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Case Report

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Primary Multi-Focal Myxopapillary Ependymoma of the Spine: One Case Report and Literature Review

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Abstract

Background: Myxopapillary ependymoma is a rare type of central nervous system neoplasm mainly located at cauda equina and filum terminale. These lesions are considered benign tumors with long-term survival. However, a more aggressive behavior with a multifocal presentation along the neuroaxisis have been reported.

Case report: Here we present a case of double primary myxopapillary ependymoma of the spinal cord in a 16-years-old male. After surgery, the clinical decision was for craniospinal irradiation. The patient suffered no complication after the procedures during six months of follow-up.

Conclusions: This presentation is rare and, so far, at the best of our knowledge, have been previously reported only 52 similar cases. We hereby present a new case and a review of the literature.

Introduction

Ependymoma is one of the least common central nervous system neoplasm (CNS) usually occurring in children and young adults. Myxopapillary ependymoma (MPE) is a subtype of ependymoma located almost exclusively at the cauda equina and the filum terminale. First described by Kernohan in 1932, it's characterized by formation of papillae and mucin production [1]. Generally, MPE is designated histologically as a grade I neoplasm [2]. However, dissemination along the cerebrospinal axis and metastasis to distant sites have been reported [3-8]. The following is a case of a young male patient presenting with low back pain and radicular symptoms caused by two primary myxopapillary ependymomas.

Case report

A previously healthy 16-years-old male was admitted to our hospital with a history of mechanical type back pain which responded well to rest. No history of trauma or infection was reported. General physical examination showed no neurological deficits.

Magnetic resonance imaging (MRI) scans revealed two well-circumscribed intradural extramedullary lesions. One located at L5-S2 level and the other at L2-L3 level. The masses enhanced homogeneously on T1-weighted gadolinium-enhanced MRI. No other tumor mass was detected elsewhere by MRI (Figure 1).

Due to the unusual features of this case, it was decided to explore the caudal lesion first and waiting for the histopathological

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report. The patient underwent open surgery consisting of a lumbar laminectomy. On opening the dura, the tumor appeared as a soft, grayish-pink, and highly vascularized mass measuring about 6 cm. The lesion appeared well encapsulated and the capsule was intact with no signs of previous rupture. A sub-total resection of the tumor was achieved. The postoperative course was uneventful.

Multiple tissue fragments, measuring 2 X 2 X 0,6 cm were submitted for histology. Tissue were fixed in 4% buffered formalin and embedded in paraffin. Conventional paraffin sections were made and hematoxylin and eosin (H&E) staining was performed. Pathological examination revealed a glial neoplasm characterized by cuboidal to elongated tumor cells arranged around blood vessels forming pseudorosettes. A mucoid matrix material had accumulated between the tumor cells and blood vassels (Figure 2).

Immunohistochemical studies were performed using the standard streptavidin-biotin-labelling technique and commercially available antibodies. Immunohistochemical staining demonstrated positive stain for glial fibrilary acidic protein (GFAP), vimentin and S-100 protein and negative stain for cytokeratins. The mucoid matrix material was readily highlighted by Alcian Blue stain (Figure 2).

A diagnosis of grade I myxopapillary ependymoma was made based on the histological appearance and immunohistochemical profile. However, the lesion showed relatively high Mib-1 labeling index up to 15%, which raised the possibility of a more aggressive behavior (Figure 2).

A second surgery on the rostral lesion was performed about one month later through lumbar laminectomy. A mass lesion, of about 1,5 cm, similar to the first was totally removed.

Multiple tissue fragments, measuring 1,5 X 1,5 X 0,3 cm, were submitted for histology. Histological examination and the immunohistochemical staining showed the same findings as those of the first operation, indicating that the two tumors were homologous (Figure 3).

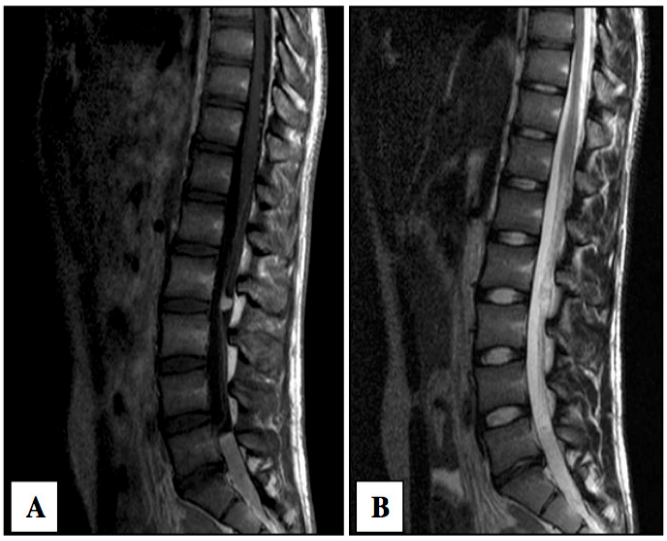

Figure 1: Preoperative MRI. Sagittal T1-weighted gadolinium-enhanced (A) and T2- weighted (B) magnetic resonance imaging demonstrating two enhancing intradural lesions. One extending from L5 to S2 and other smaller lesion at L2-L3 level.

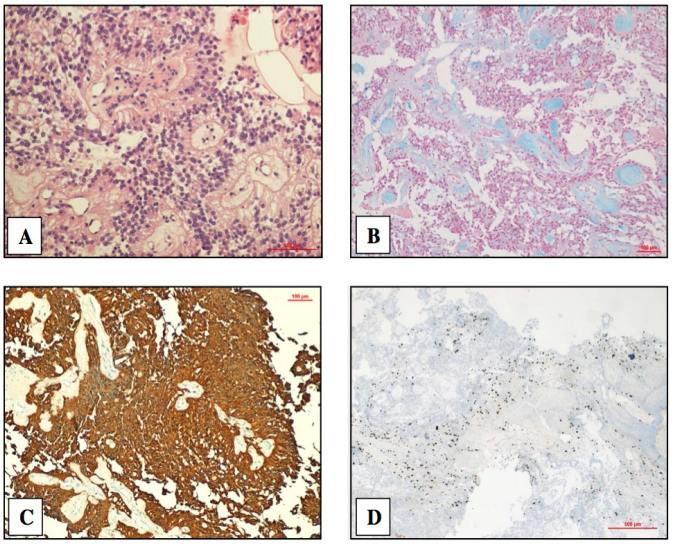

Figure 2: Histopathological findings. Tumor cell show radial processes around the vessels; Cuboidal and columnar tumor cells are arranged like pseudorosettes around blood vessels that form the cores of papillary processes (A, H&E, original magnification 20x). The perivascular myxoid material is Alcian blue positive (B, Alcian Blue, original magnification 10x). Tumor cells are diffusely positive for GFAP (C, GFAP, original magnification 10x). The tumor shows high a Mib-1 labeling index of 15% (D, Mib-1, original magnification 4x).

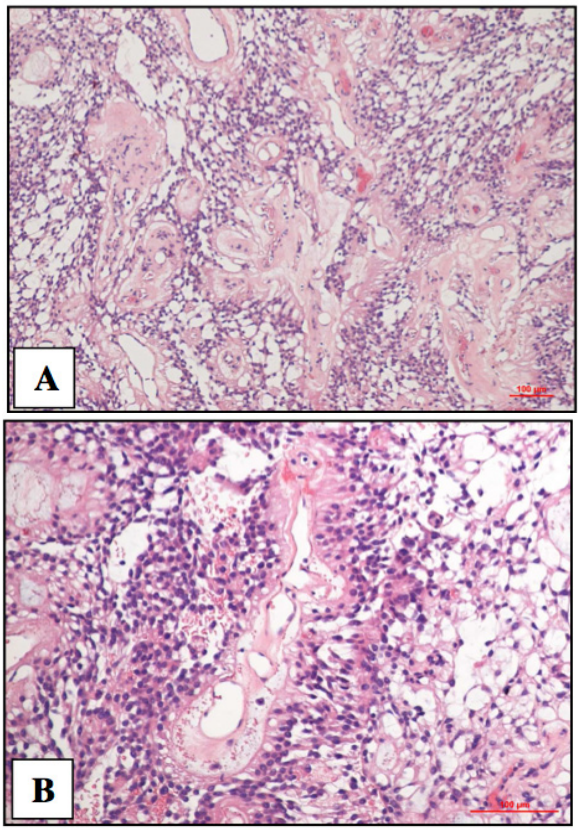

Figure 3: Histopathological findings. The tumor is formed by papillary structures surrounded by a single layer of columnar cells with round nuclei and delicate chromatin. The cores of the papillae have a central blood vessel surrounded by a mucinous/myxoid matrix. (A, H&E original magnification 10x; B, H&E original magnification 20x).

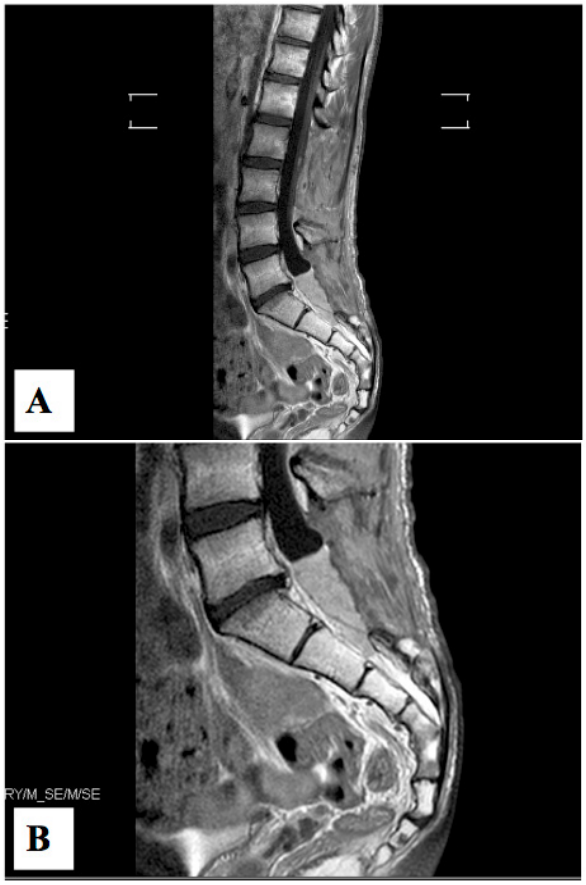

Figure 4: Sagittal T1-weighted gadolinium-enhanced magnetic resonance imaging at four months of follow up. MRI showed no signs of tumor recurrence for the rostral lesion (A) and a reduction of the residual part of the tumor localized between L5 and S2 (B).

Due to the multifocal disease, the subtotal resection of the caudal lesion and the high Mib-1 labeling index, the patient received postoperative craniospinal radiotherapy for a total dose of 39,6 Gy in 1,8 Gy per fraction. A boost was given for an additional dose of 10,8 Gy and a cumulative dose of 50,4 Gy on the preoperative extent of the disease. Radiation treatment was performed with helical Tomotherapy.

MRI examination at six months showed no signs of tumor recurrence for the rostral lesion and a reduction of the residual part of the caudal mass (Figure 4).

Discussion

MPEs are slow-growing tumor and are designated as grade I neoplasm according to the WHO classification [2]. Despite that, multiple tumors located on the spine, tumor recurrences and distant metastases have occasionally been described. At this time 52 multiple spinal cases, including our case, have been reported in literature.

Of the 52 multiple spinal cases described in literature, data about sex, age, location and treatment were available only for 29 (29/52; 55,8%) cases. Of those 24 (24/29; 82,8%) were males and 5 (5/29; 17,2%) were females. The age of the patients range from 7 to 63 years. The mean age for the available cases is 26,3 years.

On 52 multifocal spine MPE lumbar region was involved in most of the cases, following by sacral and dorsal region. In most cases (20/29; 69%) the lesions were two, in 5 cases were reported 3 lesions (5/29; 17,2%) and in 4 cases (4/29; 13,8%) there were more than three lesions.

Patients usually present with sensory disturbance, back and legs pain, bowel or bladder dysfunction and ascending neurological deficits [29].

MRI is the most common neuroimaging technique used for establishing the diagnosis in MPE but not specific signal characteristics have been identified. The tumors tend to be hypointense to isointense on T1-weighted images and hyperintense on proton-density and T2-weighted images [10].

Surgery is the treatment of choice for MPE and it must be considered also in case of multiple lesions. Of the 52 multiple cases described in literature, a gross total resection was achieved in 16 (16/28; 21,4%) patients, whereas a subtotal resection was performed in 11 (11/28; 39,3%). A biopsy was performed only in 1 (1/28; 3,6%) patient.

In 11 (11/28; 39,3%) cases radiation therapy as adjuvant treatment after resection was performed. The necessity of radiation therapy of MPE has long been debated. After partial or subtotal tumor removal, an improve in survival rates and a decrease in recurrences, have been documented using post-operative radiation therapy [11-16]. Furthermore, some authors have proposed post-operative radiation even when a gross total resection has been surgically achieved [13,17-19]. Usually the volume of radiotherapy consisted of tumor or surgery bed plus margins. In case of multifocal presentation craniospinal irradiation was suggested.

In the present case, craniospinal radiation therapy was performed after surgery because the patient had a multi-focal type of MPE, high Mib-1 labeling index and the tumor was removed in stages.

Despite extensive investigation, the histogenesis of multiple spinal MPE is still an unsolved problem. Some authors suggested that multiple lesions may be due to drop dissemination in the subarachnoid space also supported by a previous trauma that can cause haemorrhage and tumor rupture [20-24]. In our case, preoperative imaging and intra-operative findings documented two well circumscribed lesions and no evidence of dissemination was found. No history of previous trauma was reported and no sign of haemorrhage have been documented intra- operative or at microscopic examination. Given these findings, we concluded that the lesions here described were independent and not distant metastasis or dissemination along the neuraxis.

In general, the prognosis of MPE is favorable with a median survival of 19 years following a gross total removal and 14 years following a subtotal removal [25]. Despite that, a long-term follow-up is essential. In fact, Rezai et al analyzed 140 patients with surgically treated ependymoma, and reported that MPEs were 3,6 times more likely to have a shorter time to dissemination than others low-grade tumors [25].

In recent years, to better predict tumor recurrence and/or metastasis in MPEs, a variety of cell proliferation markers have been tested. For this purpose, Prayson et al analyzed Mib-1 labeling

index in 14 patients with MPE. The reported data showed that patients with higher Mib-1 labelling index had tumor recurrence [26]. Compared to these data, the Mib-1 labelling index in our case was extremely high (15%), indicating a high likelihood of tumor growth, recurrence and/or metastasis.

Conclusions

Patients with MPE are at risk for multiple lesions. Pre-operative identification of multiple MPEs is essential for considering the right surgical approach. Post-operative radiation therapy can be used to avid or reduce the likelihood of dissemination. Although more dates need to be collected, the evaluation of Mib-1 labelling index may help to identifying tumors with a high likelihood of recurrence and/or metastasis. Finally, a long-term follow-up is recommended in these patients.

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