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Letter to the Editor: New Observation

Intradural-extramedullary Spinal Cavernoma

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Cavernomas are dilated capillary vessels without intervening neural tissue.[1] Most cavernomas are intracranial; of the 5% located in the spine, most are extradural or intramedullary.[1] Intradural-extramedullary cavernomas are quite rare. Here, we present a unique case of an intradural-extramedullary cavernoma exclusively associated with a thoracic dorsal nerve root and review the clinical presentation, imaging features, surgical management, and postoperative outcomes of these rare lesions.

Case Report

A 45-year-old male patient presented with a gradual six-month history of decreased sensitivity to heat and a burning sensation to cold temperatures on his left leg, numbness, and paresthesia affecting the medial aspects of his lower extremities bilaterally, and leg weakness worse on the right side, consistent overall with a Brown-Sequard hemi-cord syndrome. He denied bowel, bladder, or sexual dysfunction. Clinical examination revealed mild thoracic myelopathy: his patellar and Achilles reflexes were increased bilaterally (3+) with two beats of ankle clonus and upgoing plantar response on the right. The remainder of the exam was unremarkable.

MRI demonstrated a 16 mm intradural-extramedullary lesion at T3-4 level, displacing and compressing the spinal cord (Figure 1A, 1B). The lesion appeared hyperintense on T1 and centrally hypointense (with surrounding high signal) on T2-weighted images, consistent with hemosiderin deposition. There was faint contrast enhancement. There was no identifiable abnormal vessels or abnormal intradural vascular enhancement seen on contrast-enhanced MR angiography.

The patient underwent a T3-4 laminectomy and complete excision of the lesion. Significant epidural venous engorgement was noted intraoperatively. Following durotomy, a dark, mulberry-like lesion separate from the spinal cord and associated with a dorsal sensory nerve root was encountered (Figure 1E). With sacrifice of this root, a complete en-bloc excision was achieved (Figure 1F).

Permanent pathology was consistent with cavernoma, demonstrating a spinal nerve root attached to the lesion (Figure 1G, 1H). The patients' symptoms completely resolved following surgery and he remained asymptomatic at 3-month follow-up (post-operative Figures 1C, 1D).

Literature Review

Intradural-extramedullary cavernomas are quite rare, including the current report, 71 cases have been reported to-date (Table 1). Fifty of these were associated with nerve roots, while in 21 cases the lesion was associated with other structures (e.g. dura), or its origin was not specified. Thirty-eight (54%) cases were located in the lumbar spine, 18 (25%) thoracic, 4 (6%) thoracolumbar, 8 (11%) cervical, and 3 (4%) with no level reported. Most patients presented between the ages of 40–59 (41%), followed by 60–79 (34%), 20–39 (20%), and rarely ≦19 (3%).

Cases reviewed from the literature demonstrated various patterns of homogenous and heterogenous enhancement on imaging, which may be attributed to mixed subacute and chronic hemorrhage.[2] Given that the classic hemosiderin ring of hypointensity was present only in some cases,[3] these lesions may be difficult to identify preoperatively without a high index of suspicion.

Ten patients (14%) presented with confirmed subarachnoid hemorrhage (SAH) and one with intramedullary hemorrhage, as might be expected for intradural-extramedullary lesions. Although an annual hemorrhage rate has not yet been reported given limited available data, a comparable reported annual hemorrhage rate for intramedullary spinal cavernomas is 2.1%.[4] All SAH cases reviewed demonstrated acute onset, whereas the unique case of intramedullary hemorrhage (Kivelet et al., 2008) displayed chronic progression (myelopathy, 5 months). However, 16 patients (23%) presented with acute onset of symptoms and 8 (11%) with a mixed acute-on-chronic presentation, indicating the true rate of hemorrhage at presentation may be higher. Comparably, in a 2014 meta-analysis, 45.4% of intramedullary cavernomas presented acutely or in a stepwise fashion, with 54.6% presenting with chronic progression.[4] Roughly half (49%) of cases reviewed here presented solely with longstanding or progressive symptoms (weeks to years). Thirty-seven percent presented primarily with

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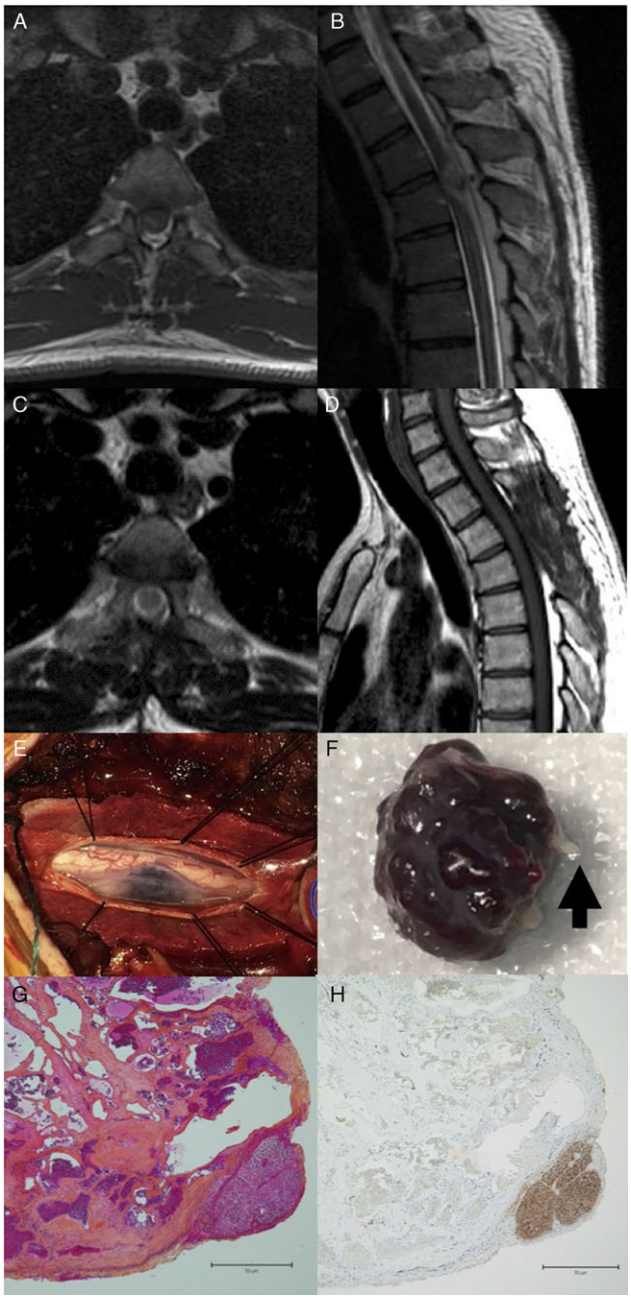

Figure 1: Thoracic spine MRI (A, B) displaying an intradural-extramedullary lesion at T3-T4. Axial T1-weighted image with gadolinium demonstrates a hyperintense lesion (A); and sagittal T2-weighted image demonstrates central area of hypointensity consistent with hemosiderin deposition (B). Post-operative imaging (C, D) on axial T2-weighted image demonstrates resolution of the previously seen T2 hyperintense signal (C); and sagittal T1-weighted image of the thoracic spine demonstrating no abnormal enhancement or suggestion of residual tumor (D). Intraoperative images (E, F) following T3-4 laminectomy and durotomy: the extramedullary lesion can be seen displacing the spinal cord to the left (E); following en-bloc excision the mulberry-like appearance of the cavernoma with an attached nerve root (arrow) (F). Pathology slides (G, H) with H&E stain (G) demonstrating multiple vascular channels with hyalinized walls of varying thickness, size and shape and no intervening neural or connective tissue. Intraluminal blood clots with both recent and remote evidence of recanalization is observed. A spinal nerve rootlet is seen attached to the lesion, highlighted by S100 protein stain (H).

radiculopathy, 21% with myelopathy, and 11% with pain only. The tendency of intradural-extramedullary cavernomas to present with radiculopathy may be a distinguishing feature of their presentation, reinforced by the fact that the majority occurred at the lumbar level.[5]

Complete lesion resection was achieved in 65 cases (92%), with the remaining having subtotal resection, nonoperative management, or no reported intervention. Of cases with nerve root involvement, 44% documented nerve root sacrifice. Good postoperative recovery (partial or complete resolution of symptoms without major complication) was documented in 80% of all cases (with 31% achieving complete resolution of preoperative symptoms), and among 90% of cases with nerve root involvement (of which 36% completely recovered).[6–8]

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Table 1: Published cases of spinal intradural extramedullary cavernous malformation associated with a nerve root (No. 1-50) and with no documented nerve root involvement (No. 51-71)

No.	Authors, Year	Age	Sex	Location/Origin	Presentation (duration)	Intervention	Outcome
Cases with confirmed nerve root involvement							
1	Hirsch et al., 1965 (cited by 68)	20	M	L2-3, root	SAH (Acute onset) Sensorimotor deficit, sphincter dysfunction (1 year)	Total + RR	No improvement
2	Pansini & Lo Re, 1966 (cited by 68)	46	M	L2, root	Radiculopathy (duration NR)	Total	No improvement
3	Heimberger et al., 1982	24	M	T2-3, root	SAH; headache, diplopia, nuchal rigidity (Acute onset, 4 episodes)	Total + RR	Improvement
4	Ueda et al., 1987 (cited by 68)	28	M	L1-2, root	SAH; back pain (Acute onset, 2 episodes)	Total	Complete recovery
5	Pagni et al., 1990 (cited by 68)	46	M	T12-L1, root	Radiculopathy (4 years)	Total + RR	Improvement
6	Ramos et al., 1990 (cited by 68)	67	F	L3, root	Hydrocephalus, cognitive dysfunction, sphincter dysfunction, and gait disturbance (3–4 months)	Total	Improvement
7	Mastronardi et al., 1991	49	F	T4, root	Myelopathy (6 months)	Total	Complete recovery
8	Mori et al., 1991	65	M	T1, root	SAH; back pain, headache, left Horner syndrome (Acute onset)	Total	Improvement
9	Bruni et al., 1994 (cited by 68)	28	M	L2, root	SAH; headache (Acute onset)	Total	Improvement
10	Cervoni et al., 1995 (cited by 68)	26	F	L1-2, root	SAH, backpain, headache (Acute onset)	Total	Complete recovery
11		32	M	L5, cauda root	Radiculopathy (3 years)	Total	Improvement
12	Makino et al., 1995 (cited by 68)	67	M	L2, root	SAH; gait disturbance, headache (Acute onset) Intermittent headaches (3 months)	Total + RR	Improvement
13	Moreno et al., 1995 (cited by 34)	63	M	T12-L1, root	Pain, motor deficit, sphincter dysfunction (Acute onset, then progressive for 7 years)	Total + RR	No improvement
14	Harrison et al., 1995	37	M	C1-C5, root	Myelopathy (Brown-Sequard syndrome) (duration NR)	Total	Improvement
15	Rao et al., 1997	60	M	L1-3, (L3 root)	Myelopathy (3 weeks)	Total	Complete recovery
16		35	F	T12, root/conus	Radiculopathy (3 years) Paraparesis (Acute onset)	Subtotal	No improvement
17	Duke et al., 1998 (cited by 68)	49	F	L4, root	Back pain (Acute onset)	Total	Improvement
18	Kim et al., 2001 (cited by 35)	65	M	L4, root	NR	NR	NR
19	Park et al., 2003	33	M	L2-3, root	Radiculopathy (1 year) Suspected SAH (unable to confirm); headache (Acute onset)	Total	Improvement
20	Nozaki et al., 2003 (cited by 68)	51	M	C5-6, anterior root	Shoulder pain (6 months) Myelopathy (2 weeks)	Total	Complete recovery
21	Flavigna et al., 2004 (cited by 68)	44	F	L4, root	Radiculopathy (4 months)	Total + RR	Improvement
22	Chung et al., 2005 (cited by 35)	52	M	L2, root	Radiculopathy (2 years)	Total + RR	Improvement
23	Rachinger et al., 2006	56	M	C7, root	Back and left shoulder pain (Acute onset, 2 weeks)	Total	Complete recovery
24	Cecchi et al., 2007 (cited by 68)	75	F	L3-4, root	Bilateral leg paresthesias; no radicular pattern (2 months)	Total + RR	Improvement
25	Caroli et al., 2007 (cited by 68)	71	M	L3, cauda root	Radiculopathy (duration NR)	Total	Improvement
26	Er et al., 2007	67	M	T12-L2, root	Radiculopathy (4 months)	Total	Improvement
27	Miyake et al., 2007 (cited by 35)	18	M	L1, root	Radiculopathy, back pain, gait disturbance (Acute onset, 5 days)	Total + RR	Improvement
28	Kivelev et al., 2008	44	M	C5-6, root	Intramedullary hemorrhage; Myelopathy (5 months)	Total	Improvement

(*Continued*)

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Table 1: (*Continued*)

No.	Authors, Year	Age	Sex	Location/Origin	Presentation (duration)	Intervention	Outcome
29	Yi et al., 2008 (cited by 42)	67	M	L2-3, root & filum	Radiculopathy (1 month)	Total	Improvement
30	Chun et al., 2010 (cited by 35)	74	F	Below L4, root	Radiculopathy (3–4 years)	Total + RR	Improvement
31	Khalatbari et al., 2011 (cited by 35)	58	M	L3-4, root	Radiculopathy (Acute onset)	Total + RR	Improvement
32		45	M	L1-2, root	Radiculopathy (Acute onset)	Total	Improvement
33	Jin et al., 2011	55	M	T12-L1, root	Sustained headaches, dizziness (3 months)	Total + RR	Complete recovery
34	Nie et al., 2012	57	F	L1, conus roots	Radiculopathy (Acute onset)	Total	Improvement
35	Popescu et al., 2013	60	F	L4, root	Radiculopathy (Acute onset)	Total	Complete recovery
36	Mataliotakis et al., 2014	79	M	L2-3, root	Radiculopathy (Acute onset, 2 weeks)	Total + RR	Complete recovery
37	Takeshima et al., 2014 (cited by 42)	44	M	L2-3, root	Radiculopathy (1 month)	Total + RR	Improvement
38	Tao et al., 2014	45	M	T3-4, root	SAH; headache (Acute onset)	Total	Complete recovery
39	Kumar et al., 2016	21	M	L3-4, root	Radiculopathy (2 years)	Total	Complete recovery
40	Yaltirik et al., 2016	13	F	L1, cauda roots	Radiculopathy (4 weeks)	Total + RR	Improvement
41	Shi et al., 2017	73	M	T11-12, cauda roots	Myelopathy (5 years)	Total	Improvement
42	Golnari et al., 2017	60	M	L2	Radiculopathy (1–2 weeks)	Total + RR	Improvement
43	Ziechmann et al., 2018	55	M	T3-4, dorsal rootlet	Myelopathy, radiculopathy (1 month)	Total	Complete recovery
44	Sun et al., 2018	51	M	L2	Radiculopathy (5 months)	Total + RR	Complete recovery
45	Henderson et al., 2018	65	F	C6, ventral rootlet	Radiculopathy (Longstanding)	Total + RR	Complete recovery
46	Apostolakis et al., 2018	77	M	L3, cauda roots	Back pain (3 years)	Total + RR	Complete recovery
47	Pétillon et al., 2018	76	F	C7-8, root	Neck pain (2 years)	Total	Improvement
48	Vicenty et al., 2019	56	M	T2, root	Myelopathy (1 year)	Total + RR	Complete recovery
49	Hilmani & Lakhdar, 2019	65	F	L2, root	Radiculopathy (6 months)	Total + RR	Complete recovery
50	McQueen et al., 2021 (Present Case)	45	M	T3-4, root	Myelopathy (6 months)	Total + RR	Complete recovery
Cases with no documented root involvement							
51	Roger et al., 1951 (cited by 68)	22	F	T11	Radiculopathy (Acute onset)	Total	Clinically worsened
52	Floris, 1958 (cited by 68)	57	M	T12	Myelopathy (8 years)	Total	NR
53	Acciarri et al., 1992 (cited by 68)	54	F	C2-3, dura	SAH (Acute onset, 2 episodes) Cervical pain (20 years, intermittent)	Total	Improvement
54	Sharma et al., 1992	63	M	T12, near conus roots	Myelopathy (2 months)	Total	No improvement
55		43	M	T5, near dorsal roots and cord	Myelopathy (2 weeks)	Total	Improvement
56-57	Ahn et al., 1992 (cited by 35)	NR	F	NR	NR	Total	NR
		NR	F	NR	NR	Total	NR
58	Choi et al., 1996 (cited by 68)	46	F	L1	Back pain (duration NR)	Total	Improvement

(*Continued*)

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Table 1: (*Continued*)

No.	Authors, Year	Age	Sex	Location/Origin	Presentation (duration)	Intervention	Outcome
59	Padovani et al., 1997 (cited by 42)	31	M	L2, cauda equina	Radiculopathy (4 months)	Total	Improvement
60		54	F	C2-3	Headache, neck pain (Acute onset)	Total	Improvement
61	Crispino et al., 2005	65	M	T1-2	Myelopathy (2 months)	Total	Complete recovery
62	Chung et al., 2008 (cited by 35)	58	F	L2	NR	NR	NR
63		50	F	T4-5	NR	NR	NR
64		59	F	L4	NR	NR	NR
65	Sulochana et al., 2012 (cited by 68)	36	M	L5-S1	Back pain (1 year)	Total	Improvement
66	Kawanabe et al. 2012	52	F	T10-12	NR	Total	Complete recovery
67	Babu et al., 2013 (cited by 42)	63	F	NR	Sensorimotor deficit, urinary incontinence (3 years)	Total	Improvement
68	Katoh et al., 2014	36	M	L1	SAH (Acute onset) Sensorineural hearing loss, headache, hydrocephalus; Chronic SAH suspected (11 years)	Total	No improvement
69	Baldvinsdóttir et al., 2017	67	M	T6-7, extending through dura	Myelopathy (Brown-Sequard syndrome) (Few months, then acute worsening)	Total	Complete recovery
70	Iacob et al., 2017	56	F	T4-5	Myelopathy (duration NR)	Total	Complete recovery
71	Chong & Poh, 2017	73	M	L1-3	Myelopathy (<7 years)	Non-operative	Stable

NR = Not reported; Total = Total resection; Subtotal = subtotal resection; RR = Nerve root resection; SAH = subarachnoid hemorrhage.
See Supplemental Materials for additional references.

Although it is not possible to draw definitive conclusions, all cases published after 2000 reported postoperative improvement, which may indicate superior outcomes with advances in surgical technique.

In considering literature review findings and insights from our own case, we provide several recommendations regarding management of these lesions. Conservative approaches, including expectant management, do not offer symptom improvement and risk further deterioration and complications associated with SAH. Endovascular treatments are unfavorable given high chances of recurrence and progression of myelopathy following embolization.[9] For symptomatic patients, we recommend prompt surgical excision. For lesions uniquely attached to a nerve root, root sacrifice may be necessary; outcomes are likely still to be favorable, with opportunity for complete resolution of symptoms.

Significance and Conclusions

Although additional data are required, intradural-extramedullary cavernomas may demonstrate a unique propensity for radiculopathy or myelopathy at presentation. Excellent postoperative outcomes in our case and others support the role of surgical resection over conservative approaches, particularly in symptomatic patients, in order to prevent further neurological deterioration.

Supplementary Material. To view supplementary material for this article, please visit https://doi.org/10.1017/cjn.2022.287.

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EFL contributed to data acquisition, manuscript preparation and revision.

YS contributed to data acquisition and manuscript revision.

LCA contributed to data acquisition and manuscript revision.

ND contributed to data acquisition and manuscript revision.

All authors have made substantial contributions to the work reported in this manuscript and all have seen and approved of the final submitted version.

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