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A patient with spinal cavernous vascular malformation: case report and review of the literature

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Abstract. – BACKGROUND: Spinal cavernous vascular malformation (SCM) is a rare type of spinal vascular malformation that can be easily misdiagnosed and overlooked, accounting for 5%-12% of all spinal vascular malformations. To date, surgical resection has been the gold standard for treating SCM, particularly in symptomatic patients. The risk of secondary hemorrhage in SCM is as high as 66%. Therefore, early, timely, and accurate diagnosis is crucial for patients with SCM.

CASE REPORT: In this report, we describe a 50-year-old female patient who was admitted to the hospital with recurrent bilateral lower extremity pain and numbness for 10 years, with recurring symptoms for 4 months. The patient's symptoms initially improved after conservative treatment but then worsened again. An MRI revealed a spinal cord hemorrhage, and after surgical treatment, the patient's symptoms improved significantly. A postoperative pathological examination confirmed the diagnosis of SCM.

CONCLUSIONS: This case, along with a review of the literature, suggests that for SCM, early surgery using techniques such as microsurgery and intraoperative evoked potential monitoring may result in better outcomes for the patient.

Key Words:
Spinal cavernous malformation, Initial modified Aminoff and Logue's scales, Surgical, Conservative, Case report.

Introduction

Spinal cavernous malformation (SCM) is a type of spinal vascular malformation that constitutes 5%-12% of all spinal vascular malformations[1]. Although the incidence of SCM is low, the advent of magnetic resonance imaging (MRI) has significantly increased the detection rate of spinal cavernous malformations[2]. The characteristic imaging changes of SCM are "popcorn"-like mixed signals surrounded by low signals due to iron-containing hematoxylin deposits[3]. While SCM may be asymptomatic, symptoms often arise due to bleeding from vascular malformations[4], which can result in recurrent and progressive neurological deficits such as decreased muscle strength, hypesthesia, pain, bowel and urinary dysfunction, and respiratory distress. Surgical resection has been the gold standard for treating SCM[1,5], particularly in symptomatic patients. According to Sandalcioglu et al[6], the risk of secondary hemorrhage in spinal cord cavernous vascular malformations is as high as 66%. Therefore, early, timely, and accurate diagnosis is crucial for patients with SCM.

Case Presentation

The patient is a 50-year-old woman who has presented with pain and numbness in both lower extremities for the past 10 years, with recurrent symptoms for the past 4 months. Four months ago, the patient was seen at another hospital for pain and numbness in both lower extremities. Further MRI examination of the thoracic and lumbar spine revealed short T1 and short T2 signals in the T10-12 plane of the spinal cord with no significant enhancement, suggesting the possibility of vascular malformation with hemorrhage. The patient was treated conservatively with medication, but the exact protocol of the medicine was unknown. After treatment, the

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patient reported relief of pain and numbness compared to before. Seven days before admission, the pain and numbness in both lower extremities worsened, especially on the right side. The patient took her own oral medication but did not experience significant improvement. She had no previous medical history and no family history of hereditary disease. On admission, physical examination showed normal muscle strength and muscle tone of the extremities, decreased deep sensation in both lower extremities, especially in the right lower extremity, decreased pain and temperature sensation in both lower extremities, especially in the left side, and an II° burn wound in the left lower leg in front of the shin. Pathological signs were negative, and Babinski's sign was suspiciously positive. The initial modified Aminoff and Logue's scales (ALS)[7,8] for gait, urination, and defecation were 2 (limited motor tolerance), 0 (normal), and 0 (normal), respectively. After admission, an MRI of the thoracic and lumbar spine was conducted, and it revealed a band of low signal in the spinal cord in the T10-12 plane with localized punctate T2-weighted (T2WI) high signal. This did not show significant enhancement after contrast enhancement, which led us to consider the possibility of vascular malformation with possible bleeding (Figure 1A). A spinal angiography was conducted, but it did not show any obvious spinal cord malformation vessels. Given the patient's worsening symptoms, we decided to proceed with surgical treatment with the aim of safely resecting the vascular malformation.

To ensure the safety and success of the procedure, we utilized skilled microscopic techniques, intraoperative somatosensory evoked potential monitoring throughout the surgery, and intraoperative ultrasound backup. During the operation, we observed a localized elevation and dark redness of the spinal cord surface in the posterior aspect of the spinal cord in the T10-11 canal plane (Figures 2A-B). There was no significant deterioration of the patient compared to the pre-operative period (Figure 1B). However, the patient experienced a temporary decrease in muscle strength in both lower limbs, with a grade of approximately 2. There was also a loss of deep sensation and occasionally urinary retention, with ALS of 5 (wheelchair required), 2 (occasional urinary incontinence or retention), and 0 (normal) for gait, urine, and defecation, respectively. A repeat MRI of the thoracic spine showed significant spinal cord edema and fluid accumulation in the operative area (Figure 1C). We determined that the spinal cord edema was a result of the surgical operation and proceeded to provide edema relief and rehabilitation treatment. At the time of discharge, the patient's muscle strength had returned to level 4, the deep sensation had returned, and urination was normal. The ASL was 3 (needing some support), 0 (normal), and 0 (normal) for gait, urination, and defecation, respectively. When the patient

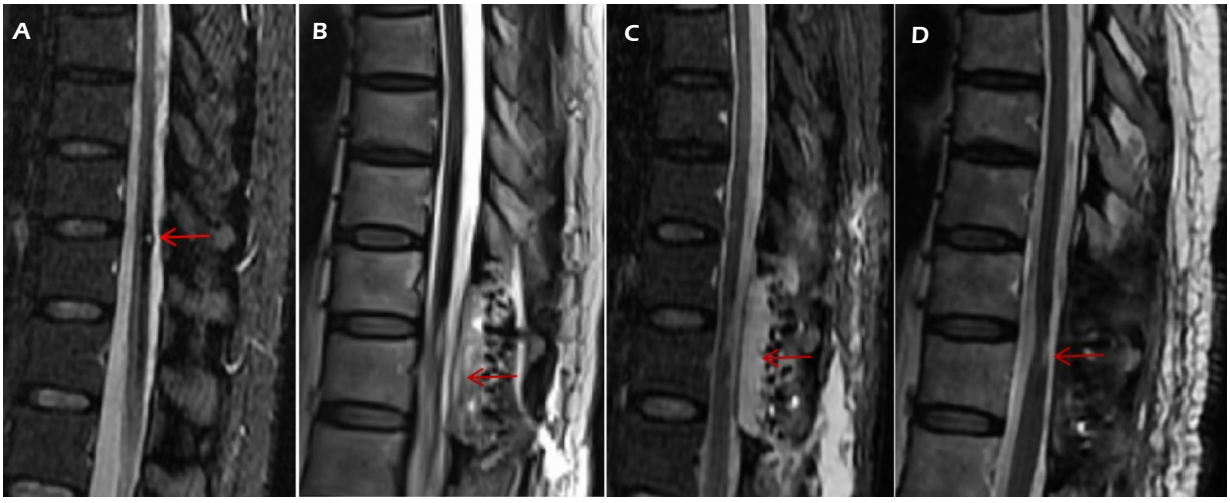

Figure 1. **A**, T2-weighted MRI study showing striped low signal and localized punctate high signal in the spinal cord, which indicates fresh hemorrhage. **B**, T2-weighted MRI study showing resection of the malformed vascular mass with slight edema in the operative area. **C**, T2-weighted MRI study showing an epidural effusion, which is a major factor in worsening postoperative symptoms. **D**, T2-weighted MRI study at 3 months post-operative shows no abnormal cord signal change and the fluid in the operated area has been absorbed.

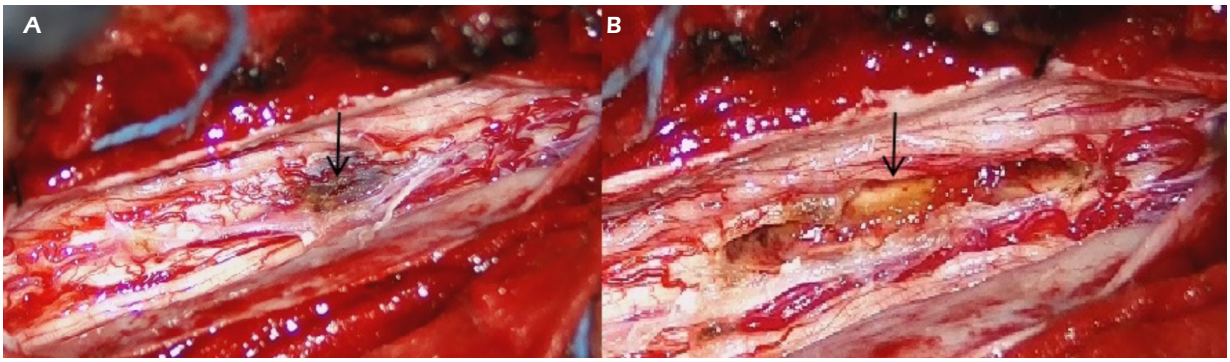

Figure 2. **A**, Intraoperative finding: locally elevated and dark red. **B**, The lesion had a relative border with the normal spinal cord, and there was no residual cavernous vascular malformation in the tumor bed after surgery.

returned to the hospital for follow-up 3 months later, muscle strength had returned to grade 5, the deep sensation had returned, and ASL was 1 (leg weakness with unrestricted walking), 0 (normal), and 0 (normal) for gait, urination, and defecation, respectively. A repeat MRI of the thoracic spine indicated significant relief of spinal cord edema in the operative area compared to the postoperative period (Figure 1D). The patient expressed satisfaction with the outcome of the procedure and provided written informed consent for the release of images.

Discussion

A cavernous vascular malformation is a vascular lesion that consists of dilated sinusoidal venous channels lined with a single layer of endothelial cells and lacking an intact vessel wall[1,9] (Figures 3A-B). The prevalence of cavernous vascular malformations of the central nervous system is 0.4%-0.6%, with the majority of these being intracranial and a predominance of supratentorial compartments[10]. Only 3-5% of lesions are located in the spinal cord[11]. However, spinal cavernous vascular malformations account for only 5-12% of spinal vascular malformations[9,12-14]. The cause of spinal cord cavernous vascular malformations is still unclear, and most scholars believe that most of them are congenital in origin, with a few acquired. Laberge-le et al[15], Liquori et al[16], and Bergametti et al[17] have suggested that it may be associated with the heterozygous loss of function of three mutations in *CCM1* (*KRIT1*), *CCM2,* and *CCM3* (*PDCD10*) in families. According to Hong et al[18], disseminated spinal cord spongiform vascular malformations may be as-

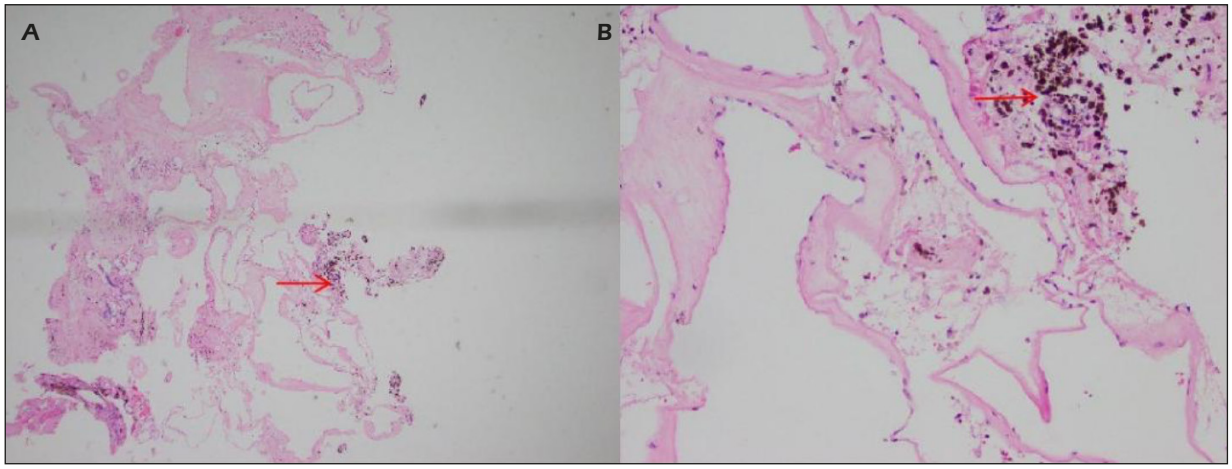

Figure 3. **A**, It consists of dilated sinusoidal venous channels that lack an intact vessel wall; Histology, Hematoxylin-Eosin (HE), 40×. **B**, The surrounding capillaries are surrounded by iron-containing hematoxylin deposits; Histology, HE, 100×.

sociated with certain central cell mutations, and *PIK3CA* mutations may lead to a higher risk of significant hemorrhage in patients with disseminated spongiform vascular malformations. In recent years, it has also been reported[19] that spinal cavernous vascular malformations can develop secondary to spinal cord radiotherapy, suggesting that acquired factors may also contribute to spinal cavernous vascular malformations. In this case, there is no family history of genetically related diseases, and a disseminated spinal cavernous vascular malformation is suspected.

There are four main clinical manifestations of spinal cord spongiform vascular malformations. The first is recurrent, intermittent neurological deterioration with varying degrees of recovery. The second is gradually progressive neurological deterioration. The third is sudden, rapid neurological deterioration, which can still have an exacerbation process of hours or days. This is different from bleeding caused by spinal cord arteriovenous malformations, which can lead to complete and permanent spinal cord paraplegia or partial paralysis[20]. The fourth manifestation is a gradual worsening of spinal cord function with mild, sudden-onset symptoms lasting several weeks to months[21]. Acute symptoms are most often caused by new bleeding, while chronic progressive symptoms may be caused by the compression of the surrounding spinal cord by the lesion or by small recurrent hemorrhages[22], hyaline degeneration, thickening of the capsule wall, or altered microcirculation[14]. The most common symptoms of spinal cord cavernous vascular malformations are motor and sensory dysfunction, such as hypesthesia and decreased muscle strength. Badhiwala et al[5] reported that out of 632 patients, 60.5% had motor deficits, 57.8% had sensory deficits, 33.8% had pain, 23.6% had urinary and/or fecal dysfunction, 0.5% had dyspnea, and 0.9% had no symptoms. Rare clinical manifestations may also be seen, such as Baldvinsdóttir et al[23] reporting a case of spinal cavernous vascular malformation presenting as spinal hemisection syndrome. In this case, the main manifestation was recurrent, intermittent neurological deterioration, which recovered with medication, but the symptoms of neurological damage worsened again on the latest admission, and we considered the possibility of rebleeding from the cavernous vascular malformation.

The main treatments for SCMs are surgical and conservative. Aggressive surgery is recommended for symptomatic patients, as it can lead to a good prognosis. Sandalcioglu et al[6] state that total excision of the lesion is the only way to prevent distant bleeding. In a meta-analysis by Fotakopoulos et al[13], 66.6% of patients underwent surgery, while 33.4% chose conservative treatment. The prognosis was better for patients who underwent surgery than for those who opted for conservative treatment. Asimakidou et al[9] suggested in their systematic review that surgical treatment within three months of symptom onset would result in a better prognosis for symptomatic patients, and prophylactic surgery in asymptomatic or incidental patients is controversial. Gross et al[14] retrospectively analyzed 352 surgically treated patients in 27 publications, of whom 91% achieved complete resection. Postoperative exacerbations were experienced by 36%, while 61% improved, 27% remained unchanged, and 12% became worse. The vast majority achieved complete resection and only 12% experienced long-term complications. Our patient opted for conservative treatment at four months of admission and experienced some symptom relief, but on the most recent admission, a re-bleeding spinal cord cavernous vascular malformation was suspected. This led to admission with a transient postoperative exacerbation, but improvement was observed over the pre-operative period after three months. This is consistent with most studies, which suggest that symptomatic spinal cord cavernous vascular malformations should be aggressively treated with surgery.

There are three main surgical approaches for the removal of cavernous hemangiomas, depending on the lesion's location within the spinal cord[1]. The first is the posterior median approach, used for lesions in the posterior median region of the spinal cord. The second is the posterior lateral approach, which is used for lesions in the posterior lateral part of the spinal cord. The third is the lateral approach, for lesions located laterally or anterolaterally to the spinal cord. Safe resection of spinal cord cavernous vascular malformations requires the use of various adjunctive techniques, such as intraoperative evoked potential monitoring, intraoperative ultrasound, and intraoperative angiography. Anson and Spetzler[22] suggest that skilled microscopic techniques and intraoperative evoked potential monitoring can facilitate the safe resection of the lesion. Liang et al's report[24] notes that intraoperative somatosensory evoked potentials and motor evoked potentials can sig-

nificantly reduce intraoperative injury to the normal spinal cord. Lunardi et al[25] suggest that the use of intraoperative ultrasound can help localize intramedullary cavernous vascular malformations. We were able to resect the lesion using our skilled microscopic technique successfully and intraoperative evoked potential monitoring, resulting in a good prognosis for the patient. We did not use intraoperative ultrasound since our lesion was located almost posteriorly medially and was easy to locate.

Conclusions

Our case is unique in that it is a kind of cavernous vascular malformation involving the spinal cord, which is a relatively rare condition. Our case highlights the importance of timely surgical intervention for symptomatic spinal cord cavernous vascular malformations, which can lead to a better prognosis for the patient. However, it is important to note that while our case had a positive outcome, it is still an isolated case, and the outcome may not always be favorable. Further research is needed in the future to understand the condition and improve treatment outcomes fully.

Conflict of Interest
The Authors declare that they have no conflict of interests.

Data Availability
The original contributions presented in the study are included in the article or Supplementary Materials; further inquiries can be directed to the corresponding author.

Ethics Approval
This study was approved by the Ethics Committee of the Mianyang Central Hospital, and the whole data and procedures complied with the principle of ethical standards (S20220421-02).

Informed Consent
Written informed consent was obtained from the patient for the publication of this case report.

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Not applicable.

Authors' Contribution
Y. Zhang and J. Wu conceived the idea and conceptualized the study. Y. Zhang, Y.-W. Huang and J. Feng collected the clinical data. Y. Zhang and Z.-P. Li drafted the manuscript. Y.-W. Huang and J. Wu reviewed the manuscript. All authors approved the final version of the manuscript before submission.

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