



Case Report

Cervical intramedullary spinal cavernoma in setting of unresolved myelopathy: A case report

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ABSTRACT

Background: Spinal cavernous malformations are rare, accounting for approximately 5–12% of all spinal cord vascular lesions. Fortunately, improvements in imaging technologies have made it easier to establish the diagnosis of intramedullary spinal cavernomas (ISCs).

Case Description: Here, we report the case of a 63-year-old male with an >11-year history of left-sided radiculopathy, ataxia, and quadriparesis. Initially, radiographic findings were interpreted as consistent with spondylotic myelopathy with cord signal changes from the C3-C7 levels. The patient underwent a C3-C7 laminectomy/foraminotomy with instrumentation. It was only after several symptomatic recurrences and repeated magnetic resonance images (MRI) that the diagnosis of a ventrally-located intramedullary lesion, concerning for a cavernoma, at the level C6 was established.

Conclusion: Early and repeated enhanced MR studies may be required to correctly establish the diagnosis and determine the optimal surgical management of ISCs.

Keywords: Cervical spine, Complex surgery, Diagnosis, Excision, Intramedullary spinal cavernoma, Laminectomy, Magnetic resonance, Myelotomy

INTRODUCTION

Spinal cord cavernous malformations (cavernomas) are rare vascular malformations (e.g., 5–12% of all such lesions).^[3,7] Although they may remain stable and asymptomatic for decades, intramedullary spinal cavernomas (ISCs) hyperpermeable borders result in a 2.1–2.5% risk for hemorrhage per year, making surgical excision the optimal approach for accessible lesions.^[1,2,5,6]

Here, we present a 63-year-old male with an ISC documented on magnetic resonance (MR) at the C6 level. Despite an original C3-C6 laminectomy/decompression with C3-C7 fusion, the patient rebelled requiring repeated intervention.

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CASE DESCRIPTION

History	A 63-year-old male presented with an 11-year history of progressive radiculopathy and ataxia involving the left upper and lower extremities, newly accompanied over the past year by a mild quadriparesis and left leg paresthesia. In 2008, MR showed cervical stenosis with C3-C7 spondylotic myelopathy, for which a C3-C7 laminectomy/instrumented fusion was performed [Figure 1]. However, in 2014, he presented with 12 months of progressive pain left-sided L5 radiculopathy. Despite a left-sided laminectomy and foraminotomy, his symptoms failed to resolve
Physical examination	In 2019 (11 years after his original 2008 fusion), he presented with recurrent cervical radiculopathy, a mild quadriparesis, ataxia, and paresthesia in the left lower extremity (the latter over the past year). The follow-up MR documented a small intramedullary nodule ventrally located at the C5-C6 level accompanied by perinodular edema extending throughout the cord; the lesion remained stable on successive MR studies over the ensuing several months [Figure 2]. Due to progressive myelopathy, gait instability, and vertigo, the patient underwent a posterior approach requiring removal of the cervical C5-C6 fusion for resection of the intramedullary lesion
Operative findings	The procedure required the utilization of ultrasound, fluoroscopy, and microsurgical technique to perform a C5-C6 midline myelotomy [Figure 3]. During resection, both the left and right somatosensory evoked potentials (SSEPs) were lost. At closure, the right SSEP normalized, but the left remained diminished/absent

Pathology	Intraoperatively, the frozen section staining demonstrated a collection of thin-walled vessels with focal fibrosis and thrombosis without intervening neural parenchyma, diagnostic for a cavernous angioma [Figure 4]
Postoperative course	Postoperatively, the patient exhibited a mild paresis in the left L4, L5, and S1 dermatomes (chronic preoperatively), and a new left-sided upper and lower extremity hemisensory deficit without hyperreflexia. The patient was able to ambulate with assistance (i.e., due to proprioception deficits) and was discharged home

DISCUSSION

Here, we described a 63-year-old male with >11-year history of radiculopathy, progressive quadriparesis, and ataxia, with recurrent cervical myelopathy despite two operations. After recurrence, the MR indicated an intramedullary lesion at the C6 level. Following secondary surgical resection with pathological confirmation, the patient sustained significant postoperative sensory deficits but no further hemorrhagic events.^[4,8]

Review of relevant literature

We conducted a review of the literature on intramedullary cavernomas, with a focus on spinal lesions. The preceding case followed a similar course as other chronically untreated ISCs, although, in the other cases, early resection was more common.^[1,2] The cervical location combined with a slowly progressive course of neurologic deterioration (e.g., motor and sensory symptoms) were typically described in other series,^[2,3,5,8] as was the typical intramedullary location of these lesions responsible for symptoms (e.g., paresthesias) and signs (differing degrees of paralysis).^[3,7] A summary of the pertinent aspects of our literature review is available in Table 1.

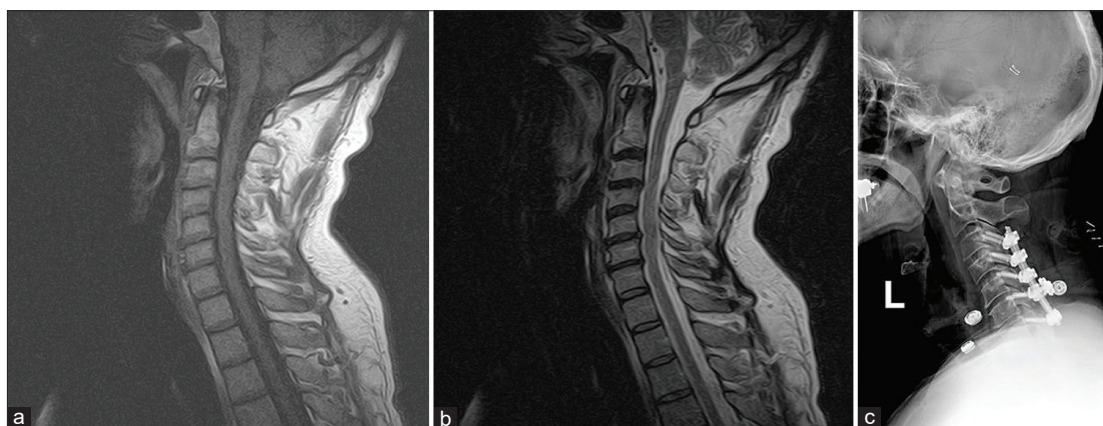


Figure 1: Perioperative imaging of the cervical spine fusion performed in 2008. (a) Preoperative T1-weighted magnetic resonance imaging (MRI) of the cervical spine. (b) Preoperative T2-weighted MRI of the cervical spine. No significant foci are noted, but canal stenosis is visible from C3-C6. (c) Postoperative X-ray demonstrated instrumented fusion from C3-C7. “L” denotes that the image taken from the left side of the patient’s body.

Table 1: Pertinent aspects of each of manuscript in our literature review.

Author	Type	# of cases	Cavernoma type	Relevant findings
Akers	Systematic review	1966	Cerebral (CCM)	Brain MRI (gradient echo/weighted sequences) should be used to diagnose/track CCMs; angiography is not recommended unless to exclude AVMs; surgical resection is not recommended for asymptomatic CCM, especially if in eloquent, deep, or brainstem, nor w/ multiple asymptomatic CCMs; resection may be warranted in singular asymptomatic CCM if accessible in noneloquent area, to prevent hemorrhage, psychological burden, or in anticoagulated patients; early resection indicated for CCMs causing epilepsy, especially if medically refractory; symptomatic accessible CCMs may be resected, if M&M of surgery equivalent to waiting ~2 years; with 2+ bleeds in brainstem CCM, resection and postoperative M&M risks may be balanced against nonoperative progression; CCM in eloquent area with unacceptable surgical risk may be ablated with radiosurgery; asymptomatic, familial, or those in accessible areas should not be radiosurgically ablated given risk of <i>de novo</i> CCM
Badhiwala	Meta-analysis	632	Intramedullary spinal (ISC)	Spinal levels: cervical (38%), cervicothoracic (2.4%), thoracic (55.2%), thoracolumbar (0.6%), lumbar (2.1%), and conus medullaris (1.7%); average size: 9.2mm; CCMs cooccurred in 16.5%, family history in 11.9%; step-wise progression (45.4%) and slowly progressive (54.6%); presenting Sx: motor (60.5%), sensory (57.8%), pain (33.8%), bladder and/or bowel (23.6%), respiratory distress (0.5%), or absent (asymptomatic; 0.9%); annual hemorrhage rate 2.1% (95% CI 1.3%–3.3%); management was surgical (89.9%) or conservative (10.1%); outcomes were better for resection versus conservative management (OR 2.79, 95% CI 1.46–5.33, $P=0.002$); better outcomes in hemilaminectomy approach (OR 3.20, 95% CI 1.16–8.86, $P=0.03$), gross-total resection (OR 3.61, 95% CI 1.24–10.52, $P=0.02$), motor Sx (OR 1.76, 95% CI 1.08–2.86, $P=0.02$); versus sensory (OR 0.58, 95% CI 0.35–0.98, $P=0.04$); superficial and deep-seated ISCs equivalent (OR 1.36, 95% CI 0.71–2.60, $P=0.36$)
Cosgrove	Case series	5	Spinal	Level: cervicothoracic (4/5) or thoracolumbar (1/5); presenting Sx: acute LE sensory (4/5) or hand weakness (1/5). Through myelography, the lesion was intramedullary in 2/5; resection subtotal (2/5) or complete (3/5)
Grasso	Case rep	1	Spinal	Cavernous angiomas are 5–12% of all vascular spinal malformations
Gross	Review	352	Intramedullary spinal (ISC)	Resection rate: 91%; transient morbidity: 36%; long-term outcomes: improved (61%), unchanged (27%), worse (12%); cranial accompanying lesions in 27%; chronic progressive deficits were the presenting Sx in 50%. Roughly equal sexes
Leep	Review		Spinal cavernoma	Trauma is the most common cause of spinal hemorrhage; atraumatic is rare and usually caused by vascular malformations (intradural AVMs or ISCs). The neurologic decline can occur after initial hemorrhage due to secondary tissue response
Steiger	Case series	15	Cerebral angioma	Female (12/15), male (3/15), presenting Sx: headache (8/15), hemiparesis (1/15), seizures (6/15); substantial hematoma occurred in 8/15 patients. Ultimate pathologic Dx: AVM (11/15), cavernous angioma (3/15), venous malformation (1/15).
Sun	Case series	10	Spinal cavernoma	Location: cervical (6/10), thoracic (4/10); 100% resected; negative outcomes: hemiparesis (1/15) resolved on f/u



Figure 2: Preoperative imaging of the intramedullary spinal cavernoma resection performed in 2019. (a) Preoperative T1-weighted magnetic resonance imaging (MRI) of the cervical spine. (b) Preoperative T2-weighted MRI of the cervical spine. A small but notable signal is visible ventrally at the level of C6 consistent with a focal nodule. A significant signal consistent with peri-nodular edema is also visible from C3-C7.

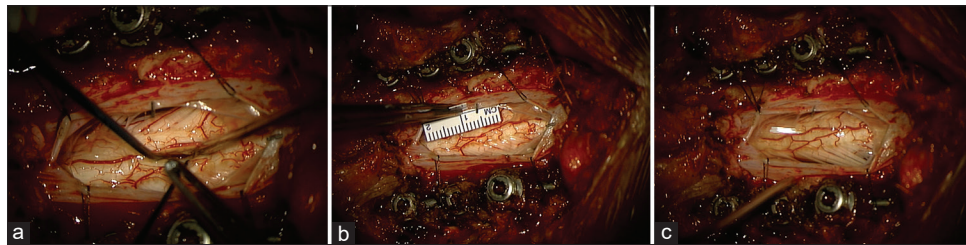


Figure 3: Intraoperative imaging of myelotomy and dural opening. (a) Color photo of operative field postmyelotomy. The screws depicted in the top and bottom of the photo correspond to instrumentation at C5, C6, and C7 (from cephalad to caudal) (b) Color photo as in a, with added ruler for the measurement. (c) Color photo of operative field postdural opening.

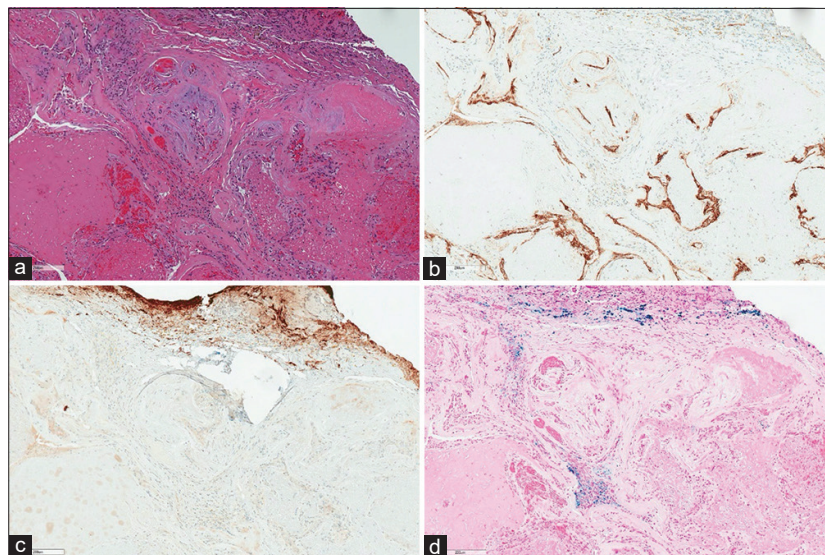


Figure 4: Histological examination of cavernous angioma. (a) Hematoxylin and eosin-stained section demonstrating a collection of thin-walled hyalinized vessels surrounded by a thin rim of central nervous system tissue. (b) Immunohistochemical stain for CD34 highlights the vascular endothelium. (c) Immunohistochemical stain for glial fibrillary acidic protein highlights the surrounding reactive astrocytosis in the rim of central nervous system tissue. Of note, there is no appreciable intervening central nervous system tissue between the vessels. (d) Iron stain highlights hemosiderin deposition (blue) indicative of remote hemorrhage. (Magnification $\times 100$; all scale bars equal $200\ \mu\text{m}$).

CONCLUSION

As demonstrated in the case presented and following a literature review of ISCs, one must maintain a high index of suspicion for cavernoma, especially when patients exhibit a progressive/stuttering course of neurological deterioration along with repeated intramedullary spinal hemorrhages.

Declaration of patient consent

Patient's consent not obtained as patient's identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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