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Adhesive arachnoiditis, subarachnoid hemorrhage, and intradural extramedullary thoracic cavernoma: illustrative case

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BACKGROUND Spinal arachnoiditis can result from various factors, including spinal subarachnoid hemorrhage (sSAH). In this paper, the authors describe a case of intradural extramedullary cavernoma with an initial presentation of subarachnoid hemorrhage leading to multilevel spinal arachnoiditis to discuss the pathophysiology and optimal treatment strategy.

OBSERVATIONS Spinal intradural extramedullary cavernoma manifesting with sSAH is a rare clinical presentation; therefore, there is no clear strategy for the management of sSAH. Spinal arachnoiditis is a result of chronic inflammation of the pia arachnoid layer due to hematomyelia. No effective treatment that interrupts this inflammatory cascade and would also prevent the development of spinal arachnoiditis has been described to date.

LESSONS Lumbar drainage could aid in sSAH management, relieve spinal cord compression, and restore the normal spinal cerebrospinal fluid circulation gradient. It could help to clear the blood degradation products rapidly and prevent early inflammatory arachnoiditis development. Mini-invasive intrathecal endoscopic adhesiolysis appears to be a reasonable approach for reducing the risk of aggravating spinal arachnoiditis with a mechanical-surgical stimulus. Whether a conservative approach should be applied in these patients with mild myelopathy symptoms is still debatable.

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KEYWORDS spinal cavernoma; arachnoiditis; subarachnoid hemorrhage; cavernous malformation; extramedullary

Spinal arachnoiditis is a rare but serious medical condition that involves inflammation of the arachnoid membrane, one of the three membranes that surround and protect the brain and spinal cord. It can occur as a result of various factors, including infection, trauma, surgery, and, in some cases, subarachnoid hemorrhage (SAH).[1] The latter refers to a bleeding that occurs in the space between the arachnoid and pia mater, two of the meninges that cover the brain and spinal cord. SAH can be caused by a ruptured aneurysm, a head or spine injury, an arteriovenous malformation, or other conditions that affect blood vessels in the brain or spine. Spinal subarachnoid hemorrhage (sSAH) accounts for less than 1% of all SAHs and is mainly caused by vascular malformations, dural arteriovenous fistulas, or intradural tumors.[2]

Spinal cavernoma is classified as a neoplastic vascular spinal lesion, because it is known to be angiographically silent.[3] It is defined as a benign vascular hamartoma composed of different thick and thin-walled sinusoid vascular channels deprived of normal neural parenchymal tissue, feeding arteries or large draining veins. Spinal cavernous malformations account for approximately 20% of intramedullary tumors,[4] and intradural extramedullary cavernomas are the least frequent in the spinal cord, with 71 cases described to date.[5] The clinical manifestation can vary from progressive to acute neurological decline, with SAH as one of the possible presentations due to superficial localization.[4,6]

In this paper we describe the case of an intradural extramedullary nerve root cavernoma with a clinical presentation of sSAH that led to adhesive arachnoiditis development and management.

Illustrative Case

A 67-year-old female with no relevant medical history presented with acute, burning, and excruciating pain in the lumbar and sacral region with pain radiating to the lower extremities lasting for 10 days. The pain was worse at night, for which the patient was taking anti-inflammatory and opioid medication, although the pain was

ABBREVIATIONS CSF = cerebrospinal fluid; Hb = hemoglobin; MRI = magnetic resonance imaging; SAH = subarachnoid hemorrhage; sSAH = spinal subarachnoid hemorrhage.

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refractory. Clinical examination did not reveal any motor or sensory deficits. Spinal magnetic resonance imaging (MRI; Fig. 1) revealed an intradural extramedullary centimetric T9 cavernoma with medullary compression and myelopathy on the same level associated with extensive SAH in the thoracolumbosacral region. MRI was repeated 3 weeks later, which showed the T9 cavernoma and partial resorption of the hemorrhage. The decision to proceed with surgery was made because of the risk of rebleeding.

A posterior minimally invasive approach was performed using an expandable tubular retractor through a right paramedian incision. Navigation was used to determine the extent of the lesion and to target the hemilaminectomy, and a crossover technique was used to expose the complete canal and the underlying dura at the level of the cavernoma. The durotomy was followed by the arachnoid plane of cleavage covered by hemosiderin. After careful microsurgical dissection, the cavernoma was found attached to the right T9 nerve root (Fig. 2), which was isolated and then divided to achieve gross-total resection. The surgical procedure was uneventful, and the patient was discharged after 3 days with relieved postoperative pain and no motor deficits.

Spinal MRI at the 3-month follow-up showed multilevel adhesive arachnoiditis from T6 to T11 with multiple arachnoid cysts. There were also flow void signals at the T6–7 and T10–11 levels with spinal cord compression and severe radiological myelopathy (Fig. 3). The patient presented with mild right radiating pain to a lower anterolateral abdominal area and the lateral thigh without any bladder or bowel symptoms and no motor or sensory deficits on the neurological examination. Given the severity of the radiological symptoms with the severe subclinical myelopathy that had progressed since surgery, we decided to treat the patient surgically with a minimally invasive two-level laminotomy and adhesiolysis using a flexible fibroscope.

A posterior T6–7 laminotomy was performed. After opening the dura, the thick and inflamed arachnoid layer with hemosiderin was opened with a scalpel and carefully dissected from the posterior surface of the spinal cord to remove traction on the spinal cord at this level and to open the subarachnoid space above and below the incision. Then, a flexible pediatric endoscope was inserted in the subarachnoid space and navigated below and above the laminotomy level, and the arachnoid cysts were opened on the posterior surface using the tip of the endoscope for blunt dissection (Fig. 4). At the T10 level, the arachnoid membrane was too thick to open it with the endoscope; above this level, the endoscope was introduced

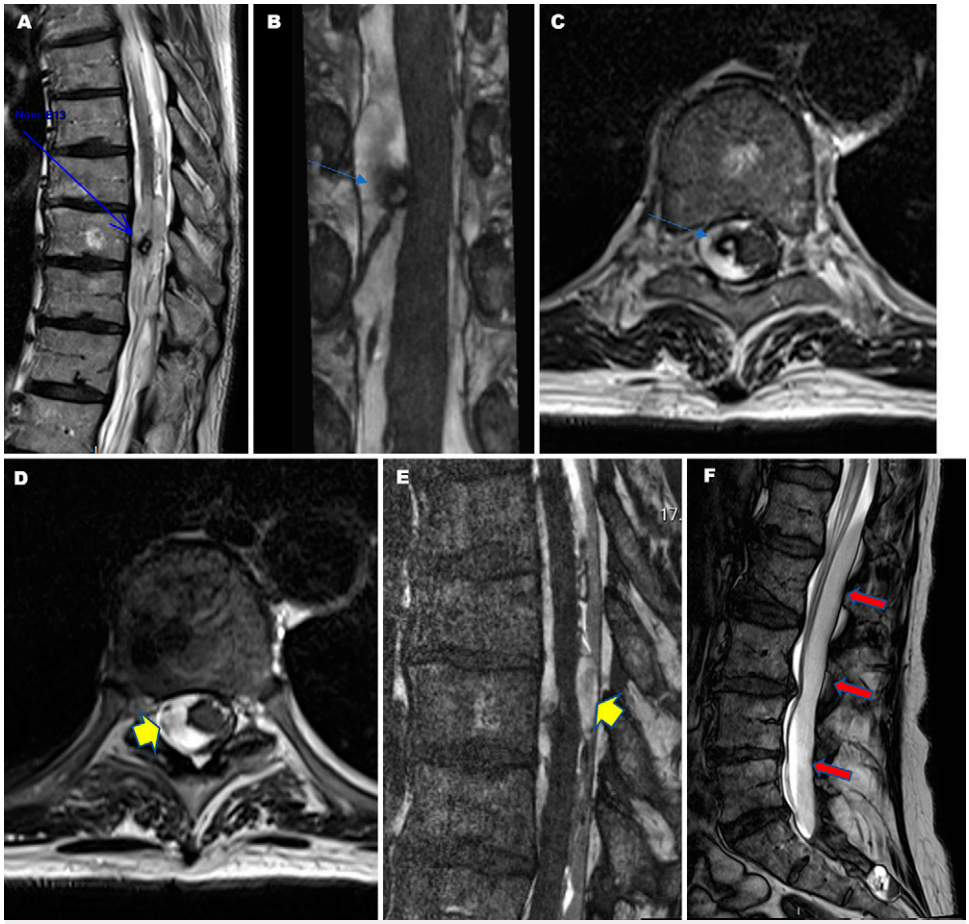

FIG. 1. Initial spinal MRI. **A:** A centimetric extramedullary cavernoma (*arrow*) at the T9 level. **B:** The cavernoma in relation to the T9 root (*blue arrow*). **C:** Axial T2-weighted imaging showing slight compression of the spinal cord on the right side. **D:** Axial MRI at the T10 level showing a thickened arachnoid pouch (*arrow*). **E:** Multiple pouches of thickened arachnoid and extensive SAH (*arrow*). **F:** Extensive blood (*arrows*) at the cul de sac on lumbar sagittal MRI.

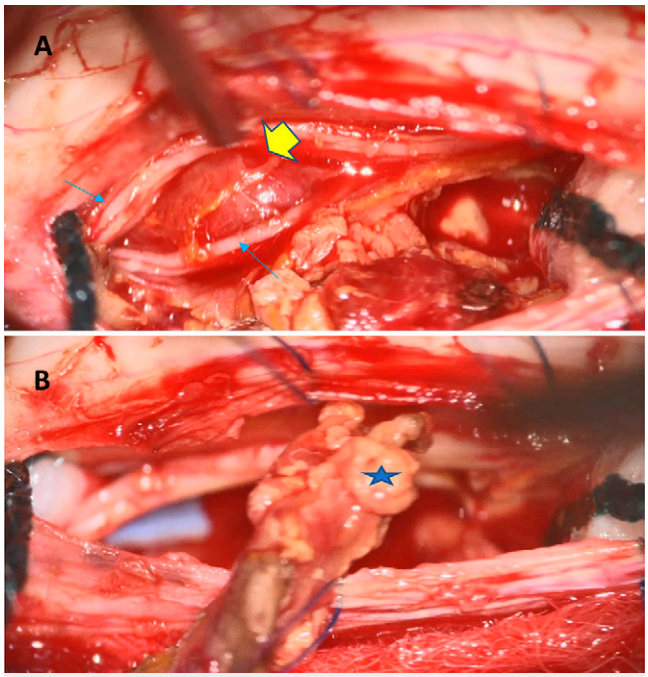

FIG. 2. Perioperative images showing a cavernoma (*yellow arrow*) attached to a splitting of the T9 root (**A**, *blue arrows*) and gross-total resection after the cavernoma was removed (**B**, *star*).

up to the T4 level. A ventricular drain was inserted above and below the laminotomy region and attached to the dura mater. The second lower incision was avoided to refrain from aggravating the arachnoiditis by mechanical factors.

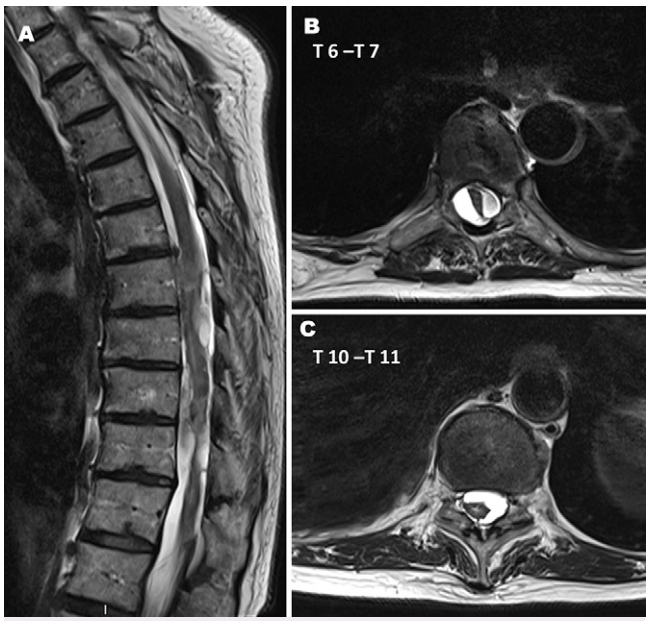

FIG. 3. Postoperative MRI (**A**) at 3 months showing multilevel adhesive arachnoiditis from T6 to T11 with multiple arachnoid cysts. Flow void signal at the T6–7 level in the axial plane (**B**) and the T10–11 level (**C**) with severe radiological T2 changes.

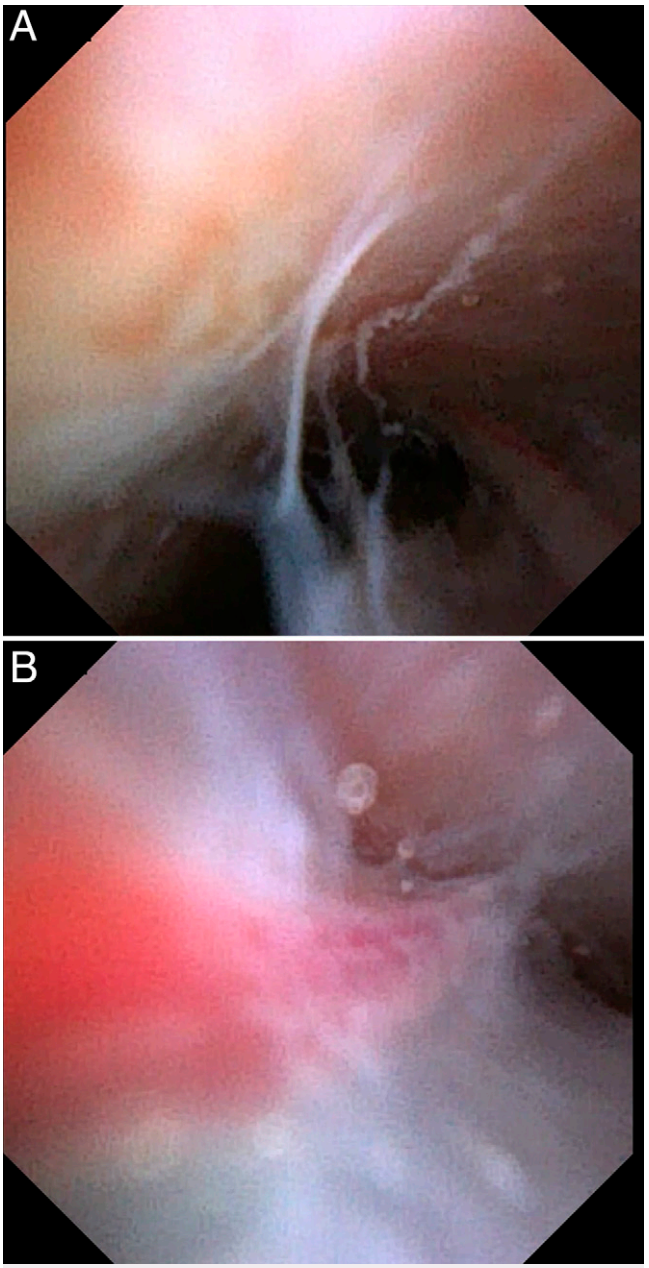

FIG. 4. Endoscopic views of blunt dissection of the arachnoid cysts with a flexible pediatric endoscope. **A:** Dorsal view of the spinal cord and severe arachnoiditis, levels T9–10. **B:** Cranial view of the ventral dura and spinal cord severe arachnoiditis.

Immediate postoperative MRI showed regression of the multiple cysts and discrete regression of the myelopathy. The patient no longer reported having any radiating pain and was discharged without any postoperative wound complications. Eight months after surgery, she presented with a favorable evolution; however, the long-term follow-up will be essential to judge the efficacy of the surgery.

Patient Informed Consent

The necessary patient informed consent was obtained in this study.

Discussion

Observations

In this paper, we present a case of thoracic spinal root cavernoma with an initial presentation of acute SAH lasting for 10 days, which led to severe arachnoiditis with mild radiological symptoms.

Spinal cavernomas are a rare entity that accounts for up to 20% of all intramedullary spinal cord tumors. Only approximately 3% of spinal cavernomas are intradural, and an intradural extramedullary presentation is the least frequent.[4] Under the electron microscope, abnormal gapping among tight endothelial junctions has been observed, which may permit the leakage of blood.[7] The data on the risk of hemorrhage of a spinal cavernoma are limited because of the differences in the definition of cavernoma hemorrhage among studies, even though the annual rate is estimated to be approximately 2.1%.[8]

The literature reports that the cumulative risk of a first bleed over a 5-year period of time for patients with spinal cavernous malformation is up to 40%, twofold higher than the rate in the cranial cavernoma patient group, which can increase to 55% after the first bleed.[9,10] Santos et al.[11] recently published a cross-sectional study in which they calculated a cumulative risk of 67% for a third hemorrhage over 5 years for spinal cavernous malformations. The indication for surgical treatment for spinal cavernomas is based on a higher rebleeding risk, complex anatomy of the region leading to higher hemorrhage or surgical treatment morbidity, and a relatively small space quickly leading to compression of neural structures. These factors lead to more aggressive treatment in mildly symptomatic patients, as in brainstem cavernoma.[12]

SAH is a known chemical factor associated with the development of adhesive arachnoiditis due to the inflammatory cascade. First, there is an acute inflammation leading to fibrinous exudate that can manifest as radical symptoms. During the adhesive phase, the cerebrospinal fluid (CSF) carries cytokines and fibrinolytic enzymes, and because of the avascular nature of the arachnoid layer, the healing process is impaired, leading to the formation of fibrinous bands and later to the arachnoid layer scarring. Arachnoid trabeculae and cyst formation disrupt normal CSF circulation, which no longer supplies the spinal cord with the necessary nutrients. Moreover, because CSF clearance from toxic waste is also diminished, leptomeninges are longer exposed to hemoglobin (Hb) degradation products (chemical trigger), and the source for ongoing inflammation is maintained. Later, atrophy of the involved nerve roots or spinal cord is observed. The impaired CSF circulation may create a pressure gradient over the obstructed area, leading to interstitial movement throughout the spinal parenchyma toward the center and the development of syringomyelia.[1]

There is no clear management strategy in cases of SAH to avoid complications such as adhesive arachnoiditis formation. Historically, conservative management was suggested in asymptomatic patients, and surgical evacuation of hematoma was commonly performed in cases of symptomatic spinal cord compression.[13] Ichiba et al.[13] described their own 5 cases of spontaneous subarachnoid hemorrhage and 19 from the literature; in that series, 1 patient was treated with lumbar drainage. It was speculated that in these situations, it might help to restore the normal CSF circulation by decompressing the spinal cord as well as by evacuating the hematoma gradually over days, as there could be a recurrent hemorrhage.[13]

There could be another explanation, as in vasospasm prevention in aneurysmal SAH or the prevention of delayed complications such as hydrocephalus, which is also believed to be initially caused by the inflammatory cascade caused by hemoglobin degradation products.[14] It has been shown that lumbar drainage in cases of aneurysmal SAH creates a ventriculolumbar gradient, improves CSF clearance from toxic Hb degradation products, and restores normal CSF flow.[15]

Finally, multiple-level adhesive arachnoiditis with multiple arachnoid cysts remains a surgical challenge, as it is associated with a worse prognosis than focal cysts because of the higher risk of recurrence due to postoperative scarring and retethering.[16] The treatment of choice is one- to two-level laminectomy with intrathecal endoscopic adhesiolysis, which allows lysis of subarachnoid adhesions and cyst fenestration below and above the level of laminectomy. As better surgical outcomes are inversely related to the number of levels explored, this minimally invasive endoscopic technique allows exploration beyond the laminectomy level.[17]

Lessons

The pathophysiological mechanism underlying SAH-induced spinal arachnoiditis appears to be the consequence of a long-lasting chronic inflammatory response of the pia arachnoid membrane induced by hematomyelia, although it is not yet fully understood.

In this illustrative case of SAH associated with a cavernous malformation, initial treatment with lumbar drainage could have been useful to rapidly remove the hematoma so it reduced the natural resolution of hematoma time, and it may have avoided adhesions within arachnoid layer formation.

Spinal arachnoiditis can manifest clinically through meningeal thickening, adhesions with cord deformity, meningeal contrast enhancement, arachnoid cyst, and syrinx formation. It is debatable whether to adopt a conservative approach in mildly symptomatic patients. Endoscopic arachnoid cyst adhesiolysis seems to be a reasonable treatment option because of its minimally invasive nature.

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Disclosures

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

Conception and design: Barges-Coll, Andriuskeviciute, Mondragón-Soto. Acquisition of data: Barges-Coll, Mondragón-Soto, Penet. Analysis and interpretation of data: Barges-Coll. Drafting the article: Barges-Coll, Andriuskeviciute, Mondragón-Soto. Critically revising the article: Barges-Coll, Andriuskeviciute. Reviewed submitted version of manuscript: Barges-Coll, Mondragón-Soto. Approved the final version of the manuscript on behalf of all authors: Barges-Coll. Study supervision: Barges-Coll.

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