Spinal intradural cystic venous angioma originating from a nerve root in the cauda equina

Case report

YUSUKE NISHIMURA, M.D., PH.D.,1,3 MASAHITO HARA, M.D., PH.D.,1
ATSUSHI NATSUME, M.D., PH.D.,1 YASUHIRO NAKAJIMA, M.D., PH.D.,1
RYUICHI FUKUYAMA, M.D., PH.D.,2 TOSHIHIKO WAKABAYASHI, M.D., PH.D.,1
AND HOWARD J. GINSBERG, M.D., PH.D.3

1Department of Neurosurgery, Nagoya University, Nagoya, Japan; 2Division of Pathology, Konan Kosei Hospital, Aichi, Japan; and 3Division of Neurosurgery, St. Michael’s Hospital, University of Toronto, Ontario, Canada

A spinal intradural extramedullary venous angioma is extremely rare and has not been previously reported. In this paper, the authors report on this entity with morphological and immunohistochemical evidence, and discuss the surgical strategy for its treatment. A 54-year-old woman presented to Nagoya University Hospital complaining of left-sided pain in the hip, thigh, and inguinal and perianal regions, with progressive worsening during the previous 2 weeks. Lumbar spine MRI showed an intradural extramedullary cyst at the level of T12–L1, which extended from the conus medullaris to the cauda equina. The cyst wall was not enhanced on T1-weighted MRI with Gd. Intraoperatively, a midline dural opening allowed the authors to easily visualize a dark-red cyst behind the spinal nerve rootlets in the cauda equina adjacent to the conus medullaris. The cyst was believed to originate from one of the spinal nerve rootlets in the cauda equina and a cluster of veins was identified on the cyst wall. The cyst was resected with the affected nerve rootlet. The surgery left no detectable neurological deficit. Based on the morphological and immunohistochemical evidence, the lesion was diagnosed as a venous angioma. No tumor recurrence was confirmed based on MRI at the time of the 2-year follow up. This is the first report of an intradural extramedullary cystic venous angioma that was successfully resected.

(http://thejns.org/doi/abs/10.3171/2013.8.SPINE121012)

KEY WORDS
- venous angioma
- intradural
- cystic lesion
- cauda equina
- oncology

VASCULAR lesions comprise approximately 6%–7% of all spinal intradural tumors.12 Spinal vascular malformations may be classified as capillary tel-angiectasias, cavernous angiomas, arteriovenous malformations, or venous angiomas.9 Venous angiomas are the most frequently encountered cerebral vascular malformations, with an incidence of up to 2.6% in a series of 4069 brain autopsies.14 However, spinal venous angiomas are extremely rare lesions. Only 2 previous reports of spinal epidural venous angiomas3/9 have been found, but a previous report on spinal intradural venous angiomas was not encountered.

We present an extremely rare spinal, intradural, extramedullary cystic venous angioma originating from a spinal nerve root in the cauda equina, which appears to be the first recorded example with a detailed assessment using MRI, intraoperative photography, and immunohistochemistry.

Case Report

History and Examination. A 54-year-old woman had a 4-year-history of low-back pain. This was followed by the development of left-sided pain in the hip, thigh, and inguinal and perianal regions, with progressive worsening during the 2 weeks prior to admission. On examination, the patient had severe left-sided pain and numbness in the hip, thigh, and inguinal and perianal regions, but no muscular weakness. She also had normal rectal tone and bladder function. Laboratory analysis revealed no abnormalities.

A lumbar spine MR image revealed a cystic mass at

Abbreviations used in this paper: EMA = epithelial membrane antigen; GFAP = glial fibrillary acidic protein; NSE = neuron-specific enolase; SMA = smooth muscle actin.
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the level of T12–L1, which was an intradural extramedullary lesion extending from the conus medullaris to the cauda equina (Fig. 1). The cyst content was slightly hyperintense on T1-weighted MRI (Fig. 1a) and isointense with CSF on T2-weighted MRI (Fig. 1b, d, and e). This lesion had no Gd enhancement (Fig. 1c). The differential diagnosis included cystic tumor and spinal cyst. Surgical exploration and resection were performed due to the progressive nature of the symptoms.

Operation. After a T12–L2 osteoplastic laminotomy and midline dural opening, a dark-red cyst was easily identified behind the spinal nerve rootlets in the cauda equina (Fig. 2a), located just lateral to the conus medullaris. The lesion was readily dissected from the conus medullaris and was not connected to it. During the dissection, the cyst was perforated and the dark red fluid leaked out, leading to complete deflation of the cyst. The fluid appeared to be old hemorrhagic blood, which was subsequently verified by pathological examination. There was no active bleeding from the cyst. The cyst was believed to originate from one of the spinal rootlets in the cauda equina because it completely encased the nerve rootlet (Fig. 2b). A cluster of dilated veins was visualized on the cyst wall (Fig. 2b) and was considered a “caput medusae” type of architecture. These vessels converged proximally on the cyst wall, where they became obliterated. Although we could not find a large draining vein like those usually found in a brain venous angioma, we speculated that occlusion of the drainage vein collecting these dilated veins occurred at this point. To preserve most of the nerve rootlet, we tried to dissect and isolate the lesion from the nerve rootlet, just as one would do during the resection of a schwannoma. Ultimately, the cyst was not able to be separated from the affected nerve rootlet, which was entirely resected (Fig. 2c) with the cyst. The cyst was successfully removed without any detectable neurological deficit.

Pathological Findings. Sections stained with H & E showed that the lesion was characterized by many irregular and cystic vessels (Fig. 3). The lumina of these structures were lined by a flattened monolayer of cells. Immunohistochemical analysis revealed that the surface-lining cells (Fig. 4a) were positive for CD31 (Fig. 4b), Factor VIII (data not shown), and vimentin (Fig. 4d), but negative for epithelial membrane antigen (EMA; Fig. 4c) and glial fibrillary acidic protein (GFAP) (data not shown), indicating that they were endothelial cells. Moreover, some
of these vessels (Fig. 5a) included smooth muscle and connective tissue in their walls, which was positive for smooth muscle actin (SMA; Fig. 5b), indicating that these blood vessels were the venous type, and not merely capillaries. These vessels appeared to develop within neural tissues as revealed by the existence of neuron-specific enolase (NSE) and GFAP-positive tissues between vessels (Fig. 5c and d). Taking all of these data into account, we diagnosed this lesion as a venous angioma.

**Postoperative Course.** Postoperatively, the patient’s neurological symptoms resolved immediately and she remains symptom free with no recurrence on MRI during 2 years of follow-up in the outpatient clinic (Fig. 6).

**Discussion**

This is the first report of an intradural spinal venous angioma. No other cases have been previously reported. Venous angiomas are extremely rare in the spinal region, but are the most frequently encountered cerebral vascular malformations. Venous angiomas are characterized by a cluster of venous radicles that converge into a collecting vein. In the brain, stenosis is commonly observed on the collecting vein of venous angiomas, and hemorrhagic or ischemic infarction around venous angiomas results from acute thrombosis of the collecting vein. The venous angioma in our case appeared to originate from the nerve rootlet and have no direct relationship with the conus medullaris, although it lay adjacent to the conus medullaris. The cluster of dilated veins was identified and converged proximally on the cyst; however, we could not find any other dilated veins outside the cyst wall on the affected nerve rootlet. We can speculate that intracystic hemorrhage caused by venous congestion of the draining vein, which collected the flow of dilated veins on the cyst wall, resulted in progressive worsening of the neurological deficit, just as in similar situations in the brain. Given the fact that venous flow of the nerve rootlets in the proximal cauda equina is primarily from the periphery toward the conus medullaris, we believed that the occlusion of the draining vein occurred proximally on the cyst.

The spinal nerve rootlets float in CSF, and their nerve root sheaths are very thin. Most all of the vessels of nerve rootlets are located inside their thin radicular sheaths, where venous angiomas should exist. Nerve root venous angiomas appear to arise from a completely different location from nerve sheath tumors such as schwannomas; schwannomas arise from the radicular sheaths of nerve roots, not from nerve root fascicles themselves. Schwannomas should be dissected off of the greater part of the nerve root, even if some nerve fascicles may be encased by tumor. However, in this presenting venous angioma originating from a spinal nerve rootlet in particular, the cyst membrane was considered to be a radicular sheath itself that was stretched by intraneurve rootlet hemorrhage, and we observed a venous cluster on the thin and dilated radicular sheath. Given these facts, there was no choice but to transect the entire affected nerve rootlet for complete resection of the tumor.

The differential diagnosis of cyst-forming entities includes not only spinal cysts, but also tumors such as dermoid cysts, epidermoid cysts, myxopapillary ependymomas, cystic astrocytomas, and cystic schwannomas. These tumors should be differentiated based on MRI findings such as the heterogeneity of the cyst content, positive en-
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hancement and irregular thickness of the cyst wall, and presence of a solid component. Intradural spinal cysts are uncommon benign formations histologically classified as arachnoid cysts, neurenteric cysts, ependymal cysts, and ventriculus terminalis. Magnetic resonance imaging findings of these cysts are very similar to each other. Our case displayed similar MRI findings to those of spinal cysts. Cystic fluid intensity is distinct on MRI depending upon the contents (protein or hemorrhage) within the cyst, even within the same pathology. Furthermore, cyst walls are similarly homogeneously thin in each pathology. Therefore, among these cystic lesions, a definitive diagnosis is extremely difficult based on MRI findings alone, but several key points would help us with a preoperative presumptive diagnosis. Key points for differential diagnosis are listed in Table 1. Arachnoid cysts are the most common intradural spinal cysts, occurring dorsal to the spinal cord and usually in the thoracic spine. The best diagnostic procedure for arachnoid cysts is myelography with delayed CT to maximize the chance of filling arachnoid cysts with contrast medium. Neurenteric cysts usually occur ventral to the spinal cord in the lower cervical and upper thoracic spine and are extramedullary. Neurenteric cysts are congenital anomalies frequently associated with unique bone anomalies, and are more likely to be found in the first or second decade of life. Ependymal cysts are well-defined lesions, usually located in the spinal cord from the cervical spine to the conus medullaris, with the cyst content isointense to CSF on MRI. The cysts do not communicate with the central canal. The term “ventriculus terminalis,” or fifth ventricle, refers to an ependyma-lined space of the conus medullaris containing CSF. This ventricle is formed during embryogenesis as a result of canalization and regressive differentiation. Cystic dilation of the ventriculus terminalis is always ovoid, with smooth walls and regular marginations, continuous with a nondilated central canal without any internal septae. Signal intensity of the intraventricular fluid closely resembles that of CSF on all sequences.

Histopathological examinations are imperative to arrive at a definitive diagnosis for these types of cases.

### TABLE 1: Summary of characteristics of spinal cystic lesions based on MRI findings

<table>
<thead>
<tr>
<th>Lesion</th>
<th>Cyst Content</th>
<th>Cyst Wall</th>
<th>Enhancement of the Cyst Wall</th>
<th>Location</th>
<th>Other Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cystic tumors</td>
<td>heterogeneous</td>
<td>irregular thickness</td>
<td>enhancement of the cyst wall</td>
<td>various places</td>
<td>presence of solid component</td>
</tr>
<tr>
<td>Arachnoid cysts</td>
<td>homogeneous</td>
<td>uniformly &amp; regularly thin</td>
<td>none</td>
<td>dorsal to the spinal cord</td>
<td>most common intradural spinal cysts</td>
</tr>
<tr>
<td>Neurenteric cysts</td>
<td>homogeneous</td>
<td>uniformly &amp; regularly thin</td>
<td>none</td>
<td>ventral to the spinal cord &amp; extramedullary</td>
<td>associated w/ bone anomalies</td>
</tr>
<tr>
<td>Ependymal cysts</td>
<td>homogeneous</td>
<td>uniformly &amp; regularly thin</td>
<td>none</td>
<td>intramedullary</td>
<td>no communication w/ central canal</td>
</tr>
<tr>
<td>Ventriculus terminalis</td>
<td>homogeneous</td>
<td>uniformly &amp; regularly thin</td>
<td>none</td>
<td>ependyma-lined space of the conus medullaris</td>
<td>ovoid &amp; continuous w/ a nondilated central canal</td>
</tr>
</tbody>
</table>

**Fig. 6.** Postoperative sagittal (a), coronal (b), and axial (c) T2-weighted MR images obtained 2 years after surgery showing no residual tumor and no recurrence.
As for the present case, H & E staining showed that this lesion was characterized by many irregular and cystic vessels with thin walls and surface-lining epithelial cells. Immunohistochemical analysis revealed that surface-lining epithelial cells were positive for CD31 and Factor VIII, with vessel walls positive for SMA, but negative for EMA, indicating that they were endothelial cells, but not arachnoidal or ependymal cells. We can rule out other cystic lesions, such as arachnoid cysts, neurenteric cysts, and ependymal cysts, by the configuration of the tumor on H & E sections as well as the fact that GFAP- and NSE-positive spinal neural tissues intervene in the lesion on immunohistochemical analysis. We could not find any direct involvement of the conus medullaris by this lesion because a clear border between these two structures was observed and readily separated intraoperatively. Thus, neural tissues constituting the tumor do not appear to originate from the conus medullaris. Although not commonly reported, infrequently associated with venous angioma are focal anomalies of neuronal migration. There is evidence that the migration anomaly is vascular in origin. Venous angiomas are widely accepted as a focal anomaly of neuronal migration. Venous angiomas are focal anomalies of neuronal migration. Venous angiomas are focal anomalies of neuronal migration. Venous angiomas are focal anomalies of neuronal migration.

A spinal, intradural, extramedullary venous angioma is an extremely rare entity, which has not been previously reported. However, because cystic dilation of the lesion caused by intrallesional congestion and hemorrhage would provoke a progressive neurological deficit and require surgical intervention, venous angioma should be considered in the differential diagnosis of spinal cystic lesions.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Nishimura. Acquisition of data: Nishimura, Nakajima. Analysis and interpretation of data: Nishimura, Natsume. Drafting the article: Nishimura. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Nishimura. Administrative/technical/material support: Haru, Natsume, Fukuyama, Wakabayashi. Study supervision: Haru, Fukuyama, Wakabayashi, Ginsberg.

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