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Case Report

Intradural extramedullary cavernous malformation with extensive superficial siderosis of the neuraxis: Case report and review of literature

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Abstract

Background: Spinal cavernous malformations usually affect the vertebral bodies and are seldom intradural. Here, we report a rare spinal intradural-extramedullary cavernous malformation associated with extensive superficial siderosis along the neuraxis in a patient with radicular complaints.

Case Description: A 60-year-old male presented with subacute headaches, intermittent fever, and acute back and radicular leg pain for 1–2 weeks. Magnetic resonance imaging revealed an intradural-extramedullary lesion just below the conus medullaris (at the L2 level). There was associated subarachnoid hemorrhage in the lumbar cistern and superficial siderosis along the entire spinal neuraxis. Following surgical resection, the patient's symptoms resolved. Histopathology of the lesion was of a cavernous malformation.

Conclusions: There are only 56 cases of spinal intradural-extramedullary cavernous malformations published in the literature; however, only 3 described superficial neuraxis siderosis as noted in this case. In the present case, slowly recurring hemorrhages of the lesion located at the conus likely contributed to the complete neuraxis superficial siderosis. Timely evaluation and treatment of these lesions is warranted to avoid further compressive and/or hemorrhagic complications.

Key Words: Cavernous malformation, extramedullary, intradural, superficial siderosis

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INTRODUCTION

Spinal cavernous malformations (cavernomas or cavernous hemangiomas) are infrequent vascular malformations which occur primarily in the vertebral body, with or without extradural extension, that constitute 5–12% of all spinal vascular abnormalities.[11] Only 3% of spinal cavernomas are intradural, with intradural-extramedullary lesions being more rare compared with intramedullary

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locations.[8] Most patients present with symptoms related to spinal cord compression. Only 2 cases of intradural-extramedullary spinal cavernomas presented with superficial siderosis. Here, we report a cavernous malformation of the cauda equina presenting with extensive superficial siderosis extending from the brain and throughout the spinal neuraxis.

CASE REPORT

History and examination

A 60-year-old male with diffuse headaches and intermittent fever over several months presented with a 1–2-week history of left-sided low back pain radiating to the buttocks and the posterior aspect of the left leg. Other than radicular complaints, he had no focal neurological deficit. Laboratory findings were unremarkable. Magnetic resonance imaging (MRI) of the spine revealed a 1.8 × 1.3 × 1.1 cm intradural-extramedullary hemorrhagic lesion below the conus medullaris (at the L2 level) along with moderate subarachnoid hemorrhage (SAH) in the lumbar cistern and superficial siderosis along the conus [Figure 1]. MRI of the brain, cervical, and thoracic spine also identified diffuse superficial siderosis, a finding likely consistent with repeat hemorrhages [Figure 2].

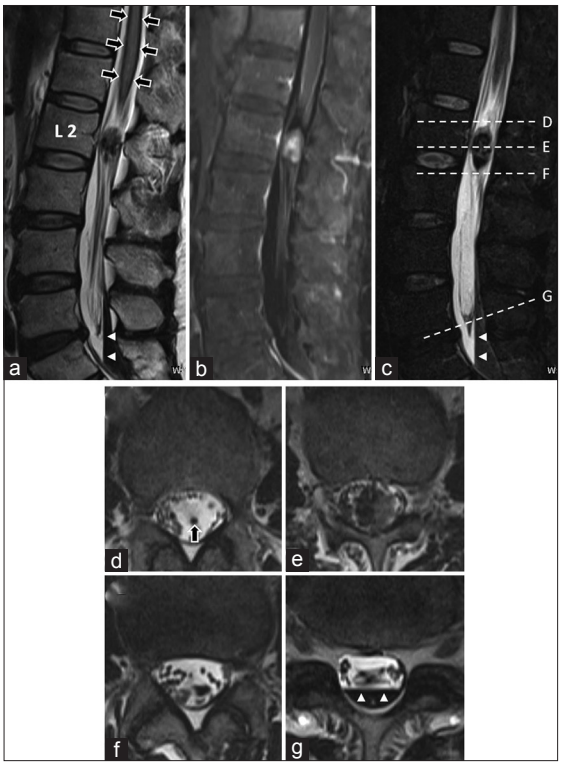

Figure 1: Lumbar spine MRI demonstrating intradural-extramedullary spinal cavernous malformation. Sagittal T2-weighted (a), fat-suppressed gadolinium-enhanced T1-weighted (b), and T2 inversion recovery turbo-spin echo images show an intradural extramedullary lesion at L2. Hemosiderin deposition is seen along the conus (arrows), and fluid-subarachnoid blood level is layered in the lumbar cistern (arrowheads). Axial T2-weighted images (d-g) corresponding to scout levels on image (c) show the nerve root proximal to the lesion (d), followed by sections in the midst (e) and immediately below (f) the lesion. Layering of subarachnoid blood is well-appreciated on axial images (arrowheads in g)

Surgery

An L1-L2 laminectomy and durotomy were performed, and a large vascular lesion with numerous small blood-filled sacs consistent with cavernoma was identified. The lesion was readily separable from the surrounding nerve roots and was dissected out of the thecal sac [Figure 3]. As a clear margin could not be distinguished between the lesion and nerve roots and because direct stimulation of the proximal and distal nerve roots [Figure 3a and b] did not elicit any motor response on neuromonitoring, the lesion was removed en bloc [Figure 3c]. The patient's headaches and back pain rapidly resolved following surgery. Six months later, the patient was neurologically intact.

Histopathology

Histopathological examination revealed that the mass was an encapsulated cavernous malformation with acute and chronic hemorrhages and focal inflammation [Figure 4].

DISCUSSION

There are 56 reports (including the present) of intradural-extramedullary spinal cavernous malformations [Table 1]. However, only 2 such cases with neuraxis siderosis have previously been reported;[3] this is the third such report.

Superficial siderosis of the central nervous system (CNS) is caused by recurrent hemorrhage into the subarachnoid space, with resultant hemosiderin deposition in the subpial layers of the brain and spinal cord. Extensive hemosiderin deposition can be seen along the surface of the neuraxis, especially along the cerebellum, brainstem, and lower

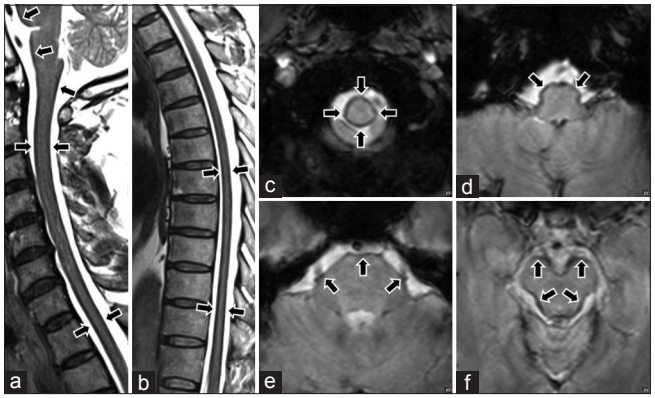

Figure 2: Brain and cervicothoracic MRI demonstrating extensive superficial siderosis. Cervical (a) and thoracic (b) T2-weighted sagittal MRI images show superficial siderosis (arrows) across the cervical and thoracic spinal cord. Intracranial involvement (arrows) is seen in the cerebellum and along the brain stem on axial gradient-echo sequences (c-f)

cranial nerves; the triad of sensorineural hearing loss, cerebellar ataxia, and pyramidal tract symptoms reflects the common sites of involvement in this disease.[5] Spinal intradural-extramedullary cavernomas are very rarely a cause of superficial siderosis. Nevertheless, our case, along with the 2 previously reported cases,[3] suggest that intradural-extramedullary cavernous malformations of the lower spine can indeed lead to intracranial superficial siderosis, warranting consideration of such lesions as part of the differential diagnosis and prompting neuraxial imaging in these patients.

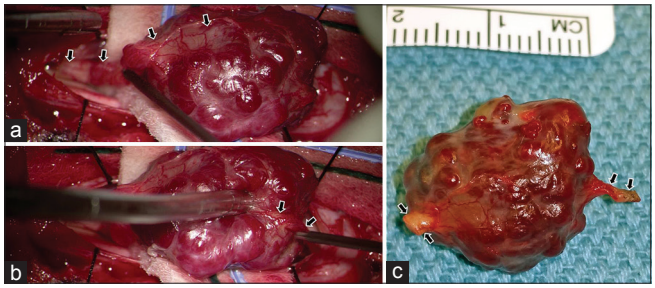

Figure 3: Intraoperative images during resection of spinal cavernous malformation. Stimulation of the proximal (a) and distal (b) ends of the involved nerve root (arrows) did not reveal any detectable motor function, and therefore the lesion was resected en bloc, along with the adjacent nerve root segments entering/exiting the lesion (c)

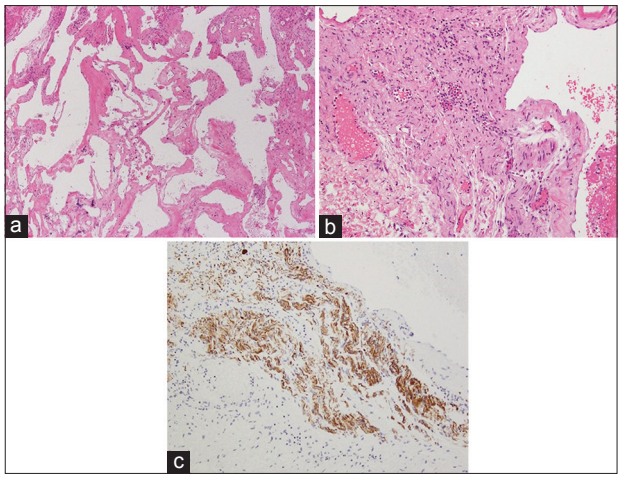

Figure 4: Sections showing a vascular lesion composed of tightly packed vascular channels with varying wall diameters and hyalinization. Vessel walls lacked any significant amount of smooth muscle or elastic tissue. Some vessel walls contained hemosiderin laden macrophages suggesting remote microhemorrhages. Scattered vessels showed thrombi at different stages of organization. These histologic features were consistent with a diagnosis of cavernous angioma. Intermediate power image (a) showing tightly packed vascular channels with vessels walls of varying diameters and some with hyalinization. Vessels walls lack elastic tissue and a significant amount of smooth muscle. High-power H and E image (b) of the cavernous angioma with entrapped nerve fibers. High-power image (c) of neurofilament staining the entrapped nerve fibers

Table 1: Published cases of spinal intradural extramedullary cavernous malformations*

No	Authors, year	Age (year)	Sex	Symptoms	Duration (month)	Level	Origin	Surgery	Improvement†
1	Roger *et al*., 1951 cited by[3]	22	F	Sciatic and back pain, sensorimotor deficit	UTD	T11	UTD	Total	No‡
2	Floris, 1958 cited by[3]	57	M	Sensorimotor deficit	96	T12	UTD	Total	UTD
3	Hirsch *et al*., 1965 cited by[3]	20	M	SAH, sensorimotor deficit, sphincter dysfunction	12	L2-L3	Root	Total (+RR)	No
4	Pansini and Lo Re, 1966 cited by[3]	46	M	Sciatic and back pain, sensorimotor deficit, sphincter and erectile dysfunction	UTD	L2	Root	Total	No
5	Heimberger *et al*., 1982 cited by[3]	24	M	SAH	13	T2-T3	Root	Total	Yes
6	Ueda *et al*., 1987 cited by[3]	28	M	SAH, back pain	36	L1-L2	Root	Total	Yes
7	Pagni *et al*., 1990 cited by[3]	46	M	Sciatic and back pain	48	T12-L1	Intraroot	Total (+RR)	Yes
8	Ramos *et al*., 1990 cited by[3]	67	F	Hydrocephalus, cognitive dysfunction, sphincter dysfunction, gait disturbance	4	L3	FT	Total	Yes
9	Mastronardi *et al*., 1991 cited by[3]	49	F	Sensorimotor deficit	6	T4	Root	Total	Yes
10	Mori *et al*., 1991 cited by[3]	65	M	SAH	86	T1	Root	Total	Yes
11	Acciarri *et al*., 1992 cited by[3]	54	F	SAH	240	C2-C3	Dura	Total	Yes
12-13	Sharma *et al*., 1992 cited by[3]	63	M	Back pain, sensorimotor deficit, sphincter dysfunction	2	T12	UTD	Total	No
		43	M	Chest pain, sensorimotor deficit, sphincter dysfunction	0.1	T5	UTD	Total	Yes
14-15	Ahn *et al*., 1992 cited by[11]	UTD	F	UTD	UTD	UTD	UTD	Total	UTD
		UTD	F	UTD	UTD	UTD	UTD	Total	UTD

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Table 1: Contd...

No	Authors, year	Age (year)	Sex	Symptoms	Duration (month)	Level	Origin	Surgery	Improvement†
16	Bruni *et al*., 1994 cited by[3]	28	M	SAH	0.1	L2	Root	Total	Yes
17-18	Cervoni *et al*., 1995 cited by[3]	26	F	SAH	0.1	L1-L2	Root	Total	Yes
		32	M	Back pain, sensorimotor deficit	36	L5	Root	Total	Yes
19	Makino *et al*., 1995 cited by[3]	67	M	SAH, hydrocephalus, sphincter dysfunction	3	L2	Root	Total (+RR)	Yes
20	Moreno *et al*., 1995 cited by[8]	63	M	Pain, motor deficit, sphincter dysfunction	84	T12-L1	Root	Total (+RR)	No
21	Harrison *et al*.,[2] 1995	37	M	Brown-Sequard syndrome	UTD	CMJ-C5	Root	Total	Yes
22	Choi *et al*., 1996 cited by[11]	46	F	Back pain	UTD	L1	UTD	Total	Yes
23-24	Rao *et al*., 1997 cited by[3]	60	M	Sensorimotor deficit	0.7	L1-L3	Root	Total	Yes
		35	F	Back pain, sensorimotor deficit	24	T12	Conus	Subtotal	No
25-26	Padovani *et al*., 1997 cited by[10]	31	M	Intermittent radicular symptoms	4	L2	UTD	Total	Yes
		54	F	Headache, Neck pain	0.03	C2-C3	UTD	Total	Yes
27	Duke *et al*., 1998 cited by[3]	49	F	Sciatic and back pain, sensory deficit, sphincter dysfunction	0.1	L4	Root	Total	Yes
28	Kim *et al*., 2001 cited by[11]	65	M	UTD	UTD	L4	Root	UTD	UTD
29	Park *et al*., 2003 cited by[11]	33	M	Back pain	12	L2-L3	Root	Total	Yes
30	Nozaki *et al*., 2003 cited by[3]	51	M	Sensorimotor deficit	6	C5-C6	Root/dentate ligament	Total	Yes
31	Falavigna *et al*, 2004 cited by[3]	44	F	Sciatic and back pain, sensorimotor deficit, sphincter dysfunction	4	L4	Intraroot	Total (+RR)	Yes
32	Chung *et al*., 2005 cited by[11]	52	M	Back pain, sensory deficit	24	L2	Intraroot	Total (+RR)	Yes
33	Crispino *et al*., 2005 cited by[8]	65	M	Back pain, sensory deficit	2	T1-T2	UTD	Total	Yes
34	Rachinger *et al*., 2006 cited by[3]	56	M	Shoulder pain	0.5	C7	Root	Total	Yes
35	Caroli *et al*., 2007 cited by[3]	71	M	Sciatic and back pain. Sensory deficit	UTD	L3	Intraroot	Total	Yes
36	Cecchi *et al*., 2007 cited by[3]	75	F	Sensory deficit	2	L3-L4	Root	Total (+RR)	Yes
37	Er *et al*., 2007 cited by[3]	67	M	Sciatic and back pain, sensorimotor deficit, sphincter dysfunction	4	T12-L2	Root	Total	Yes
38	Miyake *et al*., 2007 cited by[11]	18	M	Back pain	0.26	L1	Root	Total (+RR)	Yes
39-41	Chung *et al*., 2008 cited by[11]	58	F	UTD	UTD	L2	UTD	UTD	UTD
		50	F	UTD	UTD	T4-T5	UTD	UTD	UTD
		59	F	UTD	UTD	L4	UTD	UTD	UTD
42	Kivelev *et al*.,[4] 2008	44	M	Brown-Sequard syndrome, bladder paresis	5	C5-C6	Root	Total	Yes
43	Yi *et al*.,[13] 2008	67	M	Sciatic and Back pain	1	L2-L3	FT, root	Total	Yes
44	Chun *et al*., 2010 cited by[11]	74	F	Sciatic pain	36-48	Below L4	Intraroot	Total (+RR)	Yes
45-46	Khalatbari *et al*., 2011 cited by[11]	58	M	Sciatic and back pain	0.16	L3-L4	Root	Total (+RR)	Yes
		45	M	Sciatic and back pain	0.1	L1-L2	Root	Total	Yes
47	Jin *et al*., 2011 cited by[3]	55	M	Sensorineural hearing loss, headache, dizziness	180	T12-L1	Root	Total	Yes

Contd...

Table 1: Contd...

No	Authors, year	Age (year)	Sex	Symptoms	Duration (month)	Level	Origin	Surgery	Improvement†
48	Sulochana *et al*., 2012 cited by[3]	36	M	Back pain	12	L5-S1	UTD	Total	Yes
49	Nie *et al*.,[8] 2012	57	F	Pain, sphincter dysfunction	6	L1	Conus	Total	Yes
50	Popescu *et al*.,[11] 2013	60	F	Back pain	0.13	L4	Root	Total	Yes
51	Babu *et al*.,[1] 2013	63	F	Sensorimotor deficit, urinary incontinence	36	UTD	UTD	Total	Yes
52	Mataliotakis *et al*.,[7] 2014	79	M	Back pain, sensorimotor deficit, cramping feelings in leg	0.5	L2-L3	Root	Total (+RR)	Yes
53	Takeshima *et al*.,[12] 2014	44	M	Back pain, bilateral sciatica, neck pain	1	L2-L3	Root	Total (+RR)	Yes
54	Katoh *et al*.,[3] 2014	36	M	Sensorineural hearing loss, headache, hydrocephalus	132	L1	UTD	Total	No
55	Kumar *et al*.,[6] 2016	21	M	Back pain, radiating to leg	24	L3-L4	Root	Total	Yes
56	Present, 2017	60	M	Back pain, radiating to leg, headache, fever	0.33	L2	Root	Total (+RR)	Yes

UTD: Unable to determine; SAH: Subarachnoid hemorrhage; RR: Nerve root resection; FT: Filum terminale. *Summary of cases published to date. Of note, differences between the above table and prior reviews relate to inclusion of cases,[9] which on close review described pathologies other than strictly intradural extramedullary spinal cavernous malformations. †Postop at last follow-up (Improvement was defined as either partial or complete improvement). ‡The only case that got worse in follow-up

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Conflicts of interest
There are no conflicts of interest.

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