

Symptomatic Intramedullary Cavernous Angioma During Pregnancy. Case report

Angioma Cavernoso Intramedular Sintomático Durante Gestação. Relato de caso

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ABSTRACT

The cavernous angioma is a vascular malformation that can affect various organs, and its location in the spinal cord is unusual. We report a case of a pregnant patient who presented with pain in the lower limbs and urinary retention, diagnosed with cavernous angioma of the spinal cord and treated with surgical excision. Our case highlights this rare pathology and its initial presentation during pregnancy.

Key words: Cavernous angioma; Cavernoma; Spinal cord

RESUMO

O angioma cavernoso é uma malformação vascular que pode acometer diversos órgãos, sendo incomum sua localização na medula espinhal. Relatamos o caso de uma paciente grávida que apresentou dor em membros inferiores e retenção urinária, diagnosticada com angioma cavernoso da medula espinhal e tratada com ressecção cirúrgica da lesão. Nosso caso destaca essa rara patologia e sua apresentação inicial durante a gravidez.

Palavras-chave: Angioma cavernoso; Cavernoma; Medula espinhal

INTRODUCTION

The cavernous angioma is a vascular malformation that can affect several organs, and its location in the spinal cord is unusual. It can be asymptomatic or present symptoms in different ways, depending on its location and the type of complication. It has radiological and histological characteristics that allows its differentiation from other lesions of the nervous system. The best therapeutic option still remains as surgical resection. Diagnosis and early treatment may decrease progression to more extensive complications, and some patients may show some degree of recovery from the neurological deficit. We present a clinical case of a patient with cavernous angioma of the spinal cord, as well as discussion and conclusion regarding epidemiology, diagnosis, treatment and prognosis.

CASE REPORT

A 17-year-old female patient, pregnant (first gestation), with gestational age of 28 weeks and 5 days, without any prior comorbidities, presenting with lower limbs pain and no urinary excretion for 1 day. The pain was intense and associated with paresis of lower limbs without upper limbs involvement. She reported that 2 days before the establishment of the paresis, she only felt low back pain.

On the second day of hospitalization, there was worsening of the paresis of lower limbs, making ambulation impossible. A neurological evaluation was performed, from which were raised the hypotheses of transverse myelitis or expansive medullar process. Laboratory tests, lumbar puncture (LP) and magnetic

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resonance imaging (MRI) were performed. The last one showed images compatible with cavernous angioma of the spinal cord at T7 level (Figure 1).

On the third day of hospitalization, the patient evolved to paraplegia, loss of sphincter control and urinary retention. Decreased sensitivity, hyperreflexia and clonus in both lower limbs were also noted. The study of CSF showed xanthochromia, lipid aspect, acellular smears and scarce eosinophilic material.

She underwent a T6-78 laminectomy bilaterally. After durotomy, a midline myelotomy revealed a reddish mass. It had multiple feeders surrounding it. It was soft and hemorrhagic on manipulation. It was removed en bloc after circumferential dissection and the surrounding hematoma was evacuated (Figure 2). There was no improvement of neurological deficit already installed after the procedure.

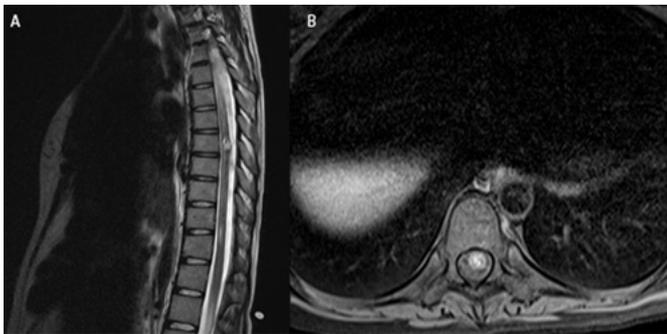


Figure 1. Magnetic resonance. **A.** T2-weighted, sagittal view. **B.** T2-weighted, axial view, demonstrating cavernous angioma on T7 level.

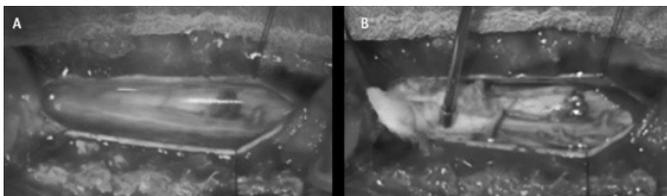


Figure 2. **A.** Operative site after T6–8 laminectomy and dura opening show no visible abnormalities on the spinal cord surface. **B.** After myelotomy, the cavernous malformation was exposed and removed completely following evacuation of the surrounding hematoma.

DISCUSSION

Epidemiology and pathophysiology

Cavernous angiomas or cavernomas can be found in the heart, kidney, liver, spleen, skin, and central nervous system. Its occurrence in the spinal cord is very rare^{1,2} and there has been an increase in incidence with the advent of MRI.

Currently, they are considered as vascular hamartomas, which by definition do not grow with mitotic activity^{1,2}. Generally, symptomatic patients are between 30 and 60 years of age, with a predominance of females (2:1)³⁻⁶. The prevalence of the disease in pregnancy is not described in the literature.

They can occur in two forms: sporadic or familial pattern. In the sporadic form, the pathophysiological mechanism is still not well understood, and it is possible that mutations in individual cell genes and exposure to radiation therapy could act as risk factors.

Regarding the familial pattern, the lesions tend to be multiple, being inherited in an autosomal dominant way, with high clinical penetrance¹⁻³. Up to the present moment, three genes whose mutations could predispose to cavernous angioma have been identified: 1) CCM1 (Cerebral Cavernous Malformation 1), located on the long arm of chromosome 7, position 21.2; 2) CCM2, located on the short arm of chromosome 7, position 13; and 3) CCM3, located on the long arm of chromosome 3, position 26.1. It is believed that both genes encode proteins involved in the development and maintenance of the structure of blood vessels. The CCM1 gene encodes the KRIT1 protein; CCM2, the Malcavernin protein; while CCM3, the protein Programmed Cell Death 10 (PDCD10)⁴.

Cavernomas occur in 0.5% to 0.7% of the population. These lesions are typically intra-axial, with 78% of the cases located in the supratentorial region and 22% in the infratentorial^{1,3}. Also, it corresponds to 3 to 15% of vascular malformations of the spinal cord^{3,5}. The frequency of occurrence of association between spinal and intracranial cavernomas is only 8%⁴.

In relation to their spinal topography, it may be cervical, thoracic or lumbosacral, with the thoracic segment being the most commonly affected⁵. In relation to the plane of the dura, it may be extradural or intradural, with the extradural lesion being subdivided into vertebral body lesions extending into the extradural space and lesions of the extradural space. The intradural lesions are subdivided into juxtamedullary and intramedullary. The injury of the vertebral body with extension to the extradural space is the most common type of lesion^{1,3}.

Histologically, cavernous angiomas are composed of irregular sinusoidal channels, with thin walls of endothelium and collagen, and scarce or absent elastic or smooth muscle fibers. There is no interposition of neural or glial tissue, which distinguishes it from capillary telangiectasia^{1,3}. The lesion may

have components of blood degradation, with hemosiderin being the most important. Hemosiderin is less abundant in extradural and vertebral body cavernomas, probably due to the more abundant vascularization at this site, as well as the absence of a blood-brain barrier in this topography, conditions that facilitate its removal. In juxtamedullary and intramedullary lesions, hemosiderin is almost always present in greater abundance^{1,3}.

Our case consisted of a young patient in relation to the most common age of the onset of malformation. The location of the lesion is one of the most unusual among the reported cases of the literature - spinal and intramedullary topography, which highlights big spectrum of this disease and the rarity of the case.

Clinical features

There are four clinical syndromes described: 1) slow and progressive spinal cord syndrome (most common); 2) acute medullary syndrome; 3) local pain; 4) radiculopathy^{1,11}.

The cavernous angiomas of the spinal cord may be asymptomatic and may present acute or chronic symptoms due to bleeding, thrombotic occlusion or medullary dysfunction by compression^{1,3}. Some authors report that there is an increased risk of bleeding in cavernous angioma during pregnancy due to increased levels of estrogen, which would cause changes in the walls of the vessels, predisposing to bleed^{4,7-10}. The main complications include subarachnoid haemorrhage, epidural hematoma, spinal cord and spinal roots compression, and ischemic changes with progressive chronic radiculomyelopathy^{1,3}.

In most patients with the acute or the chronic medullary syndrome, symptoms have been correlated with intralesional hemorrhage, whereas in cases with local pain or radiculopathy, symptoms have been correlated with progressive lesion growth¹.

Our patient presented a rapidly progressive disease, losing the spinal functions below the level of the lesion within 3 days after hospitalization. The probable etiology of this progression was an intralesional bleeding, which may be associated with the role of the pregnancy hormones over the blood vessels. It is emphasized that this is an unusual presentation of the cavernous angiomas, which, for the most part, present a slowly progressive symptomatology.

Imaging studies and lumbar puncture

The exam of choice for imaging studies is the MRI^{1,3,5,11,12}, which may show lesions with homogeneous hyperintense signal in T2, with or without hypointense ring indicating components of blood degradation, such as hemosiderin. In T1, it can present a signal of intensity similar to the marrow and muscle. The absence of the hypointense ring in the T1 and T2 weighted images is remarkable in the extradural lesion, due to the faster removal of hemosiderin in this location, in consequence the rich vascularization and absence of a blood-brain barrier in this topography.

The ovoid shape and the absence of an anatomical relationship with the adjacent intervertebral disc or with the exit of the nerve root helps to distinguish the cavernoma with disc herniation, while the homogenous signal of high intensity in T2 distinguishes it from a neurogenic tumor¹. Often the lesion extends to the intervertebral foramen, but usually it is not enlarged, which may help in the differential diagnosis with neurinoma¹⁻³.

Computed tomography may show a hyperdense area with low contrast uptake, with hypodense halo due to edema. The cavernoma can present as a structure exerting mass effect on the dura mater¹.

Cavernous angiomas are typically angiographically occult, since they do not have a large vascular supply and significant venous drainage, which makes this study very useful to rule out another pathologies^{1,5,6}.

The study of cerebrospinal fluid may reveal a slight increase in protein content and more rarely, xanthochromia⁵.

MRI has shown to be essential for the diagnosis of malformation, whereas there are no clinical signs that can confirm the etiology of the lesion. CSF has shown to be xanthochromic, which is uncommon according to the literature. However, it is noted that the rapid progression of the symptoms is possibly associated with spinal hemorrhage, which led to this aspect of the fluid.

Treatment and outcomes

The total surgical resection with microsurgical techniques is the treatment of choice^{1,3,4,6,11-14}. Total surgical resection may result in improvement (57%), stabilization (33%) or worsening (10%) of symptoms¹⁴.

Radiotherapy is reserved for non-resectable lesions, residual

lesions of partial resections or for patients with clinical contraindications for the surgical procedure^{1,3}. However, the benefits and outcomes of this technique are controversial in the literature.

In our case, the surgical procedure only resulted in stabilization of the neurological deficit.

CONCLUSION

In this context, can be concluded that cavernous spinal cord angioma is a very rare vascular malformation. However, it should be included in the differential diagnosis of spinal cord injuries. The clinical form of presentation depends on bleeding, thrombotic occlusion or compressive effect by expansion. MRI is the imaging exam of choice for diagnosis. Total surgical resection is still considered the best treatment, and radiotherapy remains restricted to particular situations, such as partial resection, contraindication to the surgical procedure, although the evidence of benefits is controversial in the literature. The improvement, stabilization or worsening of symptoms after surgery depends on the timing of the diagnosis and treated, with the best outcomes being associated with early intervention. Considering the possible propensity to bleed during pregnancy, this pathology should be suspected in the presence of spinal signs during pregnancy.

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The authors have no conflicts of interest to disclose.

Case report organized according CARE guideline (consensus-based clinical case reporting)