Multiple spinal intramedullary cavernous angiomas with bleeding episode mimicking an intramedullary tumor

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Multiple spinal intramedullary cavernous angiomas with bleeding episode mimicking an intramedullary tumor

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ABSTRACT

Intramedullary cavernous angioma is a rare vascular malformation compared to cerebral cavernous malformation. The incidence of cavernous angioma is about 3 - 5% of all central nervous system lesions, 5 - 12% of all spinal vascular lesions, and 1% of all intramedullary lesions in pediatric patients. Although intramedullary cavernous angioma has the same histological picture as cerebral cavernous angioma, the natural history, and surgical approach are different from cerebral cavernous angioma. Due to its location in the eloquent area of the spinal medulla, a slight change in the size of the lesion can affect the neurological function of the patient. We describe a case of an intramedullary cavernous angioma with hemosiderin post bleeding, located in the cervical cord which was initially misdiagnosed as hemorrhagic ependymoma. On whole spine MR imaging we also found an intramedullary cavernous angioma in the lower thoracal cord at the T12 level with mild hemorrhage. Abdominal MRI showed cavernous angiomas in both kidneys. The patient underwent surgical treatment with removal of the lesion in the cervical cord and T12 level, with histopathologic findings consistent with cavernous angioma. No malignancy was detected. Post-surgery, the symptoms gradually improved. Symptomatic intramedullary cavernous angioma tends to bleed repeatedly and being unstable. Early diagnosis and surgical treatment can prevent rebleeding and more severe symptoms.

CASE REPORT

CASE REPORT

A 6-year-old boy was referred to our hospital with symptoms of a stiff neck and pain for 2 weeks before admission. A weakness of the right limb for 4 days before admission and vomiting, no decreased consciousness, convulsions, fever, cough, or tightness were detected. The history of fall that hit the area around the head and neck was denied.

The patient also had a previous history of hypertension, which was detected 1.5 years ago and was never controlled after the diagnosis.

He was the first child with a normal history of his mother during the pregnancy, borne full term, normal delivery helped by a midwife with 2,450 g body weight, and age appropriate history of growth and development.

1.5 Tesla cerebral magnetic resonance imaging (MRI) examination has been performed, which detected no cerebral abnormality. However, there was evidence of a cervical cord lesion from C1 through C6 level with hemorrhage and cord edema which is suspicious of an intramedullary tumor with intra-tumoral hemorrhage. The examination was continued with a whole spine MRI which showed two intramedullary lesions on the cervical level and T12 level with adjacent cord edema and without contrast-enhancement (Fig. 1&2). The lesions demonstrated eccentricity with popcorn appearance and hypointense rim suggestive of hemosiderin (Fig. 3).

Because the patient also suffered from hypertension, abdominal MRI was performed with the findings of cavernous angiomas in both kidneys and hypoplasia of the left kidney (Fig. 4). There was no sign of bleeding at the lesion site in the kidney, and the patient did not complain of hematuria, thus lesions required only observational management.

Surgical exploration was performed for the cervical cord. Chronic blood was evacuated, no tumor was detected, surgical resection of cavernous angioma was also performed at the distal T12 level (Fig. 5). Histopathology examination showing old blood at both cervical and distal T12 level. No malignancy was detected, consistent with cavernous angioma (Fig. 6). Post-surgery, the symptoms gradually improved.

DISCUSSION

Etiology & Demographics:

Cavernous angiomas, also known as cavernous malformations, cavernous hemangiomas, or cavernomas, are uncommon vascular malformations of the central nervous system [1,2] They are characterized by an abnormally dilated blood vessel, outlined by endothelium that disrupts normal nerve tissue [3]. Spinal cavernous angioma can be either epidural or intradural-intramedullary [2,4]. The exact etiology of intramedullary cavernous angioma remains unknown [4–6]. It is thought to develop in compensation for the absence of normal blood vessels due to hypoplasia, aplasia, or early occlusion of the developing vein [7]. Pathologically, they consist of endothelial-lined caverns lacking appropriately formed tight junctions. The vascular leak due to poorly formed tight junctions leads to the clinical consequences of the disease [8].

This condition can be sporadic or familial. Individuals with single lesions and no affected relatives are most likely to have a sporadic disease. Multiple lesions may be of familial type due to changes (mutations) in the gene [7].

Intramedullary cavernous angioma is very rare [1,4–7]. The incidence is about 3-5% of all central nervous system lesions and 5 – 12% of all spinal vascular lesions, and 1% of all intramedullary lesions in paediatric patients [4,6,8]. More common in females and the peak age of presentation is in the fourth decade [12]. In pediatrics, the bimodal age distribution was found with peaks below 4 years and over 12 years [13].

Clinical & Imaging Findings:

Cavernous angioma may be asymptomatic or may have various clinical presentations [14]. Patients may come to medical attention due to intracerebral or spinal hemorrhage, seizure, focal neurological deficit without overt hemorrhage, or as an incidental finding [8]. Due to the location of the lesion, it will affect the neurological function of the patient, especially when the hemorrhage happened [5]. Intramedullary cavernous angioma with hemorrhage and cord edema reveals a pseudotumor and looks like a tumor with intratumorally and peritumoral hemorrhage. Neurological damage can be confused with different pathologies such as demyelination, myelitis, intramedullary tumors, and arteriovenous vascular malformations of the spine [15.

According to the literature, the location is usually in the cervical and thoracic spinal cord, with the incidence of thoracic being the most common (46%), then cervical (38%), cervicothoracic (8%), and conus being the least common (8%) [16]. They may be associated with cavernous angiomas in other organs or the central nervous system [3].

Cavernous angiomas rarely involve the kidneys. They are usually single, rarely multiple, or bilateral. They are often misdiagnosed, both clinically and radiologically. Certain cases remain asymptomatic and are detected incidentally. The most common clinical manifestation is hematuria [17].

MRI has increased the number of reported cases of cavernous angioma of the spinal cord [18]. Intramedullary cavernous angioma without bleeding is easily diagnosed with Magnetic resonance imaging [1,20]. A study revealed that features of small size, eccentric location in axial plane, minimal enhancement with contrast, and absence of edema were more frequently observed on MRI of spinal cavernous angiomas compared to hemorrhagic ependymomas ($p < 0.01$) [20].

They can have little or no mass effect unless they are complicated by hemorrhage [20]. They can be distinguished from intramedullary lesions, especially hemorrhagic ependymoma [20]. The most common growth mechanism is represented by small hemorrhages, then forming a small cavity around the blood vessels, causing the deposition of hemosiderin in the surrounding nerve tissue. Another possibility is the appearance of a nidus which increases the malformation.

Eventually, the lesion may bleed acutely with rapid expansion and acute mass effects [3].

Cavernous angiomas may have areas of internal thrombosis or bleeding of various ages. There may be conversion of hemoglobin to methemoglobin, which results in a hyperintensity focus on the T1WI. Hemosiderin can shift from the central area towards the periphery [20]. The presence of hemosiderin deposits in spinal cavernous angiomas is indicated by the presence of a hemosiderin rim at T2WI, and approximately 22-33% of spinal cord ependymomas also demonstrate a hemorrhagic "cap sign" at the tumor poles. The similar appearance of these two lesions often presents a diagnostic dilemma as well as challenge for the treatment strategy [20].

Treatment & Prognosis:

Management of stable spinal cavernous angiomas is conservative, but if the condition is unstable it requires surgery to prevent neurological damage [21]. Symptomatic intramedullary cavernous angioma tends to bleed repeatedly and is unstable. The lesion should be surgically removed to avoid further deterioration due to recurrent hemorrhages [6]. Management of renal cavernous angioma includes observation, nephrectomy, heminephrectomy, papillectomy, and embolization. In healthy patients with mild to moderate hematuria clinically and radiographically no abnormality found, observation is indicated [22].

The functional prognosis in children treated for spinal cord cavernous angioma seems better than in adults. However, surgical resection is the treatment of choice in cavernous angioma with hemorrhage because it is associated with a better prognosis [19].

Differential Diagnoses:

The characteristics of spinal intramedullary cavernous angioma on contrast enhanced MRI are small size, eccentric location in axial, minimal enhancement with contrast, and absence of edema. It can differentiate between hemorrhagic ependymomas which have a larger size, central location in axial, strong enhancement with contrast, and present edema ($p < 0.01$) [20].

The presence of diffuse or intralesional hemorrhage can cause atypical radiological features, making the diagnosis of cavernous angioma difficult [23]. In our case, we found two intramedullary cavernous angiomas in the cervical cord and distal thoracic cord near the medullary conus, no intracranial cavernous angioma was found, and we also detected cavernous angiomas at both kidneys. The cavernous angioma located in the cervical cord had recent bleeding with hemorrhage and adjacent cord edema. This condition is mimicking intramedullary tumors especially hemorrhagic ependymoma. Contrast-enhanced MRI and if needed MRA and DSA should be performed to exclude other vascular malformation. However, after contrast administration, no mass contrast enhancement was detected. And again, we found the cavernous angioma in the distal thoracic cord near the medullary conus, confirming all the findings were cavernous angiomatosis with intramedullary bleeding in the cervical cord area.

TEACHING POINT

Intramedullary cavernous angioma is rare. Magnetic resonance imaging is a useful modality for the diagnosis of intramedullary cavernous angioma. The lesion may bleed acutely, causing intramedullary hemorrhage with mass effect, which can be confused with an intramedullary tumor. Contrast-enhanced MRI should be performed to exclude other vascular malformations. Early diagnosis and surgical treatment can prevent rebleeding and more severe symptoms.

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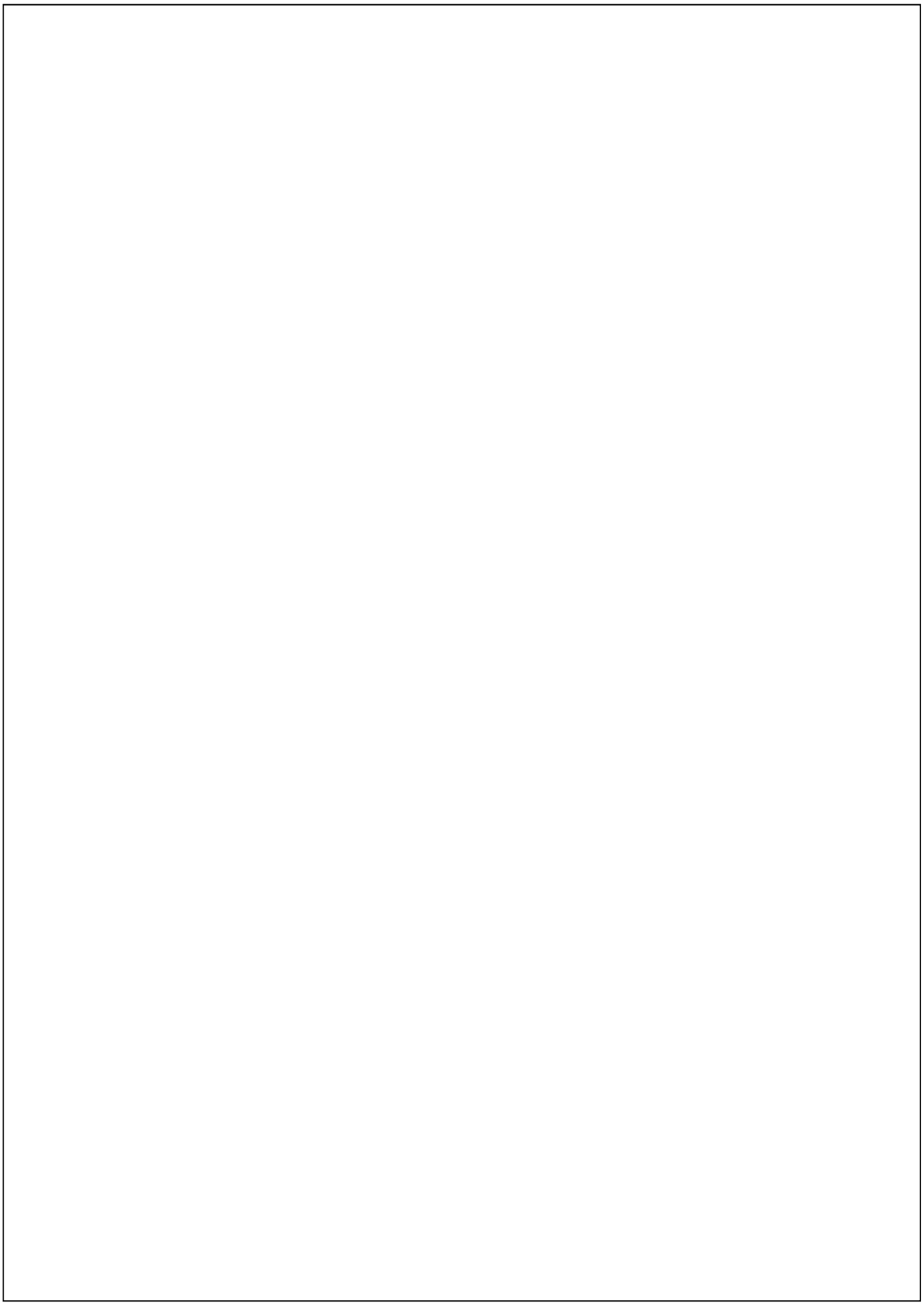

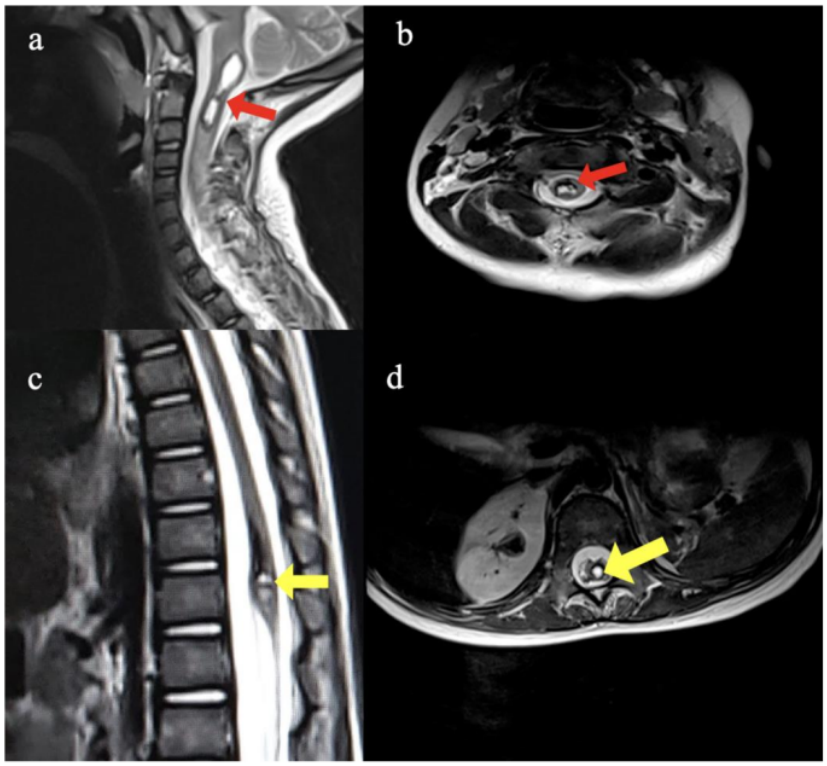

Figure 3: 6-year-old boy with spinal intramedullary cavernous angiomas.

Findings: a) Sagittal T2 TSE, b) axial T2 TSE spinal MRI in cervical level, c) Sagittal T2 TSE and d) axial T2 TSE spinal MRI in thoracic level demonstrating an eccentric lesion with popcorn appearance and hypointense rim suggestive of hemosiderin (red arrow) at the cervical and (yellow arrow) thoracic levels.

Technique: (a-d) Whole spine MRI. GE Optima 360 1.5T GE. Sagittal T2 TSE (TE: 81 TR: 4000 Thickness: 3mm). Axial T2 TSE (TE: 106 TR: 5830 Thickness: 3mm).

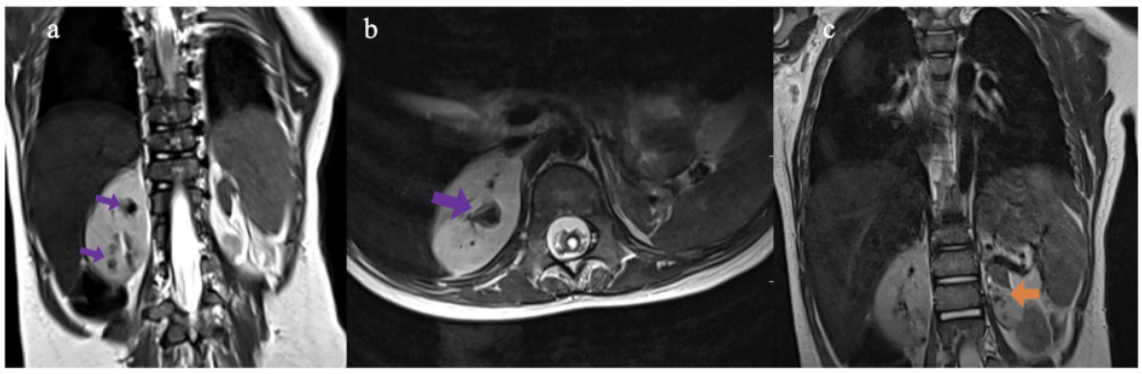

Figure 4: 6-year-old boy with spinal intramedullary cavernous angiomas.

Findings: a) Coronal T2 TSE b) axial T2 TSE and c) coronal T2 TSE abdominal MRI demonstrating cavernous angiomas in upper and lower pole of the right kidney (purple arrow), hypoplasia of the left kidney with a cavernous angioma in the upper pole (orange arrow).

Technique: (a-c) Abdominal MRI. GE Optima 360 1.5T GE. Coronal T2 TSE (TE: 81 TR: 3200 Thickness: 3mm). Axial T2 TSE (TE: 106 TR: 5830 Thickness: 3mm).

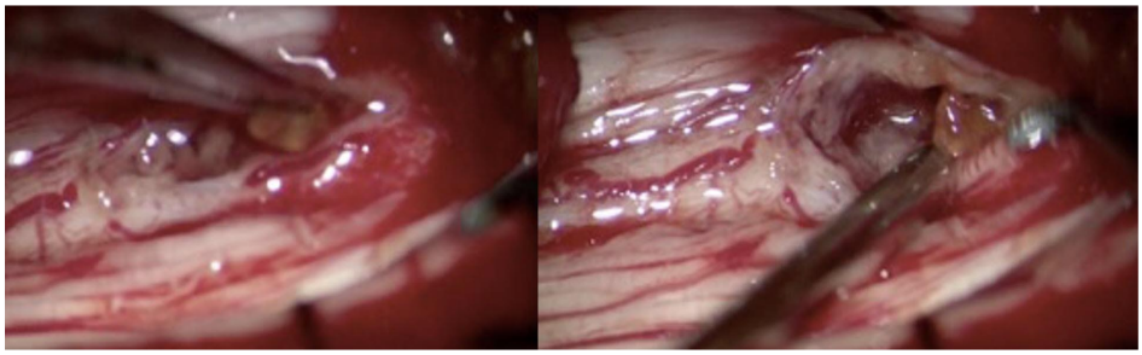

Figure 5: 6-year-old boy with spinal intramedullary cavernous angiomas. Intraoperative photograph revealing an intramedullary lesion with chronic blood.

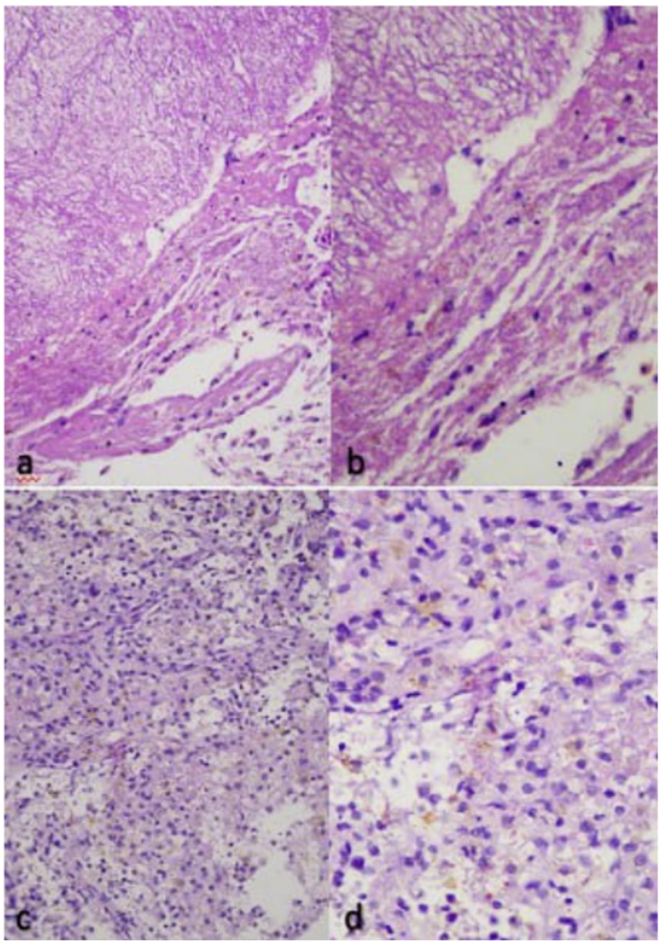

Figure 6: 6-year-old boy with spinal intramedullary cavernous angiomas. Histopathology of the surgical exited spinal intramedullary lesion.

a) Hematoxylin and Eosin stain (magnification 200x) and b) (magnification 400x) Cervical part lesion show fibrin and macrophage with hemosiderin, no malignancy was detected.
c) Hematoxylin and Eosin stain (magnification 200x) and d) (magnification 400x) Distal XII thoracic part lesion show macrophage with hemosiderin pigment and some capillary vessels, no malignancy was detected.

Etiology	• The exact etiology remains unknown. It is thought to develop in compensation for the absence of normal blood vessels due to hypoplasia, aplasia, or early occlusion of the developing vein.
Incidence	• 3 – 5% of all central nervous system lesions • 5 – 12% of all spinal vascular lesions • 1% of all intramedullary lesions in pediatric patients • Single or multiple
Gender ratio	• More common in women
Age Predilection	• The peak age: a fourth decade • In pediatric, peaks: below 4 years and over 12 years
Clinically	• Asymptomatic or may have various symptoms. • Symptom may be due to intracerebral or spinal hemorrhage: seizure or focal neurological deficit
Location	• Usually in the cervical and thoracic spinal cord • They may be associated with cavernous angiomas in other organs or the central nervous system
Imaging Finding	• Small size, eccentric location in axial, minimal enhancement with contrast, and absence of edema • Hemosiderin rim at T2WI
Treatment	• stable lesion: conservative • unstable: surgical resection
Prognosis	• Prognosis in children treated seems better than in adults • Surgical resection is a better prognosis in hemorrhagic condition
Pathology	• Consists of endothelial-lined caverns lacking appropriately formed tight junctions. The vascular leak due to poorly formed tight junctions leads to the clinical consequences of the disease

Table 1: Summary table of spinal intramedullary cavernous angioma.

Characteristics	**Cavernous Angioma**	**Hemorrhagic Ependymoma**
Size	Small size	Larger
Axial Location	Eccentric	Central canal
Edema	Absent	Present
Hemorrhagic sign	Hemosiderin rim	Hemorrhagic “cap sign” at the tumor poles
Contrast gadolinium MRI	Minimal enhancement	Strongly enhancement

Table 2: Differential diagnosis table for cavernous angioma and hemorrhagic ependymoma.

ABBREVIATIONS

C1 = Cervical 1
C6 = Cervical 6
DSA = Digital Subtraction Angiography
L1 = Lumbar 1
MRA = Magnetic Resonance Angiography
MRI = Magnetic Resonance Imaging
T1WI = T1 Weighted Image
T2WI = T2 Weighted Image

KEYWORDS

bleeding episode; cavernoma; cavernous angioma; intramedullary; magnetic resonance imaging

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