

Multifocal myxopapillary ependymoma and iatrogenic spinal cord herniation: Management options and lessons learned

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Abstract

Although myxopapillary ependymoma is a fairly common tumour of the lumbosacral spine, primary multi-focal myxopapillary ependymoma is a rare variant. Drop metastasis and leptomeningeal spread in the craniospinal axis is seen more frequently in the paediatric population, although it is unusual in adults. Surgical resection of the primary lesion remains the standard treatment. As per the authors' knowledge, to-date there is only one prior case in literature reporting iatrogenic spinal cord herniation with indentation after surgery for thoracolumbar spinal tumour. Here, we are discussing an unusual case of primary multi-focal ependymoma in a 16-year-old Asian boy, with drop metastasis and lepto-meningeal disease, who developed iatrogenic spinal cord herniation after the first surgery for the primary tumour. He presented to the Shaukat Khanum Memorial Cancer Hospital & Research Centre (SKMCH & RC), Lahore, after his first surgery. He underwent the definitive corrective surgery at SKMCH & RC where he was managed further. We discuss the management options for this patient and the lessons learned along the way.

Keywords: Myxopapillary ependymoma, paediatric, iatrogenic, spinal cord herniation.

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Introduction

Myxopapillary ependymoma is a common tumour of the lumbosacral spine.¹ Drop metastasis and leptomeningeal spread of Myxopapillary Ependymoma in the craniospinal axis are seen more frequently in the paediatric population, although it is unusual in adults.² As per the authors' knowledge, there is only one prior case in literature to-date, reporting iatrogenic spinal cord herniation after surgery for thoracolumbar spinal tumour.³ Here, we are discussing a unique case of primary multi-focal myxopapillary ependymoma with drop metastasis and leptomeningeal disease in a 16-year-old boy who developed iatrogenic spinal cord herniation with indentation after surgery of the primary tumour in the thoracolumbar spine, and was later

managed at the SKMCH & RC. Data was collected retrospectively from hospital records after consent from the patient and ERC approval.

Case Report

A 16-year-old Asian male presented at the Shaukat Khanum Memorial Cancer Hospital & Research Centre (SKMCH & RC), Lahore with backache since the past six months and bilateral lower limbs weakness since three months. He had initially sought treatment at a hospital in Southern Punjab on July 20 2020 where a Magnetic Resonance Imaging (MRI) of the whole spine with contrast was done on July 24, 2020, which showed an ill-defined diffuse mass involving the lower dorsal cord showing faint patchy enhancement and a similar intensity lesion in the lower lumbosacral canal as well. The patient underwent sub-total resection of the D11-12 lesion on August 8, 2020 at a facility in Southern Punjab. Histopathology Sections showed a neoplasm composed of cuboidal to columnar cells surrounding myxoid vascular papillary cores. Immunohistochemically, glial fibrillary acidic protein (GFAP) was positive, which is consistent with Myxopapillary ependymoma, WHO grade-I (Figure 1).

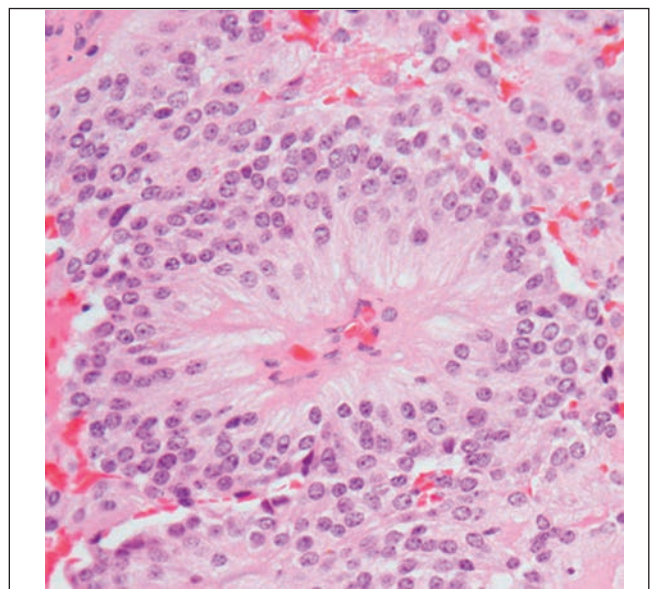


Figure-1: Histopathology Sections showing a neoplasm composed of cuboidal to columnar cells surrounding the myxoid vascular papillary cores, consistent with Myxopapillary ependymoma, WHO grade-I.

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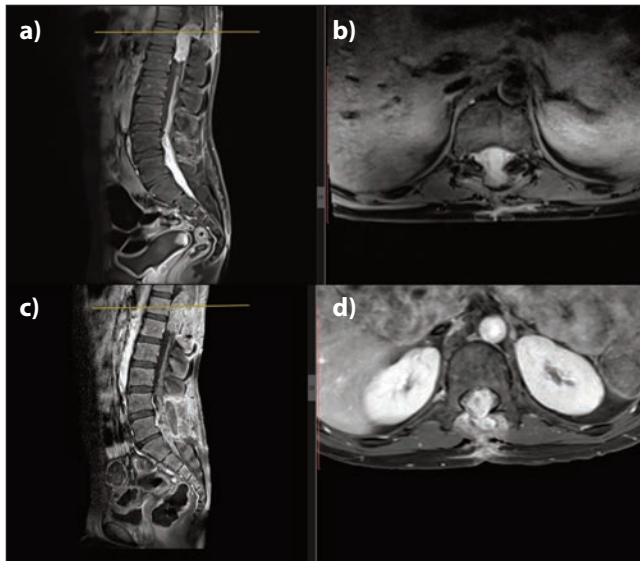


Figure-2: (a & b- above) This is the initial MRI done at our hospital on presentation, showing two intradural masses; one, residual disease in the thoracolumbar region, and another involving the lower lumbosacral canal. Another interesting finding was the herniating cord with tumour through the laminectomy defect with indentation at the lower end (see axial at level of T12). (c & d- below) After the second surgery, MRI, showing reduced herniation of the cord in the thoracolumbar region (see axial at level of T12), and excision of the lower lumbosacral lesion. The patient's neurology improved remarkably and rapidly post-operatively, the sphincter function returned, and he is now able to walk independently.

After this surgery, there was no improvement in the weakness in his lower limbs, and he was referred to our unit at Shaukat Khanum Memorial Cancer Hospital & Research Centre for further management. At the time of presentation in September 2020, he had bilateral lower limbs weakness with power of 2/5 in right lower limb and 1/5 in left lower limb, with poor bladder sphincter control.

A repeat MRI of the whole spine with contrast was done on September 09, 2020, it demonstrated an intramedullary lesion T2 high, T1 low signal, showing post-contrast enhancement extending from T11 to L1 vertebral body level, in keeping with residual disease. It also showed spinal cord and cauda equina herniating into the laminectomy wound. Similar long segment signal intensity abnormality is noted in the lower lumbosacral canal from L5 to S2 level, likely represent drop metastasis. (Figure-2, a & b).

The case was discussed in the hospital's Neuro-oncology and Spinal Multi-Disciplinary meeting and a repeat surgery was decided upon, to attempt resection of the lesions and repair the spinal cord herniation. The surgery was to be followed by radiotherapy to the cranio-spinal axis for any residual disease as well as for the dural metastasis.

The patient underwent excision of the lumbosacral space occupying lesion and thoracolumbar decompression, and reduction and repair of the spinal cord hernia on October 03, 2020.

Immediately postoperatively, the patient was moving both limbs with improved power as compared to preoperatively, with a power of at least 4/5 in the right and 3/5 in the left lower limb. The sacral bed sore rapidly healed, showing marked improvement, sphincter control was intact. Four weeks postoperatively, he received radiotherapy of 4500 cc Gys in 25 fractions.

The post-operative MRI showed reduced herniation and complete excision of the lumbosacral lesion (Figure-2, c & d).

Now, six months after treatment, the patient was able to walk independently and was kept on end of treatment follow-up with three-monthly MRI of the whole spine.

Discussion

It has been reported in literature that myxopapillary ependymoma is located exclusively in the conus medullaris and cauda equina region,³ however, the multifocal type of myxopapillary ependymoma is relatively rare.¹ Hanbali et al⁴ reported that two out of 26 patients had multifocal tumours. In our case, the tumour was multifocal with foci at the T11-L1 and L5-S2 levels.

A favourable prognosis is expected when a total resection is performed, whereas a 50-70% recurrence rate of spinal ependymoma has been reported after a subtotal resection.^{5,6} In the present case, a subtotal resection for the tumour was performed at the T10-12 region because the tumour was firmly attached to the cord, no definite plane could be identified between the tumour and conus.

In addition, subarachnoid dissemination and external metastasis has been observed occasionally.⁷ This is more commonly seen in the paediatric population. Our patient also had diffused and nodular Dural enhancement with sugar-coating appearance that might represent drop metastasis or lepto-meningeal disease.

A most interesting aspect of this case was the iatrogenic post-operative spinal cord herniation through the laminectomy defect to the extent that the cord had a kink over the ventral aspect, against the lamina at the lower border of the previous laminectomy surgery. It was decided to correct it in addition to attempting gross total resection of the tumour, as it seemed prudent to relieve the mechanical compression. The author has come across only one similar case in literature and this is the second case report of an iatrogenic cord herniation at the

thoracolumbar junction, following subtotal/ gross total resection of an intradural lesion.

The postoperative course of the present case was uneventful during the follow-up. The patient had no tumour recurrence in the brain, spinal cord, or the cauda equina. The patient had an uneventful clinical course over the four-month follow-up period.

The aim of this case report is to share our lessons with our fellow colleague and have iatrogenic cord herniation in our differential diagnosis of a patient who deteriorates neurologically after surgery for intramedullary spine tumours.

Conclusion

This case highlights that it is important to consider iatrogenic cord herniation as a differential diagnosis in cases whose neurology deteriorates after surgery for spine tumours, as it is a potentially correctable cause of neurological deterioration, and timely decompression can lead to good results.

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Conflict of Interest: None.

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