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Intramedullary cavernoma. A case report

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Abstract

The cavernoma is a vascular malformation made of well-circumscribed agglomeration of pseudo-capillaries. Bone marrow localization is rare and accounts for about 5% of spinal cord vascular lesions. Clinical symptomatology is marked by progressive bone marrow compression syndrome. The diagnosis is strongly evoked in magnetic resonance imaging and confirmed by histology. The management is essentially surgical.

We report a case of bone marrow cavernoma in a 38-year-old man seen in consultation, paraplegic for 2 weeks. The spinal cord MRI revealed a lesion opposite D11, evoking a cavernoma. The patient was operated on with total removal of the lesion; histology confirmed the diagnosis of cavernoma. The immediate post-operative follow-up was marked by the partial recovery of the deficit. We discuss, through this clinical case, the clinical, radiological and especially therapeutic aspects of the medullary cavernoma.

Keywords
cavernoma,
microsurgery,
spinal cord

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Introduction

Cavernomas are benign vascular abnormalities consisting of cavities into which blood circulates at low flow and low pressure. Intramedullary localization is unusual, represents about 5 to 12% of spinal vascular malformations and 3% of intramural vascular malformations (5% of spinal vascular malformations) (1). Unlike cerebral cavernomas, intramedullary cavernomas often have a more aggressive clinical course due to acute or rapidly progressive alteration of spinal cord functions. Asymptomatic forms are common. The diagnosis is strongly evoked by imaging, in particular by magnetic resonance, and confirmed by the anatomo-pathological study. Surgery is essential especially in symptomatic forms.

We report a case of dorsal intramedullary cavernoma and analyses in the light of literature, the clinical, radiological and surgical aspects of the spinal cavernoma.

Case report

This is a 38-year-old man patient with no significant medical history and who had a low back pain for 1 year. One year after back pain, it showed

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a total functional impotence of the lower limbs of rapidly progressive installation. We received him in consultation, coming from a neighbouring country, two weeks after the establishment of the deficit. The clinical examination found a conscious patient, a urinary tube in place (because of urine retention), paraplegic at 0/5, hypoaesthesia under umbilical region, a positive Babinski sign, sharp osteotendinous reflexes. Magnetic resonance imaging showed an irregular intramedullary lesion (previous reverse) opposite D11, in hyper signal in both T2 and T1 and surrounded by a hypo signal halo and myelopathy extended up to L1. The set evoked recent bleeding and the diagnosis of cavernoma was mentioned.

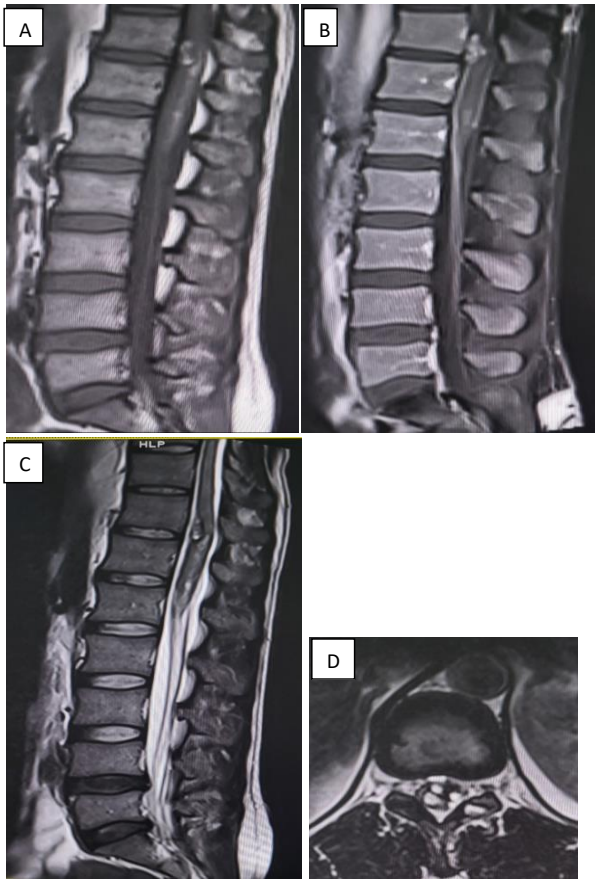
The patient was operated on, a laminectomy D11 and D12 was performed. After opening the hard mother, we discovered intramedullary in depth a globular, reddish and little hemorrhagic lesion. We realized a complete single-block removal.

The immediate postoperative follow-up at 24 hours were simple and marked by a clear improvement in neurological deficit, in particular, the removal of the urinary catheter and correct urination without retention, the improvement in the motor deficit whose rating increased to 1/5 in proximal (quadriceps) and 3/5 in distal (toes). Three months after the surgery, the patient presents an improvement, 4/5 in extension and 3/5 in flexion of the lower limbs.

The anatomopathological study confirmed the diagnosis of cavernoma

Discussion

The definition of cavernoma is histological, it is a vascular malformation made of dystrophic and ectasic blood vessels without interposition of tissue between the vessels (2). It is an unusual vascular malformation of the central nervous system. It occurs sporadically or hereditaryly. Cavernomas are generally not associated with other pathological entities. However, one case of cavernoma associated with oligodendroglioma, two cases associated with an NF1 and one case, with Klippel-Trenaumay syndrome, have been reported (3, 4). Medullary cavernomas are considered rare (5) and thoracic localisation is the most common (2, 6). The origin of these malformations is poorly known, they are said to be more common in women with a peak frequency between 30 and 60 years (6). The risk of bleeding is on average 2 to 3% each year (7). The rate of re-bleeding increases to 9 to 10% each year in case of cavernoma larger than 1cm, symptomatic patients and those with a history of haemorrhage (8).



Picture A: T1 sequence in the sagittal plane: intradural intramedullary lesion, in projection of D11-D12, in spontaneous T1 hypersignal associated with intramedullary haemorrhagic changes.

Picture B: T1 FS sequence after injection of gadolinium in the sagittal plane: absence of notable enhancement of the intramedullary intradural lesion.

Picture C: T2 sequence in the sagittal plane: the intradural intramedullary lesion 2cm in height is of heterogeneous signal with alternating zones in hypo and hypersignal T2 realizing the appearance of salt and pepper; it is associated with a range of edema in T2 hypersignal.

Picture D: SE T2 sequence in the axial plane: the intradural intramedullary lesion is lateralized to the left, pushing the marrow to the right, of heterogeneous signal with zones in hypo/hypersignal T2 in pepper and salt.

The clinic for intramedullary cavernomas is not pathognomonic. They can remain asymptomatic for a very long time or be responsible for a slowly progressing alteration of neurological functions or reveal themselves abruptly (9). Acute forms occur in

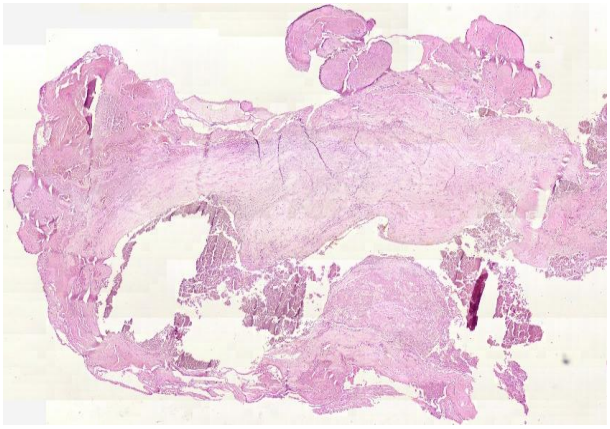
order of a hemorrhagic context while progressive forms (60%), can be reversible during the first month or be accompanied by a progressive pseudotumour worsening (10). Classical symptomatology is made of moderate spinal pain followed by muscle weakness in the lower limbs of progressive worsening, responsible for walking disorders (1, 3). In children, the clinical picture is most often acute in the form of a very rapid neurological deficit (3).

Radiological diagnosis is essentially based on magnetic resonance imaging (11). MRI currently represents the most efficient imagery for both dignostic and evolutionary monitoring (12, 13, 11). Technically, we must emphasize the interest of T2 in gradient echo (T2*). The typical aspect is that of a heterogeneous hyper signal lesion on the T1 and T2 sequences, surrounded by a hypointense peripheral area in T1 and T2 (12). Bone angiography is most often normal but retains its place in differential diagnosis especially with arteriovenous malformations (13, 14) or haemorrhagic spinal cord tumors (15). Computed tomography is less in demand and rarely poses diagnostic (13, 16).

The treatment of medullary cavernoma is neurosurgical with complete microsurgical resection of the malformation. Surgery improves the functional prognosis and very often allows a permanent cure without major sequelae (14, 5). The removal of the lesion must be done in operational theatre by bipolar coagulation of the surface, which decreases its volume and makes removal easier (17). Our case consisted of block removal without much coagulation. The basic principle is to avoid bleeding of the lesion during its removal to operate in a clean field so as not to lose sight of the border with the marrow (9). In the absence of surgical treatment, the evolution can be chronic myelopathy (14, 11). It is indeed essential in case of surgical indication to know the exact situation of the cavernoma, schematically near or in contact with the ventral surface, the lateral surface, or the dorsal surface of the marrow (18). Topography is therefore one of the determining criteria for the surgical indication and the expected clinical results (19). The superficial posterior forms make it possible to consider complete removal, most often without long-term neurological worsening. Centromedullary localisations are accessible through a classic median commissural myeloma with a transient probability of posterior cord damage. An anterior or anterolateral distribution is often respected in paucisymptomatic forms, in the case of a surgical approach (severe or recurrent deficit), a previous approach may be considered (10). This last localisation corresponds to that of our case but a later approach was enough for a complete excision.

Unanimity seems to be around abstention in asymptomatic forms while symptomatic forms hardly lend for discussion (18). However, a distinction must be made between haemorrhagic, deficit forms and only painful forms. The disappointing post-surgical results observed in painful forms make surgical indications cautious in this context (20).

Post-operative results are variable. A multicentre series (10) of 19 operated patients obtained 47% improvement, 42% aggravated and 5% unchanged. Another series (9) of 24 operated patients, did not get any improvement, however, notes 2 patients aggravated while 22 patients kept a condition similar to that observed before surgery. She recommends rapid surgery for any cavernoma associated with neurological disorders consistent with the localisation of the lesion.



Picture E: Histological appearance of a cavernous hemangioma. Hematoxylin and eosin coloring. Low magnification (x20).

Conclusion

The bone marrow localization of the cavernoma is unusual. The thoracic site is the most common. The origin of these vascular malformations is poorly known. The clinic is not pathognomonic, it is that of a rapidly progressive installation medullary compression. MRI is the most efficient examination for the diagnosis, topographical assessment and monitoring of cavernoma. The diagnosis of certainty

is histological. The management of symptomatic spinal cavernomas is surgical with complete resection of malformation followed by quality physiotherapy.

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