Intraspinal Extradural Myxopapillary Ependymoma of the Sacrum: A Case Report

Ashish Chugh, MD., Prashant Punia, MD., Sarang Gotecha, MD.

Department of Neurosurgery, Dr. D.Y. Patil Medical College and Hospital, Pimpri, Pune, Maharashtra, India

ABSTRACT

Background Data: Myxopapillary ependymoma (MPE) is a low-grade variant of ependymoma that typically occurs in conus medullaris and filum terminale of the spinal cord. Only rarely do these tumors present at an extradural location in the sacrococcygeal region and even rarer is the development of these extradural tumors inside the spinal canal. The authors report a case of an intraspinal extradural MPE of the sacrum with relevant literature review on salient features and management.

Study Design: Case Report.

Purpose: To report a rare entity at an uncommon location.

Case Report: A 12-year-old male child presented with chief complaints of low backache and radiation of pain in both lower limbs for six months. The patient also had a history of straining during micturition for one month. On examination, he was found to have mild weakness of bilateral ankle plantar flexion and knee flexion (grade 3). Ankle jerks were absent and there was evidence of sacral hypoaesthesia. Intraoperatively sacral lamina was found to be thinned out. Laminectomy was performed at L5 and sacrum, an extradural, fleshy lesion extending from L5 to S5 was identified, and gross total resection was achieved. There was no intradural expansion of the tumor.

Results: Patient had an uneventful course and was discharged home on day 4.

Conclusion: Intraspinal extradural MPE is a rare entity and should be kept in mind while diagnosing a midline lower back lesion. Treatment consists of gross total excision wherever possible coupled with radiotherapy. These tumors tend to recur locally and systemically and, thus, patients should be followed up periodically. (2020ESJ226)

Keyword: Myxopapillary ependymoma, Sacral extradural tumor, Sacrococcygeal ependymoma

Address correspondence and reprint requests: Prashant Punia, MD.
Associate professor, Department of Neurosurgery, Dr. D.Y. Patil Medical College and Hospital, Pimpri, Pune, Maharashtra, India. - E-mail: getdrprashant@gmail.com

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INTRODUCTION

Myxopapillary ependymoma (MPE) is a low-grade variant of ependymoma that typically occurs in conus medullaris and filum terminale of the spinal cord. Only rarely do these tumors present at an extradural location in the sacrococcygeal region. Even rarer is the development of these extradural tumors inside the spinal canal. These tumors closely mimic other sacral region tumors such as teratoma, chordoma, and lipoma. Management consists of a gross total resection with wide margins wherever possible and/or coupled with radiotherapy (RT). Only eight cases have been previously reported in the English literature. The authors report a case of an intraspinal extradural MPE of the sacrum with relevant literature review on salient features and management.

CASE REPORT

A 12-year-old male child presented with chief complaints of low backache and radiation of pain in both lower limbs for six months. The patient also had a history of straining during micturition for one month. On examination, he was found to have mild weakness of bilateral ankle plantar flexion and knee flexion (grade 3). Ankle jerks were absent and there was evidence of perianal sacral hypoaesthesia. MRI of the lumbosacral region revealed an expansible lytic lesion in the sacrum extending from L5 to S5 vertebral body with mild erosion of the posterior portion of the S1 vertebral body. The lesion appeared hyperintense on T2 and hypointense on T1-weighted imaging, which occupied the canal and is extended into the sacral foramina, causing compression of contained and exiting nerve roots. It measured 10.4 × 8.4 × 2.3 cms in superior-inferior, transverse, and anteroposterior dimensions and was homogenously enhanced on intravenous gadolinium contrast administration.

Keeping the above-mentioned features in mind, we made a possible diagnosis of a chordoma/nerve sheath tumor (Figure 1). The patient was scheduled for surgery, where intraoperatively, sacral lamina was found to be thinned out. Laminectomy was performed at L5 and sacrum and an extradural, fleshy lesion extending from L5 to S5 was identified. It was compressing the thecal sac and was easily separated from the dura. Decompression was done around the thecal sac and gross total excision was achieved. A frozen section procedure was performed to determine diagnosis, which was proved to be an ependymoma. In view of frozen section findings, intradural expansion of tumor was suspected and the dura was opened, and no evidence of tumor was found intradurally (Figure 2). The patient had a smooth, uneventful operative and postoperative course and was discharged from our hospital on day 4 and scheduled for outpatient clinic routine follow-up. Histopathology revealed cuboidal and elongated cells with minimal nuclear variation and mitosis. A radial arrangement of tumor cells around the fibrovascular core was observed. Cells were reactive for Glial Fibrillary Acidic Protein (GFAP) and S100. Findings were suggestive of a grade 1 MPE.

The patient was referred to an oncologist and underwent three cycles of radiotherapy, after which he was kept on regular periodic outpatient follow-up. Three years postradiotherapy, the patient showed a small recurrence at the S5 level with no complaints. Resurgery was advised for him, but the patient instead chose to wait and be monitored with sequential scans (Figure 3). All procedures performed in this study involving human participants were in accordance with the ethical standards of the institutional and/or national research committee (Dr. D.Y. Patil Medical College and Hospital) and with the 1964 Declaration of Helsinki and its later amendments or comparable ethical standards. Informed consent was obtained from all individual participants included in the study.
Table 1. Data summary of reported intraspinal extradural sacral MPE.

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<tr>
<th>No</th>
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<td>9</td>
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<td>Present case</td>
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<td>M</td>
<td>GTR + RT</td>
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</table>

STR: subtotal resection; RT: radiotherapy; GTR: gross total resection.

Figure 1. Sagittal MRI of the lumbosacral spine. (A) T2-weighted image showing lytic lesion extending from L5 to S5 segment. (B) Contrast-enhanced T1WI image showing tumor with homogeneous contrast enhancement. (C) Postoperative MRI T2WI at 3-year follow-up showing small local site recurrence at S5 level.

Figure 2. Intraoperative photographs (A) showing extradural tumor compressing the thecal sac to the right side, (B) showing no intradural extension of the tumor, and (C) showing tumor cavity after gross total excision of tumor with decompressed thecal sac and nerve root.
DISCUSSION

Ependymomas are a well-recognized and frequently encountered entity in neurosurgical practice. Location of these tumors outside the central nervous system (CNS) is rare. The first case of this entity was reported by Mallory in 1902. These extrameningeal ependymomas are even rarer when they arise from or extend intraspinally as in our patient. Only eight of these extradural ependymoma cases have previously been reported and out of these, one patient had an extradural spread from an intradural tumor. The previously reported eight similar cases comprised 4 female and 4 male patients with a mean age of 44 years. The majority of these patients presented with bowel and bladder dysfunction and were found to have a loss of ankle jerk on physical examination. Pain in the lower back and radiculopathy along sciatic nerve distribution were other complaints of the patients. Our patient’s clinical profile matches the literature as he had presented with sciatic radiculopathy and bladder complaints. Some patients may present with mass either in the lower back or in the pelvis when a significant portion of the tumor bulges out of the spinal canal either posteriorly or anteriorly, respectively. This was not the case with our patient as he had no palpable mass on examination. Rare presentation of these tumors is not unheard of; Rosario and Sison reported a case wherein the presentation was hemorrhage of iatrogenic cause (spinal anesthesia).

Four possible types of extrameningeal ependymomas were recognized, namely, 1) metastasis or tumor displaying direct extension following surgical excision from a primary CNS tumor; 2) primary ependymoma of the lower spinal cord, cauda equine, or filum terminale extending into soft tissues of the sacrococcygeal area; 3) primary abdominal, pelvic, or presacral tumor; 4) primary skin and subcutaneous tumors of the sacrococcygeal area having no demonstrable connection to the spinal cord. Our patient falls under the 2nd category, with only the filum terminale involved. The vestigial remnants of coccygeal medullary space, also called extramedullary ependymal cell rests, are responsible for the origin of this type of MPE. This ependyma-lined cleft is a derivative of the persistent caudal neural tube and is present beneath the skin of the postanal pit. Thus, these lesions may present as midline mass in the lower back and should alert the neurosurgeon. MPEs are predisposed to systemic spread, most commonly to the lymph nodes, liver, and lungs. Treatment of MPE consists of local and systemic disease control. Batic et al. recommended a wide local gross total excision, either en bloc or piecemeal, coupled with early adjuvant radiotherapy. Fourney et al. stated that presacrally extending tumors are more invasive.
and local recurrences, if any, can be managed well with repeated excisions. They further argued that whether RT is useful for extradural tumors is unclear because of insufficient data. The authors concurred with the idea of adjuvant RT as it might prove helpful to avoid local recurrence and, in turn, save the patient from a resurgery. Three cycles of the RT were given to our patient and a small local site recurrence was observed after a 3-year follow-up. Although the patient refused resurgery as he was symptom-free, he has been on regular periodic follow-up with sequential scans. Molecular profiling of the tumor should be done as a part of treatment wherever possible. Specific and recurrent chromosomal aberrations can now easily be identified and correlated with histological subtype and grade of tumor. In a study on molecular behavior of pediatric MPE, it was concluded that extraspinal MPE and MPE with anaplastic features might be phenotypic variations of spinal MPE. The authors of this study further stated that there might be different underlying biology in adult and pediatric MPE. Although molecular studies currently may not help in an extradural MPE due to its rare presentation and lack of data, they are likely to give us important information on tumor behavior in the future. Differential diagnosis of MPE consists of a chordoma, sacrococcygeal teratoma, nerve sheath tumors, dermoid cyst, and a plasmacytoma. Although features like bony erosion are seen with MPE, they are not specific. Moelleken et al. stated that bony destruction is not as common with MPE and large destructive lesions of the sacrum are rare due to MPE and more common due to metastasis. Thus, immunopositivity of tumor cells with GFAP confirms the diagnosis.

CONCLUSION

Intraspinal extradural MPE is a rare entity and should be kept in mind while diagnosing a midline lower back lesion. Treatment consists of gross total excision wherever possible coupled with radiotherapy. These tumors have the tendency to recur locally and systemically and, thus, patients should be followed up periodically.

REFERENCES


الملخص العربي

الورم البطاني العصبي خارج النخاع من عظم العجز: تقرير حالة

البيانات الخلفية: الورم البطاني العصبي (MPE) هو متغير منخفض الدرجة من الورم البطاني العصبي الذي يحدث عادة في المخروط النخاعي والنهاية الخبيطة للجلب الشوكي. نادرًا ما توجد هذه الأورام في مكان خارج الجافية في المنطقة العجزي العصعصية. والأندر هو تطور هذه الأورام خارج الجافية داخل القناة الشوكية. أبلغ المؤلفون عن حالة الورم البطاني العصبي النخاعي خارج النخاع في العجز مع مراجعة الأدبيات ذات الصلة حول السمات البازرة والإدارة.

تصميم الدراسة: تقرير حالة.

الغرض: الإبلاغ عن كيان نادر في مكان غير مألوف.

المريض و الطرق: قدم طفل يبلغ من العمر 12 عامًا شكاوى رئيسية من آلام أسفل الظهر وإشعاع من الألم 12 شهرًا. أعطى المريض أيضًا تاريخ إجهاد أثناء البول لمدة شهر واحد. عند الفحص، وجد أنه يعاني من ضعف خفيف في الانثناء الأخمصي للكاحل والانثناء الركبة (الدرجة 3). كانت نفضات الكاحل غائبة وكان هناك دليل على نقص التخدير العجزي. تم العثور على الصفيحة العجزية أثناء الجراحة لتكون ضعيفة. تم إجراء استئصال الصفيحة الفقرية في L5 والجزء L5 وتم تحديد آفة خارج الجافية تمتد من L5 إلى S5 وتم تحقيق الاستئصال الكلي الإجمالي. لم يكن هناك توسع داخل الجافية للورم.

النتائج: يحصل المريض على مسار هادئ ويبقى من المنزل في اليوم الرابع.

الخلاصة: إن خارج النخاع هو كيان نادر ويبقى أن يوجد في الاعتبار أثناء تشخيص آفة أسفل الظهر في خط الوسط. يتكون العدل من الاستئصال الكلي الكلي كما أمكن ذلك مع العلاج الإشعاعي. تميل هذه الأورام إلى التكرار محليًا وجهائيًا وبالتالي يجب متابعة المريض بشكل دوري.