Spinal Venous Vascular Ectasia With Unusual presentation of Abdominal Pain: Case Report

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BACKGROUND AND IMPORTANCE: Spinal venous vascular ectasia associated with a low-flow fistula of paravertebral location is a very low-frequency entity. It usually manifests with myelopathy; however, in the present case, the symptomatology corresponded to thoracic radiculopathy, which can be difficult to diagnose and may be the cause of chronic pain refractory to analgesic management.

CLINICAL PRESENTATION: An adult patient who consulted about a 1 year and a half of flank pain refractory to analgesic management and hypoesthesia on palpation in the left T8 dermatome, therefore, pain of radicular origin was suspected. MRI of the thoracic spine was requested, showing an image suggestive of vascular malformation at the level of T8. After identification of the lesion, the patient was taken to spinal angiography to delimit the vascular malformation, finding an image suggestive of spinal venous vascular ectasia associated with a low-flow fistula of paravertebral location at T8. Subsequently, the case was discussed by a multidisciplinary team that established endovascular embolization as the best option for treatment. Thus, achieving complete occlusion of the lesion without complications and a slow improvement of the symptoms.

CONCLUSION: Spinal venous vascular ectasia associated with a low-flow fistula of paravertebral location is a very low-frequency entity that despite manifesting with myelopathy in some cases may generate radicular irritative symptoms and is differentiated from other lesions by its unusual location in spinal angiography.

KEY WORDS: Spinal venous vascular ectasia, Abdominal pain, Spinal cord, Varices

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Spinal venous vascular ectasia (SVE) is a very low-frequency pathology that is usually related to neurological deficits such as hemiplegia, dysesthesia, and dysarthria.[1] As with any other local irritative syndrome, SVE can be related to myelopathy and radiculopathy of the segment involved. Thus, venous ectasia can cause compression and lead to compressive spinal cord syndrome.[2] It is not frequent that radicular compression leads to abdominal pain; however, it is possible whether the involved roots correspond to thoracic nerve roots.[3]

Cases of SVE are generally described in relation to an arteriovenous malformation or a fistula of spinal cord location which frequently manifests with myelopathy but rarely presents at paravertebral level and with radicular symptoms. It is considered that its symptomatology may be the result of irritation of an adjacent nerve root secondary to compression by venous ectasia.[3] In many cases, the lack of knowledge of SVE causes it to be underdiagnosed and a definitive diagnosis is often not reached.[4]

Understanding the diagnostic challenge that this pathology represents and that abdominal pain as a representation of radicular irritation is infrequent.[4] Thus, in this case, the report we describe is a case of an adult patient who presented with abdominal pain refractory to analgesic management with the unclear onset of initial symptoms of spinal venous vascular ectasia.

ABBREVIATIONS: AVM, arteriovenous malformations; **SVE,** spinal venous vascular ectasia.

CLINICAL PRESENTATION

The case refers to an adult patient who consulted with a clinical symptom of approximately 1 year and a half of evolution consisting of abdominal pain in the left upper quadrant of insidious

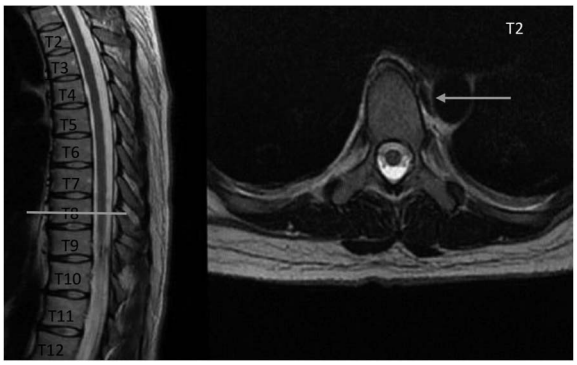

FIGURE 1. *MRI of the thoracic spine in T2. Suspicious image of left perimedullary vascular malformation at the height of T8.*

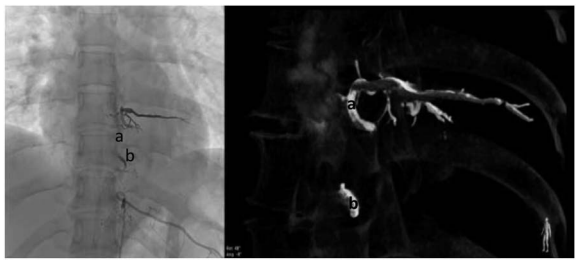

FIGURE 3. *Postembolization spinal arteriography. a: Occlusion with embolic fluid in the segmental artery. b: The use of coils to avoid the phenomenon of "reflux" of embolic material into unwanted segments of the spinal cord.*

character that was refractory to analgesic management. Initially, important pathologies such as endometriosis, porphyria, and mesenteric thrombosis were ruled out. On physical examination, the patient presented with pain of radiculopathy characteristics and hypoesthesia of the left T8 dermatome. The patient was evaluated by neurosurgery who took MRI of the thoracic spine where a suspicious image of vascular malformation at the level of T8 was identified (Figure 1). Initially, analgesic management was indicated to modulate pain, and a spinal arteriography was performed where the SVE was evidenced (Figure 2) (Video). In the spinal arteriography, it was determined that it corresponded to an SVE associated with a low-flow fistula given its paravertebral location and differed from other lesions because it was not properly located at the level of the spinal cord.

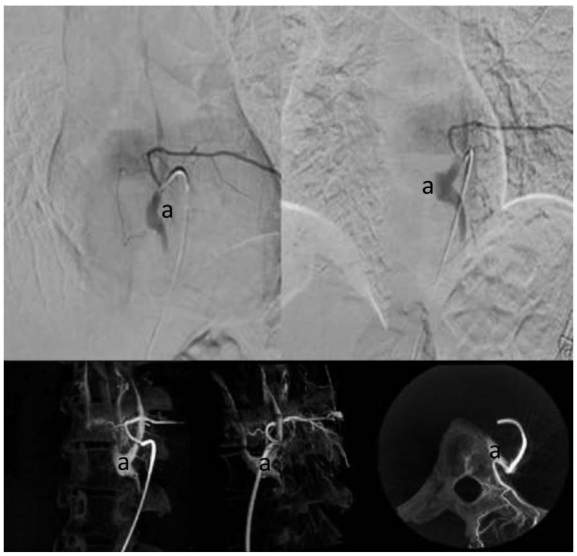

FIGURE 2. *Spinal arteriography. a: Spinal venous vascular ectasia at the level of left T8.*

The endovascular embolization of the anterolateral isolated SVE of T8 was the course of action decided in consensus, and the patient consented to the procedure. The procedure was performed where the lesion was occluded with embolic fluid without complications (Figure 3), and management of the pain was established with analgesia. Nine days after the postoperative period, the patient was discharged. At the 3-month postoperative follow-up, the patient denied the presence of any symptoms.

DISCUSSION

The SVE is a very low-frequency entity with an estimated annual incidence of 1/100 000 and prevalence of around 10/100 000 people.[5] The SVE is related to arteriovenous malformations (AVMs) or fistula and are generally asymptomatic; however, they can generate diverse symptomatology ranging from lumbar symptoms to gastrointestinal or bladder manifestations that are the result of venous hypertension which causes compressive symptoms in the compromised segment.[6]

The most frequent location of spinal AVMs is at the thoracic level and adjacent to the spinal canal; however, in the presented case, the lesion is located at the paravertebral level.[7] Anatomically, the lower thoracic region of the spinal cord is more susceptible to venous congestion because of the smaller number of venous outflow channels.[8]

The diagnosis can be made through clinical evaluation and image findings where computed tomography and MRI can be useful to identify the lesion; however, the gold standard for the diagnosis of SVE is spinal angiography because it allows to characterize it and determines the limits and anatomic location.[9] In spinal angiography, the SVE is evidenced in relation to a arteriovenous malformation or fistula feeding; the lesion generally has a closer relationship with the spinal cord and can be seen as a wide vascular bed; however, in the present case, the lesion manifested in an atypical location in the paravertebral region causing irritation of the adjacent nerve root.[10]

TABLE. Main Vascular Abnormalities of the Spinal Cord

Type of vascular abnormality	Description	
Cavernous Malformation	• Do not involve a shunt. • They are small, generally have low blood flow, and are supplied by sinusoidal blood vessels. • They are more frequent in the brain.	
Intradural arteriovenous malformation	• They are usually high-flow lesions. • May be intramedullary or have partial extramedullary components. • Usually manifests with subarachnoid or intramedullary hemorrhage causing back pain. • Glomus arteriovenous malformations are usually located in the anterior section of the spinal cord. • Juvenile arteriovenous malformations are usually high-flow abnormalities.	
Perimedullary arteriovenous fistulas	• They are generally located in the thoracic or lumbar spine. • The vein distal to the fistula is usually a venous varix.	Type I are low-flow fistulas frequently located in the conus or filum terminale and are supplied by an anterior spinal artery.
		Type II are medium or high-flow fistulas and may be supplied by anterior or posterior spinal arteries.
		Type III are high-flow fistulas.
Dural arteriovenous fistula	• These are low-flow abnormalities that are supplied by a dural branch of the intervertebral artery. • They are acquired abnormalities that can lead to spinal cord ischemia and progressive myelopathy.	

Elaborated from "A Review of Vascular Abnormalities of the Spine."[11]

At the spinal level, mainly the vascular abnormalities presented in Table can be considered.[11] In this way, the lesion presented corresponding to an SVE is best described in relation to a fistula. In the same sense, according to the classification proposed by Merland[12] where fistulas are classified depending on the flow, it can be considered that the lesion corresponds to an SVE associated with a low-flow fistula of paravertebral location; however, the classification of the lesion presented is not very clear given its atypical location.

Currently, the treatment of AVMs is based on reducing symptoms and the risk of bleeding. Therefore, treatment is determined by the location, size, and blood flow to the spine.[10] In general, all patients should have sufficient images to delimit the lesion such as MRI and angiography. Finally, the treatment route should be reached in consensus by a multidisciplinary team.[3]

Initially, the treatment can be medical, seeking to reduce the symptoms and lower back pain; however, in refractory cases, the interventions should be the elected choice of treatment. Among the interventions to choose from are the surgical approach, endovascular embolization, and radiosurgery.[13] Conventional surgery is preferred in cases where the lesion is accessible and easy to resect, whereas endovascular embolization and radiosurgery represent a minimally invasive elective approach in cases where it is desired to reduce the risk of bleeding and partial or complete postoperative neurological deficit.[14,15]

In the case of surgical approaches, given the extent of the vascular anomalies and the wide arterial and venous bed, surgical ligation can be more complex with a high risk of recurrence if all the vessels supplying the vascular abnormality are not ligated; thus, surgical ligation is appropriate in low-flow fistulas that are easily accessible surgically. On the other hand, embolization allows to be more selective regarding the occluded vessels, and the application of coils reduces the "reflux" phenomenon that may occur in adjacent segments, thus ensuring that the embolic material is located in the desired section.[11]

Thus, the case presented highlights the atypical symptomatology of abdominal pain and the presence of a paravertebral SVE as a cause of radicular irritation syndrome where the thoracic MRI and spinal angiography made it possible to characterize the lesion and differentiate it from other lesions such as AVM. Finally, endovascular embolization was chosen due to the unusual location and the risk of bleeding.

CONCLUSION

The SVE is a very low-frequency entity that if not recognized in a timely manner can be the cause of chronic pain refractory to medication and can seriously affect the patient's quality of life. This pathology can manifest as lower back pain or can generate atypical symptoms such as abdominal pain in addition to neurological symptoms such as hypoesthesia along a dermatome. Therefore, it is important to characterize this entity to guide the diagnosis and treatment in the future.

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VIDEO. Spinal arteriography shows a dilatation of a left paravertebral venous vessel at the T8 level possibly associated with a low-flow fistula located at the paravertebral level, an atypical location for this type of lesion. The SVE can be seen in the second 2 of the video in the midarterial phase.

COMMENTS

This case report overviews the rare entity of a spinal venous vascular ectasia presenting as abdominal pain. MRI revealed a T8 spinal vascular malformation, which was further characterized by spinal angiography. The lesion was in the paravertebral region and caused irritation to the adjacent nerve root. It was a low-flow fistulous lesion.

In the setting of the physical findings and initial MRI, spinal angiography was the correct diagnostic test and allowed identification of a lesion for embolization. Embolization was successful in alleviating symptoms. A wide range of embolic materials are available and due to the rarity of the lesion type, a generalized consensus has not been established. Because the lesion acts like a fistula, blocking off the fistulous connection should be the goal of treatment.

The authors elected for an endovascular approach, which is reasonable given the location. As most SVEs appear in the thoracic region, endovascular treatment is more common than open surgical ligation. It is imperative to identify the site of fistulous connection for successful treatment similar to treating a dAVF.

SVEs warrant further investigation regarding the underlying etiology, associated risk factors, and necessity for long-term follow up. Further guidelines for management can be established once the etiology is better characterized. The current study offers important teaching points and highlights a unique case. It can serve as a catalyst for discovery regarding follow up investigations.

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