Rupture of a Spinal Dural Arteriovenous Fistula as a Differential Diagnosis of a Coronary Syndrome: Case Report

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Received, March 15, 2023; **Accepted,** May 05, 2023; **Published Online,** August 11, 2023.

BACKGROUND AND IMPORTANCE: Spinal dural arteriovenous fistulas (SDAVFs) are the most common vascular malformation of the spine and can lead to progressive paraplegia if left untreated. However, because of their nonspecific clinical presentation, they are often misdiagnosed as other pathologies, such as discopathies or degenerative neuropathies, which can result in delayed diagnosis and treatment.

CLINICAL PRESENTATION: A case of a 73-year-old female with a history of acute coronary syndrome who presented to the emergency department with sudden onset chest pain suggestive of an acute myocardial infarction is presented. Further evaluation revealed a subdural hematoma at T2-T5 and T8-L5, caused by a ruptured SDAVF at the T12-L1 level. The patient required emergency decompression surgery, but because of late diagnosis, she suffered a spinal cord injury with an ASIA-A classification.

CONCLUSION: SDAVF is a disease with nonspecific initial symptoms, which can easily be mistaken for other pathologies. However, early recognition of the presence of the fistula, especially in cases of rupture, can improve prognosis and increase the chance of better outcomes. It is important to keep this condition in mind when evaluating patients with unexplained neurological symptoms and consider SDAVF as a differential diagnosis of acute coronary syndrome.

KEY WORDS: Spinal dural arteriovenous fistula (SDAVF), Chest pain and coronary syndrome

Neurosurgery Practice 2023;4(3):e00050. https://doi.org/10.1227/neuprac.0000000000000050

Spinal dural arteriovenous fistulas (SDAVFs) are the most common type of spinal vascular disease, comprising an estimated 70%–80% of all spinal vascular malformations. The incidence of SDAVF is relatively rare, with approximately 5–10 cases per million inhabitants.[1]

SDAVFs predominantly affect men, with approximately 80% of cases occurring in males, and the mean age of diagnosis is typically between 50 and 60 years. The etiology of SDAVF is not yet fully understood, and although it is believed to be an acquired pathology, the exact cause remains unclear.[2,3]

SDAVF is characterized by nonspecific initial symptoms, including venous congestion and progressive neurological symptoms, such as sensory or gait disturbances.[4] Wang et al[5] conducted a retrospective review of 326 patients and found that 265 cases (81.3%) were misdiagnosed.

This case report highlights the importance of timely and accurate diagnosis of SDAVF in patients presenting with unexplained neurological symptoms. The patient, initially misdiagnosed with acute myocardial infarction, was found to have a ruptured SDAVF resulting in ASIA-A spinal cord injury. Timely recognition of SDAVF can improve prognosis and quality of life of affected patients.

ABBREVIATIONS: SDAVF, spinal dural arteriovenous fistula.

CLINICAL PRESENTATION

A 73-year-old female patient with a medical history of dyslipidemia and coronary artery disease presented to the emergency department with sudden onset of severe, oppressive chest pain radiating to the neck and left shoulder. The patient also reported associated symptoms of diaphoresis, several episodes of emesis,

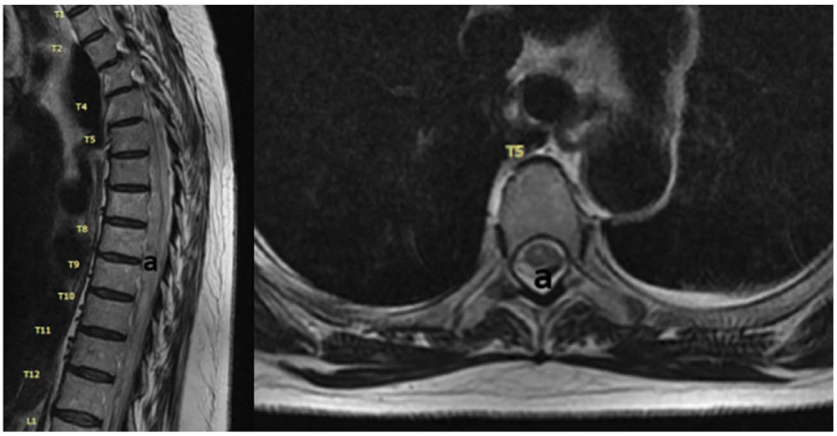

FIGURE 1. *Thoracic spine MRI in T2 sequence in the sagittal and axial view showing extensive subdural hematoma at T2-T5 and T8-L5. a: Spinal subdural hematoma.*

liquid stools, and headache. The patient reported partial symptom resolution after taking two aspirins. Physical examination showed tachypnea and hypertension, but no other significant findings.

An electrocardiogram showed bifascicular block without ST segment elevation, and laboratory tests revealed a positive percentage delta troponin, albeit with very low absolute values. A chest X-ray was unremarkable. Based on the positive delta troponin, the patient was treated with dual analgesia and triple therapy, including acetylsalicylic acid, ticagrelor, and fondaparinux.

Further investigations, including transthoracic echocardiography and computed tomography angiography, were performed by the cardiology department but did not reveal significant findings. However, the patient's condition suddenly deteriorated, with the onset of lower back pain, motor deterioration, hyporeflexia, and loss of sphincter control during hospitalization. An immediate MRI of the thoracic and lumbosacral spine revealed a spinal subdural hematoma at T2-T5 and T8-L5 levels (Figures 1 and 2), leading to the discontinuation of anticoagulant therapy and consultation with the neurosurgery department. With the patient's informed consent, an emergency decompression was performed through wide hemilaminectomies from T2 to T5 and from T8 to L5, along with duroplasty using autologous grafts without complications.

Once the patient's condition stabilized, spinal angiography was performed to investigate whether there was an underlying pathology that might have caused the bleeding. The spinal angiography revealed an arteriovenous malformation at the T12-L1 level (Video 1) (Figure 3), which was the cause of the subdural bleeding. Several hours later, an attempt was made to embolize the lesion using interventional radiology, but the SDAVF was not visualized (Video 2) (Figure 3), and the procedure was, therefore, not performed, suggesting resolution of the lesion. Despite therapeutic efforts, the patient continued to have paraplegia and was therefore diagnosed with a spinal cord injury categorized as ASIA-A. The patient received 2 days of rehabilitation therapy before discharge.

Follow-up at 10 days, 1 month, and 3 months showed no clinical improvement.

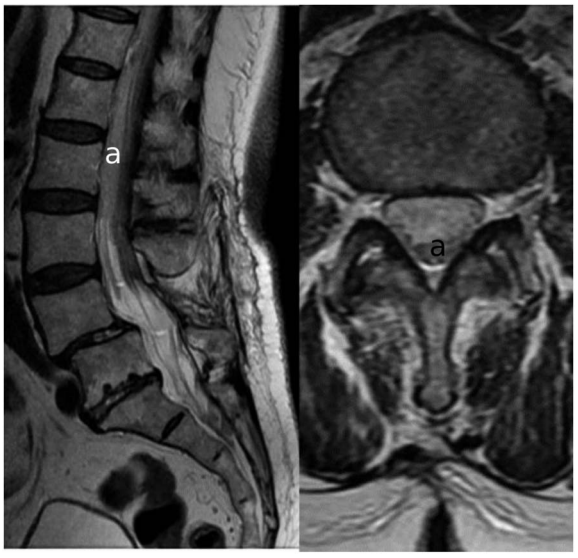

FIGURE 2. *Lumbosacral spine MRI in T2 sequence in sagittal and axial view showing extensive subdural hematoma at T8-L5. a: Spinal subdural hematoma.*

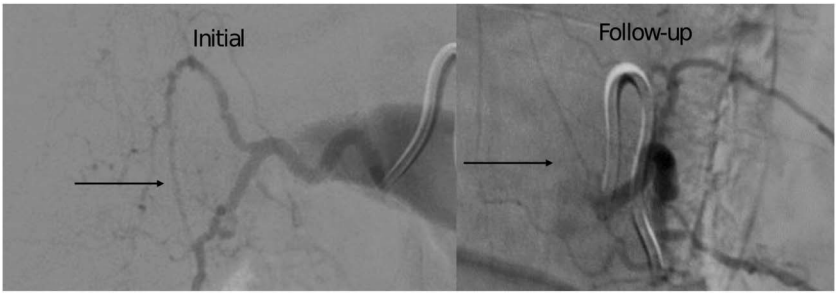

FIGURE 3. *Spinal angiography. A comparison between the initial spinal angiography and the most recent angiography reveals that the lesion is no longer detectable. Vascular malformation.*

DISCUSSION

It is important to note that SDAVFs, despite being the most common vascular pathology affecting the spinal cord, are often difficult to diagnose because of their nonspecific symptoms.[6]

The etiology of SDAVF remains uncertain although traumatic and infectious causes are commonly reported.[7] In the present case, it is likely that anticoagulant therapy facilitated the expansion of the subdural hematoma secondary to SDAVF. Consequently, the initial misdiagnosis might have contributed to the sudden onset of the subdural hematoma.

The most frequently observed symptoms of SDAVF are paraparesis (found in 95% of cases), lower limb sensory alterations, and loss of sphincter control.[8]

The atypical presentation of chest pain observed in this case is likely attributable to the subdural hematoma's proximity to the thoracic nerve roots, resulting in irritation and referred pain to the thoracic region. Moreover, the fact that this is an older patient may imply challenges in characterizing the type of pain, given that alterations in pain threshold are commonly observed in this patient population.[9]

SDAVF can be initially evaluated with a spinal MRI, which typically reveals a high–signal intensity image with a swollen spinal cord in T2 sequences. Lee et al[10] have reported that a perimedullary flow void on a T2-weighted image has a sensitivity of 95% for SDAVF. However, spinal angiography remains the gold standard for diagnosis because it allows for the precise identification of the location of the fistula.[11]

When considering the optimal treatment for SDAVF, meta-analyses like that in the study by Goyal et al[10] have shown that surgical ligation provides superior outcomes compared with endovascular treatment, despite the latter being less invasive. Endovascular treatment is associated with a higher incidence of initial failure and recurrence of the lesion.[6]

The prognosis of SDAVF depends on the timing of diagnosis. An early and accurate diagnosis can prevent secondary complications such as rupture and subsequent hemorrhage, which can cause irreversible spinal cord injury.[12]

In the literature, there are some reported cases of SDAVF with the presentation of chest pain, but these cases typically involve long-standing lancinating pain, which is not easily confused with a coronary syndrome.[13] In the case presented, the initial diagnosis was an acute coronary syndrome and it was managed accordingly. However, a more thorough physical examination led to a diagnostic hypothesis of radicular syndrome, which was confirmed by MRI and characterized by spinal angiography. Unfortunately, because of the time elapsed between diagnosis and treatment, the patient did not achieve a recovery of spinal cord function.

CONCLUSION

SDAVF presents a diagnostic challenge because of its nonspecific symptoms that can lead to spinal cord injury. It is crucial for physicians to consider SDAVF as a possible differential diagnosis in patients with unexplained neurological symptoms, even in those with acute coronary syndrome, and perform a prompt and accurate diagnosis to enhance prognosis and recovery.

Funding

This study did not receive any funding or financial support.

Disclosures

The authors have no personal, financial, or institutional interest in any of the drugs, materials, or devices described in this article.

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VIDEO 1. Spinal angiography. At the level of left T12-L1, Adamkiewicz artery originating from the radiculomeningeal branch of L1 is observed. In the radiculomeningeal artery of T12, there is a suspicious image of an epidural fistula with drainage to right paravertebral veins, which could correspond to a bleeding site.
VIDEO 2. Spinal angiography. The origin of the intercostal artery that gives rise to the branches of L1 and T12 on the left side, lumbar and intercostal, respectively, is catheterized. Adamkiewicz artery is observed from the meningeal radicular artery of L1. No alterations were observed in the T12 meningeal radicular artery. A new injection is performed with oblique projection, without evidence of epidural or dural fistula in the previous study, and therefore, embolization is not performed.