IMAGING FEATURES OF SPINAL EPIDURAL Cavernous Malformations


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SUMMARY

Cavernous angioma or cavernoma is a vascular malformation that may affect any area in the neuraxis. Epidural location is very rare and therefore seldom considered in the differential diagnosis of spinal cord compression. We report two cases of epidural cavernous angiomas. The first case is a solitary and purely epidural dorsal cavernous angioma without foraminal expansion or bone modification causing spinal cord compression in a 35 year old woman. The second case is a solitary epidural dorsal cavernous angioma with foraminal extension causing spinal cord compression in a 56 year old woman. Histological confirmation is available for both cases. We describe the MRI features of this lesion insisting on its differential diagnosis on imaging.

Key words: cavernous angioma, spine, epidural space, magnetic resonance imaging.

RÉSUMÉ

Aspects radiologiques des angiomes caverneux épiduraux

L’angiome caverneux est une malformation vasculaire qui peut siéger partout dans le système nerveux central. La localisation épidurale, en particulier dépourvue d’extension extra-rachidienne, est très rare et elle est peu considérée dans le diagnostic différentiel des masses épidurales. Nous rapportons deux observations d’angiomes caverneux épiduraux de topographie thoracique. Le premier est de siège strictement épidural révélé par une compression médullaire chez une femme de 35 ans et le second, survenue chez une femme de 56 ans, présente une extension extrarachidienne à travers un foramen intervertébral élargi.

Nous décrivons les caractères sémiologiques en imagerie par résonance magnétique de cette lésion sous ses différents aspects tout en insistant sur les diagnostics différentiels en imagerie.

Mots-clés : angiome caverneux, rachis, espace épidural, IRM.

INTRODUCTION

Cavernous angiomas or cavernomas are rare vascular malformations of the central nervous system [12]. They may develop anywhere in the neuraxis but they rarely affect the spine. At this level they can involve the vertebral body with or without epidural extension, epidural space without bony involvement, intradural extramedullary space and the spinal cord [14]. According to the literature, epidural cavernomas represent 12% of all intraspinal cavernomas and 4% of all spinal epidural tumors [3, 5, 16, 26, 33]. Recent advances in neuroradiologic imaging techniques have resulted in an increased number of cavernomas being diagnosed [1, 14, 26]. However, since the first description by Globus and Doshay in 1928 [5], only fewer than 60 cases have been reported in the literature [33]. In this paper, we report two cases of epidural cavernous angiomas without bony involvement.

CASE REPORTS

Case 1

A 35 year old woman had a history of progressive spinal cord compression. She complained of weakness and numbness in both legs lasting for a few months associated to sphincter disturbances. On admission, she was alert. The general physical examination was normal. The neurological examination revealed spastic paraparesis with hyperreflexia and prominent ankle clonus in the lower limbs. Bilateral positive Babinski signs were found. The pinprick sensation was absent below the level of T10. Urinary retention was verified. Plain film of
the thoracic spine showed no bony abnormality. Magnetic Resonance Imaging (MRI) was performed on a 1 Tesla magnet. Spin-echo (SE) T2-weighted images were acquired in the sagittal and axial planes. Axial and sagittal SE T1-weighted images were obtained before and after intravenous administration of gadolinium. MRI revealed a well defined dorsally located mass extending from T8 to T10. On T2-weighted MR images, the lesion was hyperintense, but slightly less than cerebrospinal fluid (figures 1a and 1b). On T1-weighted MR images, the mass had a heterogeneous signal with iso- and hyperintense foci relative to spinal cord and muscles (figure 1c) with marked enhancement after gadolinium administration (figures 1d and 1e).

At surgery, a soft reddish and hemorrhagic tumor lying over the dura was totally resected through a laminectomy. Histologically, it proved to be a cavernous angioma. Numerous abnormal vessels with thin walls lacking internal elastic lamina and muscularis were found (figures 1f and 1g). Postoperatively, progressive improvement of neurological symptoms was noted.

Case 2

A 56 year old woman presented walking difficulties. Clinical symptoms started one year prior to presentation with back pain radiating to the lower limbs and progressed over the following six months with developing hypesthesia and sphincter disturbances. The neurological examination showed a pyramidal syndrome of the lower limbs with a sensory level at T6. MRI was performed on a 1 Tesla magnet. It showed a posterior epidural mass extending from T4 to T6 that was T2 weighted hyperintense (figures 2a and 2b), T1 weighted iso-intense (figures 2c and 2d) with homogeneous contrast enhancement (figures 2e and 2f). This mass widened the left T5 intervertebral neural foramen and extended to the extraspinal space (figures 2b, 2d, 2f and 2g). Epidural cavernous angioma was diagnosed and confirmed by histologic study.

DISCUSSION

The embryogenesis of cavernous angiomas has long been debated and current classification defines them as congenital vascular malformations and not tumoral lesions. In fact, they are different from hemangiomas of infancy which are benign vascular tumors that always regress spontaneously. Therefore, correct terminology for these vascular malformations includes “cavernous angiomas, cavernomas or venous vascular malformations of cavernous type”, while the term hemangioma should be restricted to the common tumoral lesion of infancy [4, 13, 31].

Cavernous angiomas tend to develop between the third and sixth decade of life and may appear as sporadic or familial cases [7, 34]. Extra-axial cavernous angiomas are relatively rare lesions especially at the spinal level. According to Pia [29] they result more frequently from the extradural extension of vertebral body cavernomas and have rarely a pure spinal epidural location. At this level, they are usually in contact with the dura matter.

They are more frequently located at the thoracic level with predilection for the T2-T6 segment. The lumbosacral segment is less frequently affected [9], whereas cervical involvement is very rare [5, 9, 11, 14, 27, 34].

In the axial plane, they are usually located in the posterior part of the epidural space. This is explained by the larger size of this posterior side [37]. Foraminal extension is usually present [3, 11-13, 26, 34]. Some authors have reported extension to the paravertebral area [7, 24]. Even a pure foraminal location has been reported [5, 35].

These lesions are histologically characterized by the presence of large sinusoidal vascular spaces, lined by a thin wall, devoid of elastic and muscular tissue without intervening neural tissue, and surrounded by a fibrohyaline capsule [26, 38]. Unlike intradural lesions which are avascular, extradural lesions frequently have a rich vascularity [14].

The clinical course of these malformations is similar to that of other epidural lesions. It is commonly slow due to their progressive enlargement with spinal cord compression [14]. However, an acute onset of the symptoms may also be encountered. It can be explained by the sudden expansion of the volume of the lesions secondary to microhemorrhages and venous thrombosis within the malformations [26, 27] or by rupture of the cavernoma as the two cases reported by Koyama et al. [18] and Kubo et al. [19]. Patients may also present with radicular signs due either to intervertebral foramen extension [26, 34, 38] or to lumbar anterior and lateral localization mimicking clinically, in both cases, a disk herniation [5, 11-13, 26, 35]. These symptoms can be aggravated by some particular situations such as trauma, exercise and pregnancy [11, 33, 34].

From an imaging point of view, standard radiographs of the spine showed abnormalities such as pedicle erosion or foraminal enlargement in 41% of the reported cases [3]. Myelography usually shows an extradural block but gives no information about the nature of the lesion [11, 13, 21, 26, 38]. CT myelography confirms the extradural compression. It usually shows a homogeneous lesion with mass effect on the dura and frequent extension into the vertebral foramen which is rarely enlarged [8, 13, 38]. The CT scan can demonstrate eventual erosions of adjacent bone [34]. Spinal angiography is usually negative although intraoperative hemorrhage is frequently abundant [14].

In spite of the few cases studied in the literature, MRI is the most sensitive diagnostic imaging method and the technique of choice for a presumptive presurgical diagnosis [11, 26]. It shows the exact location and extension of the lesion and its anatomical relationship with the cord. It can even provide specific information about its nature [8, 26].

Epidural cavernous angioma usually has an oval shape, extends over 2 or more metameres, and sometimes flattens on the spinal cord [36]. But in some cases, it takes a nodular form and is located at the level of the intervertebral disc simulating a herniation...
FIG. 2. – MRI. Sagittal T2-weighted image shows a hyperintense well-defined-ovoid-epidural-lying mass extending from T4 to T6 (a). Axial T2-weighted image demonstrates the posterior location of the mass which displaces the spinal cord and extends through the left neural foramen (b). MRI. Sagittal T1-weighted image shows a homogenous epidural mass isointense to the spinal cord (c) and extending through the left foramen on the axial T1-weighted image (d). MRI. Sagittal (e) and axial (f) contrast-enhanced T1-weighted images demonstrating homogeneous enhancement of the mass. CT image. Enlargement of the left foramen (g).

FIG. 2. – IRM. Coupe sagittale T2 montrant une masse épidurale, hyperintense, bien limitée, étendue de T4 à T6 (a). La coupe axiale T2 précise le siège postérieur de la lésion qui refoule la moelle et s’étend à travers le canal de conjugaison gauche (b). IRM. Coupe sagittale T1 montrant une masse épidurale homogène isointense à la moelle (c) et s’étendant à travers le foramen gauche sur la coupe axiale T1 (d). IRM. Coupe sagittale (e) et axiale (f) T1 après injection de Gadolinium montrant un rehaussement homogène de la masse. Image tomodensitométrique. Elargissement du foramen gauche (g).
A paravertebral extension through the intervertebral foramen, as in our second case, is possible [34]. It has different imaging characteristics from intramedullary and intracerebral cavernomas [13]. In some cases, the signal can be mixed on all sequences similar to intracerebral or intramedullary lesions [26]. But usually, this lesion has a hyperintense signal on T2-weighted images due to the high content of stagnant blood [31] and a homogeneous isointense signal on T1-weighted images due to the slow blood flow [31, 38]. After gadolinium administration, the enhancement is generally mild and homogenous [26, 35] and rarely heterogeneous [1]. However, some epidural cavernomas do not change [12]. But the most striking difference is the lack of the low signal hemosiderin ring on both T1 and T2-weighted images. This is presumably caused by easier removal of blood products outside the blood-brain barrier [13, 38]. The low signal rim recognized around the lesion corresponds, according to some authors, to the dura interposed between the mass and spinal cord [34].

In some cases this lesion can be revealed by a spontaneous epidural hematoma which is hyperintense on both T1 and T2 sequences. This hypersignal is less hyperintense than fat on gradient echo sequences [13]. The differential diagnosis for extradural cavernomas includes several lesions but the MRI characteristics should be helpful to establish the diagnosis [13]. The spinal epidural meningioma is isointense to the spinal cord on both T1 and T2-weighted images with frequent broad dural attachment [10]. Schwannoma or neurofibroma is isointense to the spinal cord on T1-weighted images, hyperintense on T2-weighted sequences and the contrast enhancement is more intense. A target appearance with hyperintense rim and hypointense center is frequently seen on T2 and contrast enhanced T1 sequences. The enlargement of the inter-vertebral foramen is not a differentiating criterion because it can also be observed with cavernoma [10, 11, 34, 36, 38]. The majority of spinal angiolipomas are noninfiltrating and present as a well demarcated fusiform lesion occupying generally the posterior thoracic epidural space. Because of their lipomatous content, they are hyperintense on non-enhanced T1 and T2-weighted images [2, 38]. Rarely, there are small hypointense regions interposed through the high signal due to a high degree of vascularity. Post-contrast T1-weighted sequence shows heterogeneous enhancement that is more marked in these vascular structures. The use of fat saturation seems to better define the post-contrast enhancement and tumor margins. They can extend into adjacent neural foramina resulting in foraminal widening [30]. Epidural lipomatosis is well identified by MRI which shows hyperintense non-encapsulated epidural tissue that can be well differentiated from a hemorrhagic component by fat-sat sequence [15]. In the presence of hemorrhagic changes, the differential diagnosis with a hematoma (due to a vascular malformation or spontaneous) can be discussed. Spinal angiography is necessary to eliminate a vascular malformation. The absence of spontaneous resolution on several follow-up excludes the diagnosis of a pure hematoma. Lymphomas have low signal intensity on T1-weighted and heterogeneous hyperintense signal on T2-weighted images [25]. The multiplicity of lesions and bone involvement are usually found [5, 12, 13, 22, 31, 38]. Metastasis is the most considered diagnosis when the lesions have a posterior epidural location. In the lumbar spine, especially when the lesion occupies the antero-lateral epidural space, an extruded disk herniation should be considered in the differential diagnosis. But this fragment presents a low intensity signal on both T1 and T2-weighted MR sequences and peripheral enhancement [9, 16, 31]. This differential diagnosis seems far more difficult when in the presence of degenerative changes and posterior bulging disk adjacent to the mass.

Other rare differential diagnoses are possible: fibrosis scar [13], extra-osseous Ewing’s sarcoma of the spinal epidural space [23], epidural lymphangiomata [28, 32], epidural spread of multiple myeloma, even without evidence of vertebral bone destruction [28], epidural extramedullary haemopoiesis [6], epidural abscess or phlegmon which may occur without disc pathology [20].

Complete surgical resection of the epidural cavernous angioma is the treatment of choice, usually with good prognosis. However, severe intraoperative bleeding and anterior or intrathoracic extension can limit tumor removal. In these cases, radiotherapy may be delivered as an adjuvant to surgery. Embolization plays a less prominent role because pure epidural cavernous angiomata are often angiographically occult [12, 14, 27].

In summary, extra-axial cavernous angiomata must be considered as vascular malformations. They can arise in the spinal epidural space, so they should be included in the differential diagnosis of extra-dural spinal cord compression. The preoperative diagnosis is made easy since the MRI characteristics of this vascular malformation are known.

REFERENCES

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