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Isolated lumbar myxopapillary ependymoma Case-report

Sameh Achoura, Hajer Kammoun, Hichem Ammar, Med Dehmani Yedaes, Khaled Radhouen, Ridha Chikili

University Tunis El Manar, Tunisia

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INTRODUCTION

Spinal ependymoma is described typically as an intramedullary tumor arising from ependymal cells lining the ventricles and the central spinal canal. Intradural extramedullary (IDEM) ependymomas subtype are extremely rare. (1)

Case-report:

It's about a 32-year-old patient with no past medical history who was admitted to our neurosurgical department for non-systematized bilateral lumbocrural pain set on upon an intense sport activity limiting his ability of walking with a perimeter reduced to 50 meters deteriorating progressively over 6 months.

The patient revealed the onset of the same symptomatology 4 years ago resolving spontaneously and not explored.

On the exam, the patient had only a contracture of the lumbar paravertebral muscles and a bilateral positive Lasegue sign with no motor nor sensitive deficit.

An MRI was performed showing an intradural extramedullary medial central process in the level of L2L3 measuring 13*17*24mm recalling at first a neurinoma. (Figure 1)

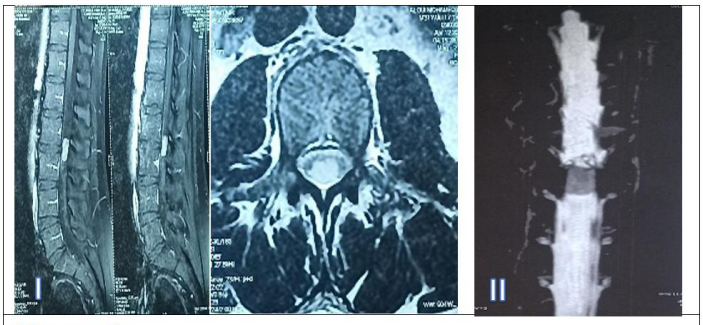

Figure 1:

I. Lumbar MRI on sagittal and axial planes:

Intradural extramedullary medial central process in the level of L2L3 homogenously hyperintense in T2 measuring 13*17*24mm filling the central canal and conflicting with its roots content

I. Myelography showing a stop frame in the level of L2L3

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The patient underwent L2 and L3 laminectomy and durotomy revealing a crumbly and bleeding tumor that was well limited and that emerged spontaneously under pressure (Figure 2) enabling us to make a complete resection. The patient evolved well and was discharged on the 2nd day after surgery.

Figure 2:
A crumbly and bleeding well-limited tumor emerging spontaneously upon durotomy

The anatomopathological exam revealed a lumbar intradural papillary proliferation presenting extensive myxoid changes consistent with a myxo-papillary ependymoma.

DISCUSSION

Between 30 and 40 cases of IDEM tumors have been described in literature to date. (2–4)

A female predominance was noted. The age ranged between 11 and 87 with an average around 45 years old. The localization was mainly thoracic. Only 3 cases had an isolated lumbar localization. (5)

According to the 2021 World Health Organization (WHO) classification of the central nervous system tumors, ependymal tumors are now classified according to a combination of histopathological, molecular features as well as anatomic site. Myxopapillary ependymoma is morphologically and histologically distinct from other ependymoma subtypes and is currently considered a grade 2 tumor rather than 1, since its likelihood of recurrence is now understood to be similar to conventional spinal ependymoma. (6)

The myxopapillary histological variant is known to have a slight male predominance and a peak age in young adults at 3rd decade. (7)

Myxopapillary ependymomas often arise from the filum terminale and are encountered in the conus medullaris area as the result of cerebrospinal fluid (CSF) seeding.

In our case, the tumor was located in the filum terminale’s area in the level of L2L3.

Concerning the prognosis, IDEM spinal ependymoma can recur, undergo anaplastic transformation, or metastasize. In these cases, gross total resection may not always be possible, and the prognosis becomes poor. However, the myxopapillary ependymoma is known to have a better prognosis than the ordinary type of ependymoma and even better in adults than in children. (8)

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CONCLUSION

IDEM isolated lumbar myxopapillary ependymoma is rare but should be suspected as a differentialdiagnosis of neurinoma with relatively a good prognosis. Early diagnosis and surgery remain the maincontributors to better neurologic outcomes. (9)

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