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Case Report

Cervical intra-extradural meningioma with en-plaque, dumbbell-shaped, and an unusual calcified pattern in a young patient

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ABSTRACT

Background: Most spinal meningiomas primarily grow in the intradural extramedullary location. Epidural meningiomas are uncommon; if detected, they usually coexist with intradural lesions. They inhere more aggressive and invasive characteristics compared with their counterparts inside the dura.

Case Description: We report a 22-year-old female who was admitted to the hospital with weakness and numbness in both lower limbs. Her cervical magnetic resonance imaging revealed an en-plaque and dumbbell-shaped lesion located from C5 to C8. After gadolinium injection, the whole mass was enhanced and unveiled two portions: intradural and extradural. The bone window of the computed tomography scan revealed calcification inside the lesion. The patient underwent tumor removal surgery. The pathology findings showed a psammomatous meningioma. After 6 months of surgery, the patient has been able to walk with walkers.

Conclusion: We should consider spinal meningioma as a differential diagnosis when encountering an extradural lesion in the cervical region. The optimal surgical treatment for young patient with epidural meningiomas is radical surgery with dura attachment removal.

Keywords: Calcification, Cervical spine, Dumbbell shaped, En plaque, Epidural meningioma

INTRODUCTION

Meningiomas are common lesions in the spine, covering about 25% of all spinal cord tumors.[31] The majority of them have been found in the extramedullary intradural location. Spinal epidural meningiomas are uncommon, involving only 2.5–3.5% of all spinal meningiomas,[25] and most of them are intraextradural meningiomas in nature.[27]

Spinal meningiomas originate from arachnoid cap cells lying within the arachnoid membrane.[32] Extradural findings, therefore, are exceptionally rare. Some authors have proposed several hypotheses for this phenomenon: ectopic arachnoidal cells proliferate around periradicular nerve root sleeves; primitive embryonic remnants of the arachnoid mater and villi anchor along the periradicular dura; or arachnoid tissue migrates into the extradural space.[37,41]

In general, epidural meningiomas have been categorized into three different growth patterns: en plaque, dumbbell shaped, and fusiform.[42] However, to the best of our knowledge, the simultaneous appearance of en-plaque and dumbbell-shaped features plus macroscopic calcification in the same epidural tumor of our patient is extremely exceptional and unique, resulting in an inconclusive preoperative diagnosis and an unpredictable surgical outcome.

CASE REPORT

A 22-year-old female was admitted to the hospital with weakness and numbness in both lower limbs. On neurological evaluation, sensory examination showed reduced pain and temperature sensations below dermatome T1 and loss of vibration sensation and proprioception in all extremities; motor examination showed 2/5 muscle strength in lower extremities and normal upper extremities strength except for a mild weakness at right fingers flexion. She also had global hyperreflexia in all extremities; ankle clonus and Babinski's sign are positive on both sides. Her cervical magnetic resonance imaging (MRI) revealed a lesion located from C5 to C8 hypointense compared to the spinal cord on the T1-weighted (T1W) image and slightly hyperintense on the T2-weighted (T2W) image. After gadolinium injection, the whole mass was enhanced and unveiled two portions: intradural and extradural [Figure 1]. The intradural part displayed an infiltrative pattern with an ambiguous border. On reviewing the coronal T1W with contrast, we noticed a dumbbell-shaped expansion at the left C6–C7 and C7-T1 intervertebral foramen [Figure 1f]. The bone window of the computed tomography (CT) scan revealed calcification inside the lesion [Figure 2]. With all these hurdles, the outcome of the surgery was unpredictable; thus, we could not inform our patient of a concrete surgical strategy. The patient underwent tumor removal surgery. During the operation, we confirmed that the tumor was macroscopic calcification, located in extramedullary, intra-, and extradural sites [Figure 3]. The tumor was completely removed followed by dura closure. The patient was discharged 5 days after surgery without any complication. She continued her rehabilitation plan and made significant improvements in muscle strength. After 6 months since the surgery, the patient has been able to walk with a walker. The pathology findings showed a psammomatous meningioma [Figure 4].

DISCUSSION

The histologic image that makes meningiomas standout among central nervous system tumors is psammoma bodies.[5] Considering neuroimaging, MRI is currently the superior diagnostic modality for suspected spinal meningiomas. We can indicate a CT scan in patients with contraindications for MRI. Indeed, CT scan has a higher

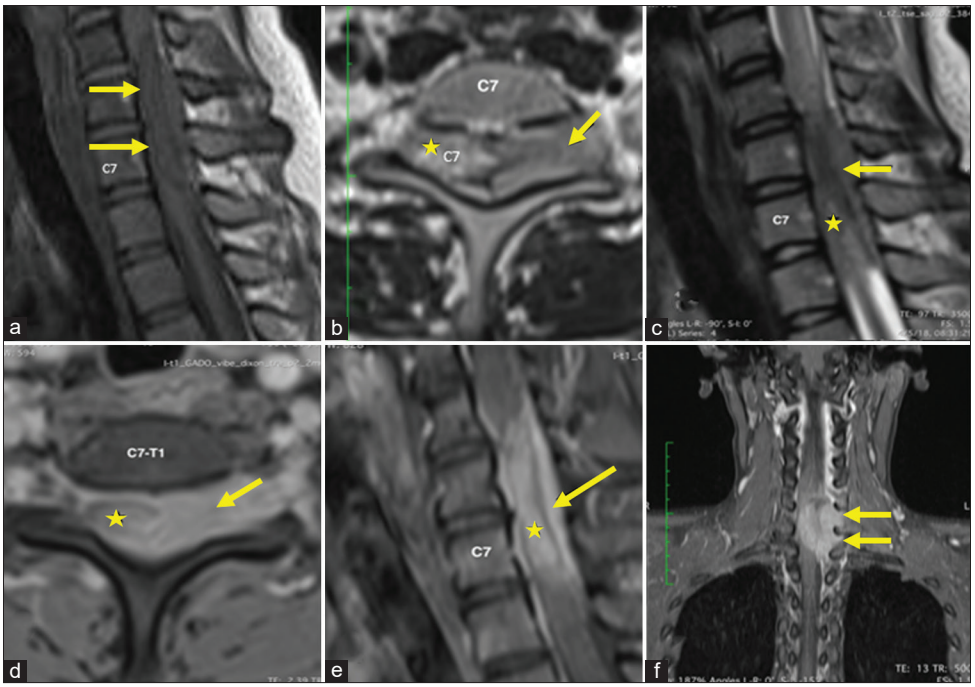

Figure 1: Magnetic resonance imaging of the cervical spine without gadolinium reveals a lesion which is hypointense (arrows) on the T1W sagittal (a) image. The lesion appears lightly hyperintense on the T2W axial (b) and sagittal (c) image with an intradural (star) and extradural (arrow) portion. Both components are enhanced after gadolinium injection on the T1W axial (d) and sagittal (e) image, which expands through the foramen on the T1W coronal image (f) with a dumbbell-shaped pattern.

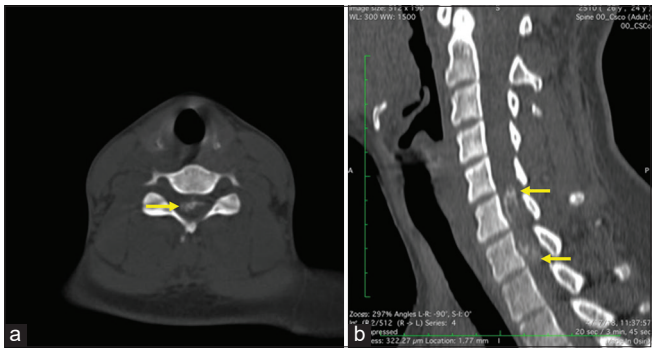

Figure 2: Computed tomography scan of cervical spine demonstrates calcification inside the lesion (arrows) on the axial (a) and sagittal (b) image.

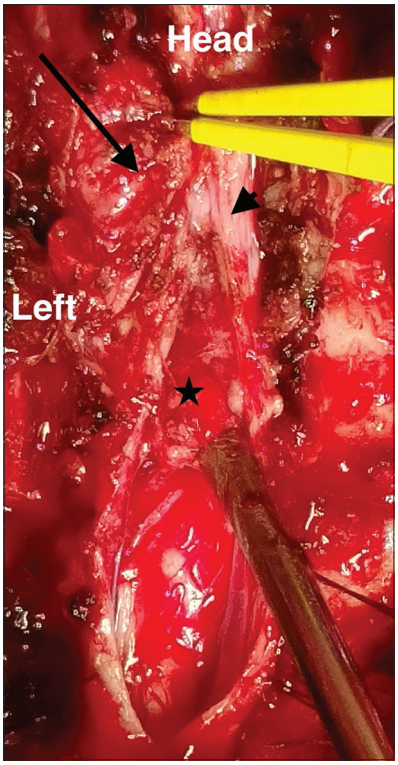

Figure 3: An extramedullary tumor with intradural (star) and extradural (arrow) portion.

sensitivity in detecting psammomatous calcifications;[26] hence, it will provide additional hints for the diagnosis in patients with a hazardous tumor appearance like in our case.

On MRI, spinal meningiomas normally show isointense to the spinal cord on T1W and T2W, and hypointense on T2W if calcified. T1W with contrast illustrates a prominent and homogeneous enhancement. Dural tails could usually be seen in this sequence. On CT scan, we could observe calcification which may assist a diagnosis of meningioma. However, it is only found in 1.0–4.6% of all spinal meningiomas.[24]

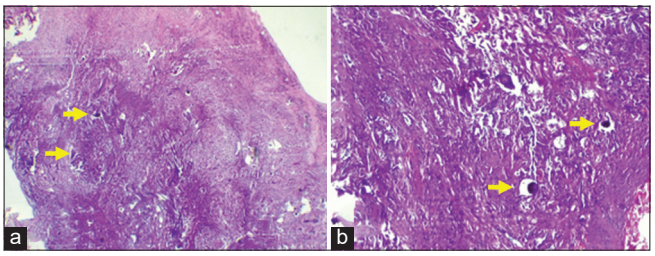

Figure 4: Histology study of the tumor revealed a psammomatous meningioma. This densely calcified tumor is characterized by the presence of numerous psammoma bodies (arrows). (a): ×40; hematoxylin-eosin stain, (b): ×100; hematoxylin-eosin stain.

Based on the location and the features of the lesion, we were concerned about either an intraextradural lesion, an extended intramedullary tumor, or metastasis. Our differential diagnoses include metastasis, ependymoma, glioblastoma multiforme, meningioma, and nerve sheath tumors (schwannoma/neurofibroma).

In spite of a negative metastatic work-up, physicians should always raise concerns about a cancerous potential when confronting an extradural lesion regardless of age. Metastases can spread throughout the vertebrae, epidural space, or spinal cord.[29] On MRI, metastasis is associated with the destruction of vertebrae and appears T1W hypointense and T2W focal hyperintense signal due to diffuse edema. T1W with contrast may display a heterogeneous enhancement.

The intradural portion has raised the concern about an extension of an intramedullary tumor. Spinal ependymomas arise from ependymal cells lining the central canal; therefore, they are strictly intramedullary in location. Due to their specific origin, they usually widen the spinal cord on the imaging. MRI reveals isointense to hypointense that would be seen on T1W and hyperintense on T2W. T1W with contrast shows a strong enhancement and is somewhat inhomogeneous. They classically have the "cap sign" (a hypointense hemosiderin rim on T2-weighted images), indicating a hemorrhage in 20–33% of cases. In contrast to intracranial ependymomas, calcification is uncommon in this entity.[18]

Despite the fact that calcified spinal glioblastoma multiforme is extremely rare,[30] we considered glioblastoma multiforme as a differential diagnosis due to the high disparity in the prognosis between this entity and other tumors. On the MRI, spinal glioblastoma shows isointense to hypointense on T1W and hyperintense on T2W. T1W with contrast demonstrates a patchy enhancement pattern.

Nerve sheath tumors are characterized by isointense or hypointense on T1W and may be heterogeneously hyperintense on T2W. The heterogeneous enhanced dumbbell-shaped expansion on the T1W with contrast has posed the likelihood of a nerve sheath tumor. However,

calcified schwannomas/neurofibromas are rare,[13] additionally it is very uncommon for these lesions to have two separated portions in and outside of the dura.

Microscopic calcification is a common finding, but gross appearance is only seen in 1–5% of all spinal meningiomas.[1] The fact that we identified this phenomenon in such young patient is even more uncommon; our case would be considered as an outlier in several previous studies (by Lunardi *et al.*, patient aged 67;[21] by Freidberg, patient aged 69;[10] and by Alafaci *et al.*,[1] patient aged 59).

Most spinal meningiomas are benign tumors; however, when they occur at younger age, the lesions are somewhat more aggressive.[6,7] The range of age at diagnosis is 14–75 years (mean 38 years), and 64.7% of patients are women.[9] This gender predominance, as highlighted by some authors, may indicate the role of sex hormones in the growth of meningiomas.[31] Authors have noted that in children and young individuals, epidural spinal meningiomas may not be as rare as in the general population,[6,39] posing an interesting question about the vulnerability of the dura in this population. The literature review identified 18 case reports of intraextradural meningioma which were confirmed intraoperatively [Table 1]. Although there have been some authors reported they found the tumor in the cervical and lumbar spine;[2,6,33,40] the majority of case, however, were thoracic (78%, 14/18 cases).[2,3,6,31,35] The mean age and the gender distribution are unable to obtain because there are 8/18 cases which the author did not disclose any the information other than the location.

In recent days, the word "dumbbell shaped" does not necessarily solely illustrate an hourglass figure, it could, however, broadly refer to a tumor which has two distinct parts connecting to each other from two different locations (i.e. intradural and epidural) but outside the paravertebral space.[15] Nevertheless, dumbbell-shaped meningioma is rare.[22,23,36] The MRI of the present case demonstrated a dumbbell-shaped tumor with a small extraspinal portion, which was confirmed intraoperatively. Unlike schwannomas which originate directly from nerve roots, meningiomas theoretically arise from the arachnoid membrane, from which they emerge to the extradural location via a narrow transforaminal space. Therefore, one could expect to detect an "imbalanced" dumbbell image from this group of meningiomas with a smaller end lying outside the dura as in our case.[4,6,12,13,22]

Table 1: Published cases of intraextradural meningioma which were confirmed intraoperatively.

Author	Intraextradural/ total	Age	Gender	Location
Soo, 1966[33]	1/2	7	Female	C1-T7
Calogero and	4/4	40	Female	T4
Moossy, 1961[6]		54	Male	L5 and sacrum
		28	Female	T9-T11
		33	Male	T7
Borghi, 1973[3]	2/118	60	Female	T3-T4
		28	Female	T5-T6
Stern *et al.*, 1980[35]	1/3	60	Male	T2-T6
Solero *et al.*, 1989[31]	8/174			"Mostly thoracic"
Yoshiura *et al.*, 1998[40]	1/1	16	Female	C2-C4
Barbanera *et al.*, 2007[2]	1/1	53	Female	C5-T1

Management and prognosis

From a surgical viewpoint it is very important to identify exactly the location and characteristics of meningiomas before surgery. As most meningiomas are benign, we can expect a good surgical outcome. However, en-plaque and calcified epidural meningiomas often severely adhere to adjacent tissues, making them extremely hard to be completely removed.[42] Even though an entire tumor has successfully been taken out, a prognosis may still be unchanged,[31] or even fatal. In a retrospective analysis of 97 cases of spinal meningioma of Levy *et al.*,[20] three out of four calcified tumors resulted in devastating consequences (e.g., paraplegia) after complete resection, the author suggested this aftermath probably due to an extra effort needed to achieve complete surgical resection, which, in turn, causes unexpected injuries to the cord.

The arachnoid, in fact, protects the cord from an expansion of meningiomas before the first surgery; from there on, it will inevitably be impaired because of scarring effects.[34] Thus, although the recurrence rate might not have been changed after each surgical attempt, recurrent meningiomas would be more invasive,[11,28,34,38] resulting in devastating CNS events.[8,14,16,38] This, indeed, emphasizes the important role of the first meningioma resection. Unfortunately, the recurrence rate after surgery of epidural meningiomas is 4 times higher than those found in intradural space.[17] Calcification potentially escalates the recurrent risk of spinal meningiomas. In Lee *et al.*'s study, five of the seven patients with calcified features had their lesions reappeared after 4 months to 4 years of follow-up.[19]

In our case, we have extracted the mass in toto followed by dura closure. Among elderly patients, the recurrence rate is generally not being affected by the management of the dural attachment (i.e., resection versus cauterization); this may be due to the latency effect.[24] Thus, in younger patients who have a longer life expectancy, we highly recommend radical surgery with dura attachment removal.

CONCLUSION

Epidural meningioma is uncommon and usually coexists with an intradural lesion; therefore, a careful preoperative

imaging evaluation has a critical role in establishing diagnosis and surgical strategy. Due to the longer life expectancy, the optimal treatment for young patients should be radical surgery with dura attachment removal.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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