Case Report

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Extradural Spinal Lymphoplasmacyte-Rich Meningioma in the Thoracic Spine: A Case Report and Literature Review

흉추에서 발생한 척수 경막 외 림프구 형질세포 과다형 수막종: 증례 보고와 문헌 고찰

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Most spinal meningiomas have an intradural or partly extradural location. The meningothelial origin is the most common pathologic type of spinal meningioma. Pure extradural spinal meningiomas are not common, and lymphoplasmacyte-rich meningioma (LPRM) is very rare. We report a case of isolated extradural spinal meningioma in the thoracic spine that was pathologically confirmed as LPRM.

Index terms Spine; Extradural Spinal Meningioma; Magnetic Resonance Imaging; Tomography, X-Ray Computed

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INTRODUCTION

Histologically, lymphoplasmacyte-rich meningioma (LPRM) is an extremely rare type of meningioma. An LPRM is a grade I tumor of the 2017 WHO classification of central nervous system tumors. Incidence of LPRM is less than 1% of all the meningiomas. LPRM was first announced by Banerjee and Blackwood in 1971, as a subfrontal tumor with characteristics of meningioma and plasmacytoma (1), including many inflammatory cells and various percentages of meningothelial elements (2). In previous reports, LPRM rarely occurred in the spine and developed only in the cervical spine (3).

We report MRI findings of pathologically confirmed isolated extradural LPRM in tho-

racic spine, a very rare type of spinal meningioma.

CASE REPORT

A 22-year-old female presented with back pain for 2 months and weakness and sensory loss for 2 weeks. The patient showed no neurologic deficit on physical examination, with motor grade five on both legs according to the medical research council (MRC) scale for muscle strength.

Thoracic spine CT demonstrated a crescent-shaped hyperdense solid mass that occupied the extradural space at the T9-10 disc level. There was no combined calcification, pressure erosion, or perilesional bony lesion (Fig. 1A). Thoracic spine MRI revealed an about 1.8-cm-sized extradural hyperintense mass on a T2-weighted image, encroaching the right side of the neural foramen of T9-10. The mass had higher signal intensity than the spinal cord on T1- and T2-weighted images and intense homogeneous enhancement in T1-enhanced images. The mass has dural tail sign on sagittal images (Fig. 1B). The spinal cord was compressed and shifted to the left side by the mass and showed an increase in signal intensity on T2-weighted images, which means compressive myelopathy. There was no cystic degeneration in the mass or discernible vascular structure. Preoperative radiologic differential diagnoses were extradural lymphoma or extradural meningioma.

The patient underwent surgical excision of the spinal mass, the day after radiologic diagnosis. T9 spinous process resection and right unilateral laminectomy was performed after midline incision of soft tissue. Then the surgeon identified a gray mass located from right posterior epidural space to right neural foraminal level. Spinal cord was displaced by the mass to anterolateral side. It was hard to dissect the mass, so intralesional curettage was done.

A pathologic examination of the mass revealed that multiple fragments of the tumor consisted of extensive chronic inflammatory infiltrates and focal meningothelial components. The meningothelial component has largely uniform cells with oval nuclei. The inflammatory infiltrates were mainly plasma cells, intermingled with occasional lymphocytes. The meningothelial cells are immuoreactive for progestin receptor and the plasma cells are inmmunoreactive for CD138 (Fig. 1C). The pathologic findings are consistent with LPRM.

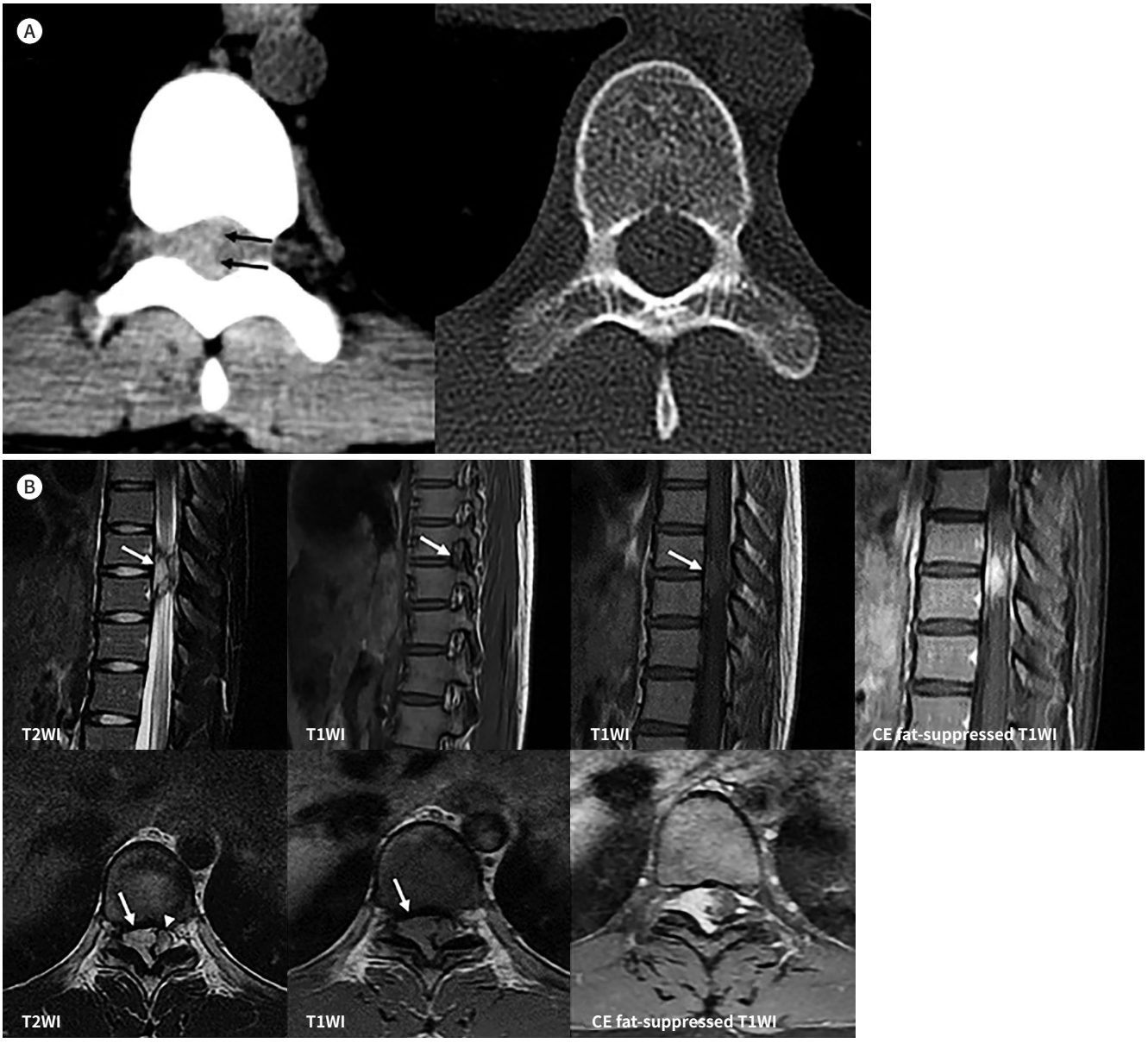
This retrospective study was approved by the local Institutional Review Board and informed consent was waived (IRB No. SCHBC 2021-09-018).

DISCUSSION

Spinal meningiomas are common spinal neoplasms, accounting for 25%–46% of primary spinal tumors (4). Meningiomas arising in the spine account for 12% of all the meningiomas (5). Most spinal meningiomas have an intradural extramedullary location but can sometimes extend to the extradural space. However, isolated extradural spinal meningiomas are rare, representing only 2.5%–3.5% of spinal meningiomas (4).

There are some hypotheses about development of extradural meningiomas. One is that extradural meningiomas arise from arachnoid cells around the periradicular nerve root at the border of the leptomeninx and dura mater. Another is that extradural meningiomas origi-

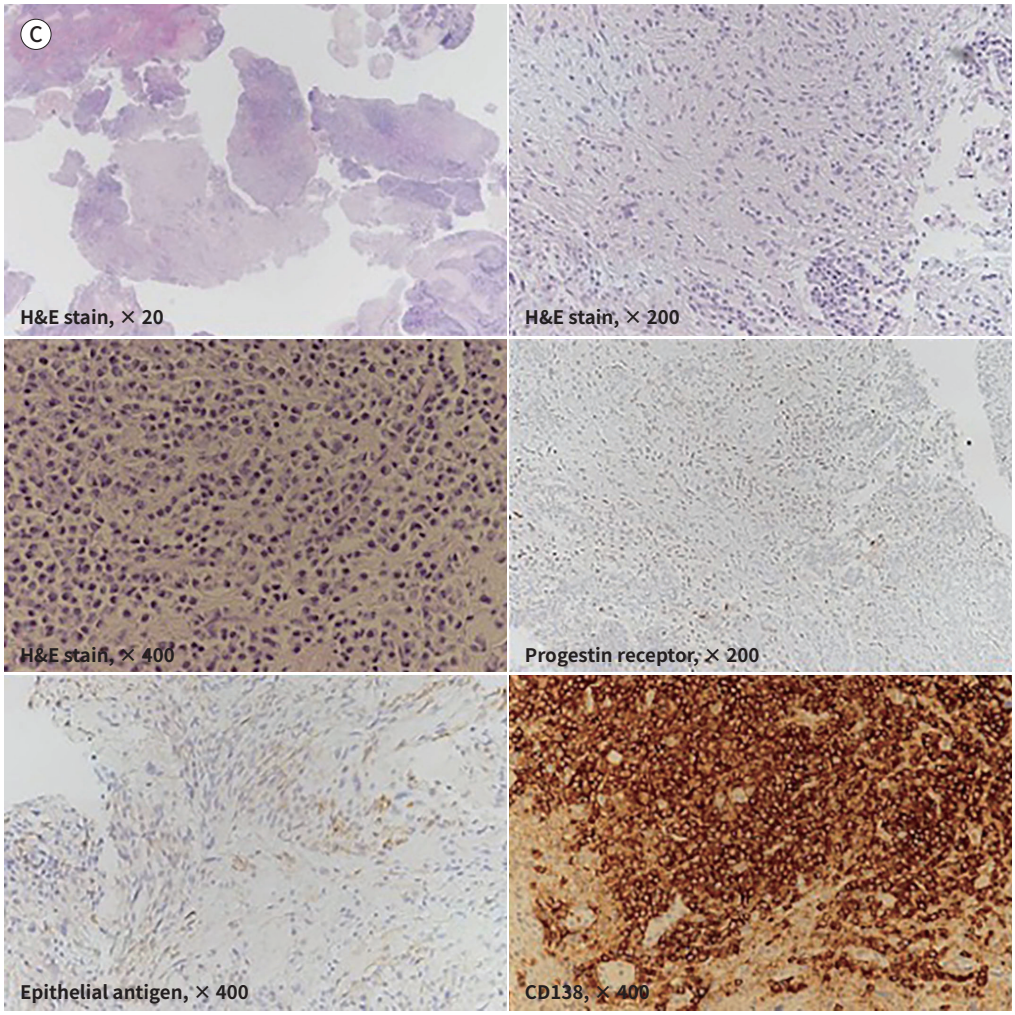
Fig. 1. Extradural spinal lymphoplasmacyte-rich meningioma in a 22-year-old female.
A. Axial non-CE CT shows a slightly hyperdense mass in the right epidural space of the T9-10 disc level, which has a crescent shape (arrows). There is no evidence of calcification or pressure erosion.
B. CE MRI shows that the mass (arrows) has a higher signal intensity than the spinal cord on the sagittal and axial T1WI. It shows a higher signal intensity than the spinal cord on the sagittal and axial T2WI. The mass encroaches the right side of neural foramen (sagittal T1WI and CE fat-suppressed T1WI). Given that the mass compressed the spinal cord to the left side, the signal intensity of the spinal cord increased on the T2WI (arrowhead). The mass shows a dural tail sign on sagittal images. The mass revealed homogeneous enhancement on the CE fat-suppressed T1WI.
CE = contrast-enhanced, T1WI = T1-weighted image, T2WI = T2-weighted image



nate from embryonic remnants of arachnoid mater and villi in periradicular dura. Finally, it is hypothesized that extradural meningiomas develop from aberrant arachnoid islets in the extradural space (4).

There are 39 reported cases of extradural spinal meningiomas since 2002 (6, 7). The pa-

Fig. 1. Extradural spinal lymphoplasmacyte-rich meningioma in a 22-year-old female.
C. Histologic examination shows that the multiple tumor fragments consist of extensive chronic inflammatory infiltrates and a focal meningothelial component (upper left). The meningothelial component shows a sheet of largely uniform cells, with oval nuclei as well as nuclear holes and a nuclear pseudoinclusion. These findings are compatible with normal meningothelial cells (upper right). The inflammatory cell components were mainly plasma cells intermingled with occasional lymphocytes (middle left). Immunohistochemical staining shows that the meningothelial cells are immunoreactive for progestin receptor (middle right), focally immunopositive for epithelial membrane antigen (lower left), and the plasma cells that are immunoreactive for CD138 are the dominant inflammatory component (lower right).
H&E = hematoxylin-eosin



tients showed female predominance, with 25 females and 14 males. The age of the patients ranged from 9 to 76 years, with a mean of 42.8 years. Of the 39 cases, 64.1% were located in the cervical spinal canal, 33.3% were in the thoracic spinal canal, and 2.5% were in the cervical and thoracic spinal canal. Most tumors showed en plaque lesion (en plaque: 61.5%, dumbbell shape: 15.4%, fusiform/oval shape: 2.5%). Histological findings of extradural spinal meningiomas are meningothelial, fibroblastic, transitional, psammomatous, angiomatous, and choroid meningioma. Of those reported cases, 53.8% were psammomatous meningiomas, 23.1% were meningothelial meningiomas, 5.1% were angiomatous type, 2.5% were fibroblas-

Table 1. Previously Reported Spinal Lymphoplasmacyte-Rich Meningioma

No.	Reference	Sex/Age	Level	Location	Treatment	Outcome
1	Mirra et al., 1983 (9)	F/39	C3-7	Subdural	Laminectomy	3 years, recurrence
2	Yamaki et al., 1997 (10)	M/22	Diffuse planum sphenoidale, parasellar to C5	Intradural	Partial resection and laminectomy	7 years, recurrent at C2-3, but others regressed
3	Yamaki et al., 1997 (10)	F/24	Multiple bilateral (clivus to spinal, C1-2)	Extradural	Subtotal resection	8 years, enlargement, 10 months later spontaneous regression
4	Zhu et al., 2013 (3)	M/25	C1-2	N/A	Total resection	37 months, no recurrence
5	Zhu et al., 2013 (3)	F/57	Foramen magnum to C1-4	N/A	Subtotal resection	33 months, no recurrence
6	Zhu et al., 2013 (3)	M/51	C3-4	N/A	Total resection	28 months, no recurrence
7	Present case	F/22	T9-10	Extradural	Excision	6 months, no recurrence

N/A was information unavailable.

tic, 2.5% were choroid, and 2.5% were transitional meningiomas. Most cases of extradural spinal meningiomas are psammomatous or meningothelial type (6). LPRM is a very rare type of meningiomas and occurs in cerebral convexities, skull base, para-sagittal area, or cerebral canal. Seven cases of spinal LPRM have been reported in the English literature, including the present case (Table 1). The presented case is the first LPRM not located in the cervical spine (3). This is the first case report of LPRM at the thoracic spine diagnosed with CT and MRI.

The epidemiology of LPRM is different from that of other extradural spinal meningiomas, which have a peak incidence in the fifth to sixth decades and female predominance (6). In contrast, LPRM occurs in younger people and has no sex predilection (4).

The exact pathogenesis of LPRM is unknown, and the origin, whether neoplastic or inflammatory, is controversial.

There are two hypotheses about the pathophysiology of LPRM. One is the result of collision of a plasmacytoma with a meningioma. Another is that meningothelial nests embedded within the inflammatory infiltrate which are hyperplastic rather than neoplastic in nature. The relationship between inflammatory lesions and meningiomas is not clear, but an inflammatory cell reaction to a meningioma produces LPRM (1, 3). In our case, LPRM was hyperdense compared to the spinal cord on CT, possibly reflecting the cellular nature of meningioma (8).

Similar to spinal meningioma, previously reported LPRM cases displayed hypo- to isointense signal intensity on T1-weighted imaging and hyperintense signal intensity on T2-weighted imaging. Such cases show strong homogeneous enhancement after administration of gadolinium, and a dural tail sign is observed in some cases (3). Most extradural spinal meningiomas share MR findings with those of spinal LPRM (6). Depending on tumor shape, epidural meningiomas can be classified into en plaque, dumbbell shaped, or ovoid/fusiform. As seen in the present case, most previously reported cases appeared as en plaque (6, 7).

When young patients have an enhanced extradural mass in the thoracic spine on MRI, extradural spinal LPRM must be distinguished as follows. Growth patterns and MR findings of lymphoma are similar to those of extradural meningioma. However, lymphomas usually are seen at the ventral side of the dura and are accompanied by a paravertebral soft tissue mass and infiltrative osteolysis of adjacent bone (7). On CT, epidural mass with higher density mass than the spinal cord is an additional clue to predict the nature of a meningioma.

Neurogenic tumors can have similar shape to an extradural mass, though they are more often dumbbell shaped rather than en plaque. In addition, neurogenic tumors have heterogeneous enhancement, while meningiomas show homogeneous contrast enhancement (7).

Extradural spinal LPRM is a very rare type of spinal meningioma. Although LPRM is a rare pathologic type of spinal meningioma, we find that imaging features of extradural spinal LPRM are similar to other types of extradural spinal meningiomas.

Author Contributions

Conceptualization, S.E.H., Y.S.Y., C.J.G.; funding acquisition, C.J.G.; resources, S.E.H., Y.S.Y., C.J.G.; supervision, C.J.G.; writing—original draft, S.E.H.; and writing—review & editing, all authors.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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흉추에서 발생한 척수 경막 외 림프구 형질세포 과다형 수막종: 증례 보고와 문헌 고찰

서은혜[1] · 차장규[1*] · 윤유성[1] · 문아림[2]

대부분의 척수 수막종은 경막 내에 위치하거나 부분적으로 경막 외에 위치한다. 척수 수막종의 가장 흔한 병리학적 유형은 수막세포종 기원이다. 온전하게 경막 외에 위치한 척수 수막종은 드물며 림프구 형질세포 과다형 수막종은 매우 드문 유형이다. 우리는 병리학적으로 확진된 흉추에서 발생한 경막 외 척수 림프구 형질세포 과다형 수막종의 자기공명영상과 컴퓨터단층촬영 영상의 소견을 소개하고자 한다.

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