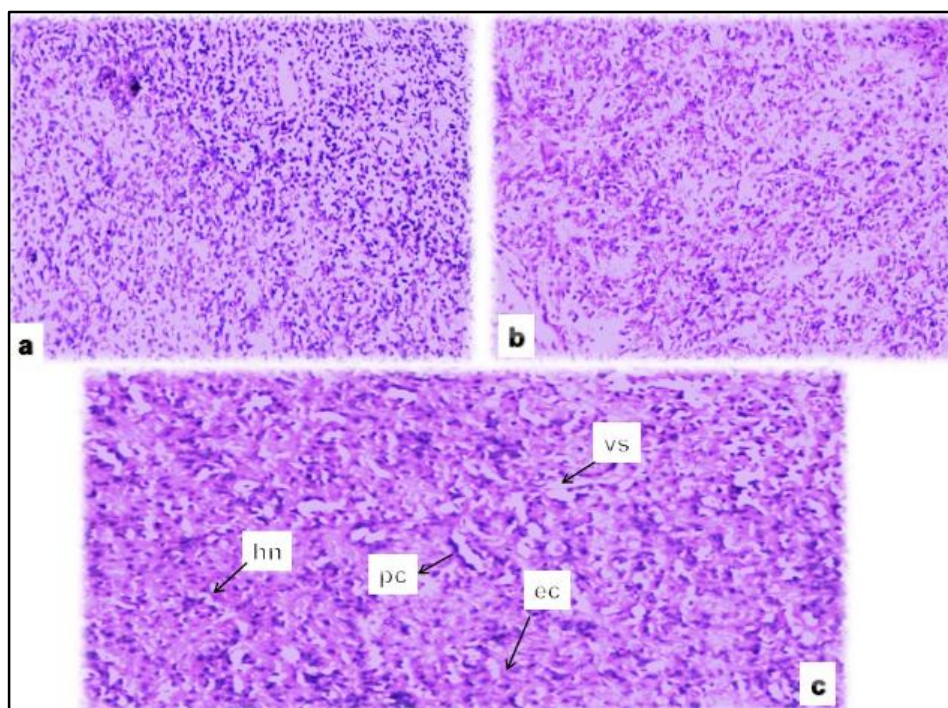


Figure 3a, 3b & 3c

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