 doi: 10.2176/jns-nmc.2024-0262

NMC Case Report Journal 12, 109-114, 2025

Thoracic Intradural Extramedullary Cavernous Malformation Mimicking Meningioma

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Abstract

Cavernous malformation, also known as cavernoma or cavernous hemangioma, is a benign vascular malformation characterized by abnormal proliferation of capillaries that lack intervening neural tissue. Spinal intradural extramedullary cavernous malformations are exceedingly rare and, therefore, are often misdiagnosed. Here, we report a rare case of thoracic intradural extramedullary cavernous malformation that mimicked meningioma on preoperative imaging. A 77-year-old male patient presented with a two-month history of myelopathy, progressive paresthesia in the lower extremities, and difficulty walking, noted during an outpatient clinic visit. Spinal magnetic resonance imaging detected an intradural extramedullary mass lesion on the dorsal side at the Thoracic (Th) 10 vertebral level, compressing the thoracic cord. Preoperative imaging revealed a uniform T1- and T2-weighted intensity signal, suggesting meningioma. Intraoperatively, the mass appeared reddish with multiple abnormal vessels and adhered firmly to the thoracic cord. The lesion was completely resected, and postoperative pathological examination confirmed the diagnosis of cavernous malformation. The patient's preoperative symptoms improved postoperatively. Thoracic intradural extramedullary cavernous malformations are extremely rare and typically present with heterogeneous intensity signals on magnetic resonance imaging. Additionally, they can cause subarachnoid hemorrhage, emphasizing the importance of considering them in the preoperative differential diagnosis, even though imaging characteristics may be atypical. Surgical gross total resection should be considered to prevent future hemorrhage and neurological deterioration.

Keywords: cavernous malformation, intradural, extramedullary, thoracic, myelopathy

Introduction

Spinal cavernous malformations (CMs) are relatively rare lesions, most occurring intracranially. Approximately 5% of CMs are located in the spine, typically presenting as extradural or intradural intramedullary lesions.[1)] These lesions account for 5%-12% of all spinal vascular malformations and only 3%-5% of spinal cord lesions.[2-4)] Among these, intradural extramedullary CMs are particularly uncommon.[5-9)] A slight male predominance has been reported, and these lesions are more frequently observed in the thoracolumbar region and filum terminale.[6,8)] On magnetic resonance imaging (MRI), these lesions are often characterized by heterogeneous signal intensity due to hemosiderin deposition, which is considered a hallmark feature.[1)] Here, we present a case of thoracic intradural extramedullary CM that mimics meningioma in the preoperative differential diagnosis, as well as review the imaging features and management strategies for such cases.

Received October 29, 2024; Accepted January 6, 2025

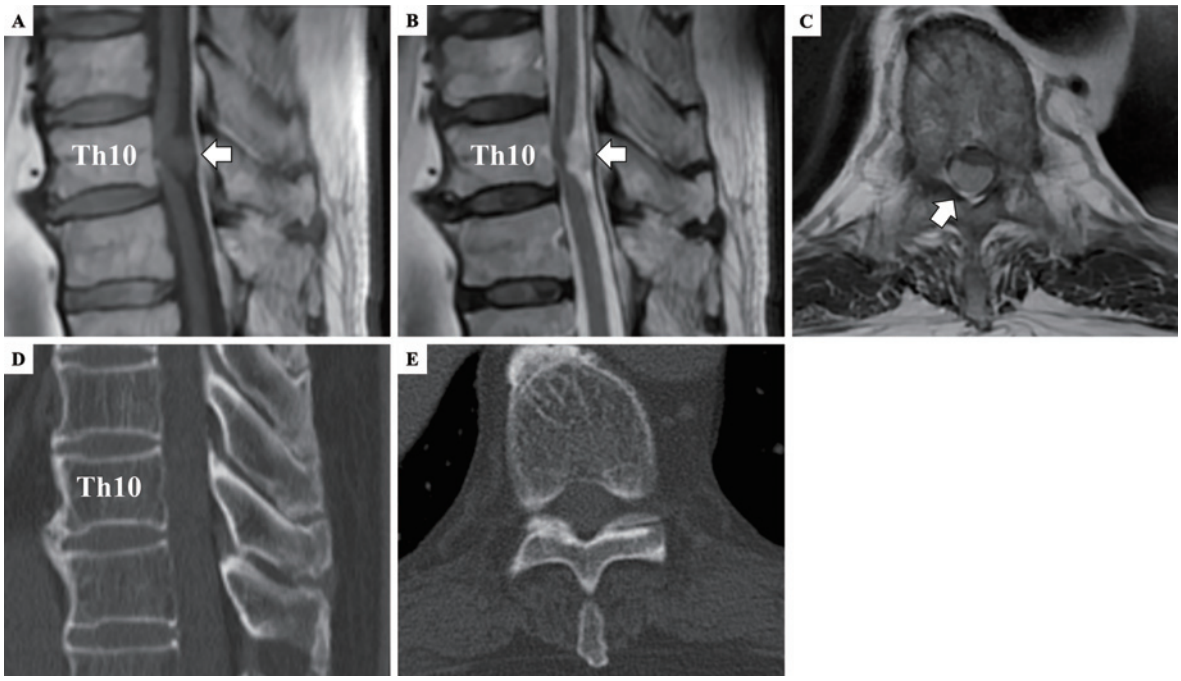

Fig. 1 Preoperative images.
Sagittal views of T1-weighted (A) and T2-weighted (B) MR images. (C) Axial view of a T2-weighted MR image at Th10. An intradural extramedullary mass lesion (arrow) is centered at Th10 on the dorsal side of the thoracic cord. Sagittal (D) and axial (E) views of CT images at Th10.
CT: computed tomography; MR: magnetic resonance; T1: type 1; T2: type 2; Th10: T helper 10

Case Report

A 77-year-old male patient presented with bilateral lower extremity numbness that had been progressively worsening over the past couple of months, along with increasing urinary retention at the outpatient clinic. Additionally, he reported difficulty in walking for a week and arrived in a wheelchair. On neurological examination, bilateral lower extremity weakness was noted, with a manual muscle testing (MMT) score of 4/5. Hyperreflexia of the patellar tendons was observed, and the patient exhibited urinary retention. Sensory examination revealed subjective numbness in the lower extremities, corresponding objective deficits in temperature and pain sensation. MRI revealed an intradural extramedullary T1-isointense, uniformly T2-high-intense signal mass lesion (arrow) centered at Th10 on the dorsal side of the thoracic cord (Fig. 1A-C). The lesion on MRI showed no heterogeneous signal intensity or cystic components. Computed tomography (CT) demonstrated no calcified lesion (Fig. 1D and E). These figures indicated a preoperative diagnosis of meningioma. The lesion robustly compressed the thoracic cord and exhibited signs of myelopathy, prompting surgical intervention. We approached the lesion using a Th10 laminectomy under the exoscope (ORBEYE, Olympus). Upon opening the dura, a dark reddish mass lesion was observed on the dorsal side of the cord in the subarachnoid space (Fig. 2A). The lesion was not attached to the dura mater but was securely adhered to the thoracic cord (Fig. 2B), and multiple abnormal red vessels were noted on the thoracic cord (Fig. 2C). Indocyanine green angiography indicated no flow within the lesion (*) (Fig. 2D). Careful detachment of the lesion from the cord was achieved, and gross total resection was performed (Fig. 2E). Pathological examination demonstrated no evidence of tumor cells within the lesion. The lesion consisted of multiple vessels containing red blood cells. Immunohistochemical analyses revealed CD31 positivity in the vascular endothelium. Additionally, alpha smooth muscle actin was positive in the smooth muscle surrounding the vessels, consistent with a diagnosis of CM (Fig. 3A-D). Postoperative contrast-enhanced magnetic resonance (MR) images showed complete lesion removal (Fig. 4A-C). The patient had an uncomplicated postoperative course and gradually recovered from his preoperative neurological deficits. Within a week, motor strength in his lower extremities improved to MMT 5/5, and his preoperative numbness, as well as deficits in temperature and pain sensation, completely resolved. Normal urinary function was also restored. He was discharged walking independently.

Discussion

CMs are low-flow vascular lesions characterized by ab-

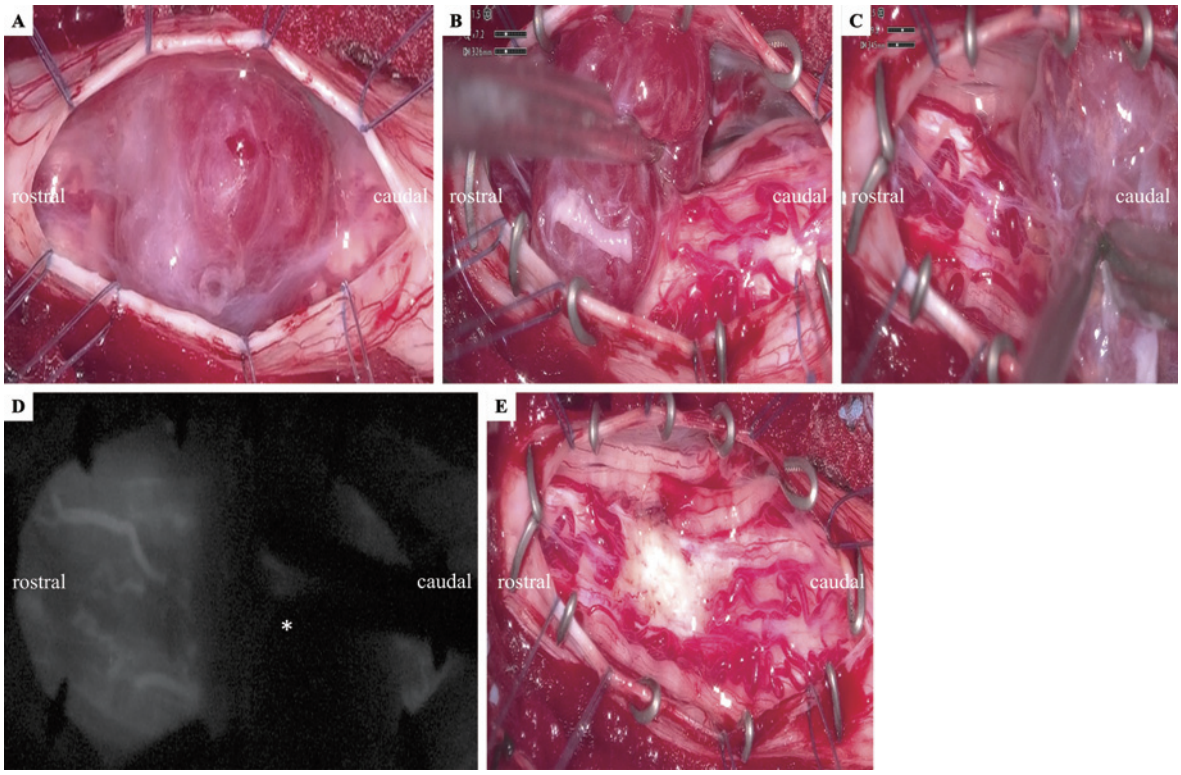

Fig. 2 Intraoperative findings.
(A) A reddish mass lesion located in the subarachnoid space. (B) The lesion was firmly adhered to the thoracic cord. (C) Multiple tortuous vessels were observed on the thoracic cord. (D) Indocyanine green angiography revealed no stain on the lesion (*). (E) Gross total resection was achieved.

normally structured, endothelium-lined sinusoidal chambers with sluggish blood flow, giving them a mulberry-like appearance. These lesions lack mature vascular mural elements and are devoid of intervening neural tissue.[10] CMs are more frequently encountered in the brain, whereas their epidemiology in the spinal cord is less well documented, with spinal CMs accounting for approximately 5% of intramedullary lesions in adults.[11] The thoracic cord is the most predominant site for spinal CMs.[12] Cranial intradural extramedullary CMs, though rare, have been reported in locations such as the intraorbital and sellar regions.[13,14] However, spinal intradural extramedullary CMs are extremely rare, with their incidence and exact prevalence remaining unknown. Patients with intradural extramedullary CMs frequently present with progressive myelopathy, and back pain through subarachnoid hemorrhage is often reported as the initial symptom.[6-8,15-17] Notably, hemorrhage from these malformations occurs in more than 25% of cases.[18]

Intradural extramedullary CMs typically demonstrate characteristic features on MRI due to the presence of mixed subacute and chronic hemorrhage. A combination of high and low signal intensities circumscribed by a hypointense border on MRI, indicating hemosiderin deposition, strongly suggests CMs.[19] These lesions are not typically visible on spinal angiography, similar to spinal intramedullary CMs.[14] Contrast enhancement is rare and generally minimal when present.[12] In this case, a preoperative contrast-enhanced imaging study was not performed because the plain MRI demonstrated uniformly iso-intensity on T1 weighted imaging (T1WI) and uniformly high intensity on T2 weighted imaging (T2WI). Based on these findings, the lesion was assumed to be a meningioma, and surgical intervention was planned accordingly. Additionally, the typical low-intensity rim commonly observed on T2WI and the presence of abnormal vascular structures were absent, which led to CMs not being considered in the preoperative differential diagnosis. Had a preoperative contrast-enhanced MRI been performed, minimal enhancement might have highlighted atypical imaging features inconsistent with meningioma or schwannoma, which typically show more pronounced enhancement. These atypical MRI findings highlight the importance of recognizing that CMs can exhibit a wide spectrum of signal intensity patterns on MRI.

Regarding imaging, MRI, with its characteristic features, enables accurate preoperative diagnosis. The most prevalent intradural extramedullary tumors are schwannomas (15%-50%) and meningiomas (20%-25%).[20,21] In this case, the lesion's uniform signal intensity on MRI, without pe-

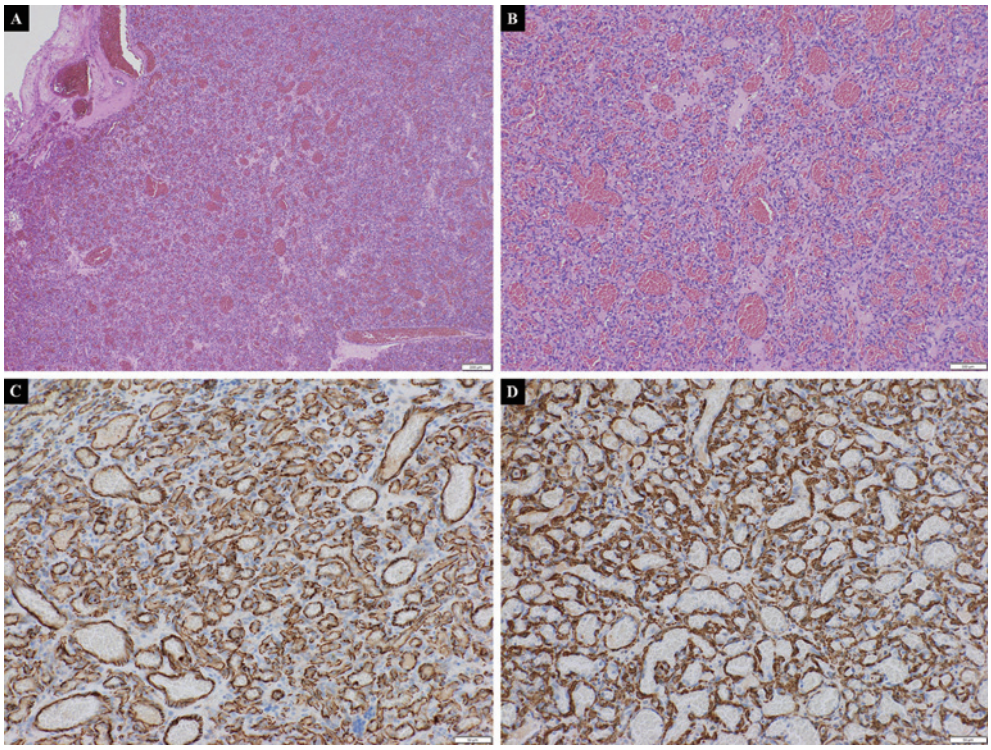

Fig. 3 Histopathological investigations.
(A) No signs of tumor cells (H&E staining × 40). (B) Accumulation of multiple vessels containing red blood cells (H&E staining × 100). (C) CD31 staining was positive in the vascular endothelium (CD31 immunostaining × 200). (D) Positively stained smooth muscle surrounding vessels (αSMA immunostaining × 200).
αSMA: alpha smooth muscle actin; H&E: hematoxylin and eosin

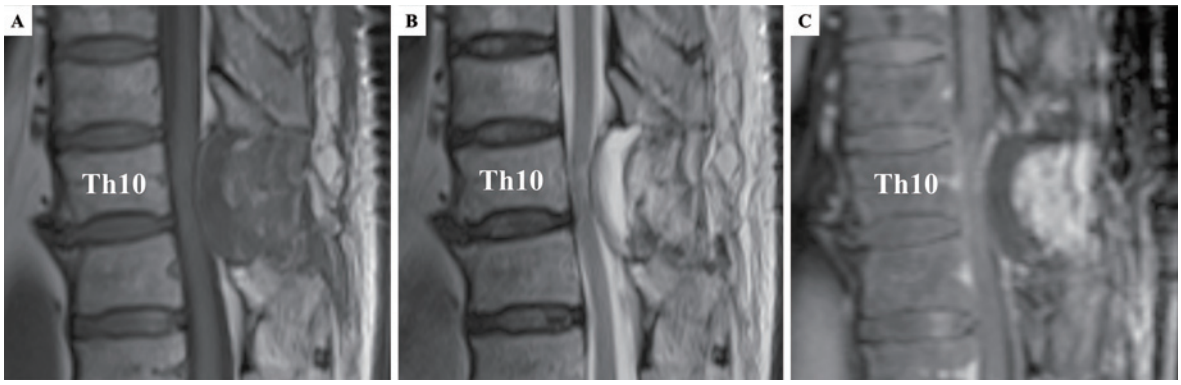

Fig. 4 Postoperative images.
Sagittal views of postoperative MRI showing T1-weighted (A), T2-weighted (B), and T1-weighted contrast-enhanced (C) images, confirming complete lesion removal.
MRI: magnetic resonance imaging; T1: type 1; T2: type 2

ripheral hemosiderin deposition, suggested meningioma as the likely diagnosis. Meningiomas typically appear as well-circumscribed lesions with a broad dural base, iso- to hypointense on T1WI, and slightly hyperintense on T2WI.[22] Schwannomas, another common differential diagnosis, are well-circumscribed lesions that appear iso- to hypointense on T1WI and hyperintense on T2WI, often with heterogeneous signal intensity due to hemorrhage or cystic degeneration. Solitary fibrous tumors, though rare, are also considered in the differential diagnosis. These tumors are isointense on T1WI with variable T2WI signals, and calcification or hemorrhage is uncommon.[23,24] This case highlights the importance of recognizing atypical MRI features, which may complicate distinguishing CMs from more

common intradural extramedullary tumors.

Intradural extramedullary CMs are well-demarcated lesions that compress the spinal cord, causing myelopathy and occasionally hemorrhage, resulting in hematomyelia. Predicting the natural history of intradural extramedullary CMs is difficult due to their rare occurrence. Initial symptoms may include low back pain, sciatica, and myelopathy. They may acutely cause hemorrhagic transformation, even when small in size and clinically silent at first, thereby producing neurological deficits. In rare cases, they can cause acute massive hemorrhage with significant mass effects rapidly. Lumbar puncture may be considered when minor bleeding is suspected. As demonstrated in this case, preoperative imaging may not always reveal typical features, necessitating the inclusion of CMs in the differential diagnosis. Gross total resection is frequently attainable in these scenarios, even when the lesions are adherent to nerve roots.[6] In this case, the lesion was firmly attached to the dorsal aspect of the thoracic spinal cord, suggesting its origin from small vessels within the spinal cord. Postoperative hemorrhage is a potential risk with partial resection, making gross total resection with meticulous microsurgical technique the optimal treatment. Since these tumors are venous anomalies surrounded by abnormal vasculature, careful detachment from surrounding structures and avoiding incision into the tumor itself are critical to minimizing bleeding and ensuring safe, en bloc removal.

Informed Consent

Informed consent has been obtained from the patient in this case report.

Disclaimer

Author Haruhiko Kishima is one of the Editorial Board members of the Journal. This author was not involved in the peer-review or decision-making process for this paper.

Conflicts of Interest Disclosure

All authors have no conflict of interest.

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