Case Report

Lumbar myxopapillary ependymoma mimicking neurofibroma

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Objective: To report a case of lumbar myxopapillary ependymoma in whom neuroradiological and surgical findings strongly suggested neurofibroma.

Clinical presentation: The patient presented with a 2 year history of progressive monoparesis and monoparesia of his right leg. He reported having minimal fecal and urinary incontinence.

Intervention: Total resection of the tumor was achieved by total L1-L2 laminectomies. There was no attachment to the spinal cord and dura.

Conclusion: In spite of contemporary sophisticated neuroradiological facilities, we may still have diagnostic difficulties in some spinal tumors.

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Introduction

Ependymomas are rare tumors derived from ependymal cells in the spinal cord. They represent 2% of all primary tumors of the central nervous system.¹ Thirty to 60% of all spinal gliomas, and 40 to 60% of them occur in the lumbar region.² In this report, we describe a case of lumbar primary myxopapillary ependymoma in which the radiological and surgical findings were consistent with neurofibroma. This case serves as a reminder that, even with the current sophisticated neuroimaging methods available, a significant proportion of spinal tumors still present diagnostic challenges. The preoperative macroscopic appearance of this tumor also led to the tentative diagnosis of neurofibroma, and this underlies the importance of obtaining a conclusive histopathologic diagnosis.

Case report

A 46-year-old man presented with a 2 year history of progressive monoparesis and monoparesia of his right leg. He reported having minimal fecal and urinary incontinence. Neurological examination revealed mild paresis of the right quadriceps and iliopsoas muscles, and absence of the Achilles reflex on the right.

Lumbar spinal magnetic resonance imaging (MRI) revealed a 2.5 × 1.6 × 1.5 cm intradural extramedul-

lary mass at L1-L2. The lesion showed intermediate signal intensity on T1-weighted (W) and T2W images (Figure 1), and was homogeneously enhanced after intravenous contrast injection (Figure 2). On the basis of the MRI findings, our differential diagnosis for this intradural extramedullary mass lesion included Schwannoma, meningioma, and neurofibroma.

After performing standard laminectomies at the L1 and L2 vertebra, we opened the dura exposing a circumscribed, encapsulated, pinkish mass dorsal to the filum terminale. The mass appeared to have rootlets at its base, but did not seem to be attached to the spinal cord or dura mater. The distal portion was not continuous with the rootlets of the filum terminale, and was easily elevated by coagulating and dividing a few feeders. We achieved total resection by dividing the proximal feeders and freeing the weak rootlet attachments.

Histopathological examination identified the tumor as a myxopapillary ependymoma (Figures 3 and 4). Cranial and whole spinal axis MRI examination revealed no other mass lesions. At 6 weeks post-surgery, the patient's urinary and fecal incontinence had resolved completely and there was improvement with respect to his monoparesis.

Discussion

Ependymoma of the lumbar region is usually histologically benign pathology, and the myxopapillary subtype is most common to this area.^{1–6} Since

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Figure 1 (A, B) A sagittal T1W image demonstrates and intradural extramedullary mass at L1-L2 of intermediate signal intensity (A) A sagittal T2W image shows the intradural mass displacing the filum terminal anteriorly, and reveals no expansion of the spinal cord (B)

prognosis is directly related to the extent of surgical resection, it is important to treat ependymomas aggressively with surgery.^{7,8} 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.^{1,2,4,5,9,10} Even after gross total resection, the recurrence rate for myxopapillary ependymoma is 10 to 19%^{2,9} so it is important to follow these patients closely with MRI scans.^{6,11}

In the case presented here, preoperative radiological evaluation and preoperative observation of the mass

led to a tentative diagnosis of neurofibroma. Macroscopic features, such as the tumor's encapsulation, the apparent lack of attachment to the spinal cord and 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 patient survival. This case is a reminder that, even in the face of today's sophisticated neuroimaging methods, certain spinal tumors are still diagnostically challenging.

Figure 2 (A, B, C) Images after contrast enhancement. Homogenous enhancement of the intradural extramedullary mass lesion at L1-L2 in sagittal (A), coronal (B), and transverse (C) images. Note the lack of foraminal or extradural extension

Figure 3 Myxopapillary ependymoma. Histological examination showed mucinous material (upward curved arrow), vascular hyalinization (straight arrow), and microcyst formation (downward curved arrow). (HE, $\times 115$)

Figure 4 The myxopapillary ependymoma immunohistochemically stained with glial fibrillary acidic protein (GFAP). (Immunoperoxidase, $\times 230$)

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