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Case Report

Extradural spinal meningioma: Revisiting a rare entity

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Abstract

Spinal meningiomas are mostly intradural in location although at times these are associated with some extradural extensions. Purely extradural spinal meningiomas (EDSMs) are however, extremely rare and when present, may cause diagnostic dilemma preoperatively. Only seven cases of pure EDSM have been reported till date. In this paper, we describe two cases of EDSM affecting the cervical spine and present their clinical profiles, radiological findings, operative management, and follow-up data, along with a review of the literature.

Key words: En plaque, extradural, meningioma, pathology, spinal, surgery

INTRODUCTION

Spinal tumors broadly fall into two divisions, namely, extradural and intradural tumors. Further, intradural tumors can be either intra- or extramedullary in location. Common intradural extramedullary tumors are schwannomas, neurofibromas, and meningiomas while metastases form the bulk of the extradural lesions. Spinal meningiomas are usually intradural in location. However, very rarely (2.5-3.5%), these tumor scans be found lying entirely outside the dura.[1] A review of the literature revealed only seven cases of extradural spinal meningioma (EDSM) reported till date. However, all these cases have thoracic spine involvement. Here we report two cases of EDSM involving the cervical spine. We discuss their clinic-radiological findings and operative and follow-up details and discuss our cases in the light of relevant literature.

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CASE REPORTS

Case 1

Clinical and radiological details

A 50-year-old man presented to the emergency services with complaints of neck pain associated with tingling and burning paresthesia of all the four limbs for 8 months and progressive spastic quadriparesis with bladder disturbances for the past 1 week. Neurological examination revealed bilateral lower limb weakness [Medical Research Council (MRC) Grade 1/5] with normal power in both the upper limbs. Deep tendon reflexes were exaggerated in all four limbs. Sensory examination showed graded sensory loss below C4 level bilaterally.

Magnetic resonance imaging (MRI) showed an extradural

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en plaque lesion extending from C1 to C4, part of which was traversing through the right C2/3 intervertebral neural foramen, encasing the vertebral artery without its luminal compromise. On T1-weighted imaging, the lesion was iso- to hypointense, and it was isointense on T2-weighted imaging with intense contrast enhancement [Figure 1]. Presumptive diagnosis before surgery was extradural malignant spinal tumor (such as lymphoma or malignant peripheral nerve sheath tumor) with a rare possibility of chronic granulomatous pathology.

Operative intervention

After C2-C4 laminectomy, the tumor was found to be a soft, fleshy, vascular mass on the right side of the spinal canal. It was easily separable from the underlying dura with weak attachments at places. A near-total excision of the mass was performed coagulating the dural attachments. However, a small remnant, encasing the vertebral artery was intentionally left behind. Intradural exploration was done but no tumor extensions were seen. As the case was taken on an emergency basis, frozen section sampling was not possible. Later, it was revealed from the histopathological examination that the mass was a meningothelial meningioma [World Health Organization (WHO) Grade I].

Postoperative course

The patient became completely symptom-free after surgery and showed significant clinical improvement with improvement in lower limb muscle strength (+1/5 to +4/5) over a period of 2 weeks. MRI at 6 months follow-up showed no tumor recurrence [Figure 1].

Case 2

A 41-year-old gentleman presented with complaints of tingling sensation in the right hand, with progressively increasing spastic quadriparesis of 4 months duration. The patient also noticed neck movement restriction and straining during micturition for 2 months. Neurological examination revealed bilateral upper limb power of 3/5 MRC, and lower limb power of 4/5 MRC. Sensory examination showed graded sensory loss below C6 level bilaterally.

MRI showed an extradural en plaque lesion extending from C3 to C7 on the left side, showing T1 hypointensity and T2 hypointensity with intense contrast enhancement. There was no evidence of any transforaminal extension or transforaminal widening [Figure 2].

Operative intervention

After laminectomy, the tumor was found to be extending from C3 to C7 level. The mass was reddish white, highly vascular, and soft in consistency. This was present more on the left side without any ventral extension. Complete excision of the tumor was performed. No intradural extension was seen following intradural exploration. Histopathological examination revealed meningothelial meningioma.

Postoperative course

Significant alleviation in spasticity and improvement in motor power was seen at follow-up of 6 months (MRC Grade 5/5 bilateral). Postoperative MRI at 6 months and 24 months did not reveal any residual mass [Figure 2].

DISCUSSION

Meningiomas, aptly named by Harvey Cushing, arise contiguously from the meninges. The origin seems to be the arachnoid cap cells in the arachnoid cell rests present within the meninges.[2,3] They are the second most common intraspinal neoplasms after nerve sheath tumors. A well-established female preponderance (4:1) and predisposition of thoracic spine typifies spinal meningiomas (with around 80% of the tumors occurring in the thoracic spine).[4] Most begin manifesting themselves during the fifth or sixth decade of life. Spinal

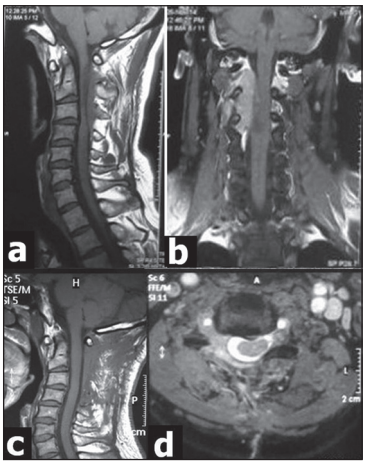

Figure 1: (a) Sagittal T1W shows isointense extradural lesion from C2 to C4 (b) T2 coronal shows lesion on right side and pushing the cord to the left (c and d) Postoperative images show no evidence of residual tumor

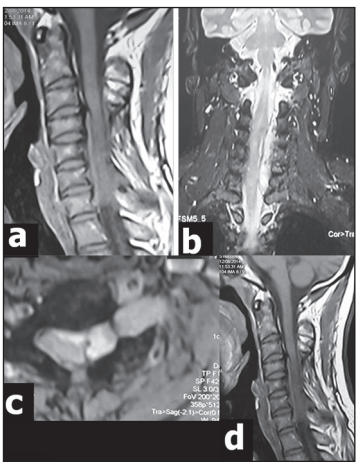

Figure 2: (a) Sagittal T1 weighted image showing isointense extradural lesion extending from C2 to C4 (b) shows intense contrast enhancing mass extending from C2 to C4 mainly on the left side. Axial contrast images (c) shows lesion extending along the foramen and is purely extradural (d) Postoperative images 4 years later reveal no recurrence

meningiomas commonly occur as ventral/ventrolaterally located intradural extramedullary tumors, arising from arachnoid cap cells near the ventral nerve root.[5] Approximately 10% of intradural spinal meningiomas may have an extradural extension but exclusively extradurally located meningiomas are extremely rare.[4,6-9] Only seven cases of EDSMs have been described in the literature [Table 1].

The cases reported till now show a female predilection and susceptibility of the patients in their fourth or fifth decade of life. Although ourpatients presented in their fifth decade of life, they were both male, unlike in previously published reports. Moreover, all previous reports show a thoracic spine involvement unlike our patients. Our cases were also unique in that the tumor was located posteriorly/posterolaterally unlike usual meningiomas.

Normally a time lag exists between symptom onset and presumptive diagnosis. Common presentations are pain, sensory/motor deficits, and sphincter weakness. Usually back pain/neck pain augurs motor and sensory changes, with sphincter problems occurring last.[10]

In both instances, patients suffered from mild paresthesias in all four limbs for a few months before spiraling into acute decompensation, which manifested as spastic quadriparesis, with urinary incontinence. This resulted in the patients becoming bedridden and catheter-dependent. Posterior/posterolateral location of the tumor would have compressed the posterior columns first, leading to early occurrence of paresthesias before the development of weakness of corticospinal tract origin. A review of the reported cases shows that the symptoms tend to be very subtle and longstanding. This is expectedly due to indolent growth rates of meningiomas. However, as seen in our patients, en plaque rather than focal growth pattern and transforminal growth allowing "natural decompression" of the cord must have contributed to the late onset of symptoms.

Pathogenesis and pathology

Multiple competing theories exist to explain the genesis of exclusively extradural meningiomas. One theory contends that they originate from ectopic arachnoid cells, around the periradicular nerve root sleeves, at the junction of the dura and spinal leptomeninx.[6] Another postulates that the thin periradiculardura may have vestigial embryonic remnants of the arachnoid mater and villi. This may produce extradural meningiomas close to the root.[4] A less likely theory suggests that islands of arachnoid tissue migrate into extradural space, eventually producing meningiomas.[4,6]

Common histological patterns are meningothelial, fibroblastic, transitional, and psammomatous, with most case reports demonstrating the meningothelial and psammomatous types.[11]

Histopathological analysis of our cases revealed a meningothelial pattern (WHO Grade I) in both the cases. Immunohistochemistry revealed a low KI-67 index (2-3% in Case 1, 1-2% in Case 2), suggestive of a low-grade lesion, with vimentin and EMA positivity, classical of meningioma.

Imaging

MRI is the best imaging modality for spinal meningiomas. A precise demarcation between the tumor and its cord relations is necessary for surgical intervention. Typically the lesion looks iso- or hypointense on T2-weighted images, in contrast to most hyperintense epidural tumors, barring lymphomas, which

Table 1: Reported cases of extradural spinal meningiomas

Reference	Age/sex	Level	Examination	Intervention	Histopathology	Extent of excision
Suzuki[15]	58 years/M	T10-T11-T12	Normal exam. Abnormal mass on the chest radiograph	T11-T12 hemilaminectomy+thoracoscope	Fibroblastic meningioma	Complete excision
Santiago[4]	42 years/M	T2-T3	Paraparesis	T2-T3 laminectomy	Psammomatous meningioma,	Complete excision
Shrestha[16]	45 years/F	T9-T10	Normal exam	T9-10 total laminectomy+ thoracoscopic surgery	meningioma	Complete excision, no complaints
Buchfelder [17]	76 years/F			Laminectomy+ thoracotomy	Meningioma	Complete excision
Kim[18]	50 years /M	T6-T7	Paresthesia at T6	hemilaminectomy of T6	Meningothelial meningioma	Complete excision, no complaints
Restrepo[19]	57 years/F	C7-T2	Loss of strength in the lower extremity, increase in reflexes	Total laminectomy + adjuvant RT	Psammomatous meningioma, infiltration	Complete excision, no recurrence
Can Yaldiz[20]	48 years/F	C7-T2	Hypoesthesia under T1	T1 total laminectomy	Psammomatous meningioma	Complete excision
Our case 2015	50 years /M	C2-C4	Hypoesthesia	C2-C4 laminectomy	Meningothelial meningioma	Near-complete excision, no recurrence
Our case 2011	41 years/M	C3-C7	Paresthesia and spastic quadriparesis	C3-C7 laminectomy	Meningothelial meningioma	Complete excision, no recurrence

can be hypointense in up to 50% of the cases.[7,10] Our cases showed a similar radiological presentation but with foraminal widening and transforaminal extension in Case 1.MRI also shows vertebral artery encroachment as seen in our first case. With both cases showing extradural sheet-like tumor growth, T2 hypointensity, and strong enhancement, a suspicion of infiltrative lesion-like lymphoma or granulomatous lesion was considered. Hence, intraoperative pathological examination using either frozen section examination or squash cytology is very important.[12]

Treatment

Gross total tumor resection has been advised, along with resection of the part infiltrating the bone or paraspinal space. For posterior locations, a posterior laminectomy or hemilaminectomy is recommended. Anterior locations are approached *via* laminectomy extension toward the facets, with minimal displacement of the cord. An anterior approach using posterolateral thoracotomy is another alternative. Spine stabilization is necessary if posterolateral approach compromises the facet joints.[13,14] Prognosis after surgery is highly dependent on the extent of excision and evidence of invasion on histology.[13]

As the pathogenesis suggests that EDSMs arise from the dural root sleeve and not from its external surface, the dura need not be excised. The flip side is that the dura has to be opened to rule out any intradural extension of the meningioma,[4] which in our cases also did not reveal any intradural component.

CONCLUSION

Pure spinal extradural meningioma is a rare entity. As we have shown, it can affect the cervical spine, can be located posterolateral to the dural tube, usually presents with en plaque growth, and may have transforaminal extensions as well. Preoperative presumptive diagnosis can be extremely difficult and requires a high index of suspicion. These lesions are amenable to complete surgical extirpation and do not require excision of dural attachments. The surgical outcome is satisfactory. More such cases need to be reported in the future for gaining further insight into this rare disease.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Milz H, Hamer J. Extradural spinal meningiomas. Report of two cases. Neurochirurgia (Stuttg) 1983;26:126-9.
2. Cushing H. The meningiomas (dural endotheliomas): Their source, and favoured seats of origin. Brain 1922;45:282-316.
3. Buetow MP, Buetow PC, Smirniotopoulos JG. Typical, atypical, and misleading features in meningioma. Radiographics 1991;11:1087-106.
4. Tuli J, Drzymalski DM, Lidov H, Tuli S. Extradural en-plaque spinal meningioma with intraneural invasion. World Neurosurg 2012;77:202.e5-13.
5. Santiago BM, Rodeia P, Cunha E, Sa M. Extradural thoracic spinal meningioma. Neurol India 2009;57:98.
6. Zevgaridis D, Thomé C. Purely epidural spinal meningioma mimicking metastatic tumor: Case report and review of literature. Spine (Phila Pa 1976) 2002;27:E403-5.
7. Dagain A, Dulou R, Lahutte M, Dutertre G, Pouit B, Delmas JM, *et al.* Extradural spinal meningioma: Case report. Neurochirurgie 2009;55:565-8.
8. Roux FX, Nataf F, Pinaudeau M, Borne G, Devaux B, Meder JF. Intraspinal meningiomas: Review of 54 cases with discussion of poor prognosis factors and modern therapeutic management. Surg Neurol 1996;46:458-64.
9. VargasMI, Abu Eid M, Bogorin A, beltechi R, Boyer P, Javier RM. Les méningiomes rachidiens extraduraux: Données IRM à propos de deux observations. J Neuroradiol 2004;31:214-9.
10. Levy Jr WJ, Bay J, Dohn D. Spinal cord meningioma. J Neurosurg 1982;57:804-12.
11. Gezen F, Kahraman S, Canakci Z, Bedük A. Review of 36 cases of spinal cord meningioma. Spine (Phila Pa 1976) 2000;25:727-31.
12. Savardekar A, Chatterjee D, Chatterjee D, Dhandapani S, Mohindra S, Salunke P. Totally extradural spinal en plaque meningiomas — Daignostic dilemmas and treatment strategies. Surg Neurol Int 2014;5(Suppl 7): S291-4.
13. Frank BL, Harrop JS, Hanna A, Ratliff J. Cervical extradural meningioma: Case report and literature review. J Spinal Cord Med 2008;31:302-5.
14. Yamada S, Kawai S, Yonezawa T, Masui K, Nishi N, Fujiwara K. Cervical extradural en-plaque meningioma. Neurol Med Chir (Tokyo) 2007;47:36-9.
15. Suzuki A, Nakamura H, Konishi S, Yamano Y. Dumbbell-shaped meningioma with cystic degeneration in the thoracic spine: A case report. Spine (Phila Pa 1976) 2002;27:E193-6.
16. Shrestha R, Chen H, Siqing H. Dumbbell meningioma of the thorocic spine resected by combined posterior and thoracoscopic approach. Nepal Journal of Neuroscience 2012;9:41-3.
17. Buchfelder M, Nomikos P, Paulus W, Rupprecht H. Spinal-thoracic dumbbell meningioma: A case report. Spine (Phila Pa 1976) 2001;26:1500-4.
18. Kim MS, Eun JP, Park JS. A dumbbell-shaped meningioma mimicking a schwannoma in the thoracic spine. J Korean Neurosurg Soc 2011;50:264-7.
19. Restrepo CS, Herrera DA, Lemos JA. Extraforaminal meningioma with extrapleural space extension. AJR Am J Roentgenol 2006;186:1314-6.
20. Yaldiz C, Asil K, Ceylan D, Erdem S. A dumbbell-shaped meningioma in the thoracic spine: A case report. J Spine 2014;3:168.