Case Report

Conus medullaris hematomyelia associated with an intradural-extramedullary cavernous angioma

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A unique case of a 50-year-old woman with a *conus medullaris* hematomyelia associated with a low thoracic intradural-extramedullary cavernous angioma localized 2 cm above is reported. The patient had a 2-month history of progressive paraparesis, hypoesthesia of legs, and bowel and bladder disturbances. The symptoms worsened acutely during the last days before admission. A thoraco-lumbar MRI showed a space-occupying lesion at T10–T11 (vertebral interspace associated with a hematomyelia localized about 2 cm below. A T10–L1 laminectomy was performed and complete removal of both lesions was obtained with microsurgical technique.

A non-traumatic hematomyelia should always prompt the suspicion of a spinal AVM or, more rarely, of a cavernous angioma. The possible anatomical and clinical correlations of this unusual association are discussed.

Keywords: cavernous angioma; hematomyelia; intradural; extramedullary neoplasm; MRI; spinal surgery

Introduction

Cavernous angiomas or cavernomas represent 5–18% of intracranial vascular malformations¹ and 5–12% of spinal ones.^{1,2} These are usually localized at the level of the vertebral body with or without involvement of extradural space, whereas an exclusive extradural localization is rare.² About 3% are intradural, usually localized within the spinal cord.² An intradural-extramedullary cavernous angioma has been only occasionally described in the literature.^{2–4}

We report a new case of intradural-extramedullary cavernoma, at the level of the thoracic spinal cord and associated with a ‘satellite’ hematomyelia.

Case Report

A 50-year-old white woman had a 2-month history of motor and sensory deficits of the lower limbs. She ran an acute course during the last days before admission, with progressive inability to stand up, marked reduction of lower limbs sensitivity, and bowel and bladder dysfunction. On admission, a severe spastic paraparesis (left more than right) was observed: she was unable to raise up the limbs against gravity. Moreover, she had bilateral painful, thermic, and

tactile hypoesthesia (right more than left), with superior level at the umbilical transverse line, and severe sphincteric disturbances.

A dorsal-lumbar MRI (Figure 1a and b) showed a space-occupying lesion with inhomogeneous contrast enhancement at T10–T11 vertebral interspace, with the typical MR pattern of cavernous angioma, associated with an hyperintense lesion, with the typical MR pattern of blood, with maximum longitudinal diameter of 1.5 cm, localized about 2 cm below, at the upper portion of the *conus medullaris*. Spinal angiography was unremarkable.

A T10–L1 laminectomy was performed. The dura mater was opened in the midline and on the left side a roundish, red-blue, juxtamedullary mass, adhering to the ventral root of a spinal nerve (presumably T11) was found. The lesion was carefully dissected from the adjacent nervous structures and *en bloc* removed with microsurgical technique. After a linear incision of *conus medullaris* at the posterior longitudinal sulcus, about 2 cm below the extramedullary mass excised, an intramedullary spinal cord hematoma surrounded by gliotic tissue was found and removed.

On histological examination the extramedullary mass appeared to be a cavernous angioma (Figure 2). During the postoperative period an improvement of motor and sensory disturbance was observed. On discharge, she was able to move her legs against

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gravity and there was an improvement of hypoesthesia. Sphincteric disturbances unchanged.

Three months after operation she was able to walk with aids but the sphincteric functions were still impaired.

Discussion

In a recent review of spinal axis cavernous malformations, Harrison *et al*² confirm that intradural-extramedullary cavernomas are extremely rare. After our personal case previously reported,³ only one other case has been described in the available literature.² In all cases the malformation involved the thoraco-lumbar tract of the spinal cord, with tight relationship with a spinal nerve root.²

In the case observed by Ueda *et al*,⁴ the clinical picture began with a subarachnoid hemorrhage, but the present case is the first report of an intradural-extramedullary cavernous angioma associated with a satellite hematomyelia. Therefore, the peculiarity of the present case mainly resides in the possible combined pathogenesis of both pathologies.

The thrombosis or compression of a spinal cord vein may produce a centromedullary hemorrhagic infarction, with development of a transection syndrome, similar to the circulatory disturbances typical of subacute necrotizing myelopathy described by Foix and Alajouanine, caused by an extra- or intramedullary AVM.⁵ In our case the chronic compression exerted by the cavernous malformation on the ventral root exit zone, exacerbated by its volumetric increase due to recurrent intralesional bleeding, could have stressed the adjacent radicular and perimedullary venous system above the capability of supply ensured by the wide anastomotic net. Thus, it is possible that a hemorrhagic infarction developed within the underlying spinal cord when the hemodynamic venous compliance became inadequate. This interpretation seems to have anatomical support⁶ and is in accordance with the rapid onset and worsening of the clinical picture.

Figure 1 (a, b) Thoraco-lumbar MRI. T2-weighted, sagittal image. Space-occupying lesion, with inhomogeneous contrast enhancement, at T10–T11 vertebral interspace, with the typical pattern of a cavernous angioma. About 2 cm below, in relation to the upper portion of the conus medullaris, the image shows a hematomyelia with maximum longitudinal diameter of 1.5 cm

Figure 2 Histopathological photomicrographs. The intradural-extramedullary mass appeared to be a cavernous angioma

Notwithstanding our interpretation, however, it is not possible to exclude that the *conus medullaris* hematomyelia was produced by a tiny unidentified vascular malformation.

The post-operative improvement of motor and sensory deficits and the substantial lack of recovery of sphincteric disturbances observed in our patient appears to confirm the different role played by the extramedullary compression and by the centro-medullary hemorrhage.

In conclusion, a non-traumatic hematomyelia should always prompt the suspicion of a spinal AVM or, more rarely, of a cavernous angioma.

References

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