

Intradural Extramedullary Arachnoid Cyst Presenting as Arteriovenous Malformation in the Thoracic Spinal Cord

Chris Conner, B.S., and Amir Paydar, M.D.

We report the case of a 57-year-old man who was admitted on the basis of worsening paraplegia and incontinence. This patient had developed similar neurologic symptoms 15 previously but fully recovered after conservative medical therapy with corticosteroids. At that point, he was diagnosed with a thoracic spinal arteriovenous malformation (AVM); yet, no focal spinal lesion was identified. During this second presentation, the patient was initially worked up at an outside institution, where MR imaging revealed serpiginous flow voids along with anterior displacement or atrophy of the cord at the T5-T6 level. Edema of the mid and lower thoracic cord was also noted. While under our care, spinal arteriography and CT angiography failed to conclusively detect an underlying AVM. The patient underwent a second MR study, which showed no major interval change in the pathology. Subsequently, CT myelography of the thoracic spine demonstrated a large intradural extramedullary arachnoid cyst (or arachnoid adhesions resulting in the formation of an entrapped cystic collection). The flow voids that were seen on MR imaging were attributed to venous congestion and dilation arising from chronic compression by the cyst. Cord edema was also found to be secondary to increased interstitial pressure in the setting of this cord compression superiorly. Since surgery was not indicated for this patient at the time of diagnosis, no surgical or pathohistologic assessment was obtained to further characterize the nature of this cystic collection.

Citation: Authors_Initials. Intradural Extramedullary Arachnoid Cyst Presenting as Arteriovenous Malformation in the Thoracic Spinal Cord. *Radiology Case Reports*. [Online] 2009;4:263.

Copyright: © 2009 The Authors. This is an open-access article distributed under the terms of the Creative Commons Attribution-NonCommercial-NoDerivs 2.5 License, which permits reproduction and distribution, provided the original work is properly cited. Commercial use and derivative works are not permitted.

Abbreviations: AVM, arteriovenous malformation; CT, computed tomography; MRI, magnetic resonance imaging

Chris Conner, B.S., and Amir Paydar, M.D. (Email: apaydar@utmem.edu), are at the University of Tennessee College of Medicine, Memphis, TN, United States of America.

Published: May 31, 2009

DOI: 10.2484/rcr.v4i2.263

Case Report

A 57-year-old man presented with worsening incontinence and paraplegia of the lower extremities. The patient reported that he had been diagnosed with a spinal AVM at the T4 level 15 years previously after experiencing similar symptoms that remitted with the administration of systemic corticosteroids. No records of the patient's original diagnosis were able to be found. The patient had been free of neurological dysfunction

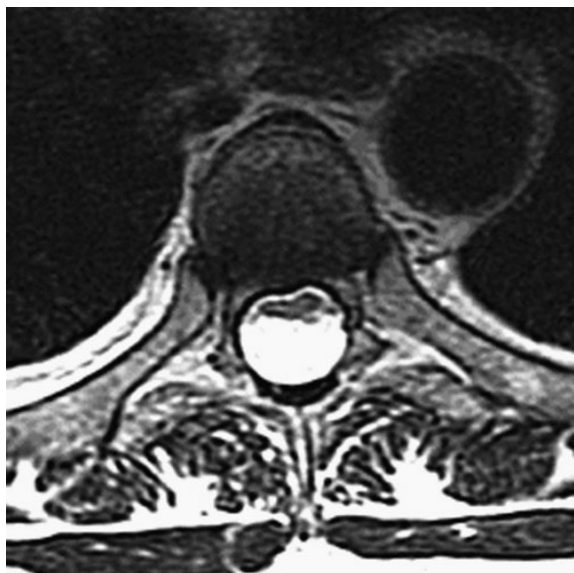


Figure 1. 57-year-old man with extramedullary intradural spinal arachnoid cyst. Axial T2-weighted MR image shows a deformed cord at the T5-T6 level where the AP cord diameter is 3mm. Comparatively, the AP cord diameter above and below this level is 6mm.



Figure 2. 57-year-old man with extramedullary intradural spinal arachnoid cyst. Sagittal T2-weighted MR image shows numerous serpiginous flow voids in the mid and lower thoracic canal (arrows). Mild central cord edema of the lower thoracic cord is also noted (arrowheads).

since the diagnosis but had experienced difficulty walking during the last year with his symptoms worsening dramatically the week prior to admission. Physical exam demonstrated increased tone in the lower extremities and significant lower extremity weakness as well as decreased sensation in the lower extremities and inguinal region.

The patient was initially evaluated at an outside institution, where spinal MR imaging was done. This study revealed intradural serpiginous flow voids typical of spinal AVMs, edema of the mid and lower thoracic cord, and anterior displacement or possible atrophy at the T5-T6 spinal level. Once the patient was transferred, interventional radiologists at our institution performed spinal arteriography to make the definitive diagnosis of spinal AVMs and possibly embolize them as a form of treatment. However, during this procedure no AVM lesions were detected in the intradural space. CT angiography was also performed, failing to show any evidence of an underlying AVM. At this point the patient underwent a second MR study without contrast enhancement and which again showed marked anterior displacement or atrophy of the cord (Fig. 1), as well as serpiginous

flow voids in the thoracic subarachnoid space and cord edema (Fig. 2). Notably, however, this follow-up MR study suggested a reduction in cord edema, possibly due to pulse corticosteroid therapy.

To further evaluate the etiology of the thoracic spinal cord compression and the signal voids seen in the subarachnoid space, a thoracic CT myelogram with multiplanar reformatting was performed after the intrathecal injection of 10mL Omnipaque-240 contrast (Fig. 3). This study showed no vascular anomalies, but did show a hypodense epidural ovoid filling defect in the thecal sac posterior to the cord at the level of maximal cord compression. The filling defect measured 11mm anteroposteriorly (AP) x 17mm transversely (RL) x 33mm superoinferiorly (SI). At this point, we made the diagnosis of an intradural extramedullary spinal arachnoid cyst (or arachnoid adhesions resulting in the formation of an entrapped cystic collection), causing secondary cord compression. Since surgery was not indicated for this patient at the time of diagnosis, no surgical or pathohistologic assessment was obtained to further characterize the nature of this cystic collection.

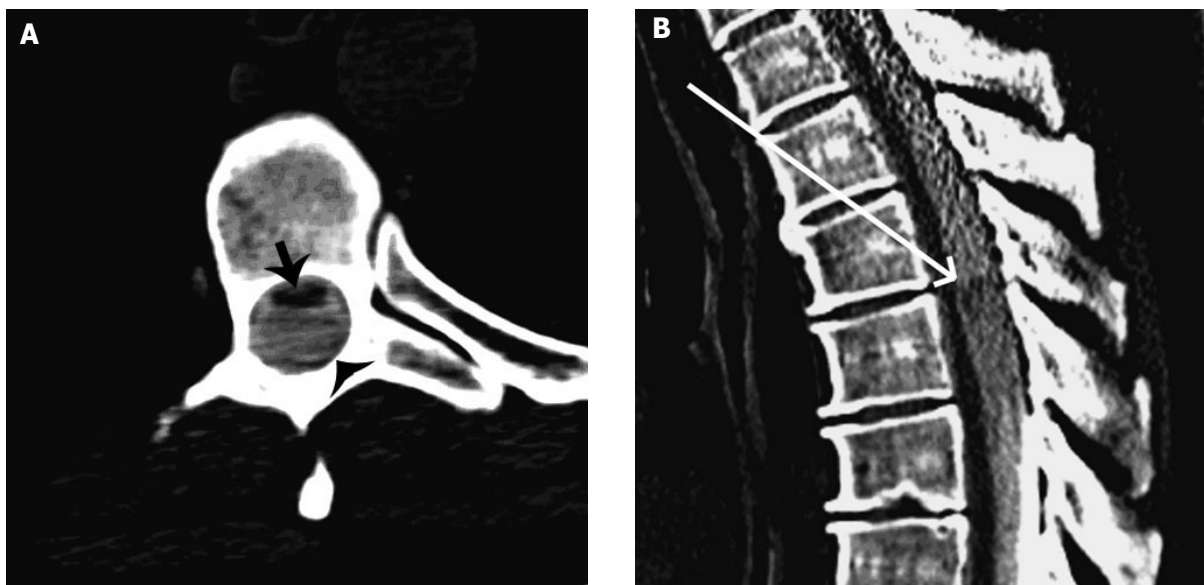


Figure 3. 57-year-old man with extramedullary intradural spinal arachnoid cyst. **A**, Axial CT myelogram post intrathecal Omnipaque-240 contrast injection shows an intermediately dense epidural ovoid filling defect (arrowhead) in the thecal sac, causing anterior displacement of a deformed thoracic spinal cord (arrow). **B**, Sagittal reformatted CT myelogram shows the same filling defect in the sagittal plane at the T6 level.

Discussion

Intradural extramedullary arachnoid cysts commonly present as incidental findings on MR imaging, with the vast majority presenting in adolescence or early adulthood [1]. These cysts are most often found at the mid-thoracic level dorsal to the spinal cord. Congenital lesions are thought to originate from the septum posticum that divides the subarachnoid space down the midline from the cervical to the lumbar region [2,3,4]. They usually develop from an enlarging diverticula with an ostia that can remain open, work as a one way valve, or completely seal the cysts contents from the surrounding CSF [2]. Cystic collections resulting from injury or an inflammatory processes, such as arachnoid adhesions, arachnoiditis, or subarachnoid hemorrhage can also present with a similar appearance on imaging [2,5,6]. Because they are often asymptomatic, treatment is often conservative with surgical intervention usually reserved for functional deficits arising from spinal cord compression [7].

Because intradural extramedullary arachnoid cysts often contain contents similar to the surrounding

cerebrospinal fluid, they can similarly have long T1 and T2 relaxation times and can appear isointense to the surrounding cerebrospinal fluid [7,8]. This can make their detection with conventional MR studies difficult, though some change in signal intensity may be seen with MR and CT studies in a minority of cases in which the cyst contains hemorrhagic or proteinaceous contents [9]. Because of this limitation, the MR studies that our patient underwent did not fully identify a distinctive mass or fluid collecting with secondary mass effect on the cord at the level of maximal compression. Although the possibility of the presence of a displacing mass was not fully ruled out, cord atrophy related to chronic myelopathy and spinal AVMs was initially considered as the most likely finding.

The diagnosis of the arachnoid cyst in this case was further complicated by an atypical presentation on imaging: concomitant presence of intradural serpiginous flow voids typical of spinal AVMs. A retrospective study of spinal AVMs in patients with unexplained myelopathy showed that while flow voids were only seen in 50 percent of MR studies with an underlying spinal AVM, the presence of flow voids was highly suggestive of an

AVM compared to other causes ($p=0.010$) [10].

Presence of cord edema in the mid and lower thoracic levels was another MR finding that needed to be addressed in this case. We initially perceived this edema as a possible sign of chronic myelopathy. Previous research by Lee and colleagues have found that chronic, progressive myelopathy can present with spinal intradural AVMs [11]. This distinct association between myelopathy and presence of AVMs also supported the idea that this presentation might have been another case of chronic myelopathy (with resultant cord edema) in the setting of spinal AVMs.

Despite these preliminary considerations, however, follow-up investigations with both conventional spinal angiography and CT angiography failed to support the diagnosis of an underlying AVM. While spinal angiography is indicated in all patients that have both unexplained myelopathy and flow voids visible on MR imaging, CT myelography should be considered in cases where angiography is negative [9,10]. Prone-supine myelography has been found to be a more sensitive imaging modality for spinal dural AVMs than spinal angiography and can often detect these lesions in cases where angiography may show no underlying pathology [9]. Therefore, CT myelography was subsequently performed to further investigate the cause of the signal voids on MR. This study conclusively confirmed the findings on angiography by showing no further evidence of an underlying AVM. In other words, a chronic lesion other than an AVM was the most probable etiology for the signal voids found on our MR images.

Concurrently, CT myelography also happened to help us determine the cause of the cord deformity visualized at the T6 level. This imaging modality revealed an intermediately dense epidural ovoid filling defect in the thecal sac posterior to the cord defect at the T6 level. The myelographic findings in this case are highly suggestive of an intradural extramedullary arachnoid cyst that may or may not have communicated with the surrounding CSF. Considering that the patient's neurologic symptoms waxed and waned throughout the past two decades, this lesion may have fluctuated slowly in size, imposing various degrees of anterior compression on the thoracic cord at different time periods.

We concluded that the signal voids seen on MR imaging were likely the result of prominent venous engorgement and hypertension in the spinal venous plexus due to chronic obstruction by the spinal arach-

noid cyst superiorly. This congestion most likely caused spinal venous plexus dilation over the span of many years, later presenting as vessels with flow voids on MRI. Furthermore, we deduced that the central cord edema in the mid and lower thoracic spine was due to chronic interstitial pressure in the intramedullary parenchyma as a result of venous outflow compression.

Acknowledgment

We would like to thank Dr. Kent Hutson (Chief of Dept. of Radiology at Erlanger Hospital in Chattanooga, TN) for introducing us to this case and assisting in the preparation of this paper.

References

1. Kumar K, Malik S, Schulte P. Symptomatic spinal arachnoid cysts: report of two cases. *Spine*. 2003 Jan 15;28(2):E25-9. [PubMed]
2. Silbergleit R, Brunberg J, Patel S, et al. Imaging of spinal intradural arachnoid cysts: MRI, myelography and CT. *Neuroradiology*. 1998 Oct;40(10):664-8. [PubMed]
3. Andrews B, Weinstein P, Rosenblum M, et al. Intradural arachnoid cysts of the spinal canal associated with intramedullary cysts. *J Neurosurg* 1988; 68(4):544-9. [PubMed]
4. Khosla A, Wippold F 2nd. CT myelography and MR imaging of extramedullary cysts of the spinal canal in adult and pediatric patients. *AJR Am J Roentgenol*. 2002 Jan;178(1):201-7. [PubMed]
5. Osenbach R, Godersky J, Traynelis V, Schelper R. Intradural extramedullary cysts of the spinal canal: clinical presentation, radiographic diagnosis, and surgical management. *Neurosurgery*. 1992 Jan;30(1):35-42. [PubMed]
6. Beall D, Googe D, Emery R, et al. Extramedullary intradural spinal tumors: a pictorial review. *Curr Probl Diagn Radiol*. 2007 Sep-Oct;36(5):185-98. [PubMed]

Intradural Extramedullary Arachnoid Cyst Presenting as Arteriovenous Malformation in Thoracic Spinal Cord

7. Secer H, Anik I, Celik E, et al. Spinal hydatid cyst mimicking arachnoid cyst on magnetic resonance imaging. *J Spinal Cord Med.* 2008 Jan; 31(1): 106-8. [PubMed]
8. Beall D, Googe D, Emery R, et al. Extramedullary intradural spinal tumors: a pictorial review. *Curr Probl Diagn Radiol.* 2007 Sep-Oct;36(5):185-98. [PubMed]
9. Gilbertson J, Miller G, Goldman M, et al. Spinal dural arteriovenous fistulas: MR and myelographic findings. *AJNR Am J Neuroradiol.* 1995 Nov-Dec;16(10):2049-57. [PubMed]
10. Strom R, Derdeyn C, Moran C, et al. Frequency of spinal arteriovenous malformations in patients with unexplained myelopathy. *Neurology.* 2006 Mar 28;66(6):928-31. [PubMed]
11. Lee J, Koyanagi I, Hida K, et al. Spinal cord edema: unusual magnetic resonance imaging findings in cervical spondylosis. *J Neurosurg.* 2003 Jul;99(1 Suppl):8-13. [PubMed]