

Intramedullary capillary hemangioma of the thoracic spine: case report and review of the literature

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Abstract

Capillary hemangiomas are benign vascular neoplasms. When associated with the spine, these growths frequently involve the vertebral body, but rarely have they been reported to occur as intradural lesions, while even more rarely occurring in a true intramedullary location. We report a rare case of an intramedullary capillary hemangioma of the thoracic spinal cord and a review of the literature.

Introduction

Capillary hemangiomas are benign vascular neoplasms typically encountered in the skin. They occur most frequently in childhood as cutaneous or subcutaneous lesions characterized histologically by nodules of capillary-sized vessels lined by flattened endothelium.¹ As the most common primary tumor of the spine, capillary hemangiomas are frequently encountered as interosseous lesions. The occurrence of these tumors in the central nervous system, however, is exceedingly rare and they are even less frequently encountered as intramedullary lesions.² Despite the high prevalence of vertebral body hemangiomas, extraosseous extension causing neurological impingement remains uncommon.

In the present case, we report an intramedullary capillary hemangioma of the thoracic spinal cord, an exceedingly rare presentation with few previously reported cases existing in the literature.^{2,6}

Case Report

The patient is a 47-year old right-handed man with several months of non-specific mid-back pain, lower extremity hyperesthesia, and

a more recent two week history of gait difficulties. His past medical history is significant for a prior left-sided intracerebral hemorrhage with consequent hemiparesis three years ago.

The patient's baseline deficits associated with his prior left hemisphere stroke included dysarthria, right facial weakness, and a dense right hemiparesis. New deficits included 3-4/5 strength of the left lower extremity and a sensory level just above the nipples consistent with approximately a T3 spinal level. Reflexes were 3+ in the bilateral lower extremities. Babinski reflex was present bilaterally.

MRI of the thoracic spine both before and after the administration of intravenous Gadolinium demonstrated what initially appeared to be a homogeneously enhancing intradural, extramedullary lesion displacing the cord anteriorly with significant cord deformation (Figure 1). Our differential diagnosis included: nerve sheath tumor, meningioma, and ependymoma.

The patient was offered an excisional biopsy of the lesion. T2 and T3 laminectomies were performed, and the lesion location was confirmed using intra-operative ultrasound. Intradural exploration revealed a highly vascular, partially encapsulated cherry-red lesion that appeared to arise from the dorsal aspect of the spinal cord. In contrast to MRI findings, the tumor was found to be intramedullary in location. Overlying the tumor was a region of thickened arachnoid, but no demonstrable dural attachment. Several involved nerve roots were coagulated and divided. Further dissection revealed an extremely friable exophytic tumor arising from the region of the dorsal root entry zone of T3. Intra-operative frozen section was consistent with a possible hemangiopericytoma. Post-operative MRI showed no evidence of residual tumor.

On histopathology, the tumor grossly consisted of a partially circumscribed lesion with a brownish-tan appearance. H&E staining demonstrated a hypercellular, vascular neoplasm composed of cytologically bland spindled cells with minimal cytoplasm. These cells were arranged in vague lobules of short irregular spindle cells and dilated vascular spaces. Immunohistochemical findings are explained in Figure 2. Further evaluation demonstrated dilated, compressed vascular channels and focal extramedullary hematopoiesis. In total, the findings were consistent with capillary hemangioma.

Post-operatively the patient had good functional recovery. At three months follow-up the patient had only minor residual lower extremity dysesthesia with return of baseline motor function. The patient has remained stable 36 months post resection.

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Discussion

Capillary hemangiomas involving the spinal cord are extremely rare, and are most frequently extramedullary in location.^{1,3} We are aware of only a handful of previously reported cases of intramedullary spinal cord capillary hemangiomas (Table 1). Notably, all but one previously reported case involved the thoracic spine or the conus.

MRI is the gold standard imaging modality for intramedullary lesions.¹³⁻¹⁶ Many such lesions can be safely removed with proper imaging, surgical planning and technique. In the case of intramedullary capillary hemangiomas, however, on MRI the lesion can be interpreted as extramedullary,^{5,8,10} and the original differential diagnosis often fails to include capillary hemangioma,^{2,8,11} as was the case with our patient. As previously discussed by Roncaroli *et al.* and Nowak *et al.*, the treating surgeon must be cognizant of intramedullary capillary hemangiomas to avoid misdiagnosis and potential excessive treatment.^{1,2} Although extremely rare, capillary hemangiomas of the spinal cord have a benign clinical course. Glial based tumors, such as ependymomas and astrocytomas, account for over 90% of intramedullary tumors.¹⁷ The distinctive gross appearance of capillary hemangiomas intra-operatively often eliminates the more common glial based lesions from the differential diagnosis. The remaining possibilities include lesions ranging from: hemangiopericytoma, hemangioblastoma, cavernous angioma, hemangioendothelioma, arteriovenous malformations, venous angiomas, and capillary telangiectasias. Fortunately, histopathological analysis reliably distinguishes these lesions for diagnosis.

Although related most closely to the category of vascular malformations, capillary hemangiomas that appear within the spinal cord parenchyma may have their origins elsewhere.

Table 1. Cases of intramedullary capillary hemangiomas previously reported in English.^{2,4-12}

Author/year	Patient age/sex	Imaging	Tumor size/location	Symptoms/	Treatment deficits	Outcome	Recurrence
Roncaroli 2000	42 M	not reported	size not reported; T11	1.5-year h/o abdominal pain, leg weakness	surgery	recovered	none reported
	50 M	not reported	size not reported; T11	1-year h/o LBP, proximal leg weakness bilaterally, L-1 sensory level, loss of patellar reflex	surgery + radiotherapy	little improvement 2 years post-op	none reported
	53 M	not reported	size not reported; conus	2 year h/o UMN, LMN signs bilateral lower extremities	surgery	leg weakness 1.5 years post-op	none reported
	64 M	not reported	size not reported; T10	2 year h/o bilateral leg pain, leg weakness	surgery	recovered	none reported
Ianelli 2005	3 months, M	CT: dilated ventricles. MRI: T4-7 intramedullary enhancing lesion	size not reported; T 4-7	Abnormal head circumference; wide, full, tense anterior fontanel with diastatic sutures. Mild irritability, lethargy apparent. No other neurological signs	surgery	neurologically intact 1 year post-op; no focal deficits	none reported
Kelleher 2005	57 M	MRI: T9 & T10 extramedullary enhancing lesion	2.4x1.0x0.5 cm; T9-T10	7-months h/o thoracic pain; 2-week h/o bilateral lower limb weakness, decreased lower limb DTRs	dexamethasone and surgery	spastic gait 3 months post-op	none reported
Mawk 1987	1 month, not stated	CT: discoid mass in R buttock. Myelography: vascular cord lesion with complete myelographic block at L1-2	size not reported; conus	apraxia of legs, hemangiomas malformation involving the skin and soft tissue of the buttock	surgery	rapid recovery	none reported
Andaluz 2002	41 M	MRI: intradural, extramedullary enhancing mass	2x1 cm, conus medullaris	3 month h/o LBP radiating to thighs. Decreased L flexor strength (4+/5), bilateral patellar and Achilles areflexia	surgery	recovered	none reported
Nowak 2000	63 F	MRI: intradural extramedullary enhancing mass	maximum diameter 1.0 cm; T12-L1	3 year h/o intermittent Lasègue's sign; hypesthesia and activity related lumbosciatalgia L thigh	surgery	residual paresis of <i>L tibialis</i> anterior, 14 months post-op	none reported
Abe 2004	65 M	not reported	size not reported; T5	2 month h/o lower limb weakness, paraparesis	surgery	little improvement 7 years post-op	none reported
Roncaroli 2000	74 M	MRI: enhancing nodules on cauda equina, lower thoracic spinal cord, conus medullaris	lower thoracic cord, conus, cauda	9-month h/o bilateral leg weakness	surgery	no changes 1 year post-op	no new hemangiomas
Shin 2000	66 F	MRI: intradural mass at T8-T9; intramedullary and extramedullary components	1.3x2 cm; T8-T9	8-month h/o LBP, weakness of the lower limbs, paraparesis, sensory abnormality	surgery	recovered	no new hemangiomas
Hida 1993	50 M	MRI: Mass from C3-T1	not reported; C3-T1	tetraparesis, upper extremities hyporeflexia, lower extremities flexion spasm, decreased pain/tactile sensation below C3, bladder/bowel dysfunction	surgery	recovered to baseline	not reported

Although several patients had imaging studies suggesting "extramedullary" lesions, they were ultimately found to have intramedullary capillary hemangiomas. h/o = history of; UMN = upper motor neuron; LMN = lower motor neuron; LBP = lower back pain; DTR = deep tendon reflexes; R = right; L = left.

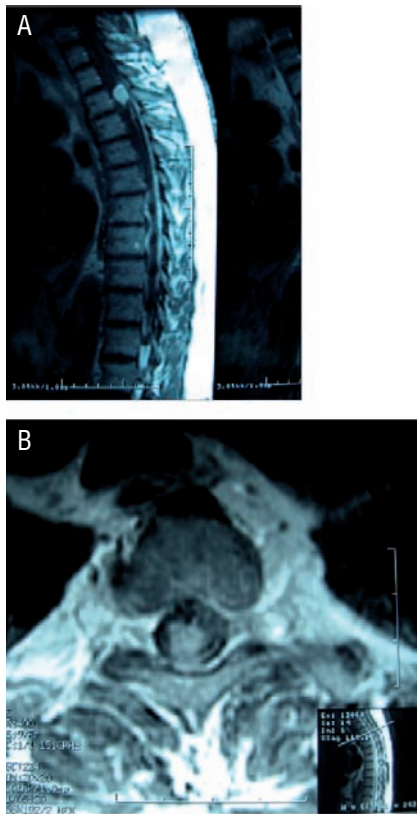


Figure 1. (A) Sagittal T1-weighted MR image with Gadolinium revealing a homogeneously hyperintense mass involving the upper thoracic spine. (B) Axial T1-weighted MR image with Gadolinium demonstrating a centrally located lesion.

Similar to intramedullary lipomas (which account for 1% of intramedullary lesions), capillary hemangiomas of the spinal cord may represent a form of dysembryogenesis. Rather than being a true neoplasm, these lesions more likely arise from inclusion of mesenchymal tissue into the neural tube during primary neurulation.¹⁸⁻²⁰ The available literature clearly demonstrates the benign nature of intramedullary capillary hemangiomas, and gross total resection appears to be curative with good outcomes and no reported recurrences (Table 1). Given the rarity of this lesion, its benign course and straightforward therapy, surgeon familiarity is important to ensure patients receive appropriate diagnosis and treatment.

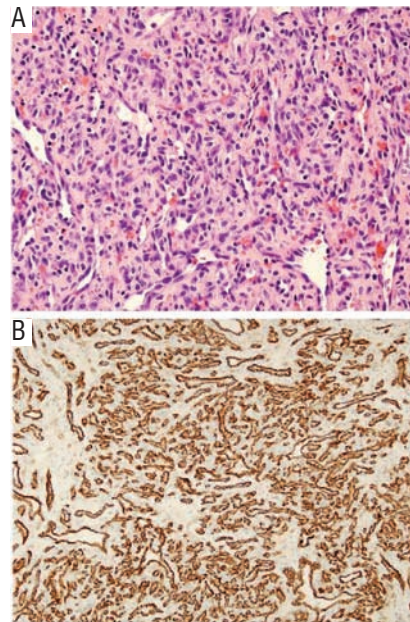


Figure 2. (A) Photomicrograph of H&E stain shows a hypervascular tumor composed of thin and irregular capillary vessels and minimal cytoplasm. Magnification X 200. (B) Immunohistochemical staining revealed the tumor cells to be strongly and diffusely immunoreactive for CD31, CD34, and CD99. Scattered S-100 protein and BCL-2 positive cells were present. Epithelial membrane antigen (EMA) and inhibin stains were negative and MIB-1 labeling was brisk, but with rare mitoses. Magnification X 200.

References

- Nowak DA, Widenka DC. Spinal intradural capillary haemangioma: a review. *Eur Spine J* 2001;10:464-72.
- Roncaroli F, Scheithauer BW, Krauss WE. Capillary hemangioma of the spinal cord. Report of four cases. *J Neurosurg* 2000;93:148-51.
- Holtzman RN, Brisson PM, Pearl RE, Gruber ML. Lobular capillary hemangioma of the cauda equina. Case report. *J Neurosurg* 1999;90:239-41.
- Iannelli A, Lupi G, Castagna M, et al. Intramedullary capillary hemangioma associated with hydrocephalus in an infant. *J Neurosurg* 2005;103:272-6.
- Kelleher T, Aquilina K, Keohane C, O'Sullivan MG. Intramedullary capillary haemangioma. *Br J Neurosurg* 2005;19:345-8.
- Mawk JR, Leibrock LG, McComb RD, Trembath EJ. Metameric capillary hemangioma producing complete myelographic block in an infant. Case report. *J Neurosurg* 1987;67:456-9.
- Abe M, Tabuchi K, Tanaka S, et al. Capillary hemangioma of the central nervous system. *J Neurosurg* 2004;101:73-81.
- Andaluz N, Balko MG, Stanek J, et al. Lobular capillary hemangioma of the spinal cord: case report and review of the literature. *J Neurooncol* 2002;56:261-4.
- Hida K, Tada M, Iwasaki Y, Abe H. Intramedullary disseminated capillary hemangioma with localized spinal cord swelling: case report. *Neurosurgery* 1993;33:1099-101.
- Nowak DA, Gumprecht H, Stolzle A, Lumenta CB. Intraneural growth of a capillary haemangioma of the cauda equina. *Acta Neurochir (Wien)* 2000;142:463-7; discussion 7-8.
- Roncaroli F, Scheithauer BW, Deen HG. Multiple hemangiomas (hemangiomatosis) of the cauda equina and spinal cord. Case report. *J Neurosurg* 2000;92:229-32.
- Shin JH, Lee HK, Jeon SR, Park SH. Spinal intradural capillary hemangioma: MR findings. *AJNR Am J Neuroradiol* 2000;21:954-6.
- Brotchi J, Dewitte O, Levivier M, et al. A survey of 65 tumors within the spinal cord: surgical results and the importance of preoperative magnetic resonance imaging. *Neurosurgery* 1991;29:651-6.
- Rothwell CI, Jaspan T, Worthington BS, Holland IM. Gadolinium-enhanced magnetic resonance imaging of spinal tumours. *Br J Radiol* 1989;62:1067-74.
- Scotti G, Scialfa G, Colombo N, Landoni L. Magnetic resonance diagnosis of intramedullary tumors of the spinal cord. *Neuroradiology* 1987;29:130-5.
- Goy AM, Pinto RS, Raghavendra BN, et al. Intramedullary spinal cord tumors: MR imaging, with emphasis on associated cysts. *Radiology* 1986;161:381-6.
- Miller DJ, McCutcheon IE. Hemangioblastomas and other uncommon intramedullary tumors. *J Neurooncol* 2000;47:253-70.
- Muraszko K, Youkilis A. Intramedullary spinal tumors of disordered embryogenesis. *J Neurooncol* 2000;47:271-81.
- Tihan T, Chi JH, McCormick PC, et al. Pathologic and epidemiologic findings of intramedullary spinal cord tumors. *Neurosurg Clin N Am* 2006;17:7-11.
- Warf BC, Scott RM, Barnes PD, Hendren WH. Tethered spinal cord in patients with anorectal and urogenital malformations. *Pediatr Neurosurg* 1993;19:25-30.