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A Case of Thoracic Extradural Chordoid Meningioma: Focusing on Radiologic Features

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Chordoid meningioma, an uncommon subtype of meningioma, occurs rarely in the spine. In this case report, the authors present a case of spinal chordoid meningioma in a young female patient, and include a detailed description of imaging findings and a literature review.

Keywords: Extradural tumor; Extradural meningioma; Chordoid meningioma

Case Report

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INTRODUCTION

Chordoid meningioma, World Health Organization (WHO) grade II meningioma, is an uncommon pathologic subtype of meningioma. It is usually located supratentorially, and spinal chordoid meningioma is even more uncommon. Because of its rarity, there are very few reports in the literature on spinal chordoid meningioma, and no one has studied the imaging findings associated with it.

Here, we report a case of spinal chordoid meningioma in a young female patient, with a detailed description of radiologic findings.

CASE REPORT

A 26-year-old female patient without any medical history visited the outpatient clinics in our hospital. She complained of aggravating weakness in both legs and gait disturbance, and said that the weakness started two months ago and was more severe on her right side. Neurological examination revealed intact muscle power in both legs, but spastic gait was observed especially in her right leg, and an abnormal sensory level was detected along the T5 dermatome.

On CT scans, a 1.5 × 1.7 × 2.2 cm, well-defined osteolytic lesion without sclerotic margin was seen in the T3 vertebra, extending to the right side of the vertebral body, pedicle, and lamina (Fig. 1). Thoracic spine MRI was performed. There was an irregular-shaped mass involving the T3 vertebra and the right epidural space at the T2-3 level (Fig. 2). Narrowing of the spinal canal and displacement of the spinal cord to the left side were seen (Fig. 2f, g). The mass showed heterogeneously high signal intensity on T2-weighted image (Fig. 2a, f), iso- to hypo-intensity on T1-weighted image (Fig. 2b), and

intense enhancement with fuzzy margin on gadolinium-enhanced T1-weighted image (Fig. 2c, g). Diffusion restriction was not observed on diffusion-weighted image (DWI) (Fig. 2d), and the lesion revealed a high ADC (apparent diffusion coefficient) value on the ADC map (Fig. 2e). Brain MRI was also performed and there were no abnormal findings (not shown). Considering its extradural location, the radiologic differential diagnoses were Ewing's sarcoma, lymphoma, and metastasis. Although rare, spinal epidural hemangioma was also included in the differential diagnoses, due to hypervascularity of the mass.

For spinal cord decompression and histopathologic diagnosis, surgical removal of the tumor was advised. Under general anesthesia, total laminectomy was done at the T2 and T3, and the extradural tumor was removed.

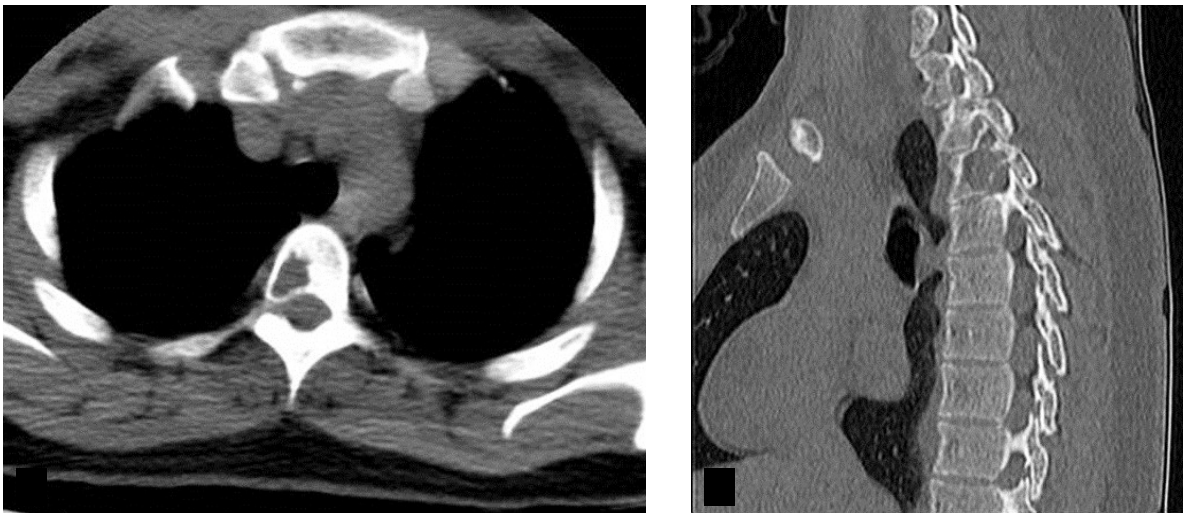
On histologic examination, the mass was composed of cords or trabeculae of eosinophilic epithelioid cells with a mucoid matrix, resembling chordoma (Fig. 3a, Hematoxylin & Eosin [H&E] stain, × 100). Chordoid areas were interspersed with more typical meningioma tissue (Fig. 3b, H&E stain, ×150). Tumor cells showed immunohistochemical reactivities for vimentin (Fig. 3c, ×100) and epithelial membrane antigen (EMA) (Fig. 3d, × 100). Histologically-mimicking lesions, such as chordoma, extraskeletal myxoid chondrosarcoma, poorly differentiated metastatic carcinoma, and ependymoma were excluded, based on negative immunohistochemical staining for pancytokeratin, S100, and glial fibrillary acid protein (GFAP). The final diagnosis was spinal chordoid meningioma.

After the surgery, the neurologic symptoms improved, but residual tumor was suspected on MRI (Fig. 4). During 3 months of post-operative follow-up, adjuvant radiation therapy was not conducted, and the patient did not show any neurologic symptoms.

DISCUSSION

Chordoid meningioma is an uncommon subtype of meningioma, comprising less than 1% of all meningiomas (1-4). The majority of chordoid meningiomas are located in the supratentorial area, and spinal chordoid meningioma is rare (3). Only 12 cases of spinal chordoid meningioma appear in the English literature (1-6) (Table 1). Even though not all of the data were available, it appears that spinal chordoid meningioma is more common in young patients, without male or female predominance, and commonly involves the cervical spine. The authors did not focus on the radiologic findings of spinal chordoid meningioma, and many did not report a detailed location of the tumor (e.g., intradural), an important factor for an imaging diagnosis of spinal tumor. For these reasons, preoperative radiologic diagnosis of spinal chordoid meningioma is still complicated, and this is the first case report of spinal chordoid meningioma which focused on its radiologic features.

In this case, chordoid meningioma, even meningioma, was not considered in the differential diagnosis from radiologic



a b

Fig. 1. Axial (a) and sagittal (b) CT scans show a well-defined osteolytic lesion without sclerotic margin, involving the T3 vertebral body, right pedicle and lamina.

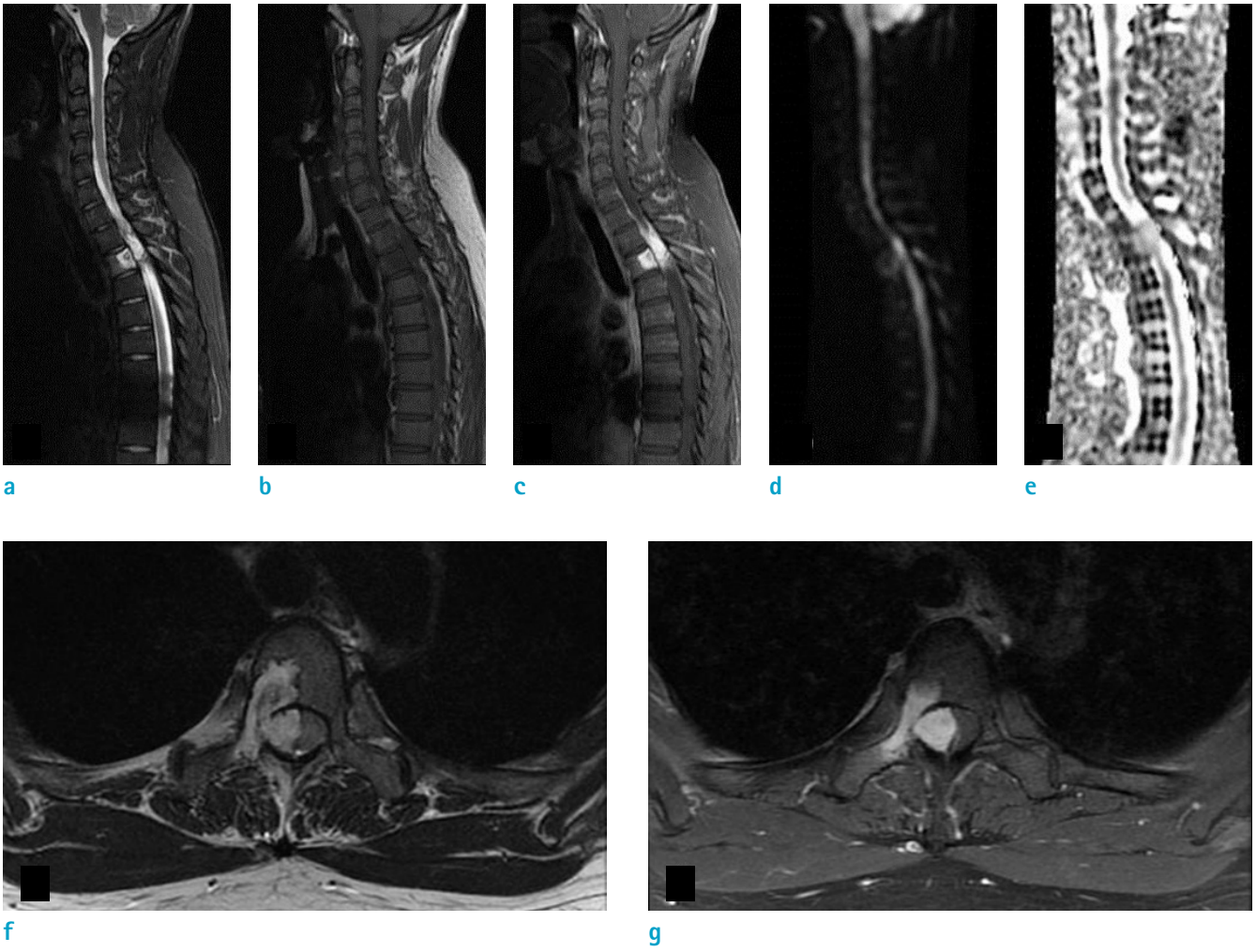


Fig. 2. Sagittal fat suppressed T2-weighted image (a), sagittal T1-weighted image (b), sagittal gadolinium-enhanced T1-weighted image (c), sagittal diffusion-weighted image (d), sagittal apparent diffusion coefficient map (e), axial T2-weighted image (f), and axial gadolinium-enhanced T1-weighted image (g). There is an irregularly shaped mass with high signal intensity on T2-weighted images (a, f) and intermediate to low signal intensity on T1-weighted image (b). The mass is located in the extradural space of the T2-3 level, and invades the posterior components of the T3 vertebra. Due to the mass effect, the spinal cord is displaced to the left side. On gadolinium-enhanced T1-weighted image (c, g), the mass shows heterogeneous, but avid enhancement with fuzzy margins. The mass shows slight hyperintensity on diffusion-weighted image (d), and apparent diffusion coefficient map (e).

studies, due to its extradural location. Extradural tumors are most commonly metastases, and usually involve multiple levels in patients with a known underlying malignancy. Lymphoproliferative disorders, such as lymphoma were also included in the differential diagnoses because they show variable imaging findings - osteolytic, sclerotic or mixed patterns on CT scan - and possibility paraspinal, vertebral and epidural involvement. However, they also commonly show multiple spinal lesions. Spinal epidural hemangioma is rare, but it can appear as a hypervascular dumbbell-shaped mass (7). However, common findings of spinal epidural hemangioma, such as lobulated contour and T2 hypointense rim were not seen this patient (8). At first, Ewing's sarcoma was suspected in this case because it preferentially occurs in adolescence and is common in the posterior element of the vertebrae. Both sclerotic and lytic lesions can be seen on CT, and paravertebral and epidural components commonly accompany the lesions. Due to the rarity of the extradural spinal meningioma, it is not the preferential diagnosis.

Extradural spinal meningiomas are reported in only

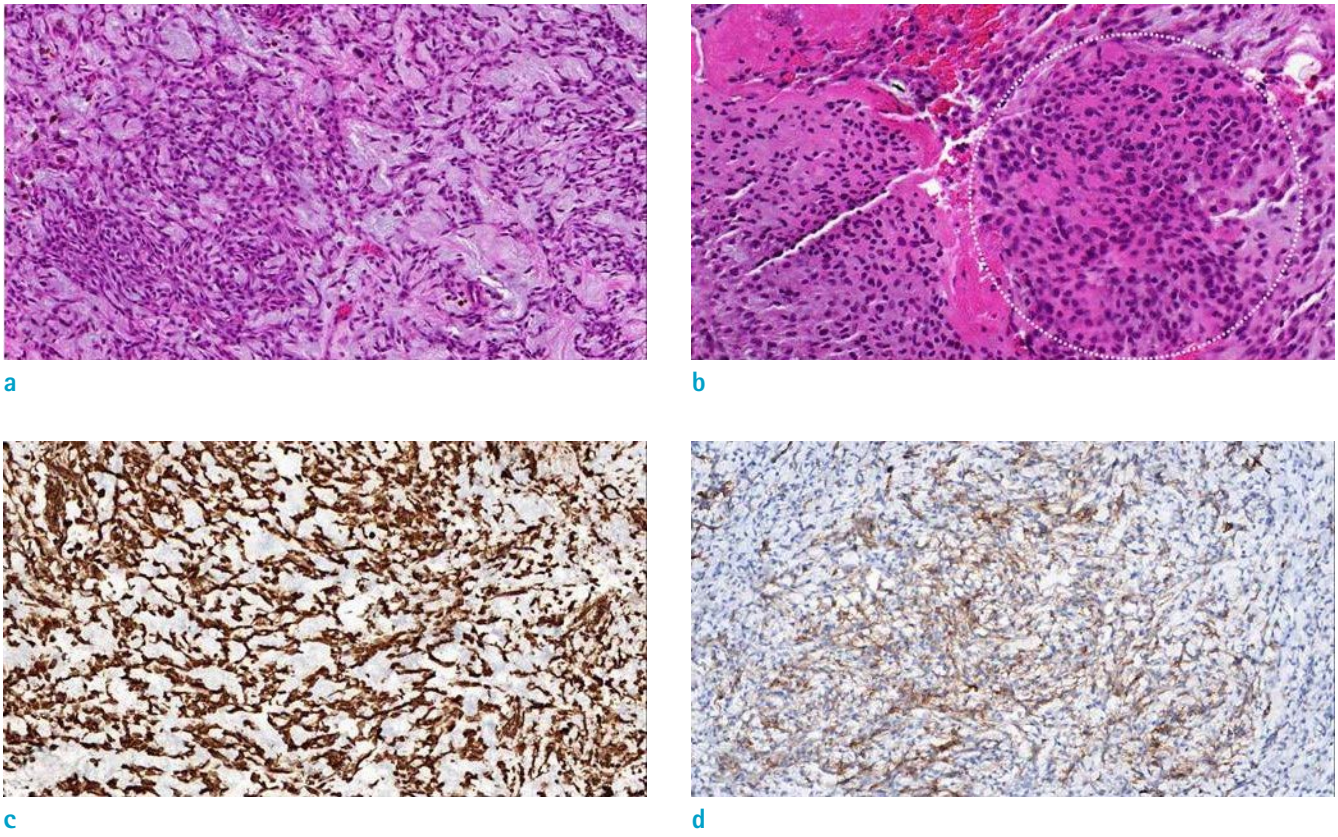


Fig. 3. Histopathologic findings. (a) Chords or trabeculae of eosinophilic epithelioid tumor cells in mucoid matrix (Hematoxylin & Eosin [H&E] stain, × 100). (b) Typical meningothelial meningioma-like focus (dotted circle, H&E stain, × 150). (c) Immunohistochemical staining for vimentin highlights the ribbon-like architecture (× 100). (d) Positivity for EMA is typically patchy in meningioma (× 100).

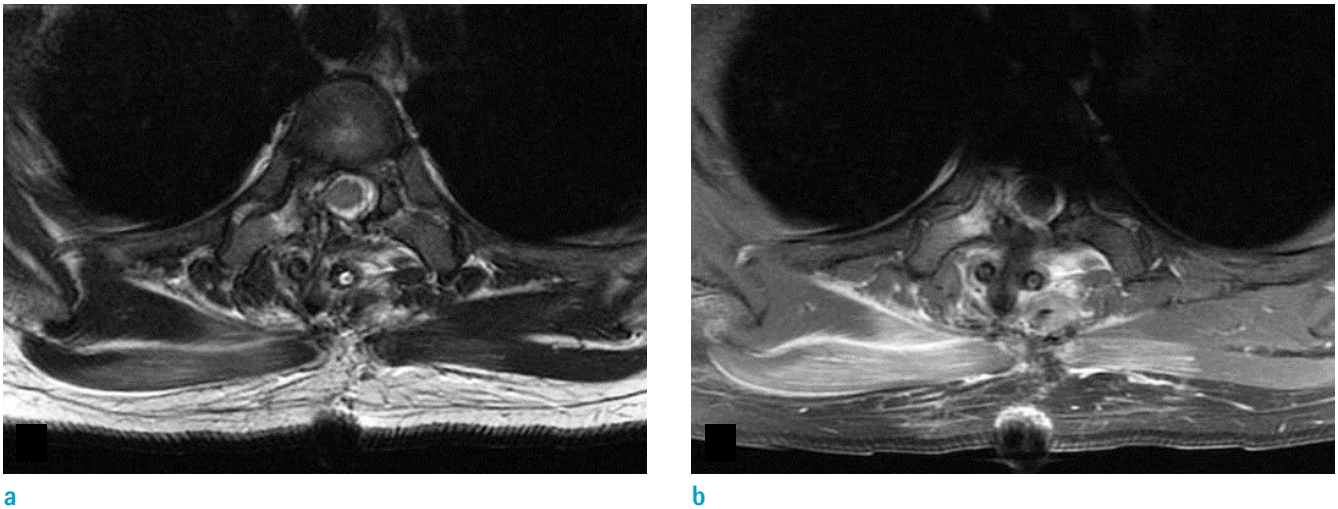


Fig. 4. On post-operative MRI, ill-defined hyperintense areas are noted in the T3 vertebra on axial T2-weighted image (a), and enhancing lesion in the same area on axial gadolinium-enhanced T1-weighted image (b), suggesting residual tumor.

Table1. Reported Cases of Spinal Chordoid Meningioma

Authors (Ref. No.)	No.	Sex	Age	Level	Location
Sadashiva N, et al., 2018 (1)	1	M	25	C3-5	IDEM
	2	F	36	Foramen magnum to C3	NA
	3	M	27	C1-2	IDEM
Tulloch I, et al., 2018 (2)	4	F	45	T5-7	ED
Yang Y, et al., 2016 (3)	5	NA	NA	Cervical	NA
	6	NA	NA	Cervical	NA
	7	NA	NA	Thoracic	NA
	8	NA	NA	Thoracic	NA
	9	NA	NA	Lumbar	NA
Wu L, et al., 2015 (4)	10	F	12	C2-3	IDEM + ED
Ibrahim A, et al., 2005 (5)	11	M	26	C2-3	IDEM
Couce ME, et al., 2000 (6)	12	F	20	C2	NA
Present case	13	F	26	T2-3	ED

M = male; F = female; IDEM = intradural extramedullary; ED = extradural; NA = not available

2.5-3.5% of all spinal meningiomas (9). The proposed pathogenesis of extradural meningiomas are that they originate from: 1) ectopic arachnoid cells around the periradicular root sleeve where the spinal meninx fuses with the dura, 2) vestigial remnants of the superficial layers of the embryonal arachnoid mater and villi which might be contained in the periradicular dura, and 3) island or arachnoid tissue migrating into the extradural space (9).

Previous reports have suggested that MRI findings in cases of cerebral and spinal chordoid meningioma are not different from other meningiomas (1, 3). There have been no reports of radiologic findings in spinal chordoid meningioma, but one article reported characteristic MRI findings in intracranial chordoid meningioma (10). Meningiomas showed high signal intensity on T2-weighted image, intermediate signal intensity on T1-weighted images, avid enhancement on gadolinium-enhanced T1-weighted image, and variable signal intensity on diffusion-weighted image. The size, location, signal characteristics, and contrast enhancement were not significantly different between chordoid meningiomas and other meningiomas, but ADC value and normalized ADC value were significantly elevated in chordoid meningiomas (10). The authors suggested that the high ADC value and normalized ADC values resulted from unique tumoral architecture and cytologic characteristics of chordoid meningioma: mucoid extracellular matrix allowing relatively free extracellular water motion, and increased diffusivity of water due to prominent cell vacuolization. Similar MRI findings were observed in our case. The signal characteristics and contrast enhancement of the tumor was similar to those of other spinal meningiomas, but the tumor characteristically showed a high ADC value on the ADC map, probably due to its unique histologic features.

Chordoid meningioma is classified as a WHO grade II meningioma because of its aggressive behavior and high recurrence rate (2). Surgical removal of the tumor is the mainstay of treatment and complete excision is important to minimize local recurrence (2-4). The role of adjuvant radiation therapy is controversial (3, 4).

In conclusion, this is the first case report of spinal chordoid meningioma, focusing on radiologic manifestations. However, preoperative radiologic diagnosis of spinal chordoid meningioma was difficult. The meningioma was located in the extradural space of the thoracic spine, and showed a high ADC value. The radiologic characteristics of spinal chordoid meningioma may be further elucidated by more cases and organized analysis. This case adds to that knowledge.

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