Dorsal Spinal Epidural Cavernous Angioma; A Case Report

Abstract
Spinal cavernous angiomas are lesions formed by vessels lined by closely clustered endothelial cells. They are common in the vertebral body and less common in an intradural location. However, these are very rare in the extra-osseous and epidural region. Less than 100 cases have been reported. Here, we report a case of dorsal spinal extradural cavernous angioma in a 52-year-old man who presented with back pain and difficulty in walking. Magnetic resonance imaging brain showed D7–D8 (thoracic) extradural spinal lesion, enhancing homogeneously on contrast administration. He underwent D7–D8 hemilaminectomy and tumor decompression. The tumor was extradural, tightly adherent to the dura, and highly vascular. He recovered completely after surgical removal with no recurrence 2 years after removal. He was not administered adjuvant radiotherapy. In this article, we review the literature regarding clinical features, imaging findings, and outcome of spinal epidural cavernous angioma.

Keywords: Cavernous angioma of the spine, epidural spine tumor, magnetic resonance imaging spine, spinal tumor, spine tumor surgery, vascular spine lesion

Introduction
Cavernous angiomas are common lesions of the central nervous system characterized by abnormally dilated blood vessels lined by a thin endothelium. In the spine, a common location is the vertebral bodies. Intradural extramedullary and intramedullary cavernous hemangiomas are less frequent lesions, while purely epidural locations are uncommon.[1] Epidural cavernous angiomas represent 12% of spinal axis cavernous malformations.[2]

The earliest case was reported in 1978 by Decker et al.[3] They are indistinguishable from that of a schwannoma, which is a much more common lesion. Here, we report a case of thoracic epidural cavernous angioma who presented with chronic backache.

Case Report
A 52-year-old man presented with back pain since a month. He had imbalance while walking on an irregular surface. It was not associated with paraparesis or urinary retention. Magnetic resonance imaging (MRI) of the spine showed a lobulated, homogeneously contrast-enhancing lesion at the thoracic D7–D8 epidural region. It was hyperintense on T2-weighted image and isointense on T1-weighted image. Indentation of the dorsal spinal cord was seen. Displacement of the cord is seen anterior and to the left side. No obvious cord signal changes were seen. The lesion shows no extension to neural foramen [Figure 1]. He underwent D7–D9 hemilaminectomy and decompression of extradural lesion. The tumor was extradural, highly vascular, and tightly adherent to the dura mater. There was no extension outside the spinal canal. The tumor was partially removed as it was highly vascular, and hemostasis was achieved with activated thrombin and Surgicel Fibrillar. Postoperatively, he had weakness in right quadriceps muscles which gradually improved over 2 weeks. He was not treated with adjuvant radiotherapy. Histopathology examination of section showed variably sized vessels lined by endothelial cells [Figure 2]. The vessels ramified between lobules of adipose tissue. Vascular endothelial cells are highlighted with CD34 [Figure 3]. A few of the vessels showed fibrin thrombi in the tumor. No mitosis or necrosis was seen. It
was reported as hemangioma of the extradural space at the D7–D8 dorsal spine. Postoperative MRI spine performed after a year showed no recurrence of the lesion [Figure 4]. He has not received any postoperative radiotherapy. The patient is asymptomatic 2 years after the operation.

**Discussion**

Cavernous angiomas of the spine are benign and are vascular hamartomas representing a dysplasia of the vessel-forming mesoderm. Harrison *et al.* postulated that primordial vessels may lose their capacity to differentiate, resulting in a cavernous malformation.[4]

Microscopically, cavernous malformations are composed of closely opposed sinusoidal vascular spaces. The walls consist of an innermost single layer of endothelial cells surrounded by adipose tissue; elastic fibers or smooth muscle cells are absent.[5]

We reviewed the literature of 46 case reports (including our case). The first case we found was reported by Decker *et al.* in 1978 [Table 1].

The average age of patients was 50 years. The youngest was 13 years old and eldest was 79 years old. There were 27 males and 19 females with slight male dominance.

Clinical symptoms were suggestive of the spinal cord or nerve root compression. Pain was the most common
Symptom. Cases presented as neck pain, interscapular pain, or back pain depending on the location of the lesion. They were also associated with numbness and radicular pain along the dermatomes of involved roots. Twenty cases presented with spastic paraparesis and urinary frequency or retention.

Thoracic spinal cord was the most common location. In 27 cases, thoracic spinal cord was involved. In our case, dorsal D7 and 8 epidural space was involved. The cervical spine was involved in 6 cases. The lumbar spine was involved in 11 cases. The sacral epidural space was involved in 1 case.

Radiological features

MRI finding showed an epidural lesion of varying sizes. It is hyperintense on T2-weighted image and isointense on T1-weighted image. Homogeneous contrast enhancement was seen with gadolinium administration. The lesion was completely extradural but tightly adherent to the dura mater. Hemorrhage was seen only in one case reported by Khalatbar et al.[21]

Dumbbell-shaped, extraforaminal extension was seen in more than 10 cases. These features resemble MRI findings of schwannoma.[22] Therefore, it is difficult to diagnose on radiological imaging alone.

Surgery

All patients underwent surgery either laminotomy or laminectomy. The tumor was extradural, purple or mulberry like, soft, and highly vascular. All surgeries were planned except few. An emergency surgery was performed for hemorrhage into the lesion in one case reported by Khalatbar et al. Li et al. reported four cases where surgery was done on an emergency basis for sudden onset of paraparesis and urinary retention. The pathology for sudden neurological deterioration was explained by Lee et al.

Cavernous angiomas do not grow by mitotic activity but have the propensity to enlarge by thrombosis and bleeding, causing a spectrum of neurological syndromes ranging from radiculopathy to sudden spinal cord dysfunction.[24]

From the surgical point of view, it is very important to understand that the extra-axial cavernous angiomas behave like tumors and not like vascular malformations.[19]

Surgical decompression is the mainstay of treatment. Radiotherapy was not given as a primary mode of treatment. Only in two cases, adjuvant radiotherapy was given few months after surgery as symptoms persisted or worsened (reported by Sohn et al. and Padovani et al.).

Fukushima et al. reported recurrence of the lesion in his case. He operated thrice over a period of two decades.

Outcome

Spinal epidural angioma has a favorable outcome with total or subtotal removal. Complete recovery was noted in 19 patients. Partial recovery was seen in 8 patients. Our case had right knee extensor weakness in the postoperative period. He recovered within 2 weeks with no neurological deficits.

Conclusion

This observation and a review of the literature highlights that spinal epidural cavernous angioma presents as compressive spinal cord pathology with symptoms of backache, weakness in legs, difficulty in walking, or retention of urine. MRI spine with contrast helps in preoperative diagnosis of angioma. It may present with sudden neurological deficits due to hemorrhage which warrants emergency decompression. Surgical decompression is the mainstay of treatment. There is no role of adjuvant radiotherapy. As the tumor is highly
### Table 1: Review of literature

| Author                  | Age (years) | Sex  | Symptom                           | Location            | Size                | Surgery                                           | Recovery |
|-------------------------|-------------|------|-----------------------------------|---------------------|---------------------|--------------------------------------------------|----------|
| Minh et al., 2005       | 18          | Male | Neck pain, paresthesia           | Cervical C6-T1      | 1.5×4.5             | C5-T2 laminoplasty                               | CR       |
| Yunoki et al., 2015     | 77          | Male | Back pain                        | Lumbar L2-L3        | Dumbbell shaped     | L2-L3 extra- and intracanalicular approach       | CR       |
| Uchida et al., 2010     | 75          | Male | Both leg pain and numbness 3 years| T11-T12 thoracic   | Extending to left   | T11-T12 partial laminectomy and medial foraminotomy | CR       |
| Talacchi et al., 1998   | 66          | Male | Spastic paraparesis, leg pain, and numbness ×3 years | T5-T7 thoracic     | Purple color       | Laminctomy at T5-T6                             | CR       |
|                         | 69          | Male | Back pain, right leg pain, numbness, weakness since 3 weeks | T6-T8 thoracic     | Mulberry, extending to right foramen             |         |
|                         | 61          | Male | Back pain, spastic paraparesis   | T8-T10              | Pushing spinal cord | T8-T10 laminotomy                              | PR       |
|                         | 72          | Female | Spastic paraparesis, urinary retention, paresthesia | Thoracic T4-T6      | Extending to left T5 foramen and pushing spinal cord to right | T4-T6 laminotomy | PR       |
|                         | 44          | Female | Spastic paraparesis, neck, right arm thoracic pain | Thoracic T5-T8      | Oval extending to right paravertebral space through T6-T7 foramen | T5-T8 laminotomy 2nd surgery (thoracotomy) for right paravertebral lesion | PR       |
| Rovira et al., 1999     | 51          | Female | Back pain and right sciatica 3 months | Lumbar L3-L4        | Extending to left L3-L4 foramen | L3-L4 laminotomy | PR       |
|                         | 16          | Male | Left sciatica 1 year             | Lumbar L5-S1        | Associated with L5-S1 Grade 1 spondylolisthesis L4 lateral recess extension | L5-S1 laminotomy | PR       |
|                         | 19          | Female | Left leg pain, pollakiuria       | Lumbar L3-L4        | L4 lateral recess   | L3-L4 Laminotomy                                | CR       |
| Padovani et al. 1982    | 75          | Male | Spastic paraparesis             | Thoracic T3-T6      | Extending to left T5 foramen | T3-T6 laminotomy, received radiotherapy | CR       |
| Morioka et al. 1986     | 50          | Male | Paresthesia in legs and abdomen below T6 | Thoracic T2-T3      | Extending to left paravertebral space through left L5 foramen | T2-T4 laminectomy and left thoracotomy | CR       |
| Decher et al. 1978      | 65          | Female | Right abdomen and leg pain       | Lumbar L1-L2        | Extending to right L1 foramen | L1-L2 right hemilaminectomy and foraminotomy | CR       |
| Lanotte et al. 1994     | 65          | Male | Neck pain since 6 months         | Thoracic T1-T2      | Extending across left T1 foramen to paravertebral space | Thoracic left T1-T2 laminotomy | NA       |
| Franz et al. 1987       | 23          | Male | Spastic paraplegia              | Thoracic T1-T2      |                    | Thoracic T1-T2 laminotomy                      | NA       |
| Li et al., 2015         | 79          | Male | None                             | T6-T7               | NA                  | T6-T7 laminotomy                                | NA       |
|                         | 56          | Male | Spastic paraplegia, urine retention | T2-T4              | NA                  | T2-T4 laminotomy                                | NA       |
|                         | 42          | Male | None                             | T7-T8               | NA                  | T7-T8 laminotomy                                | NA       |
|                         | 15          | Male | Spastic paraplegia, urine retention | C6-T2              | NA                  | Emergency C6-T2 laminotomy                      | NA       |
|                         | 35          | Male | Spastic paraplegia, urine retention | T2-T4              | NA                  | Emergency T2-T4 laminotomy                      | NA       |
|                         | 68          | Female | Spastic paraplegia, urine retention | L2-L3, T2-T4        | NA                  | L2-L3 laminotomy                                | NA       |
|                         | 66          | Male | Spastic paraplegia, urine retention | T2-T4              | NA                  | T2-T4 laminotomy                                | NA       |
|                         | 67          | Male | Spastic paraplegia, urine retention | T3-T4              | NA                  | T3-T4 laminotomy                                | NA       |
|                         | 24          | Male | Spastic paraplegia, hyperesthesia | C6-C7              | NA                  | Emergency C6-C7 laminectomy                     | NA       |

Contd...
vessel, hemostatic agents are required during surgery. The tumor may extend into paravertebral space through the intervertebral canal. This may require additional thoracotomy for tumor removal. Clinical recovery is complete after surgery in most of the cases.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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