Case Report

Intradural extramedullary capillary hemangioma of lower thoracic spinal cord

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Abstract

Capillary hemangiomas are benign tumors and tumor like conditions commonly involving skin and mucus membrane of head and neck region. They are extremely rare in the spinal cord. We report a 35-year-old male presenting with gradual progressive paraparesis over a period of 4 months. Magnetic resonance imaging showed a hypo- to isointense intradural mass at the level of D12 vertebral body on T1-weighted images and homogenous enhancement on gadolinium contrast. Complete surgical resection revealed intradural extramedullary tumor, which on histopathologic examination showed characteristics of capillary hemangioma. At 1.5 years followup patient was asymptomatic.

Key words: Capillary hemangioma, intradural extramedullary tumor, lower thoracic spine

Introduction

Hemangiomas are benign tumors and tumor like conditions.[1] They are uncommon in the spinal cord.[2-8] Their source in the cord may be the meningeal coverings and the vasa nervosum.[6] On the basis of their histopathologic characteristics, they have been differentiated into capillary and cavernous hemangiomas.

Chung *et al*.[9] stated that capillary and cavernous hemangiomas are well-defined, slow-growing vascular tumors, if confined to closed compartments like the skull or spinal cord may behave as space occupying lesions (SOL), and may present with myelopathy/radiculopathy.

Capillary and cavernous hemangiomas at times may be friable, with presentation of sudden neurological compromise due to bleeding. Cavernous angioma has recurrent bleeding tendencies (44% risk in intracranial location).[10] The natural history of spinal cord capillary hemangiomas is not well studied. There is a significant risk of bleeding, hence enbloc excision of the same is suggested.[4,11,12] Common spinal cord tumors like schwannoma and meningioma have similar magnetic resonance imaging (MRI) features to that of capillary hemangioma.[13] We report a rare case of intradural extramedullary spinal cord capillary hemangioma with a brief review of literature [Table 1].

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Case Report

A 35-year-old male presented with mid back pain, progressive weakness in both lower limbs since 4 months, with difficulty in walking since last 2 weeks. On examination of spine, there was no deformity and tenderness. On neurological examination, there was spasticity in both lower limbs, with grade 4 power in both hips and grade 2 in both knee and ankle muscle groups and hypoesthesia below L3. Knee and ankle reflexes were exaggerated, with positive Babinski's sign. MRI screening showed a well-circumscribed intradural mass of 2.78 × 1.28 cm at the level of D12 vertebral body, which was isointense on T1-weighted images [Figures 1a and b] and hyperintense on T2-weighted images [Figures 1c and d] with homogenous enhancement on gadolinium T1-weighted images [Figure 1e]. Surgical resection was performed through posterior midline approach. D11 and D12 laminectomy, with midline duratomy, was performed. Tumor mass was separated from dura, spinal cord, nerve roots and was excised completely. On gross appearance, a reddish brown, multilobular, spongy oval mass of 2.4 × 1.3 × 1.2 cm size was identified [Figure 2]. Histologically, it was aggregates of closely packed, thin-walled capillaries, blood filled and lined

Table 1: Review of literature on intradural extramedullary capillary hemangioma of spinal cord

Author	Age/sex	Site and size	Clinical features	Special investigation	Treatment	Prognosis
Chung *et al*., 2010[18]	47 years/M	T6-7 and 1 × 1.3 × 1.5 cm	BP, B/L radiculopathy, sensory impairment below T7, increased DTR, and ankle clonus		Complete removal of the mass	Gradual sensory improvement
Seyed Miri *et al*., Jan 2009[12]	20 years/M	L3	LBP with cauda equina syndrome		En bloc excision done to reduce blood loss	Weakness improved, urogenital system marginally improved, no recurrence in 1 year
Ji hoon Shin *et al*., May 2000[4]*	66 years/F	T8-9 (EM+IM) and 1.3 × 2 cm	Grade 3 paraparesis	IMH – factor VII related antigen+ve	Incomplete excision due to significant bleeding	Marked neurological improvement, MRI at 6 months – no residual tumor
Abdullah *et al*., Aug 2004[7]	32 years/M	T10 and 1.7 × 1.4 × 1 cm	LBP, left>right lower limb weakness, left-side Babinski's sign, T9 sensory level	Arteriography – lobular hypervascular supply by T11 intercostal artery	In toto excision, nerve root sacrificed	Gradual neurological improvement, no recurrence
Hanakita *et al*., 1991[21]	58 years/M	L1–L2	LBP with cauda equina syndrome	Angiography – feeding vessels and capillary blush		
Nowak *et al*., 2000[14]	63 years/F	T12–L1	LBP with hypoesthesia, activity related lumbosciatalgia of left thigh		Mass excised	Residual paresis left tibialis anterior on, 14-month followup: No recurrence
Hyeon Yu *et al*., 2006[17]	48 years/M	T6-7 and 1 × 1 × 0.7 cm size	Sudden onset back pain, paraparesis and hypoesthesia below T8		Mass was totally removed along with dorsal root	Neurologically improved, no recurrence at 2 months
Ghazi *et al*., 2006[22]	42 years/M	L3-4 and 2 × 1.5 cm	Headache, pulsatile tinnitus, transient visual disturbances, LBP with right lower limb radiculopathy	Tumor cells +ve for CD31 and CD34	Tumor+nerve root excised	Symptoms resolved completely, mild plantar flexion weakness
Andaluz *et al*., 2002[5]	41 years/M	Conus medullaris and 2 × 1 cm	LBP with radiculopathy, absent knee and ankle reflexes		Excision	Neurologically recovered, no recurrence
Alakandy, 2006[19]	60 years/M	T9	LBP, paraparesis, numbness in right limb and below the left knee	CD31 and 34 +ve on IHC	Total excision	Neurologically improved
Fukayama *et al*., 2010[20]	34 years/M	L4 and 1 × 1 × 1 cm	LBP, right ankle reflex absent, sensory loss over left foot		Mass was excised	No recurrence
Bo yoon choi *et al*., April 2001[23]	28 years/M	L1 and 1.5 × 1 × 1 cm	LBP with left gluteal pain, paraparesis		Mass was excised totally	Not commented
	52 years/M	T5-6 and 1.5 × 0.5 × 0.5 cm	Claudication, paraparesis, hypoesthesia in both lower limbs		Mass was excised totally	
	51 years/M	T4-5 and 2 × 1 × 1 cm	Claudication and radiating pain to both lower limbs with hypoesthesia below T5		Mass was excised totally	
Case 1						
Case 2						
Case 3						
H. Bozkus *et al*., 2003[11]	55 years/M	T8 (IM+EM)	B/L leg numbness, decreased sensation below T12, Babinski's positive	Spinal angiography done before revision surgery	Primary surgery – excision of the intradural EM part of the tumor	Revision surgery done for residual tumor
	37 years/F	T5-6, (IM+EM)	B/L leg numbness, flaccid paraparesis, urine retention, indifferent Babinski's, hyporeflexia		Revision surgery – complete excision of the IM part	Complete excision after revision showed favorable neurological improvement
Case 1*					Complete excision of IM+EM mass done	Small intradural residue seen at 2 months
Case 2*						Favorable outcome

BP - Back pain, LBP - Low back pain, IM - Intramedullary, EM - Extramedullary, B/L - Bilateral, DTR - Deep tendon reflex, IHC – Immunohistochemistry. *Extramedullary+intramedullary capillary hemangioma. Note: All cases are confirmed on histopathology as capillary hemangioma. MRI findings of capillary hemangioma in Table 1 are similar (T1W – hypo- to isointense, T2W – iso- to hyperintense, T1 contrast – homogenous strong enhancement)

by flattened endothelium and the vessels were separated by scant connective tissue stroma. There was no cellular atypia [Figure 3]. The patient had uncomplicated postoperative course. Patient was followed over a period of 3 months where he showed neurological improvement (of paraparesis and pain). On 1.5 years of followup, the patient showed complete neurological recovery and he is asymptomatic.

Discussion

15% of the CNS tumors are located in the spinal cord. Of these, 2–7% are of vascular origin.[2] Among these lesions, capillary hemangiomas in the spinal cord are rare and only a few cases have been reported[13] [Table 1]. The most frequent sites of these tumors are around the cauda equina and conus.[3] Features of capillary hemangioma on MR images are: Isointense lesion on T1-weighted images, hyperintense relative to the spinal cord on T2-weighted images, and homogenous, strong enhancement on contrast-enhanced T1-weighted images[6,8,14,15] (which is similar to our case). The differential diagnosis for intradural extramedullary tumors includes hemangioma, meningioma, schwannoma, hemangioblastoma, metastasis and paraganglioma.[13,16] Among these, intradural extramedullary lesions, schwannoma and meningioma are the most common neoplasms commonly occurring at the thoracic level. Schwannomas are frequently seen in middle age with equal sex predilection, while meningioma is seen at the age of 50–70 years with M:F=1:5. Both show similar MRI features, with the exception of the following: Schwannoma frequently shows cystic changes or necrosis, while meningioma shows characteristic dural attachment with dural tail sign on the contrast-enhanced study.[9,17]

Abdullah *et al.*[7] reported a similar case with MR features of enlarged perimedullary veins indicating a vascular tumor and suggested preoperative spinal angiography. Spinal angiography is a useful investigation to differentiate non-vascular tumors from vascular tumors, and thus prevents the risk of intraoperative bleeding.

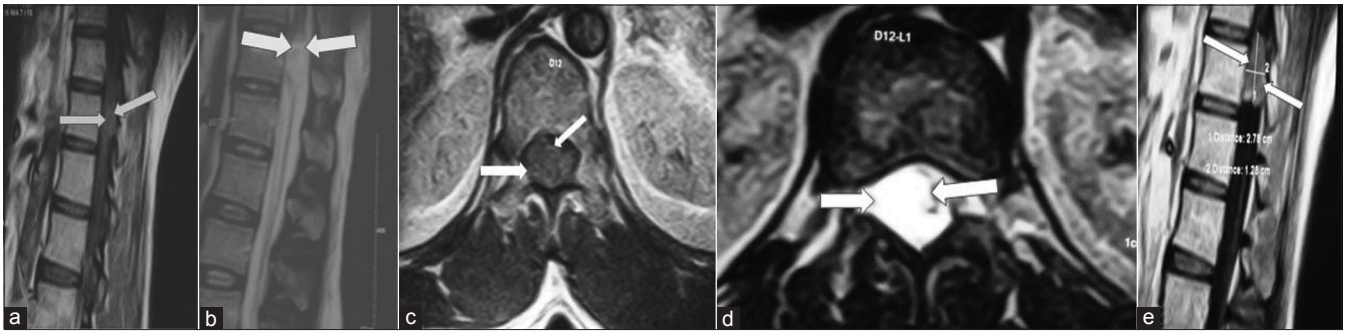

Figure 1: (a) T1-weighted sagittal image demonstrates isointense mass at the level of D12 vertebral body. (b) T2-weighted sagittal image demonstrates hyperintense oval nodular mass (c) T1-weighted axial image shows well-circumscribed intradural isointense mass (d) T2-weighted axial MR image demonstrates a hyperintense solid tumor on the right side of the thecal sac, causing cord compression (e) T1W sagittal MRI with gadolinium contrast showing 2.78 × 1.28 cm, well-defined and homogenously enhancing mass at the level of D12 vertebral body

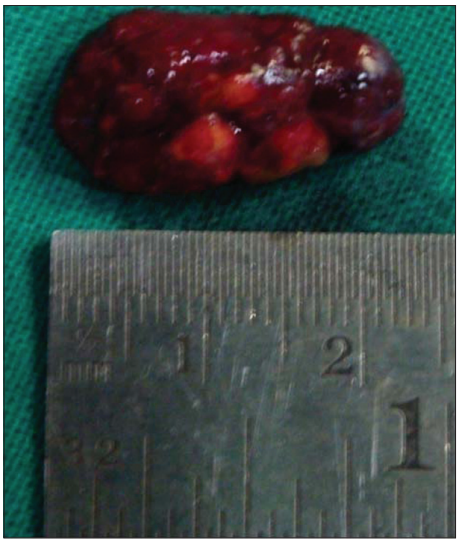

Figure 2: Peroperative photograph showing multilobular vascular tumor of 2.4 × 1.3 × 1.2 cm

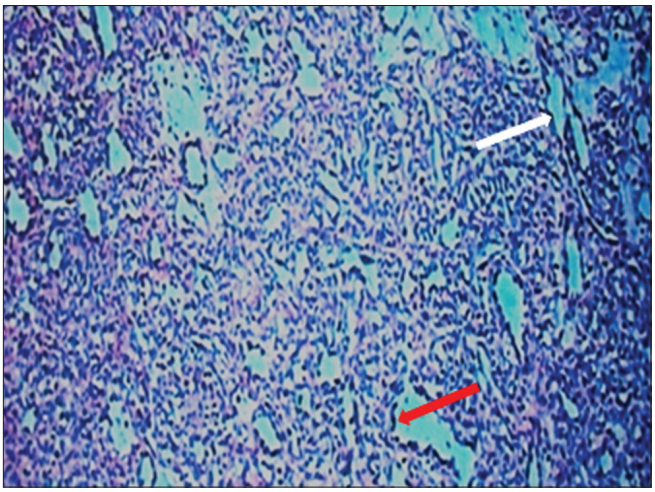

Figure 3: Histological appearance of capillary hemangioma (H and E stain) showing small, thin-walled capillaries lined by flat endothelium (red arrow) and slit-like vascular space (white arrow) separated by fibrous septae. No cellular atypia seen

Capillary and cavernous hemangiomas arise from the same cell type. They can be differentiated on histopathology by vessel size:[13] capillary hemangioma is composed of small, closely packed capillaries, while cavernous hemangioma is composed of blood-filled sinus like spaces. Both show distinct demarcation from the surrounding parenchyma, unlike other vascular lesions such as AV malformations and telangiectasias which interdigitate into the parenchyma forming the differentiating features. Capillary and cavernous hemangiomas show similar MR characteristics.[18] They may differ in their clinical presentation. Cavernous hemangiomas may have variable presentation due to variable size. They may present in four major clinical patterns: Acute episodes of step wise deterioration, slow progression, acute onset with rapid deterioration, and acute onset with gradual decline. They are common in 3rd–6th decade of life, with M:F ratio of 1:2. Capillary hemangiomas are common in 5th–6th decade, with M:F ratio of 1:1. They are commonly located near conus medullaris or attached to nerve roots of cauda equina with a variable onset of presentation[2] (as low back pain, myelopathy, radiculopathy or cauda equine syndrome). Complete surgical resection is the treatment of choice for intradural extramedullary hemangioma,[14,19] with no recurrence.[5,7,12,14,17,20]

Schwannoma and meningioma are common intradural tumors of the spinal cord and it may be difficult to differentiate them clinicoradiologically from capillary hemangioma. Awareness of this rare condition is important to avoid untoward intraoperative event.

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