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# Sacral extradural spinal meningioma with recurrence; a case report

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## Abstract

Extradural spinal meningiomas are very uncommon, and just a scattering of clinical reports on this condition have been published. Despite being benign, epidural meningiomas exhibit a higher recurrence rate compared to typical meningiomas. In this case report, we described a case of sacral extradural spinal meningioma with several episodes of recurrence and discussed the clinical characteristics, radiological features, and management, along with follow-up details. A 19-year-old woman with back discomfort and lower extremity numbness. Magnetic resonance imaging (MRI) revealed a tumor on the left posterolateral S1–S2 spinal cord. L5–S3 hemilaminectomy and foraminotomy were performed. After three years, the patient had bilateral L4–S1 laminectomy and radiotherapy for tumor recurrence. Two years later, the patient presented with pain and paresthesia in the lower limbs and was diagnosed with a tumor recurrence once more. Due to sacral area adhesion from earlier surgeries and radiation, the patient received CyberKnife radiation instead of tumor resection. No complaints or signs of recurrence were detected during follow-up examinations. Extradural spinal meningiomas are uncommon and can occasionally be misidentified as malignant metastatic tumors both before and during surgery. This misclassification could potentially lead to alterations in the recommended surgical approach. Although epidural spinal meningiomas are often benign, their long-term prognosis is uncertain, and the recurrence rate is higher than ordinary meningiomas. Hence, lengthy monitoring is necessary to identify any recurrence risk (like our case).

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## Introduction

Spinal meningiomas are primarily classified as primary spinal tumors, constituting approximately 25–46% of all primary spinal tumors and accounting for 7.5%–12.7% of all meningioma cases (1,2). Most spinal meningiomas present as intradural extramedullary tumors in the thoracic region, while meningiomas that arise extradural in the sacral area are sporadic (3). Extradural spinal meningiomas present a considerable challenge for spine surgeons because they are frequently misdiagnosed as metastatic spinal tumors during preoperative imaging and intraoperative assessments (4). Therefore, establishing an accurate diagnosis prior to surgery is crucial since it will determine the extent of the operation and impact the patient's prognosis.

There have been relatively few case reports of sacral epidural meningiomas so far, and data on the tumors' features and management strategies are limited. In addition, in most previous cases, the duration of follow-up was short, and not enough data on tumor recurrences were documented (5-8). Hence, in this study, we discuss a case of extradural spinal meningioma (S1–S2) that recurred three times during a 7-year follow-up period.

**Key point**

- Extradural spinal meningiomas can be rare and are occasionally misdiagnosed as malignant metastatic tumors, potentially affecting the recommended surgical approach.
- While epidural spinal meningiomas are typically benign, their long-term prognosis remains uncertain.
- Prolonged follow-up is essential to detect any potential recurrence and manage the condition effectively.

## Case Presentation

A 19-year-old lady complained of lower back pain that remained for over two months, with right leg radiation and numbness in the posterior region of both lower limbs that lasted two weeks. During the physical examination, the patient displayed no neurological impairment, scoring a motor grade of five on both legs based on the Medical Research Council (MRC) scale, and

exhibited intact deep tendon reflexes.

The lumbosacral spine spirals computed tomography (CT) showed no abnormalities; however, the lumbosacral spine magnetic resonance imaging (MRI) revealed an elongated structure with intermediate signal intensity on T1 and T2 sequences that extended bilaterally into the neural foramina. The contrast-enhanced T1-weighted phase revealed a 42×34×12 mm homogeneous enhancement at the level of the S1–S2 vertebrae, with distinct margins and extensive base attachments adjacent to the Dural membrane, resulting in severe sacral spinal canal stenosis. There was no evidence of cystic degeneration or evident vascular structure in the mass. Before the surgery, the radiologic differential diagnosis included schwannoma, neurofibroma, or extradural meningioma.

Two months following the radiographic diagnosis, the patient had surgical excision of the spinal tumor. After a midline incision was made in the soft tissue, L5 to S3 hemilaminectomy and foraminotomy were performed under general anesthesia. The surgeon observed a grayish mass that extended from the right posterior epidural space to the neural foraminal level on the right side, involving the S1–S2–S3 nerve roots. Choroid meningioma grade II/III WHO classification was verified by postoperative pathology, immunohistochemical EMA(+), PR(+), S-100 (-), cytokeratin (-), CK8 (-), CK19 (-), and CEA staining (-).

After surgery, the neurological status improved, and the pain decreased. Urinary and bowel incontinence occurred on the third postoperative day and resolved two months later. There was no evidence of tumor recurrence on the third, sixth, and twelfth-month postoperative follow-up MRIs (Figure 1).

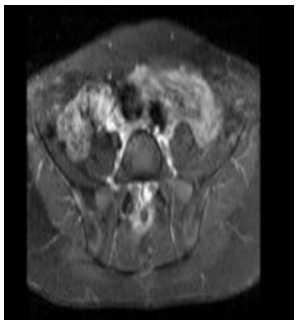

**Figure 1.** Post-operative MRI of the lumbosacral vertebrae; there is no evidence of tumor recurrent.

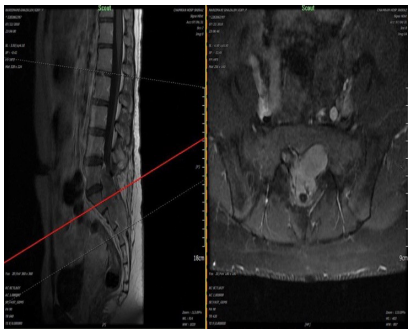

**Figure 2**. MRI of the lumbosacral vertebrae on the first tumor recurrence revealed hyposignal T1 in the spinal canal at the L5–S3 vertebrae level, isosignal in T2-weighted. The contrast-enhanced T1-weighted phase exhibited a homogeneous enhancement.

The patient presented three years after surgery with lower back discomfort radiating to the left leg and minor urine hesitancy. The lumbosacral spine MRI revealed a lobulated extradural mass lesion measuring approximately 85×30×29 mm at the L5–S3 level, which was isosignal in the T2-weighted image and iso to hyposignal in the T1-weighted image and displayed considerable homogeneous enhancement following gadolinium administration (Figure 2).

A CT-guided procedure yielded two Tru-Cut biopsies with stromal tissue of uncertain origins that morphologically matched the stromal component of the primary meningioma. The immunohistochemistry results of Tru-Cut biopsies were negative for chromogranin, CD31, CD34, S-100, SMA, and synaptophysin. On the other hand, the expression of EMA, vimentin, and Ki67 (5%+) was positive. Overall, the pathologic findings are compatible with a recurrence of meningioma. On the basis of the findings of the Tru-Cut biopsy, the patient was hospitalized and underwent total sacral lesion excision, total sacrectomy, and bilateral L4-S1 laminectomy. The resected lesion had immunohistochemistry positivity for NSE, P63, PR, and Vimentin. EMA and GFAP were mildly stained; however, Ki67 (1-2%) was relatively low. Subsequent negative staining was seen for Chromogranin, Oligo-2, S100, CK, SMA, CD34, and CD117. The preceding evidence supported the diagnosis of recurrent choroid meningioma.

She was released on day 12 postoperatively, and her symptoms were resolved at her follow-up appointment. Afterward, she received 27 radiation treatments. At 6- and 12-months post-surgery, there was no indication of residual or recurrent tumor on MRI.

Two years later, the patient presented with bilateral radicular discomfort and paresthesia in the lower extremities. A lumbosacral MRI showed the presence of a 47*37*15 mm extradural enhancing mass lesion on the posterior and left lateral side of the S1–S2 vertebral bodies that demonstrated contrast enhancement after gadolinium injection. To assess the probability of tumor recurrence, a whole-body fluorodeoxyglucose positron emission tomography (FDG-PET) was performed, revealing heterogeneously low-grade FDG-avid soft tissue density in the posterior sacral region, along with a local active lesion in the left lateral process of S1 vertebrae (Standardized uptake values [SUV] = 2.57). Concerning the potential additional utility of Somatostatin receptor PET/CT (SSTR-PET/CT) in resected/irradiated meningiomas, a Gallium Dotatate PET was performed, which revealed a high Ga-

Tate-avid soft tissue lesion posterior to L5–S1 vertebrae (SUV max = 5.21) and meningioma recurrence was established (Figure 3).

Due to significant adhesion in the sacral area caused by earlier surgeries and radiation, the patient was ineligible for tumor excisional surgery; instead, she underwent five CyberKnife radiation treatments (Accuray, Sunnyvale, CA). A one-year follow-up examination revealed no more complications with the patient (Figure 4).

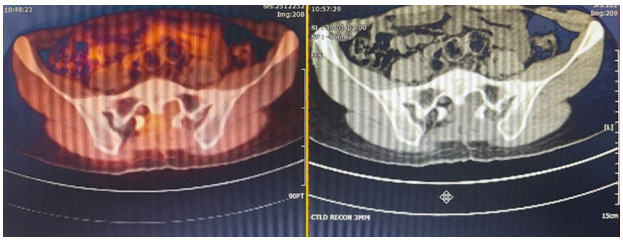

**Figure 3**. The whole-body fluorodeoxyglucose positron emission tomography (FDG-PET) after the 2nd episode of tumor recurrence exhibited low-grade FDG-avid soft tissue density in the posterior sacral area, as well as a local active lesion in the left lateral process of the S1 vertebra.

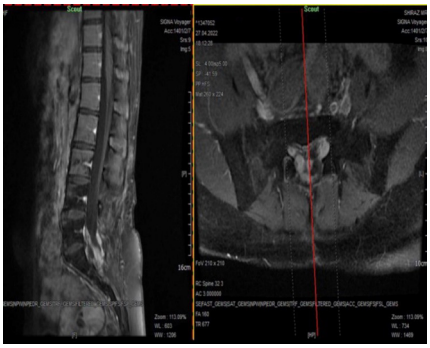

**Figure 4**. Last MRI revealed no more tumor recurrence.

## Discussion

Meningiomas are slow-growing tumors of the central nervous system that form from arachnoid granulations containing meningothelial cells (9). The majority of meningiomas are found in the brain, however spinal meningiomas make up approximately 1.2% to 12.5% of all meningiomas (10). Spinal meningiomas can occur anywhere along the spinal cord, with approximately 70% in the thoracic spine, 25% in the cervical spine, and 5% in the lumbar spine (11).

Most spinal meningiomas are intradural since they originate from arachnoid cap cells or, less often, dural fibroblast, arachnoid dural border cells, and arachnoid barrier cells (12). Although up to 10% of intradural meningiomas may spread, resulting in intradural-extradural tumors, extradural-origin spinal meningiomas are exceedingly uncommon, with a reported prevalence of just 3.5% (13,14). The establishment of extradural-origin spinal meningiomas is rather paradoxical since there is generally no arachnoid extradural (15). There are many theories regarding the pathophysiology of extradural meningiomas. Several studies indicate that the migration of arachnoid tissue islands into the extradural space may result in the development of extradural meningioma. Some scientists hypothesize that anomalous arachnid islands may exist in extradural space (16). Others have postulated that invasion of arachnoid villi into the dura mater and subsequent detachment of the main arachnoid layer may lead to the emergence of extradural meningioma (17,18). In addition, the existence of vestiges of the surface layer of fetal arachnoid and nerve root villi may be an additional reason for extradural meningioma (19).

The presentation of extradural meningioma does not significantly differ from other lesions in the same region. Taking into consideration the existing literature, in almost all cases, patients initially experience back or radiating root pain, followed by sensory alterations and sphincter dysfunction (7,8). In our case, the patient presented with back pain accompanied by radiation to the right leg and bilateral paresthesia in the lower extremities, which aligns with the findings reported in the literature.

MRI is often regarded as the optimal diagnostic technique (20). T1-weighted isosignal and T2-weighted isosignal/hypersignal were frequent radiologic results, and contrast application mostly reveals a homogeneous enhancement along with the dorsally or ventrally situated epidural meningioma in the dural sac (11).

Intraspinal masses signal probable clinical symptoms necessitating as soon as possible surgical excision of all malignancies for spinal canal decompression (21). The primary clinical determinant of recurrence is the extent of tumor resection, young age, higher pathologic grade, and calcifications (22); therefore, complete removal should be the first objective in order to eradicate the lesion completely and avoid its recurrence (11,23). Even while performing full excisional surgery on certain tumors, such as the one reported in this case report, is difficult owing to the closeness to the nerve root in the foramen, it is often carried out using isolated posterior procedures (24). Despite being a benign tumor, epidural meningioma demonstrates a higher recurrence rate compared to typical meningiomas (25). The World Health Organization (WHO) classifies epidural meningiomas into three grades: benign (grade I), atypical (grade II), and malignant (grade III). Malignant meningiomas have a higher probability of recurrence compared to atypical and benign types, with recurrence rates ranging from 50% to 94%, while atypical meningiomas have recurrence rates between 29% and 52%, and benign meningiomas have lower rates ranging from 7% to 25% (16). As it is difficult to provide radiation to meningiomas without exposing the spinal cord, postoperative adjuvant radiotherapy for meningiomas is controversial. Due to the low mitotic rate and recurrence risk of WHO grade I epidural meningiomas, following the resection of WHO grades II and III meningiomas, adjuvant radiation is generally recommended (26,27).

Adjuvant radiotherapy is typically considered when total resection of the tumor is not feasible due to its location or the patient's medical condition. Additionally, it is administered after total or subtotal resection to prevent early recurrence. Furthermore, in cases where surgical treatment poses significant medical risks, reoperation should be performed upon early recurrence, followed by radiotherapy (11). Several recent studies have indicated stereotactic radiotherapy, such as CyberKnife, for the management of spinal meningioma growth and recurrence (28). The CyberKnife system provides a high total dosage of radiation, a precise target volume, and a short treatment period, which reduces the radiological complications risks and considers it a desirable therapeutic choice (29).

## Conclusion

In conclusion, extradural spinal meningiomas are uncommon tumors that might be mistaken for metastatic disease. They are often misidentified preoperatively and intraoperatively as malignant metastatic tumors, which might affect the surgical strategy and increase morbidity. Moreover, they are mostly benign but have a greater recurrence rate than regular meningiomas. Due to conflicting data, extradural meningiomas' long-term prognosis and recurrence risk remain undetermined. Our patient encountered two recurrences in 5 years of follow-up, indicating the necessity of prolonged surveillance.

### Authors' contribution

**Conceptualization:** Majid Reza Farrokhi, Abbas Khosravifarsani.
**Data curation:** Majid Reza Farrokhi, Abbas Khosravifarsani, Mavlonov Jaloliddin Begijonovich.
**Investigation:** Majid Reza Farrokhi, Abbas Khosravifarsani, Mavlonov Jaloliddin Begijonovich.
**Project administration:** Abbas Khosravifarsani.
**Supervision:** Majid Reza Farrokhi.
**Writing–original draft:** Abbas Khosravifarsani.
**Writing–review and editing:** Majid Reza Farrokhi, Mavlonov Jaloliddin Begijonovich.

### Conflicts of interest

The authors declare that they have no competing interests.

### Ethical issues

The case report adhered to the guidelines set forth in the World Medical Association Declaration of Helsinki. Additionally, we obtained written informed consent from the patient for publication as a case report. Ethical issues (including plagiarism, data fabrication, double publication) have been completely observed by the authors.

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