Case Report

Spinal Clear Cell Meningioma: Atypical Clinical and Radiological Manifestations

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Meningioma has many subtypes with clear cell meningioma being more aggressive than other variants of meningioma and one of the rarest. We report a case of spinal clear cell meningioma that occurred in a 25-year-old lady who presented with the inability to be in the supine position. A magnetic resonance image showed an intradural mass extending from L1 to L4. Near complete excision was done. The patient had motor weakness postoperatively which improved gradually. A histopathological study showed a clear cell meningioma. In a differential diagnosis of any space-occupying lesion of the spine, clear cell meningioma should be considered though it is a rare form of meningioma due to its potential to recur. An accurate follow-up is warranted.

1. Introduction

Meningioma is a tumor arising from the meningothelial cells in the arachnoid villi. It is the commonest primary non-gial brain tumor which compromises around 13 to 19% of all primary intracranial neoplasm. [1]

The predominant site of involvement is the intracranial cavity (60%), while the involvement of the spinal canal constitutes 12% of all the cases of meningioma. 25% of all spinal cord tumors are spinal canal meningiomas [2–4].

In general, spinal tumors are classified to intradural and extradural tumors. Intradural tumors are further subdivided into intramedullary and extramedullary spinal tumors [5–7].

In most cases, spinal meningiomas confounded to the intradural space (90%), while 5% are extradural and the remaining 5% are both extra- and intradural [1, 8].

Spinal meningiomas are diagnosed based on the clinical manifestation and the imaging study findings. The most common clinical manifestation of spinal meningioma is a localized back pain, which is usually not radiating. The modality of choice in diagnosing spinal tumors is the magnetic resonance imaging (MRI) [9].

When a meningioma has an unfamiliar location and radiological characteristics with the absence of symptoms, the diagnosis becomes difficult. This is a case report of a spinal meningioma with atypical radiological features in a 25-year-old female.

2. Case Presentation

This is a case of 25-year-old Saudi lady who is a known case of epilepsy and with an intellectual disability since infancy. She presented to the clinic with a history of kyphosis for three years. She was unable to sleep in the supine position and had to sleep in the sitting position for three years. However, she was able to walk independently and had no sphincter disturbance. There was no history of loss of appetite or weight loss. She had a history of bilateral lower limb lymphedema for several years. The neurological examination at the time of presentation was normal.

The patient was investigated with a lumbar spine magnetic resonance imaging (MRI). It showed a large intrathecal mass extending from L1 to L4. This mass was hypointense on both T1- and T2-weighted images. The lesion was
approximately 10 × 5 × 4.3 cm in diameter, and it was widen-
ing the spinal canal with scoloping of the vertebral bodies
extensively seen at L3. Also, a bilateral dilatation of neural
foramina was noted. This lesion was heterogeneously
enhancing after injection of contrast (Figures 1 and 2).

The patient was further investigated with a CT scan and a
brain and whole spine MRI. No other lesions were seen in the
spine and brain MRI. The CT scan showed an intradural spi-
nal lesion extending from L1 to L4 widening the spinal canal
and causing scalloping of vertebral bodies. The lesion is also
extending to the corresponding neural foramina with widen-
ing of the foramina. A posterior extension was noted mainly
at the L3 level (Figure 3).

Preoperatively, the radiologic provisional diagnosis was
spinal myxopapillary ependymoma with possibility of schwan-
nonoma, neurofibroma, or meningioma (less likely).

The patient underwent a subtotal tumor resection and
fixation. Using a posterior midline approach, a laminectomy
from the lower edge of L1 to L3 was performed with fixation
on T12, L1, and L4 on both sides (Figure 4).

The mass appeared to be intradural, extending to the
extradural space mainly at the L3 level. The dura was opened
proximal and distal to the lesion, and it was noted that the
nerve roots were scattered over the tumor surface and invad-
ing the tumor (Figures 5(a) and 5(b)). The main bulk of the
spinal cord was bushed anterolateral toward the left side.
Intraoperative monitoring was used throughout the opera-
tion, and tumor debulking from within the capsule was done.
Almost a complete tumor resection was achieved under an
operating microscope by CUSA (Figure 5(c)). Only tumor
capsule was left as it was impossible to differentiate it from
the thecal sac.

Postoperatively, the patient was complaining of right
lower limb weakness: ankle dorsiflexion and planter flex-
ion (1/5), knee flexion and extension (2/5), and hip flexion
and extension (1/5). The power improved gradually with
physiotherapy. Upon discharge, she was able to stand
and walk few steps using a walking frame. Additionally,
she was discharged on Foley’s catheter as she was having
urinary retention.
Microscopically, the tumor characterized rounded cells with a moderate amount of clear cytoplasm in a patternless arrangement and extensive perivascular and interstitial collagenization; whorl formation was very vague and focal. In addition, there are areas showing angiomatous-like features (Figures 6(a) and 6(b)). The cells demonstrated periodic acid-Schiff-positive, diastase-sensitive intracytoplasmic glycogen granules (Figure 6(c)). Immunostains show positive EMA (Figure 6(d)), S100 (rare cells), and CD99 (weak), but negative chromogranin, CD10, CAM5.2, inhibin-A, GFAP, S100, pankeratin, and synaptophysin staining. CD34 and CD31 decorate the collagenized blood vessels. The MIB-labeling index was 2%. The final pathological diagnosis was clear cell meningioma (WHO Grade II).

3. Discussion

Spinal meningiomas account for 12% of all meningiomas. Most of them are confined within the intradural space; having an extradural extension is uncommon. Spinal meningioma accounts for 25% of all primary spinal cord tumors [10], with females being the predominant patients accounting for approximately 80% [11]. Meningiomas located in the lumbar spine are the least common site of involvement (4%), as opposed to thoracic (80%) and the cervical spine (16%) [2]. In all the studies concerning the spinal meningioma, the thoracic region was reported as the most common sit of involvement in a literature review except for two studies, where the common site was the lumbar region [12, 13].

Clinically, most of the cases presented with motor and sensory symptoms. The most commonly reported symptom was radicular, or funicular back pain (75%), followed by limb weakness (33.3%), hypoesthesia, paresthesia (15.5%), and sphincter dysfunction (10.7%) [13–15]; conversely, our patient was asymptomatic with no motor weakness, sensory changes, or any neurological deficit. However, she was walking while keeping her trunk in a flexed position with the inability to lie in the supine position. In a study analyzing 194 patients with spinal meningioma, a total of 26 patients were reported to be asymptomatic [16, 17].

Radiologically, the MRI showed a large lobulated intrathecal lumbar expansile hypointense mass in both T1 and T2 with heterogeneous enhancement extending between L1 and L4 levels extending to corresponding neural foramina with a significant widening and attached to conus medullaris and the cauda equina. Unlike our case, other reported cases showed similar signal intensity to the spinal cord on T1- and T2-weighted images, with homogeneous strong contrast.

Figure 3: A lumbar spine CT scan: axial and sagittal views.

Figure 4: X-ray AP view and intraoperative image post-intrapedicular screw insertion and laminectomy.
enhancement [2, 18]. Dural tail sign was found in most cases of extradural en plaque lesion suggesting the dura-based origin [18]. Nevertheless, our patient did not present any signs that clarify the dura origin of the tumor. It was challenging to radiologically diagnose the lesion as a meningioma since it could easily be mistaken with spinal myxopapillary ependymoma neurofibroma or schwannoma.

The histological diagnosis of clear cell meningioma is challenging as it lacks cellular whirling and other meningeal features identified in typical meningiomas [19]. In addition, the extensive stromal and perivascular collagen is considered degenerative in nature and reflects the longstanding process, as in our case, it can obscure the cellular features adding to the diagnostic challenge of this tumor. The main histological differential diagnoses include other meningioma variants such as microcytic or angiomatous meningioma; however, PAS cytoplasmic reactivity and the round cell morphology highlighted by EMA are seen in clear cell meningioma.

Figure 6: (a) Clear cell meningioma exhibiting sheets of round to polygonal cells arranged in a haphazard manner alternating with areas with thick bulky interstitial collagenization (H&E, 100x). (b) High magnification image shows lesional cells with clear cytoplasm (H&E, 400x). (c) The cytoplasm contains glycogen demonstrated by periodic acid-Schiff (PAS; 400x). (d) Weak membranous immunoreactivity for epithelial membrane antigen (IHC, 400x).
meningioma but not in the other variants [20]. In addition, obscuring the cellular features by collagen adds to the challenge in diagnosis. Other differential diagnoses such as clear cell ependymoma, hemangioblastoma, and metastatic carcinoma can be ruled out by the negative glial fibrillary acidic protein, vascular markers, and keratins (including CAM2.5), respectively [21, 22]. Although the MIB-labeling index was 2%, data show no association between the MIB-labeling index and recurrence or outcome of these tumors. Higher MIB labelling has been described in recurrences [21].

4. Conclusion

We describe here a rare case of spinal intradural extramedullary meningioma with extradural extensions at the lumbar level with atypical presentation and radiological characteristics. However, meningiomas should be considered a differential diagnosis in a slow growing lesion as they may occur in rare locations with variable imaging findings and presentation.

Conflicts of Interest

The authors declare that there are no conflicts of interest regarding the publication of this paper.

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References

[1] S. Jain, I. Sundar, V. Sharma, R. Goel, and K. Prasanna, “Multiple spinal and cranial meningiomas: a case report and review of literature,” Asian Journal of Neurosurgery, vol. 10, no. 2, pp. 132–134, 2015.

[2] T. K. Maiti, S. C. Bir, D. P. Patra, P. Kalakoti, B. Guthikonda, and A. Nanda, “Spinal meningiomas: clinicoradiological factors predicting recurrence and functional outcome,” Neurosurgical Focus, vol. 41, no. 2, p. E6, 2016.

[3] W. Hong, E. S. Kim, Y. Lee et al., “Spinal Extradural Meningioma: A case report and review of the literature,” Journal of the Korean Society of Radiology, vol. 79, no. 1, 2018.

[4] C. Hartmann, J. Boström, and M. Simon, “Diagnostic and molecular pathology of meningiomas,” Expert Review of Neurotherapeutics, vol. 6, no. 11, pp. 1671–1683, 2006.

[5] A. Oikonomou, T. Birbilis, G. Daskalogiannakis, and P. Prassopoulos, “Meningioma of the conus medullaris mimicking neurofibroma—possibly radiation induced,” The Spine Journal, vol. 11, no. 2, pp. e11–e15, 2011.

[6] J. Mizutani, M. Fukuoka, S. Tsubouchi et al., “A rare case of lumbar sacral meningioma: nondural attachment and possible enlargement by orally administered sex steroid,” Spine, vol. 27, no. 16, pp. E377–E381, 2002.

[7] R. Buerki, C. Horbinski, T. Kruser, P. Horowitz, C. James, and R. Lukas, “An overview of meningiomas,” Future Oncology, vol. 14, no. 21, pp. 2161–2177, 2018.

[8] C. L. Solero, M. Fornari, S. Giombini et al., “Spinal meningiomas: review of 174 operated cases,” Neurosurgery, vol. 25, no. 2, pp. 153–160, 1989.

[9] J. B. Wood and S. M. Wolpert, “Lumbosacral meningioma,” American Journal of Neuroradiology, vol. 6, no. 3, pp. 450–451, 1985.

[10] S. M. El Khamary and I. A. Alorainy, “Case 100: Spinal Epidural Meningioma,” Radiology, vol. 241, no. 2, article Case 100: SPINAL epidural meningioma, pp. 614–617, 2006.

[11] G. Bettaswamy, P. Ambesh, K. K. Das et al., “Extradural spinal meningioma: revisiting a rare entity,” Journal of Craniovertebral Junction and Spine, vol. 7, no. 1, pp. 65–68, 2016.

[12] J. Li, S. Zhang, Q. Wang et al., “Spinal clear cell meningioma: clinical study with long-term follow-up in 12 patients,” World Neurosurgery, vol. 122, pp. e415–e426, 2019.

[13] H. Zhang, L. Ma, C. Shu, L. Dong, Y. Ma, and Y. Zhou, “Spinal clear cell meningiomas: clinical features and factors predicting recurrence,” World Neurosurgery, vol. 134, pp. e1062–e1076, 2020.

[14] H. S. Lee, S. H. Lee, Y. S. Chung, H.-J. Yang, Y.-J. Son, and S. B. Park, “Large spinal meningioma with hemorrhage after selective root block in the thoraco-lumbar spine,” Korean Journal of Spine, vol. 10, no. 4, pp. 255–257, 2013.

[15] M. Wostrack, E. Shiban, T. Obermueller, J. Gempt, B. Meyer, and F. Ringel, “Conus medullaris and cauda equina tumors: clinical presentation, prognosis, and outcome after surgical treatment,” Journal of Neurosurgery: Spine, vol. 20, no. 3, pp. 335–343, 2014.

[16] Y. Yeo, C. Park, J. W. Lee et al., “Magnetic resonance imaging spectrum of spinal meningioma,” Clinical Imaging, vol. 55, pp. 100–106, 2019.

[17] L. Hua, H. Zhu, J. Deng et al., “Clinical and prognostic features of spinal meningioma: a thorough analysis from a single neurosurgical center,” Journal of Neuro-Oncology, vol. 140, no. 3, pp. 659–647, 2018.

[18] M. K. Demir, Ö. Yapıcıer, Z. O. Tokta, A. Akakin, B. Yılmaz, and D. Konya, “Ossified-calcified intradural and extradural thoracic spinal meningioma with neural foraminal extension,” The Spine Journal, vol. 16, no. 1, pp. e35–e37, 2016.

[19] A. Oviedo, D. Pang, J. Zovickian, and M. Smith, “Clear cell meningioma: case report and review of the literature,” Pediatric and Developmental Pathology, vol. 8, no. 3, pp. 386–390, 2005.

[20] F. Alameda, J. Lloreta, M. D. Ferrer, J. M. Corominas, E. Galtiò, and S. Serrano, “Clear cell meningioma of the lumbo-sacral spine with chordoid features,” Ultrastructural Pathology, vol. 23, no. 1, pp. 51–58, 1999.

[21] S. Zorludemir, B. Scheithauer, T. Hirose, C. Houten, G. Miller, and F. Meyer, “Clear cell meningioma: a clinicopathologic study of a potentially aggressive variant of meningioma,” The American Journal of Surgical Pathology, vol. 19, no. 5, pp. 493–505, 1995.

[22] M. Takanori, I. Kenji, T. Tsunoe, and Y. Toshihiko, “Clear cell meningioma of cauda equina in a 10-year-old child,” 1970, Retrieved March 28, 2021, https://core.ac.uk/display/228805612?source=3.