Lumbosacral myxopapillary ependymoma with intracranial dissemination

A 54-year-old man presented with a backache for two years and progressive paraparesis. There was no past medical history. MRI showed a lesion extended from L2 to the sacrum with craniospinal dissemination. Surgery failed because of relationship with the nerves and led to neurological disturbances, mainly sphincter dysfunction.

Imaging Findings:

MRI showed a huge expansile intradural extramedullary mass extending from D11 to the sacrum. The lesion was isointense to the spinal cord on T1W images (Fig. 1), hyperintense on T2W images (Fig. 2) and intensely enhancing (Fig. 4). There were T1, T2 and T2* hypointensities in the mass related to intratumoral haemorrhage (Fig. 3). The lesion leads marked widening of the lumbosacral part of the spinal subarachnoid space and scalloping of the corresponding vertebrae.

In the dorsal level, and intracranially (fourth ventricle, right cerebellopontine angle, temporal subarachnoid space) other masses were found related to dissemination along CSF pathways (Fig. 5).

There were also T2* hypointensities in the cerebellum and the cerebral cortex related to superficial siderosis (Fig. 6).

Discussion:

- Myxopapillary ependymoma is a subtype of ependymoma that occurs most of the time in the conus medullaris and filum terminale [1,2,3]. Myxopapillary ependymomas are highly vascular neoplasms characterised by abundant supporting fibrous connective tissue stroma, and mucin secretion by the tumour cells [1,2]. Intratumoral cysts, haemorrhage and reactive syrinx in the spinal cord can occur.

- Because presenting complaints are nonspecific, sometimes mimic discogenic pain and because most ependymoma of the cauda equina region grow slowly, the diagnosis is often delayed and is established after more than 2 years [2,4,5]. The use of MRI most of the time for symptoms attributed to a discogenic origin, allows approaching the diagnosis. - The MRI findings in this tumour are nonspecific, but some findings are suggestive. The classic MR imaging findings of myxopapillary ependymoma are: an intradural extramedullary location, expansion of the spinal canal, extensive tumour span, isointensity relative to the spinal cord on T1-weighted MR images, hyperintensity on T2-weighted MR images, intratumoral haemorrhage, intense enhancement, dissemination of tumour along CSF pathways, and superficial siderosis of the brain and spinal cord [1,2,6]. The differential diagnosis of myxopapillary ependymoma in the region of the cauda equina includes nerve sheath tumours, other ependymoma, astrocytoma,
metastases, meningioma, paraganglioma, lymphoma, gangliogioma, haemangioblastoma. Myxopapillary ependymomas with sacral destruction can be confused with chordoma, osseous metastases, aneurysmal bone cyst, giant cell tumour [2,4]. Furthermore, MRI is important to define the extension of tumour and its relationship to the intraspinal structures. This is all the more important as the surgery is less curative in multifocal tumour and tumour extend outside the thecal sac [4].

-Myxopapillary ependymoma is encapsulated in most cases and a total resection allows a favourable prognosis. However, a total resection in cases with involvement of the cauda equina or the filum terminale is difficult. A 50-70 % recurrence rate of spinal ependymoma has been reported after a subtotal resection [3]. To assess prognosis, adjuvant radiation therapy was recommended for cases without complete resection [3,5].

-Teaching point: Myxopapillary ependymoma must be kept in mind in case of:
  An intradural extramedullary lumbosacral huge tumour with no specific presenting complaints causing scalloping of the vertebral bodies with hypointensities in particular in rim shape and intensely enhanced.
In this typical location the main differential diagnoses are sacral tumour; chordoma or metastasis.
The earlier myxopapillary ependymoma is diagnosed, the more complete the surgical resection is and the better the prognosis is. For incomplete resection some advise radiotherapy.

**Differential Diagnosis List:** Myxopapillary ependymoma of the lumbosacral region resulting in superficial siderosis and dissemination along CSF pathways., Chordoma, Metastatic tumours, Nerve sheath tumours

**Final Diagnosis:** Myxopapillary ependymoma of the lumbosacral region resulting in superficial siderosis and dissemination along CSF pathways.

**References:**

Figure 1

Description: Expansion of lumbosacral spinal canal and scalloping of the posterior aspect of vertebral bodies (*). Origin: Department of Radiology, National Institute of Neurology, Tunis, Tunisia
Description: Intradural lumbosacral tumour predominantly hyperintense filling the spinal canal leading to scalloping of vertebral bodies. Hypointensities related to haemorrhage are seen inferiorly (arrow). Dissemination along CSF pathways with dorsal lesion. Origin: Department of Radiology, INN, Tunis, Tunisia.
Figure 3

**Description:** Hypointensities mainly in inferior part of the tumour related to haemorrhage. **Origin:** Department of Radiology, INN, Tunis, Tunisia
Figure 4

Description: Contrast enhanced T1 weighted images show intense enhancement of tumour. Origin: Department of Radiology, INN, Tunis, Tunisia
Figure 5

Description: Dissemination along CSF pathways with tumours in the fourth ventricle, foramen of Luschka, right cerebello pontine angle. Origin: Department of Radiology, INN, Tunis, Tunisia
Figure 6

Description: Hypointensities related to superficial siderosis due to the deposition of haemosiderin after episodes of haemorrhage from tumours. Origin: Department of Radiology, INN, Tunis, Tunisia