Case Report

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

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Received: 13 March 17  Accepted: 05 May 17  Published: 13 June 17

Abstract

Background: Spinal cavernous malformations usually affect the vertebral bodies and are seldom intradural. Here, we report a rare spinal intradural-extradural 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-extradural 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-extradural 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, extradural, intradural, superficial siderosis

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-extradural lesions being more rare compared with intramedullary...
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 [Figure 1]. MRI of the brain, cervical, and thoracic spine also identified diffuse superficial siderosis, a finding likely consistent with repeat hemorrhages [Figure 2].

**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
craniocerebellar 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-extradural cavernomas are very rarely a cause of superficial siderosis. Nevertheless, our case, along with the 2 previously reported cases,\(^3\) suggest that intradural-extradural 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.

| 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 |
| 14-15 | Ahn et al., 1992 cited by\(^11\) | UTD | F | UTD | UTD | UTD | Total | UTD | UTD |

Table 1: Published cases of spinal intradural-extradural cavernous malformations*

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 angiomma. 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 angiomma with entrapped nerve fibers. High-power image (c) of neurofilament staining the entrapped nerve fibers.

Contd...
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| 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 |
| 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[3] | 63 | M | Pain, motor deficit, sphincter dysfunction | 84 | T12-L1 | Root | Total (+RR) | No |
| 21 | Harrison et al., 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] | 35 | F | Back pain, sensorimotor deficit | 24 | T12 | Conus | Total | Yes |
| 25-26 | Padovani et al., 1997 cited by[3] | 31 | M | Intermittent radicular symptoms | 4 | L2 | UTD | Total | Yes |
| 27 | Duke et al., 1998 cited by[3] | 54 | F | Headache, Neck pain | 0.03 | C2-C3 | UTD | 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 | Total (+RR) | (Intraroot) | 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[3] | 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 defect | 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 |
| 42 | Kivelev et al,[4] 2008 | 44 | M | Brown-Sequard syndrome, bladder paresis | 5 | C5-C6 | Root | Total | Yes |
| 43 | Yi et al.,[10] 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 | 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 |
| 47 | Jin et al., 2011 cited by[3] | 45 | M | Sciatic and back pain | 0.1 | L1-L2 | Root | Total | Yes |

Contd...
Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES

1. Babu R, Owens TR, Karikari IO, Moreno J, Cummings TJ, Gottfried ON, et al. Spinal cavernous and capillary hemangiomas in adults. Spine 2013;38:E423-30.
2. Harrison MJ, Eisenberg MB, Ullman JS, Oppenheim JS, Camins MB, Post KD. Symptomatic cavernous malformations affecting the spine and spinal cord. Neurosurgery 1995;37:195-204.
3. Katoh N, Yoshida T, Uehara T, Ito K, Hongo K, Ikeda S. Spinal intradural extramedullary cavernous angioma presenting with superficial siderosis and hydrocephalus: A case report and review of the literature. Intern Med 2014;53:1863-7.
4. Kivelev J, Ramsey CN, Dashti R, Porras M, Tyninen O, Hernesniemi J. Cervical intradural extramedullary cavernoma presenting with isolated intramedullary hemorrhage. J Neurosurg Spine 2008;8:88-91.
5. Kumar N. Superficial siderosis: Associations and therapeutic implications. Arch Neurol 2007;64:491-6.
6. Kumar V, Nair R, Kongwad LL, Menon RG. Cavernous haemangioma of the cauda equina: Case report and review of literature. Br J Neurosurg 2016 [Epub ahead of print].
7. Mataliotakis G, Perera S, Nagaraju S, Marchionni M, Tzerakis N. Intradural extramedullary cavernoma of a lumbar nerve root mimicking neurofibroma. A report of a rare case and the differential diagnosis. Spine J 2014;14:e1-7.
8. Nie QB, Chen Z, Jian FZ, Wu H, Ling F. Cavernous angioma of the cauda equina: A case report and systematic review of the literature. J Int Med Res 2012;40:2001-8.
9. Ortner WD KH, Pilz P. Ein zervikales kavernoeses angiom. Fortschr Roengenetstr 1973;118:475-6.
10. Padovani R, Acciarri N, Giulioni M, Pantieri R, Foschini MP. Cavernous angiomas of the spinal district: Surgical treatment of 11 patients. Eur Spine J 1997;6:298-303.
11. Popescu G, Mihaela Sandu A, et al. Cauda equina intradural extramedullary cavernous haemangioma: Case report and review of the literature. Neurol Med Chir 2013;53:890-5.
12. Takeshima Y, Marutani A, Tamura K, Park YS, Nakase H. A case of cauda equina cavernous angioma coexisting with multiple cerebral cavernous angiomas. Br J Neurosurg 2014;28:544-6.
13. Yi Y Z-XD, Dong Z, Gang Y, Wen-Yuan T. Cavernous Malformation of the Cauda Equina-a Case Report. Ceska a Slovenska Neurologie a Neurochirurgie. 2008;72(6):575-579.