Lumbar extradural dumbbell cavernous hemangioma: A rare lesion

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ABSTRACT

A 52-year-old female presented with slowly progressive left lower limb polyradiculopathy. MRI of the lumbar region revealed an extradural dumbbell mass at L3 vertebral level, isointense on T1W and hyperintense on T2W images with homogenous contrast enhancement and extending into paraspinal region through left L3/4 foramen. L2 to L 4 left hemilaminectomy and excision of intraspinal part of tumor was performed. Histopathological examination revealed presence of cavernous hemangioma. This case is reported because of its rarity, unusual dumbbell shape of lesion and difficulty in making a preoperative diagnosis without a coexisting bone lesion.

Key words: Cavernous hemangioma, dumbbell, extradural, schwannoma

Introduction

Cavernous hemangioma is a benign vascular malformation and belongs to a wide and continuous overlapping spectrum of hamartomas.^[1] The cavernous angiomas occur most commonly intraaxially in cerebral hemisphere, brainstem and cerebellum. However, they can occur in any part of the neruraxis including spine.^[2,3] Most cavernous haemangiomas occur in the vertebral body and may extend into the epidural space. Purely extradural cavernous hemangiomas without any vertebral body involvement is extremely rare and accounts for only 4% of all extradural spinal tumors and 12% of haemangiomas occurring extradurally.^[4,5] We herein present a rare case of extradural dumbbell lumbar (L3) cavernous hemangioma with paraspinal extension.

Case Report

A 52-year-old female presented with insidious onset progressive low backache with pain radiating to whole

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of left lower limb associated with paraesthesias for two months. She had difficulty in walking due to pain. There were no symptoms suggestive of bladder or bowel involvement. General examination was unremarkable. Upper limbs were normal with no sensorimotor deficits. Left lower limb power was 4/5 in hip flexion, extension, abduction and adduction, 4/5 in knee flexion and extension, subtle weakness in ankle flexion and extension and great toe extension with hypotonia. Right side lower limb power was normal. The reflexes of left lower limb were sluggish whereas on right side they were brisk (normal). Sensory examination revealed impairment of all the modalities of sensations (pin prick, touch, temperature, vibration- impaired upto anterior superior iliac spine) from L2 to S4 dermatomes on left side. There was no spinal tenderness or deformity. Rest of examination was unremarkable. Magnetic resonance imaging of lumbar spine revealed an irregularly shaped, well defined extradural lesion at L3 vertebral level indenting the body, displacing the thecal sac posterolaterally and extending into left paraspinal region through L3- 4 neural foramen. Lesion was isointense on T1W, hyperintense on T2W images with strong homogenous enhancement on Gadolinium contrast study [Figure 1]. Provisional preoperative diagnosis of schwannoma was considered. Though the clinical findings and radiology were not concordant, the patient was suggested surgical intervention for the presence of enhancing mass lesion noted on the imaging with guarded prognosis.

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The patient underwent left hemilaminectomy from L2 to L4. There was an extradural, brownish red, highly vascular mass located anterolateral to thecal sac at L3 vertebral body with well defined capsule around it. It was extending into left paraspinal region through L3/4 neural foramen. Left L3 nerve root was pushed superiorly by the tumor and it was not arising from any neural tissue. It was highly vascular and bleeding heavily on taking a small biopsy. The capsule was then coagulated to control the haemorrhage. It was dissected clear from the L3 nerve and the dura keeping the dissection over the coagulated capsule. The spinal component of the lesion was excised in toto until the neural foramen, where it was coagulated and was cut sharply leaving behind the extra-foraminal portion. Perfect haemostasis was achieved using bipolar cauterisation, cross checked by Valsalva manoeuvre. Scalloping of L3 vertebral body was noted following tumor excision.

Microscopic examination revealed numerous dilated vascular channels of variable sizes lined by a single layer of flattened epithelial cells and filled with blood elements. Interspersed thick hyalinised blood vessels and mature adipose tissues were noted [Figure 2]. The histological picture was characteristic of cavernous hemangioma. Radiotherapy was administered to the residual paraspinal portion of tumor [Figure 3]. For radiotherapy, the patient was immobilised in a vacuum locking device and CT simulation was done in the treatment position. CT images were fused with the preoperative and post operative MR images. Margin of 1 cm was added to the post contrast T1W MRI to obtain the clinical target volume; planning target volume was obtained by adding a margin of 0.5 cm to the clinical target volume. Dose was prescribed to the planning target volume. Three dimensional conformal radiotherapy was planned on ECLIPSETM planning system (Varian Medical systems, Palo alto, CA) on linear accelerator (Clinac iX-3665) using 6 MV X rays. The dose prescription was 40 Gy, 2 Gy per fraction, 20 fractions given five days a week over five weeks.

Discussion

Cavernous hemangioma is a benign vascular malformation, also known as cavernous malformation, cavernous angioma or cavernoma. The cavernous angiomas occur in two forms: sporadic and familial. Ten per cent of all cavernomas are familial.^[6]

The cavernous hemangiomas occur throughout the neuroaxis including both intracranial and spinal compartments. According to location, its incidence in supratentorial, infratentorial and spinal compartments is 80%, 15% and 5% respectively.^[7]



Figure 1: Shows irregularly shaped well defined extradural lesion (arrows), hypointense on T1W (a), hyperintense on T2W (b) images with homogenous contrast enhancement (c, d, e) and paraspinal transforaminal extension (d, e)



Figure 2: (a, b) The proliferating and anatomizing capillary channels lined by endothelial cells (H and E, ×100)



Figure 3: Shows postoperative changes (arrow) in spinal canal and residual tumor in paraspinal region

Purely extradural spinal cavernous hemangiomas are rare and the extension through intervertebral foramen into extraspinal region (dumbbell) is still rarer. In that case, it can be confused with commonly diagnosed nerve sheath tumors.^[8] The onset of symptoms in these spinal cavernomas may be acute, progressive or remittent, depending on the biological behavior of the tumor. Lumbar extradural hemangiomas behave differently, not only from intramedullary cavernous hemangiomas but also from extradural cavernomas of other spinal locations.^[9] Purely extradural hemangiomas should be included in the differential diagnosis of lumbar extradural soft-tissue lesions.^[10]

In contrast to intracerebral cavernous hemangiomas, the patterns of density on CT scans and signal intensity on MR images are more homogenous in extradural hemangiomas. Another point of differentiation is the absence of low signal rim (hemosiderin) which is usually seen in intraaxial spinal cord cavernomas. As seen in present case, extradural cavernous hemangiomas are usually isointense on T1W and hyperintense on T2W images and show homogenous contrast enhancement because of the presence of sinusoidal channels.^[11]

The lesion should be differentiated from other epidural neoplastic or inflammatory conditions, such as meningioma, neurofibroma, lymphoma, hemorrhagic vascular mass, granuloma and angiolipoma.^[1,11]

Complete and in toto excision of the lesion should be attempted for clinical improvement; however, in case of subtotal excision, adjuvant radiotherapy is advised.^[12] Radiosurgery as primary or adjuvant therapy is increasingly becoming an option as advances in radiosurgical equipment are enabling safe and accurate targeting of lesions.^[13]

Conclusion

Though purely extradural cavernomas are rare in occurrence, they should be considered as possible

differential diagnosis of dumbbell lesions of spine in view of operative nuances (especially massive haemorrhage) and management thereof.

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