

Primary Intradural Extramedullary Myxopapillary Ependymoma

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We report a rare case of primary intradural extramedullary myxopapillary ependymoma of the spinal cord. A 45-year-old woman was admitted to the author's institution with a history of progressive paraparesis (grade IV/IV) with back pain. Neurologic examination revealed decreased sensation below T12 sensory dermatome level. Magnetic resonance imaging (MRI) revealed an intermediate enhanced intradural extramedullary tumor extending from T12. Total resection of the tumor was achieved by T12 laminectomy. Intraoperatively, there was no finding of attachment to rootlet and dura. Histopathological examination identified the tumor as a myxopapillary ependymoma. To the best of my knowledge, this is the first reported case of primary intradural extramedullary ependymoma in Korea.

KEY WORDS : Intradural extramedullary · Myxopapillary ependymoma.

Introduction

Although ependymoma has been well described as a frequent intramedullary tumor or as a tumor arising from the conus medullaris or filum terminale, the intradural extramedullary location of this glial lesion is exceptional^{1,3-5,7}. The authors present the case of an unusual ependymal location and discusses a method of surgical resection and a pathophysiologic hypothesis.

Case Report

A 45-year-old woman with a 1 year history of back pain and weakness of lower limbs (Grade IV/Grade IV) was presented. Neurological examination showed decreased sensation below T12 dermatome level. However, she denied any difficulties with bowel or bladder function.

Lumbar spinal magnetic resonance imaging (MRI) revealed a 1.2 × 0.6 × 0.6 cm intradural extramedullary mass at T12. The lesion showed intermediate signal intensity on T1-weighted sagittal and axial images after intravenous contrast injection (Fig. 1). Based on the MRI findings, our differential diagnosis for this intradural extramedullary mass lesion included nerve sheath tumor and meningioma.

After performing standard laminectomy at the T12, we opened the dura exposing a circumscribed, encapsulated, dark pinkish mass without infiltration of the spinal cord. Under microscopic vision, the mass appeared to have adhesion rootlets at its base, but it did not seem to be attached to the rootlets or dura mater. Microscopic features, such as the tumor's encapsulation, the apparent lack of attachment to rootlets or dura mater suggested that this tumor was atypical type of intradural extramedullary mass lesion included nerve sheath tumor and meningioma. We achieved total resection by separating adhesion

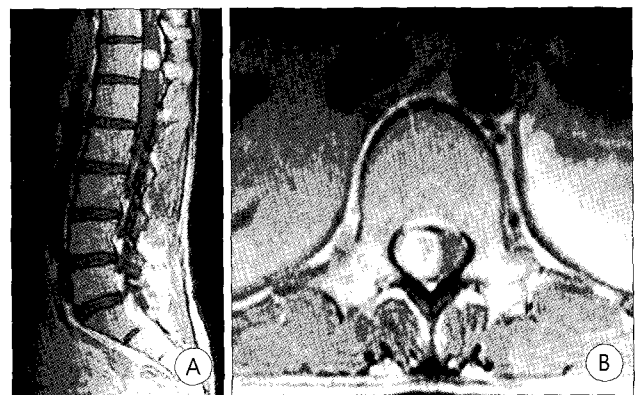


Fig. 1. A, B : T1-weighted magnetic resonance images with gadolinium enhancement shows an intermediate enhancing mass at T12 level.

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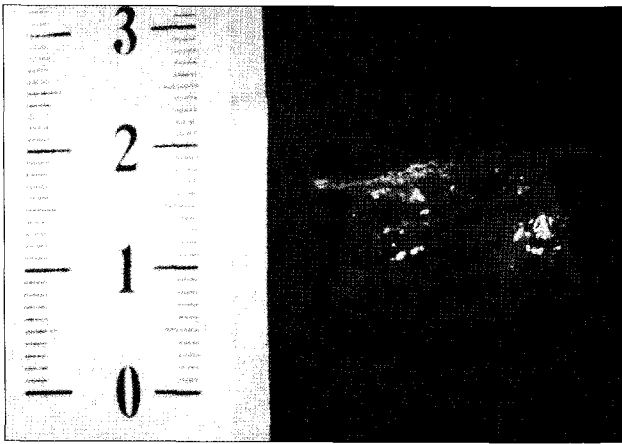


Fig. 2. Surgical specimen shows encapsulated and dark pinkish mass.

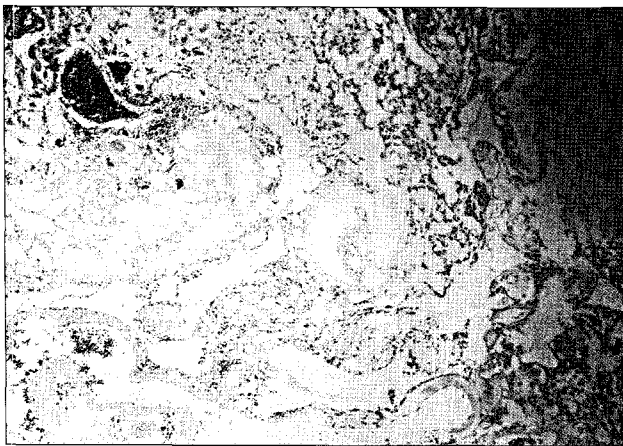


Fig. 3. Histopathological findings of the myxopapillary ependymoma. There are pseudopapillary or reticular structures formed by cuboidal or flat cells with regular nuclei, and there is an abundant amount of stroma showing conspicuous myxoid change (H&E, X100).

without difficulty (Fig. 2).

Histopathological examination identified the tumor as a myxopapillary ependymoma. The tumor cells were characterized by pseudopapillae or reticular structures formed by cuboidal or flat cells with regular nuclei, and there was an abundant amount of stroma showing myxoid change (Fig. 3). Cranial and whole spinal axis MRI examination revealed no other mass lesions. The postoperative outcome was favorable. The patient has had a good clinical recovery and no recurrence after 6 months of follow up evaluation.

Discussion

Location of ependymoma outside the central nervous system has been exceptionally reported in the literature located particularly at the cauda equina or the filum terminale²⁾. Clinical presentation may be pain, followed by a progressive history of medullary compression explainable by the extramedullary location of the lesion.

In the presented case, preoperative radiological evaluation and observation of the mass led to a tentative diagnosis of nerve sheath tumor or meningioma. Macroscopic features, such as the tumor's encapsulation, the apparent lack of attachment to the spinal cord, histopathologic findings of ependymal lining that indicated its origin and the absence of any neoplastic process within the brain or spinal cord suggested that this tumor arose from ectopic ependymal cells. Pathology is of the utmost importance for predicting the patient's survival rate.

Myxopapillary ependymoma is slowly growing tumor that may attain large size before clinical detection, occasionally filling and expanding the spinal canal. Histologically, myxopapillary ependymoma is a benign (WHO grade 1) tumor with a peak incidence of the 4th decade of life and is generally confined to the lumbosacral spinal cord. Although myxopapillary ependymomas are Grade I tumors with a tendency for slow growth and local recurrence, they are capable of spread within the central nervous system and extraneural metastasis^{11,12)}.

Since prognosis is directly related to the extent of surgical resection, it is important to treat ependymomas aggressively with surgical resection. Yet there is controversy over the benefit of radiation therapy (RT). Although in the past, RT was considered standard adjuvant therapy, today most authors advocate using RT only in cases of recurrence^{1,2,6,8-10)}. Even after gross total resection, the recurrence rate for myxopapillary ependymoma is 10 to 19%. Thus, it is important to follow these patients closely with MRI scans.

The precise histogenesis of the ependymoma written in this report could not be absolutely determined. The encapsulated feature of these tumors without relation to the central nervous system and without other medullary or cerebral locations exclude the hypothesis of ependymal metastasis into the intradural space, and such feature indicates a primary extramedullary ependymoma. The authors of the present study agree with Cooper et al.³⁾, who presumed in their 15 cases of primary extramedullary glioma with one ependymoma that these tumors developed from heterotopic glial cells located in the intradural extramedullary space. This hypothesis was supported by the descriptions of extraspinal ependymomas located in the subcutaneous tissue.

Conclusion

We report a rare case of primary intradural extramedullary myxopapillary ependymoma with a review of related articles.

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