Case Report

Advances in Neurology and Neuroscience

Intramedullary Cavernoma: Case Report and Literature Review

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Submitted: 21 Aug 2019; **Accepted**: 10 Sep 2019; **Published**: 31 Dec 2019

Abstract

Introduction: *Cavernomas are benign vascular anomalies consisting of cavities where the blood circulates at low flow and at low pressure. Intramedullary localization is unusual, represents approximately 5 to 12% of spinal vascular malformations and 3% of intra-dural vascular malformations (5% of medullary vascular lesions).*

Observation: *A patient, aged 59, consulted for the abrupt installation of moderate back pain followed by predominant muscle weakness in the two lower limb of progressive worsening, responsible for gait disorders. The patient reported thermal hypoesthesia and heaviness of the two lower limbs that had been evolving for two years. The examination found a dorsal spinal cord compression syndrome. On the MRI, there were abnormalities of intramedullary signal of the dorsal (D11) spinal cord with bleeding stigmas suggestive of intramedullary cavernomas.*

Conclusion: *The management of the medullary cavernoma is essentially neurosurgical with complete microsurgical resection of the malformation. In the absence of surgical treatment, evolution can be to chronic myelopathy or neurological aggravation.*

Keywords: Cavernoma, Intramedullary, Vascular malformation

Introduction

Cavernoma or cavernous angioma is a vascular malformation consisting of a well-circumscribed agglomeration of pseudo-capillaries. *Intramedullary* localization is rare, it accounts for about 5% to 12% of spinal vascular malformations and 3% of intra-dural vascular malformations (21). It can be asymptomatic for a long time, or be responsible for a progressive or sudden alteration of the medullary functions [1]. His diagnosis based on magnetic resonance imaging (MRI) and pathology. Surgery is not always be stripped of complications. We report a case of dorsal *intramedullary cavernoma* and discuss clinical and radiological aspects as well as surgical indications.

Observation

It is a 59-year-old female patient with no history of any particular pathological condition, who presented with D10-D11 back pain with weakness in both lower limbs and progressively progressive for 2 years. This symptomatology was complicated by sphincteric disorders like urinary incontinence and constipation. The clinical examination revealed a spastic paraparesis rated at 2/5, with an umbilical sensory level. Patellar and Achilles osteotendinous reflexes were alive in both lower limbs. Dorso-lumbar magnetic resonance imaging visualized the presence of an *intramedullary* oval lesion opposite D-11 in T1, a heterogeneous hyper-signal in T2, surrounded by a T1 hypo-signal border (Figure 1 and 2). The diagnosis of *cavernoma* has been strongly suspected.

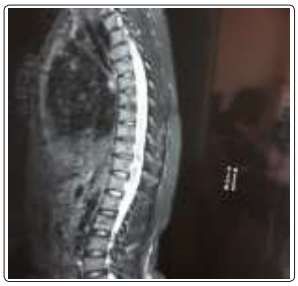

Figure 1: T2 Spinal Cord MRI, Intramedullary

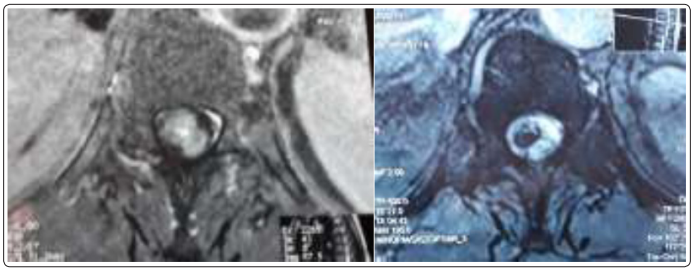

Figure 2: T1 Spinal CORD MRI with spinal cord compresssion by intramedullary process who appear heteregenous With hemorragic stigmata

Surgery was being indicated in front of a gradual worsening of neurological symptomatology. Through a laminectomy of D10-and D11, an opening and suspension of the dura mater, the tumor appeared in the form of a greyish-red, firm, non-aspirating, and slightly haemorrhagic arch with intradural hematoma who be evacuated (Figure 3). Under operative microscopy, the total excision of the lesion it was performed with sacrifice of a rootlet because the *cavernoma* infiltrated her. Postoperative follow-up was marked by a partial worsening of the motor deficit. Pathological examination confirmed the diagnosis of *cavernoma*. The patient was transferred to a specialized rehabilitation center for additional care and have good clinical result.

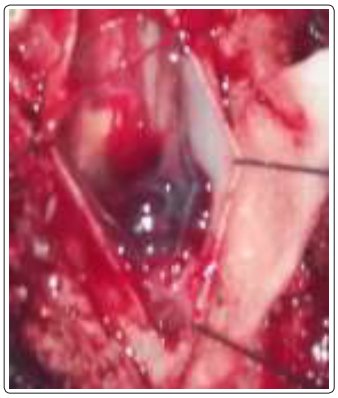

Figure 3: Per Procedure Microscopique View

Discussion

The *cavernoma* of the central nervous system may be single or multiple, rarely *intramedullary*. There are sporadic forms and familial forms with a genetic predisposition.

Etiopathogeny and Epidemiology

Intramedullary localization is extremely rare [2, 3]. Since its first description in 1912, only a few isolated cases have been published in the literature [4-6]. It accounts for about 5 to 12% of spinal vascular malformations and 3% intra-dural vascular malformations [3, 6-8]. Lefranc in a series of 368 tumors operated intramedullary, noted 24 cases of *intramedullary cavernoma* or 6.5% of cases [9]. The exact origin of these malformations is unknown, but they would seem more common in women with a peak of frequency between 30 and 60 years, of which about 55% of the cases are located on the dorsal level [8]. Shapes almost half of the all cases [10]. In our case, it is 59 old female patient and the level is Dorsal (D11).

Clinical Aspects

Clinical manifestations of *intramedullary cavernoma* depend on the lesional level, most often thoracic. The clinical presentation of *intramedullary cavernomas* is variable, they can remain a long time asymptomatic or manifest as a sudden or progressive neurological deficit [11]. The acute manifestation in the form of paraplegia or quadriplegia is often related to intra-tumor bleeding either spontaneous because of minimal trauma, physical exertion or during pregnancy [12-15].

Indeed almost 50% of patients presents with chronic rachialgia, root or root pain [16]. Barnwell, about in a series of 7 cases, highlighted the emergence of essentially sensory deficits and worsening progressive and noted the frequent occurrence of Brown-Sequard syndrome [17]. Hemorrhagic recurrence the absence of treatment seems to be the rule, in the end the direct toxic effect of hemosiderin around the cavernoma or disturbances of the surrounding microcirculation were also incriminated [18].

Paraclinical Diagnosis

Indeed, MRI is currently the exam of choice to evoke strongly the diagnosis of *cavernoma* in its typical form [11,13,19]. In spinal MRI, the diagnosis of *cavernoma* can take several aspects. Zabramski described in 1994 an MRI classification of cavernomas in four types [20]. The type I correspond to hypersignal in T1 and T2 related to recent hemorrhage and rich subacute thrombosis phenomena in methemoglobin. Type II is the most characteristic of these lesions is represented in T2 by a hyper and a hyposignal signal by a hypo peripheral halo signal; aspect said "popcorn or bee niche". The type III corresponds to a hypointense lesion in T1 and T2. Type IV manifests as an isosignal lesion T1 and T2, visible only in gradient echo. Differential diagnosis can still arise with a tumor *intramedullary*, especially when it is haemorrhagic (metastasis of melanoma) [15,18]. In that case, the use of gadolinium makes it possible to differentiate them; medullary tumors clearly strengthening, so that the *cavernome* little or not [19]. The dorso-lumbar magnetic resonance imaging in our case has visualized the presence of an *intramedullary* ovarian lesion opposite D10-D11 in iso-signal in T1, hypersignal heterogeneous in T2, surrounded by a border in hypo-signal T1 classified type II according to the classification of Zabramski [20].

Management

The management of *cavernomas* is essentially surgical as soon as the patient presents a clinical symptomatology with incomplete neurological deficit. In case of sudden paraplegia, it is sometimes it is preferable to postpone the intervention to avoid further marrow aggression surgical [5,7]. In case of accidental discovery or simple back pain, medical treatment and Clinical and radiological surveillance seems more appropriate. The surgical technique is the same as for *Intramedullary* tumor processes. After a laminectomy and dural opening centered on the area pathological, it is necessary to evacuate the hematoma and to excise the malformation under operating microscope; the first Moreover, the surgical procedure has the advantage of allowing a histological confirmation of the suspected diagnosis [10,21-23]. In our case after laminectomy and opening Dura mater we evacuate the hematoma and excise the lesion under microcope. Clinical forms of progressive progression should also benefit from surgical treatment because of the risk of sudden clinical deterioration. As in our case, the majority of authors insists on the possibility of transient postoperative worsening related to surgery [18,24]. The problem of preoperative scarring of the lesion can arise. The use of preoperative ultrasound can be of great help [25]. The realization of evoked potentials Intraoperative somaesthesia allows instantaneous quantification of the neurological risks of gesture; in our case we don't use it because we didn't have it.

Conclusion

Intramedullary cavernoma is rare. His diagnosis can currently be strongly suggested by MRI, Clinical forms of progressive progression should also benefit from surgical treatment, nevertheless the indications must be codified and the risks clearly explained to patients.

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