Image Report

Image report: Extensive disseminated thoracolumbosacral myxopapillary ependymoma

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ABSTRACT

Background: Myxopapillary ependymoma occurs more frequently in adults, but is found in the first two decades of life in around 8–20% of patients. Tumors are usually benign with low likelihood for dissemination.

Case Description: We describe a case of a 13-year-old boy who presented with progressive kyphosis and bilateral weakness of the lower limbs. MRI shows a thoracolumbosacral intradural tumor with invasion of sacral neural foramina and dissemination to the cervicothoracic region. The patient received T10-L5 laminectomy with subtotal tumor resection. Pathological examination revealed myxopapillary ependymoma. After surgical resection, the patient underwent physical therapy with whole spinal radiotherapy for disease control.

Conclusion: Spinal myxopapillary ependymomas are usually benign and slow-growing tumors. This case illustrates an extensive and disseminated myxopapillary ependymoma.

Keywords: Disseminated, Invasive, Spinal myxopapillary ependymoma

INTRODUCTION

Intramedullary spinal cord tumors in childhood are rare. They account for only 4–6% of all CNS tumors in this age group. Among the various types of spinal cord tumors, astrocytoma and ependymoma are usually the two most common in childhood. Intramedullary spinal cord tumors in childhood are rare. They account for only 4–6% of all CNS tumors in this age group. Myxopapillary ependymomas occur more frequently in adults, but are found in the first two decades of life in around 8–20% of patients. Tumors are usually benign with low likelihood for dissemination. In this report, the authors have described a rare case of spinal myxopapillary ependymoma with invasive characteristics in which the tumor extended from the lumbosacral to the thoracic region and also showed dissemination to the cervicothoracic area.

CASE REPORT

A 13-year-old boy presented in 2018 with progressive kyphosis, without back pain or weakness. The patient visited orthopedic and pediatric doctors and was initially investigated for enthesitis secondary to juvenile idiopathic arthritis. However, serological workup was negative for HLA B27, rheumatoid factor, and ANA.
MRI pelvis in 2019 revealed posterior vertebral scalloping and widening of the neural foramina, which was initially misdiagnosed as dural ectasia. He was, therefore, followed up by pediatric rheumatology as an outpatient.

The patient gradually developed bilateral lower limb weakness and urinary incontinence and after two years presented to the neurosurgery outpatient department. Physical examination revealed kyphosis of the thoracolumbar spine. Upper limb power was Grade V throughout. Lower limb power was Grade II proximally (iliopsoas and quadriceps) and Grade I distally (tibialis anterior and extensor hallucis longus). Per rectal examination found loose anal sphincter tone and decreased perianal sensation. MRI whole spine revealed a large, expansile intradural mass that completely occupied the lumbosacral spinal canal. The tumor demonstrated T1WI hypointensity and T2WI hyperintensity as compared to the spinal cord and showed avidly homogenous enhancement postgadolinium contrast, extending from T10 to S3 and causing compression at the lower thoracic cord. The mass was not separate from conus medullaris. In the lumbar region, the mass extended into the neural foramina on both sides from L1-2 to S2-3 and the posterior vertebral bodies showed marked scalloping with mild kyphoscoliosis [Figure 1]. Postgadolinium scan shows multiple enhancing intradural lesions scattered throughout the cervical and thoracic regions causing some degree of cord compression, without obvious compressive myelopathy [Figure 2]. The impression after MRI was extensive disseminated thoracolumbosacral myxopapillary ependymoma from T10 to S3 vertebral level. The patient underwent T10 to L5 laminectomy; intraoperatively, the tumor was soft and suctionable, and was of grayish-purple color [Figure 3]. Subtotal resection was performed, with residual tumor extending beyond the sacral neural foramina. The conus medullaris and nerve roots were carefully preserved with the aid of intraoperative neurophysiological monitoring. Postoperative MRI spine after 48 h showed residual tumor in levels S1 to S3, especially in bilateral neural foramina [Figure 4]. Postoperative MRI brain showed a few nonspecific foci of white matter change in both frontal lobes but no evidence of intracranial tumor dissemination. Pathological examination of the resected tissue confirmed a diagnosis of myxopapillary ependymoma [Figure 5].

At follow-up 2 months postdischarge, the patient demonstrated reduced clinical weakness following rehabilitation. Power in the lower limbs was improved to Grade IV both proximally and distally. As of July 2020, the patient is undergoing whole

**Figure 1:** Preoperative MRI whole spine: (a) a large expansile enhancing intradural mass completely occupying the lumbosacral spinal canal, extending from T10 to S3 level with posterior vertebral scalloping, causing cord compression. (b) This mass in the lumbar region extends into the neural foramina at all levels, with expansion of, and extension out of, the foramina on both sides from L1-2 to S2-3 level.

**Figure 2:** Preoperative MRI cervicothoracic spine shows multiple enhancing intradural lesions scattered throughout cervical and thoracic levels causing some degree of cord compression; the dissemination demonstrated here is not typical of MPE.

**Figure 3:** Intraoperative T10-L5 laminectomy + subtotal tumor resection (intracapsular technique); the tumor is characteristically greyish in color and suctionable.
Figure 4: Postoperative MRI at 48 h shows residual tumor in levels S1 to S3 and in bilateral neural foramina from level L1 to S3.

Figure 5: Pathological examination: (a) sheets of tumor cells are presented among a myxoid background (b) elongated tumor cells with cytoplasmic processes extending through myxoid material to reach a blood vessel.

DISCUSSION

Myxopapillary ependymomas are typically benign and slow-growing tumors. However, despite being low-grade tumors, MPEs have potential for dissemination within the spinal canal.[3] The incidence of disseminated disease is thought to have been underestimated in the pre-MRI era. Fassett et al. (2005) reported a case series of five patients with MPE, 4 of which (80%) had disseminated CNS disease at initial presentation. In combination with five other case series of pediatric patients with MPE, 58% of the patients who underwent screening were found to have disseminated disease. This shows that, in pediatric patients, MPEs may spread throughout the CNS through cerebrospinal fluid. Therefore, MRI imaging of the whole CNS is recommended to detect tumor dissemination. The initial treatment strategy for these patients was tumor resection. Patients who underwent subtotal resection were additionally treated with radiotherapy.

In the lumbosacral region, the majority of MPEs arise from intradural filum terminale. Extradural ependymoma that arises around or in the sacral area is very rare.[3] The extent of tumor resection correlates with the outcomes after MPE resection. Abdulaziz et al. reported a retrospective review of 107 patients undergoing resection for MPE; the extent of each resection was defined as being either “en bloc (no capsular violation),” “gross-total resection with capsular violation (GTR),” or “subtotal resection.” There was no recurrence in the en bloc group, where recurrence rates for GTR and STR were 15% and 45%, respectively.[1]

The tumor in our report showed extensive thoracolumbar and sacral involvement, with dissemination to the cervicothoracic spinal region. Therefore, the surgical treatment was subtotal tumor resection. After surgical resection, the patient required radiotherapy for disease control.

CONCLUSION

Spinal myxopapillary ependymomas are typically benign, slow-growing tumors. However, this image report shows a case of spinal MPE with extensive and invasive behavior, extending thoracolumbosacrally with perisacral involvement and dissemination to the cervicothoracic spinal region; the patient required multimodality treatment including surgical (subtotal) resection and radiotherapy.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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