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by Eko Agus Subagio

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CASE REPORT

Rupture of the recurrent thoracic intramedullary spinal cord cavernoma

Galih Indra Permana,
Eko Agus Subagio,
Muhammad Faris,
Abdul Hafid Bajamal

Department of Neurosurgery, Universitas Airlangga – Dr. Soetomo General Academic Hospital Surabaya, East Java, Indonesia

Abstract

The intramedullary spinal cord cavernoma is a rare vascular disease that occurs within the spinal cord or intramedullary and accounts for about 5%-12% of all pathology in the spinal vascular disease. We report a clinical progression of the disease and the evolution of the hemorrhage every year until the patient underwent a surgical procedure. A 63-years old male with progressive weakness of the lower extremity since one month before admission, worse in the left side. He also complained of loss of sensation at the level of the T4 and below it. The patient suffered three times rebleeding before undergoing surgery. Magnetic resonance imaging was showing intramedullary spinal cord cavernoma and a histology examination exhibited the characteristics of a cavernoma. Intramedullary spinal cord cavernoma is a rare disease that should be treated before rebleeding occurs and enlargement of the lesion. Postoperative neurological function in intramedullary spinal cord cavernoma patients is determined by the preoperative neurological status.

Keywords: *Intramedullary spinal cord cavernoma, Rebleeding, Vascular malformation, Progressive weakness*

Introduction

Intramedullary spinal cord cavernoma is a rare vascular disease and contributes to about 5%-12% of all vascular diseases in the spinal cord.[1] The prevalence of cavernoma in the brain and spinal cord is approximately 1.9 cases/ 100,000 person/ years with 3%-5% of lesions in the spinal cord. About 57%-73% of spinal cavernoma involved the thoracic spine, while 27-38% were cervical.[2] They are rarely found in the lumbar region. Men and women in the fourth and fifth decades are mostly affected with no sex predilection. The intramedullary spinal cord cavernoma is usually a solitary lesion, even though 27% are associated with cavernous intracranial malformation. The most common clinical sign and symptom is progressive myelopathy.[3] Subarachnoid hemorrhage and hematomyelia have also been found in the intramedullary spinal cord cavernoma. Severe neurological deficit or progressive symptoms are considered to require surgical management. The recurrence of hemorrhage during conservative treatment of intramedullary spinal cord cavernoma is regarded as an indication for surgery, even though symptoms with acute onset are sometimes relieved over time. The annual rate of

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Corresponding author:

Eko Agus Subagio
Department of Neurosurgery, Airlangga University – Dr. Soetomo General Academic Hospital.
Mayjen Prof. Dr. Moestopo Street No. 6-8, 60286. Surabaya, East Java, Indonesia
easnsurg@yahoo.com

bleeding is estimated at 2.5%.[4] We presented a case of the intramedullary spinal cord cavernoma with three times recurrence hemorrhage before surgery. This case report focuses on the clinical, neuroradiological, and management principles of the thoracic intramedullary spinal cord cavernoma.

Case Report

History and Examination

A 63-years old male patient was admitted to the neurosurgery department with progressive weakness of both lower extremities one month ago. Initially, he complained of liability of the left lower extremity two years ago but didn't wish for surgical treatment because there was no disturbance in daily living activity. The motoric function of the left L2 to S1 is 4 point and 5 point on the right side with a D American Spinal Injury Association Impairment Scale (AIS) grade.[5] While, the motoric function in the upper extremity is 5 point. A year after that, the patient underwent weakness of the right lower extremity and didn't want surgery because of no limitation in daily living activity. A few months later, the patient came with progressive weakness of both lower extremities. The motoric function of the left and right side from L2 to S1 is 4 point with a D AIS grade.[5] Patient also complained of sensitivity loss in the thoracic region as high as T4 level and below it with the grade 1 sensory impairment.[5] Patient unable to control urination and defecation since three weeks before admission. Increased Patellar and the Achilles reflexes were found in the patient. Pathological signs, the Babinski and the Chaddock, and clonus also found in the patient indicate an upper motor neuron sign.

Neuroimaging

MR imaging demonstrated lobulated intramedullary lesion at the level of thoracic 3 with the measurement of 1.1x1x1.8 cm. This lesion caused slight cord expansion. The T1 and T2 weighted imaging sequences showing hyperintensity with dark rim (Figure 1) and blooming artifact at the T2*GRE sequences indicate the blood product, with spinal cord edema at the level thoracic 2 to 4. There was no contrast enhancement at the postcontrast imaging. This description leads to an intramedullary spinal cord cavernoma. Compared with the previous MR imaging, two years and a years before, they showed no significant enlargement of the lesion (Figure 2).

Pathological Examination

The gross mass appears as a mass with a brownish color and solid elastic consistency. The microscopic shows a large bleeding area and fibrin with the invasion of the inflammatory cells, mononuclear and polymorphonuclear (PMN). On the edge of this lesion are irregular blood vessels with thin walls. There was no sign of malignancy. This histology represents a cavernous angioma (Figure 3).

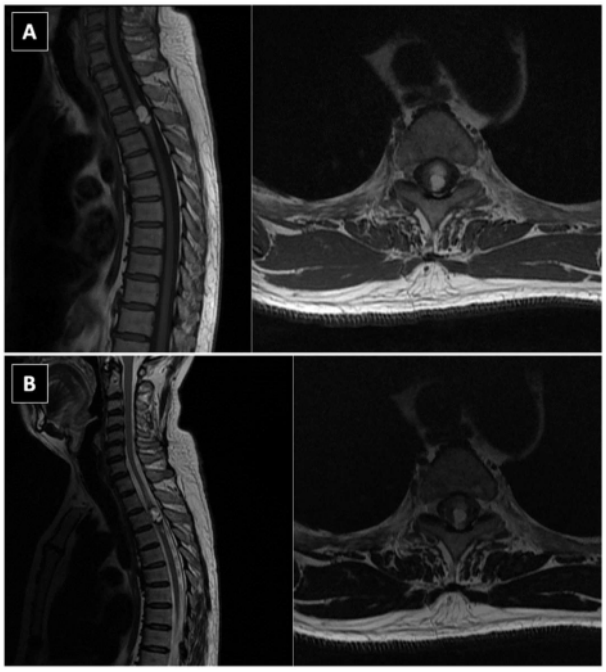

Figure 1. *Axial dan sagittal MR imaging showing hyperintensity with surrounding hypointense rim in the T1-weighted image (**A**) and T2-weighted image (**B**)*

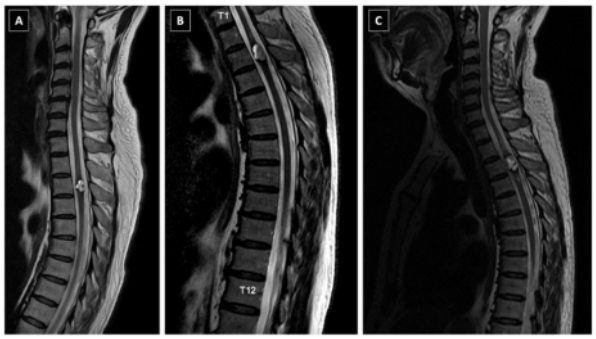

Figure 2. *Sagittal MR imaging T2-weighted image showing hyperintensity with surrounding hypointense rim in every year's evaluation, from 2019 (**A**), 2020 (**B**), and 2021 (**C**). There is no significant lesion enlargement.*

Operation and Postoperative Course

Total laminectomy of the thoracal 3, cavernoma resection and pedicle screw of the thoracal 2, 3, and 4 were subsequently performed.

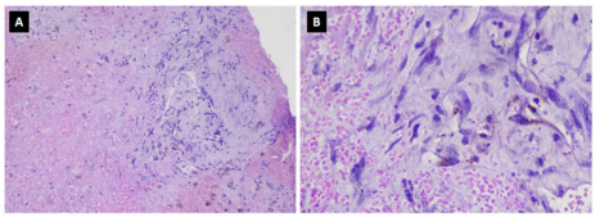

Figure 3. *Pathological anatomy of the cavernous angioma demonstrated a large bleeding area and fibrin with the invasion of the inflammatory cells, mononuclear and PMN. On the edge of this lesion are irregular blood vessels with thin walls. There were no signs of malignancy. Haematoxylin and eosin stain (**A**) ×10 and (**B**) ×40.*

At the surgery, we can identify the cavernoma at the dorsal of the myelum with the dark purplish color (Figure 4a). Dorsal myelotomy was performed, and cavernoma separated from myelum with blunt dissection and microvascular dissection. The clot hematoma intramedullary and yellowish tissue indicated gliosis and hemosiderin staining (Figure 4b and 4c). After cavernoma was resected entirely, dural closure with watertight stitches and then pedicle screw was performed at the level of thoracic 2, 3, and 4. Postoperative, there is no worsening of the neurological deficit. The patient evaluated clinical examination and MR imaging in 3 months postoperatively.

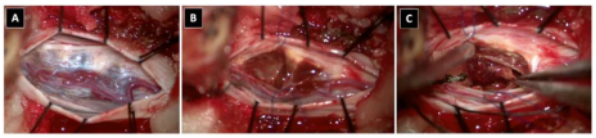

Figure 4. *Intraoperative image revealing dark purplish color at the dorsal of the myelum (**A**). After myelotomy, there was a cavernous angioma lesion intramedullary (**B**). The lesion was carefully dissected out, and there was a clot hematoma inside this lesion (**C**).*

Discussion

Intramedullary spinal cord cavernoma is a rare disease and accounting for only 5% of all spinal cord lesions, 5-12% of all spinal vascular tumors, and 5% of all of the nervous system cavernous malformation. About 57%-73% of spinal cavernoma involve the thoracic spine, while 27%-38% are cervical. They are rare in the lumbar region.[6,7] Men and women in the fourth and fifth decades are mostly affected with no sex predilection. The annual rate of bleeding is estimated at 2.5%.[4] A spinal cord cavernoma is a mulberry-shaped vascular anomaly that consists of the abnormal group of tiny sinusoidal-type capillaries and venules in the spinal cord. Cavernoma is a progression lesion that can transform over size and shape over time.[2] A characteristic of cavernoma is hemosiderin deposition, caused by minor silent bleeding that makes the cavity around the blood vessels, causing hemosiderin accumulation around the neural tissue. Hemosiderin hoarding can be demonstrated as a hypointense signal ring around the lesion in the MRI.[1,8] This study showed a case of intramedullary cavernoma with the rebleeding that occur three times every year. Intramedullary spinal cord cavernoma is a rare condition with rebleeding occurring every year, making these cases rarely happen and interesting to study.

The mode of presentation ranges from acute neurologic deficits due to frank hemorrhage within the intramedullary spinal cord cavernoma to a progressive decline in neurologic function due to the associated hyalinization and gliosis. The majority of lesions are dorsally located, making sensory deficit more common (65%).[9] Motor weakness, pain, sphincter dysfunction, and involuntary movement are possible presentations, respectively, in a decreasing order. Spinal arteriovenous vascular malformation, myelitis, intramedullary tumor, and demyelinating pathologies can cause progressive neurological deterioration, besides intramedullary spinal cord cavernoma. The initial clinical presentation was reported as subarachnoid hemorrhage and hematomyelia, even though the most frequent production is progressive myelopathy caused by the progressive expansion and bleeding of the malformation.[10] Hemosiderin has a neurotoxic effect that alters surrounding microcirculation and plays a vital role in the progression of clinical symptoms in intramedullary spinal cord cavernoma. The myelopathy can be manifest as upper motor neuron signs in this patient.

Magnetic resonance imaging (MRI) is the gold standard for diagnosis. Intramedullary spinal cord cavernoma appears as mixed intensity lesions surrounded by a hypointense rim of hemosiderin on Tl- and T2-weighted images.[11] Spinal cavernoma does not enhance with contrast and is angiographically occult. Hence, MRI of the entire neuraxis is recommended because 27% of spinal cavernoma have an associated intracranial cavernous malformation. The result of MRI showed an intramedullary lesion with the characteristic imaging of a cavernoma. Based on the MRI, cavernoma can be composed into four groups (Table 1).[12] Type I emerge hyperintense on both T1- and T2-weighted sequences show the subacute hemorrhage and hemosiderin core. Type II demonstrated the classic "popcorn" lesions. This type II has mixed signal intensity on both T1 and T2 sequences caused by multiple loculated hemorrhages of various stages enclosed by gliotic margins. The gliotic peripheral rim shows decreased signal intensity on T2 sequences. Type III shows chronic resolved bleeding within an isointense core. Type IV represents small capillary telangiectasias

that can only be seen on gradient resonance echo sequences.[13] Our cases have a typical type I spinal cavernoma MRI with hyperintensity in the T1- and T2-weighted imaging with hypo-intensity around the lesion indicates hemosiderin rim.

Table 1. *Classification of MRI for cavernous malformation*

Type	MRI Characteristic*	Pathological Characteristics
Type I	T1: hyperintense core T2: hyper- or hypointense core with surrounding hypointense rim	Subacute hemorrhage, surrounded by a rim of hemosiderin-stained macrophages & gliotic brain
Type II	T1: reticulate mixed signal core T2: reticulated mixed signal core with surrounding hypointense rim	Loculated areas of hemorrhage & thrombosis of varying age, surrounded by gliotic, hemosiderin stained brain; in large lesion, areas of calcification may be seen
Type III	T1: iso- or hypointense T2: hypointense with a hypointense rim that magnifies the size of the lesion GE: hypointense with greater magnification than T2	Chronic resolved hemorrhage, with hemosiderin staining within & around the lesion
Type IV	T1: poorly seen or not visualized at all T2: poorly seen or not visualized at all GE: punctate hypointense lesions	Two lesions in the category have been pathologically documented to be telangiectasias

*T1 and T2 show T1 and T2 weighted sequences; GE: Gradient Echo sequences

Observation is usually reserved for patients who are asymptomatic or patients who are unable to undergo surgery due to other comorbidities. Some may argue, however, that even in asymptomatic patients, it is recommended to proceed with resection given that recurrent hemorrhages may lead to the development of neurologic deficits in the long run.[14] The surgical approach is tailored to the location of the lesion—complete removal of the intramedullary spinal cord cavernoma lesion with no neurologic sequelae. The posterior approach is best suited for dorsally located lesions but could also be utilized for lateral and ventral lesions. This approach is well tolerated by the patients and familiar to neurosurgeons.[2] It also allows for minimal blood loss and is less time-consuming. Surgical resection is indicated in patients with progressive neurologic deficits, exophytic lesions, and those with acute presentation and bleeding within the intramedullary spinal cord cavernoma to avoid the posthemorrhagic effect of bleeding.[15] We performed total laminectomy to get a comprehensive view of the surgical field. Cavernoma was carefully resected totally, and clot hematoma was also evacuated. Our dissection target until the margin of gliosis tissue and yellowish stained hemosiderin tissue. A pedicle screw was performed to improve the stabilization of the thoracic region. Our goal of surgery was reached with no residual mass and no worsening of the neurological function postoperatively.

Conclusion

Intramedullary spinal cord cavernoma is a rare disorder that is recommended to be treated early before rebleeding occurs and enlarge the lesion. Postoperative neurological function in intramedullary spinal cord cavernoma patients is determined by the preoperative neurological status. Complete removal with no neurologic sequelae of the intramedullary spinal cord cavernoma is achieved as the goal of the surgery to prevent a recurrence.

Acknowledgment

The author declares that they have no conflict of interest.

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