Subarachnoid Hemorrhage Caused by an Extramedullary Intradural Foramen Magnum Cavernous Malformation

Hemorragia Subaracnoidea Secundária a Cavernoma Intradural Extramedular no Forame Magno

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ABSTRACT

Objective: Pure subarachnoid hemorrhage (SAH) secondary to spine cavernous malformation is rare. Method: Female, 41 year old, with a massive intraventricular and SAH secondary to a extradural intradural foramen magnum cavernous malformation. The patient developed a sudden headache and drowsiness without neurological deficits. A CT scan revealed a massive SAH without intraparenchymal blood. Angiogram was negative for aneurysm or vascular malformation. A MRI showed an extramedullary intradural mass in the foramen magnum. She underwent a neurosurgical resection of the mass. Results: No complications due to resection and the pathological test revealed a cavernous malformation. Conclusion: This is the second case of foramen magnum cavernous malformation reported in literature. It should be considered a differential diagnosis of SAH with negative angiogram and surgical treatment usually provides good outcomes.

Key words: Cavernous Malformation, Foramen Magnum, Subarachnoid Hemorrhage, Extramedullary

INTRODUCTION

Ruptured cerebral aneurysm is the most common cause of nontraumatic subarachnoid hemorrhage (SAH)³. SAH patients with absence of proof aneurysm during the initial angiography may suffer a variety of diseases³⁵. Cavernous malformations (CM) are angiogram-negative vascular malformations usually supratentorial and intraparenchymal. When located in the spine they are usually found intramedullary and cause symptoms of medullar or radicular disfunction⁴.

Intradural extramedullary cavernous malformations (IECM) are extremely rare, even more at the foramen magnum location. About one third of IECM reported in literature was presented as SAH⁶⁹.

We report the second case of an extraparenchymal foramen magnum cavernous malformation and the first that was presented with a massive intracranial subarachnoid hemorrhage.

CASE PRESENTATION

A 41-years-old black woman presented with a severe sudden onset headache and vomits that began 20 hours before her admission. On neurological examination the patient was...
drowsy, with signs of meningism and no other neurological deficits.

A computed tomographic scan revealed a massive SAH with intraventricular blood and a clot that spanned the foramen magnum and a segment of the upper spinal canal (Fig 1). She was submitted to an angiogram that was negative for intracranial aneurysm or vascular malformation. Although, an area of early filling and slow washout of the epidural venous plexus at the left posterior canal margin of C1 and C2 was shown (Fig 1). Magnetic resonance imaging (MRI) was performed 7 days after the ictus and showed a loculated mass with a homogeneous enhancement component that displaced the medulla slightly anterior and to the right (Fig 1).

Ten days after the ictus episode the patient was submitted to a suboccipital craniotomy and C1 laminectomy in prone position. After the opening of the dura, an encapsulated bluish mass was visualized on the posterior surface of cervical cord and was entirely extraparenchymal (Fig 2). The mass was totally microscopically resected in one piece from spinal cord and arachnoid adherences.

The patient had an uneventful postoperative course and was discharged asymptomatic five days after the procedure. A MRI performed 6 months after the surgery evidenced a gross total resection (Fig 3).

The surgical specimen was composed of dilated vascular channels with thin walls lacking both smooth muscle and internal elastic lamina. Histopathology confirmed the diagnosis of cavernous malformation (Fig 3).
DISCUSSION

The prevalence of CMs involving the CNS in the general population is about 0.5%. Spinal CMs are rare and account for 5%–12% of all spinal vascular malformations and about 5% of all spinal cord lesions. Cavernous malformations in the spinal region have been described as originating in the vertebral bodies, epidural space exclusive of bone, the intradural extramedullary space or the intramedullary compartment4,6,9,10,11.

There are approximately 30 cases of IECM reported in literature. These lesions most commonly occur in the lumbar region, followed by the thoraco-lumbar, thoracic, and cervical regions and only one in the foramen magnum region as observed in our case6,9,11.

Cavernous malformations have a wide range of symptoms. The classic symptoms of brainstem and spinal cavernous malformations depend on their size and location. Symptoms are radicular if nerve root compression is present, or myelopathic if there is spinal cord or brainstem compression. Furthermore, symptom onset can be acute, subacute or chronic. Acute symptoms usually occur in patients with intralesional or extralesional hemorrhage1,6.

Babu et al., in a retrospective review, observed that 34% of IECM has SAH as first presentation11. This natural history of IECM seems to be different from the one of intraparenchimal cavernomas5,10. SAH may occur because of the propensity for subarachnoid hemorrhage as the result of the direct communication of intraparenchimal extradural lesions with the subarachnoid space. It has also been postulated that the restriction of movement because of adherence to the spinal roots may tend these lesions more likely to bleed. Our patient presented with a massive SAH with ventricular bleeding5,6,9.

Radiological diagnosis could be challenging. Most frequent lesions located at foramen magnum are meningiomas, schwannomas, metastasis, chordomas or aneurysms7,8. Tumors located at foramen magnum usually present as homogeneous enhancing masses in CT and or MRI, but only rarely manifest as SAH. Other vascular malformations or aneurysms can be ruled out by a negative angiogram as in our case.

Cavernous malformations must be considered as differential diagnosis when initial digital subtraction angiography (DSA) is negative in patients after SAH. Brain and cervical MRI should be included in the work up of these patients. Intracranial or spinal cord CM usually present associated intraparenchymal hemorrhage along with SAH, but could manifest solely as SAH. Intradural extramedullary cavernous malformations present with SAH in about one third of the cases3,9.

Treatment of cavernous malformation in general remains controversial. Size, location, presence of hemorrhage, history of seizure and age are some factors considered for surgical decision5,7. Intradural extramedullary cavernous malformations seems to have a different natural history that their intraparenchimal counterpart with a major tendency to bleed. Surgical treatment of these lesions results in an improvement of neurological function in the majority of patients. Resection of IECM can be performed safely, with the majority of patients experiencing improved neurological outcomes as shown in our case and should be considered the first choice of treatment of these lesions6,9,11.

CONCLUSION

Intradural extramedullar cavernous malformation located at foramen magnum is very rare. It has a tendency to bleed and cause SAH as first clinical manifestation. It should be considered as differential diagnosis of SAH with angiogram-negative. Surgical treatment of these lesions usually affords good outcomes and should be considered first line of treatment.

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