Clinical Case Report

Medicine®

OPEN

Lumbar intraspinal microcystic/reticular schwannoma

Case report and literature review

Congcong Liu, MD[a], Lianqi Yan, MD[a,b,c], Qing Liu, MD[d], Jing Li, MD[d], Hongtao Jin, MD[e], Jingcheng Wang, MD[a,b,c,*], Youwen Deng, MD[f,d,*]

Abstract

Rationale: Microcystic/reticular schwannoma (MRS) is a rare histological variant of schwannoma which was initially described in 2008 with a predilection for the visceral organs. This distinct tumor had been reported to mainly affect gastrointestinal tract, subcutaneous and soft tissue, various glands and head and neck region. However, MRS involving spine is extremely rare.

Patient concerns: The authors report the first case of MRS occurring in the lumbar (L) spinal canal of a 40-year-old male who presented with continuous pain and numbness in both feet for 2.5 years. Physical examination revealed weakness of lower extremities and hyperalgesia of both feet.

Diagnoses and interventions: The findings of pre-operative investigation were suspicious for either a schwannoma or a spinal meningioma. Accordingly, total laminectomy, complete tumor resection, instrumentation and spinal fusion were performed. Post-operative histopathologic examination revealed a well-encapsulated neoplasm with reticular and microcystic growth pattern. Antoni A and Antoni B regions, Verocay bodies and hyalinized blood vessels were observed. And cytologic atypia, necrosis or mitosis was absent. Immunohistochemically, the tumor cells showed strong and diffuse positivity for S-100 as well as SOX 10. Therefore, a histopathological diagnosis of MRS was finally made.

Outcomes: The patient remains well with no evidence of recurrence at a 22-month follow-up.

Lessons: This is the first case of MRS which is located in the L spinal canal. Awareness of this distinctive entity is helpful in preventing diagnostic pitfalls and making correct treatment strategies.

Abbreviations: EMA = epithelial membrane antigen, EMC = external mucosal chondrosarcoma, GFAP = glial fibrillary acidic protein, L = lumbar, MRS = microcystic/reticular schwannoma.

Keywords: differential diagnosis, lumbar spine, microcystic, reticular, schwannoma

1. Introduction

Schwannomas are benign and generally nonrecurring tumors that usually arise in adults, with no sex predilection.[1] The common anatomic distributions of these neoplasms are the subcutaneous tissue of distal extremities and the head and neck region. There are several morphologic variants of schwannoma, including conventional, cellular, plexiform, melanotic and microcystic/reticular schwannoma (MRS).[2,3] MRS is a rare histological variant of schwannoma which was initially described in 2008 with a predilection for the visceral organs. Besides, the tumors also involve rare anatomic sites including retroperitoneum, cerebellopontine angle, mediastinum and cervical vertebra.[2,4,5] Herein the authors report a unique case of MRS which located in the L spinal canal, a unique site which has never been reported to date, without bone erosion. In addition, the clinical and pathological features of 36 cases of MRS that reported previously were also summarized.

2. Case report

2.1. Clinical findings

This study was approved by the Ethics Committee of The Second Xiangya Hospital of Central South University. And written informed consent was obtained from the patient. A 40-year-old male presenting with pain and numbness in both feet for 2.5 years was admitted. The pain was continuous and severe, being irrelevant to posture. The patient had been treated with pharmaceuticals and physiotherapy as L disc herniation in local hospital. However, the condition deteriorated and the patient began to walk unsteadily 4 months ago. The patient had no spinal

Editor: N/A.

The authors have no funding and conflicts of interest to disclose.

[a] Department of Orthopedics, The Second Xiangya Hospital, Central South University, Changsha, Hunan, [b] Department of Orthopedics, Clinical medical college of Yangzhou University, [c] Orthopedics Institute, Subei People's Hospital of Jiangsu Province, Yangzhou, Jiangsu, [d] Department of Spine Surgery, The Second Xiangya Hospital, Central South University, Changsha, Hunan, [e] Department of Pathology, Shen Zhen People's Hospital, Second Clinical Medical College of Jinan University, Shenzhen, Guangdong, [f] Department of Emergency Medicine, The Second Xiangya Hospital, Central South University, Changsha, Hunan, China.

[*] Correspondence: Jingcheng Wang, Department of Orthopedics, The Second Xiangya Hospital of Central South University, No. 139, Middle of Renmin Road, Changsha 410011, Hunan, P.R. China (e-mail: sbyywjc@csu.edu.cn); Youwen Deng, Department of Emergency Medicine, The Second Xiangya Hospital of Central South University, No. 139, Middle of Renmin Road, Changsha 410011, Hunan, P.R. China (e-mail: drywdeng@csu.edu.cn).

Medicine (2018) 97:39(e12474)

Received: 19 April 2018 / Accepted: 24 August 2018

http://dx.doi.org/10.1097/MD.0000000000012474

condition, neuropathy or any related condition before. And the familial history was uneventful. Physical examination revealed weakness of lower extremities and hyperalgesia of both feet, without pathologic reflex. Laboratory tests including 12 tumor markers revealed no positive findings.

Computed tomography showed isthmus spondyloschisis in the fifth L vertebra, mild posterior protrusion of intervertebral discs of L2/3, L3/4, L4/5, and L5/S1. Magnetic resonance imaging revealed a spindle lesion in the L spinal canal from the level of T12 to L3 (Fig. 1A–C, E, and F). The lesion showed equal signal on T1-weighted images (T1WI) (Fig. 1A) and mixed-signal on both T2-weighted images (T2WI) (Fig. 1B, E, and F) and fat-suppression sequence images (Fig. 1C). Besides, heterogeneous enhancement was also detected. Initial diagnosis was made as a primary neoplasm in the intradural extramedullary space, probably a schwannoma or a spinal meningioma.

Total laminectomy from L1 to L3, tumor resection, pedicle screw fixation from T12 to L3 and posterolateral spinal fusion was performed. And the final histopathological diagnosis of MRS was made according to paraffin sections. Neither radiotherapy or chemotherapy nor biotherapy was performed. The patient gained a follow-up of 22 months with no evidence of recurrence (Fig. 1D, G).

2.2. Histopathological findings

2.2.1. Microscopic features. In the present case, the tumor was circumscribed and well-capsulated (Fig. 2A). Mucous network, cystic structure, and hyalinized blood vessels were

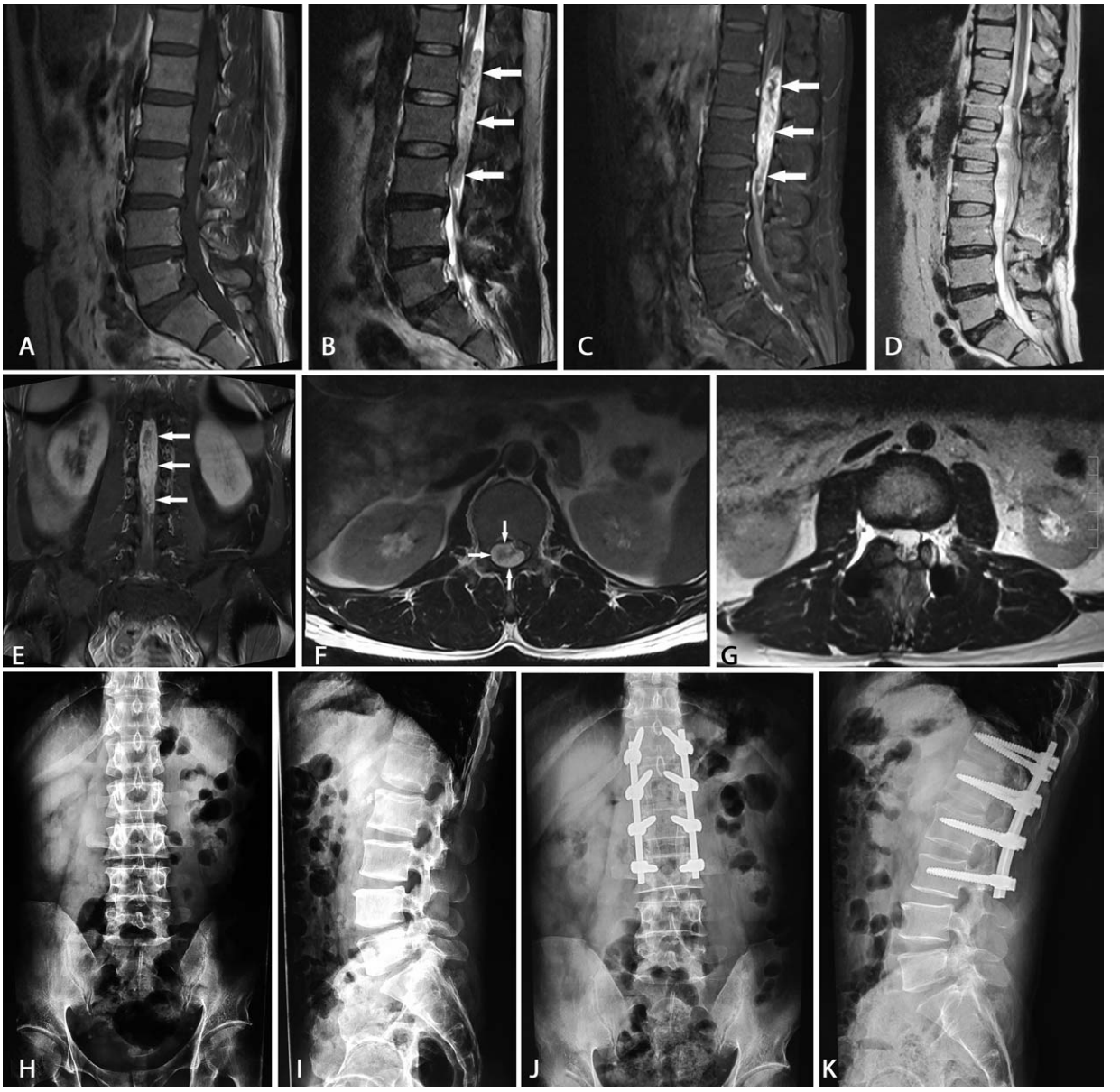

Figure 1. Pre-operative and final follow-up magnetic resonance images as well as radiographs of lumbar spine. Pre-operative sagittal, coronal and horizontal plane of magnetic resonance images revealed a spindle mass with well-defined demarcation in the lumbar spinal canal. The lesion showed equal signal on T1-weighted images (A) and mixed signal on both T2-weighted images (B, E, and F) and fat-suppression sequence images (C). Bone destruction was absent while isthmus spondyloschisis in the fifth lumbar vertebra was detected on pre-operative anteroposterior and lateral radiographs of lumbar spine (H and I). At the final follow-up, no sign of tumor recurrence was observed on magnetic resonance images of lumbar spine (D and G). And no evidence of instrumentation loosening or breakage was found (J and K).

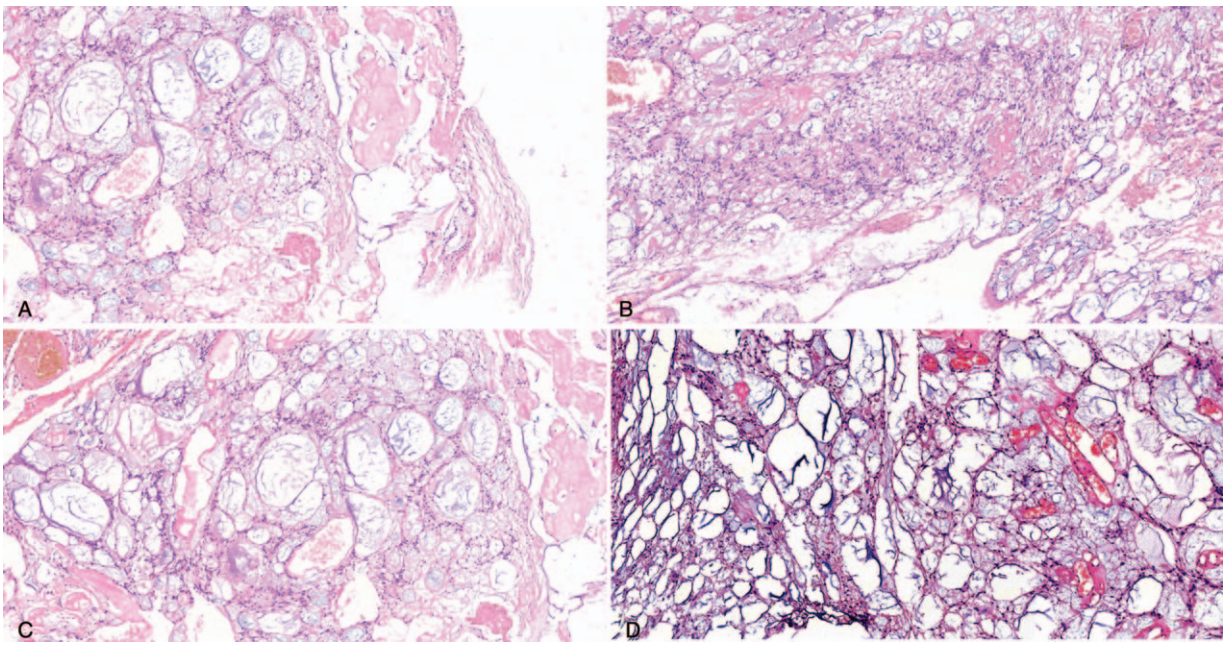

Figure 2. Microscopic findings (hematoxylin-eosin staining). (A) A fibrous capsule was found at the periphery (×200). (B) Alternately distributed Antoni A and Antoni B regions were detected (×100). (C) Microcystic arrangement of tumor cells in partial areas (×100). (D) Tumor cells are arranged in reticular structures, with prominent myxoid matrix (×100).

detected. Both Antoni A and Antoni B areas could be found and the latter was the main part (Fig. 2B). The structure of palisading nucleus known as Varocay bodies were also observed (Fig. 2C and D). In addition, cytologic atypia, necrosis or mitosis was absent.

2.2.2. Immunohistochemistry. Tumor cells showed diffuse and strong nuclear as well as cytoplasmic positivity for S-100 (Fig. 3A), nuclear immunoreactivity for SOX 10 (Fig. 3B), cytoplasmic immunoreactivity for glial fibrillary acidic protein (GFAP) (Fig. 3C) and Vim. Besides, cytokeratins (Fig. 3D),

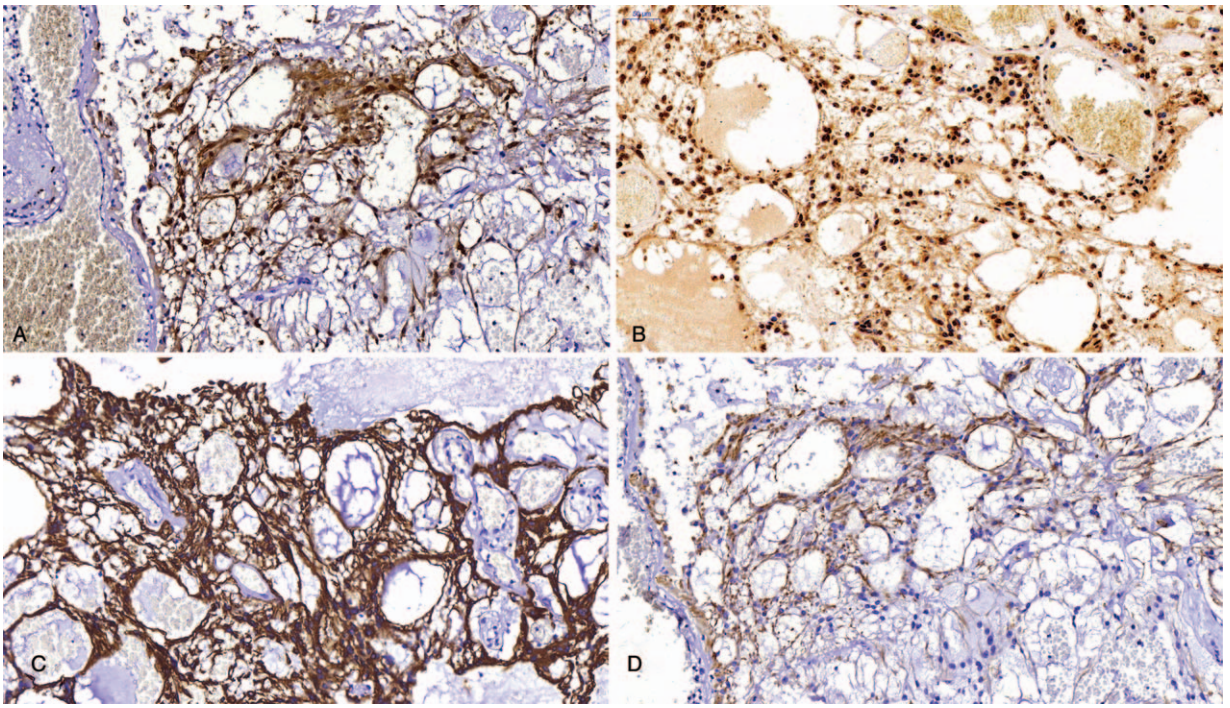

Figure 3. Immunohistochemical staining. A: Strong and diffuse positivity for S-100 was detected in cytoplasm and nucleus of tumor cells (×200). (B) Tumor cells show nuclear positivity for SOX 10 (×200). (C) Cytoplasm of tumor cells is immunoreactive for GFAP (×200). (D) Negative cytokeratin staining within the tumor cells (×200).

CD31, CD34, epithelial membrane antigen (EMA), Syn, CgA, CEA staining were performed with negative results.

3. Discussion

Schwannomas are benign mesenchymal tumors which derive from the cells of Schwann that form the neural sheath.[6] Anatomically, the distribution of schwannoma is wide, and the majority of tumors located at the distal extremities or the subcutaneous tissue of the head and neck region.[7] There are several morphologic variants of schwannoma, including cellular schwannoma,[8,9] ancient schwannoma,[10] plexiform schwannoma,[9,11,12] epithelioid schwannoma,[13,14] glandular schwannoma,[9,15] melanotic schwannoma,[11,16] hybrid schwannoma/perineurioma[17] as well as hybrid schwannoma/neurofibroma.[2,18] Generally, schwannomas are considered as benign and nonrecurring tumors, but rare cases of malignant schwannomas had also been reported.[19]

Conventional schwannomas are usually well encapsulated. They show 2 different morphological components defined as Antoni A and Antoni B areas in different ratios. Antoni A area refers to a cellular component which consists of spindle-shaped cells with ill-defined eosinophilic cytoplasm and wavy basophilic nuclei.[2] Verocay bodies are defined as cellular regions surrounded by nuclear palisades.[2,8] But Verocay bodies are not pathognomonic for schwannomas since they have also been demonstrated in several other tumors. The adhesive property of laminin may result in the tight arrangement of Antoni A areas.[20] Compared to the tight arrangement of Antoni A regions, hypocellular Antoni B regions contain more loosely organized components including lipid-laden histiocytes, lymphocytes, and myxoid stroma.[21] Occasionally, the size of Antoni B regions can be scant or absent. A common feature of both Antoni A and Antoni B regions is the presence of hyalinized thick-walled vessels.

Although not completely specific, S-100 protein which is especially prevalent in the Antoni A areas is a reliable diagnostic marker for schwannoma.[19,22] Schwannoma cells show strong nuclear and cytoplasmic positivity for S-100 and the perineurial capsule can be stained with EMA. S-100 is a highly acidic protein which is found in many neural crest tumors and may be related to the ionic regulation of nervous tissue.[19]

Increasing cases of MRS have been described gradually since 2008. The authors reviewed 36 cases of MRS previously reported in the literature (Table 1). Fourteen cases arose in the digestive tract, including 4 in the colon, 3 in the small intestine, 2 in the stomach, 2 in the cecum, and 1 each in the esophagus, rectum and

Table 1

Clinical features of 36 cases.

Case No.	Age(yr)/ Sex	Site	Size(cm)	Growth pattern	Final follow-up status	References
Digestive tract						
1	39/F	Esophagus	3.5	Unencapsulated	ANED at 27	Gu et al[7]
2	72/F	Stomach	2.0	Unencapsulated	ANED at 24	Liegl et al[2]
3	63/F	Stomach	1.9	Unencapsulated	ANED at 60	Chetty et al[3]
4	67/F	Mid-jejunum	2.2	UA	ANED at 2	Agaimy et al[27]
5	93/F	Jejunum	1.6	Unencapsulated	ANED at 7	Liegl et al[2]
6	78/M	Small intestine	0.8	Focal infiltration	UA	Liegl et al[2]
7	43/F	Meso-appendix	4.0	Encapsulated	ANED at 10	Tang et al[23]
8	68/M	Cecum	0.4	Focal infiltration	ANED at 24	Liegl et al[2]
9	67/F	Cecum	1.0	Focal infiltration	ANED at 12	Agaimy et al[27]
10	32/F	Ascending colon	1.4	Focal infiltration	UA	Lee et al[28]
11	70/F	Sigmoid colon	0.7	Unencapsulated	UA	Kienemund et al[29]
12	70/F	Sigmoid colon	1.3	UA	UA	Kienemund et al[29]
13	61/M	Sigmoid colon	0.7	Unencapsulated	ANED at 24	Trivedi et al[6]
14	73/F	Rectum	0.85	Unencapsulated	ANED at 36	Liegl et al[2]
(died of metastatic colon carcinoma)						
Subcutaneous and soft tissue						
15	50/F	Right arm	2.0	Encapsulated	ANED at 6	Liegl et al[2]
16	55/M	Right forearm	6.0	Encapsulated	ANED at 2	Chaurasia et al[30]
17	30/F	Upper arm	0.7	Partially Encapsulated	ANED from 26 to 60	Luzar et al[25]
18	55/M	Right upper arm	1.0	Partially Encapsulated	ANED from 26 to 60	Luzar et al[25]
19	56/F	Back	0.5	Encapsulated	UA	Liegl et al[2]
20	11/M	Upper back	8.8	Unencapsulated	ANED at 3	Liegl et al[2]
21	28/M	Back	0.5	Partially Encapsulated	ANED from 26 to 60	Luzar et al[25]
22	26/M	Left Masticator space	6.8	Unencapsulated	ANED at 19	Lau et al[31]
Glands						
23	62/M	Pancreas	5.0	Unencapsulated	UA	Liegl et al[32]
24	41/M	Pancreas	2.5	Unencapsulated	ANED at 7	Shen et al[33]
25	53/M	Left adrenal gland	23.0	Focal infiltration	ANED at 3(then lost to follow-up)	Liegl et al[2]
26	31/F	Adrenal gland	4.0	Encapsulated	ANED at 4	Zhou et al[4]
27	60/M	Adrenal gland	7.0	Unencapsulated	ANED at 4	Xie et al[5]
28	59/F	Parotid gland	2.8	Unencapsulated	UA	Pang et al[34]
29	34/M	Submandibular gland	4.5	Unencapsulated	UA	Lau et al[31]
Others						
30	76/F	Bronchus	3.0	Unencapsulated	Died of postoperative complications	Liegl et al[2]
31	22/F	Frontal lobe	1.8	Unencapsulated	ANED at 36	Pearson et al[35]
32	61/F	Right mandible	5.0	Focal infiltration	UA	Yin et al[36]
33	28/M	Right neck	13.0	Encapsulated	ANED at 5	Gong et al[24]
34	22/F	Palate	4.0	Encapsulated	UA	Guo et al[37]
35	35/M	Cervical spine	3.5	Unencapsulated	UA	Li et al[38]
36	40/M	Lumbar spinal canal	8.5	Encapsulated	ANED at 22	Our case

ANED = alive with no evidence of disease, UA = unavailable.

meso-appendix. Eight cases arose in the subcutaneous and soft tissue, including 4 in the arm, 3 in the back, and 1 in the masticator space. Seven cases arose in various glands, including 3 in the adrenal gland, 2 in the pancreas, 1 in the parotid gland and submandibular gland, respectively. One case each was found in the frontal lobe, right mandible, bronchus, submandibular gland, right neck, palate, and cervical spine. The present report describes the first case of MRS detected in the L spinal canal. The size of MRS ranged from 0.4 to 23.0 cm (averaged 3.8 cm). Seventeen masses were circumscribed but unencapsulated. Eight masses were circumscribed and encapsulated while 3 masses were circumscribed and partially encapsulated. Six masses were circumscribed with focal infiltration. The tumor which measured 8.5*2*0.8 cm in the present case was circumscribed and well-capsulated. The average age of 36 cases was 51 (ranged from 11 to 93). MRS occurred almost equally in female and male, with a female/male ratio of 1.1: 1. And a 40-year-old male was described in the present case. However, predilection for females exists among patients who developed MRS in the digestive tract, with a female/male ratio of 3.7:1. Most of MRS which located at parenchymal organs were detected incidentally during operations for other conditions or during routine imaging examinations.[23] Tumors arising in profundus soft tissue usually presented with enlarged masses.[24] Cutaneous cases presented as painless slightly raised nodules.[25] Besides, individual patients manifested as epigastric pain, indigestion, obstructive sensation during swallowing, or recurrent upper lobe pneumonia.[2,7] Postoperative follow-up which ranged from 2 to 60 months were gain in 24 cases with no recurrences observed. The patient in the present case gained a follow-up of 22 months with no evidence of recurrence.

MRS differs from classic schwannoma in histological morphology.[1,2,5] Spindle-shaped cells with obvious myxoid stroma may lack palisading structures.[2] And there is also a lackage of hyalinizated thick-walled blood vessels.[2] There are 2 distinct morphological components as well as the transition areas. Microcystic appearance is formed when the mucus composition is relatively few, whereas reticular appearance is formed with major mucus composition.[15] Cell atypia can be found but nuclear atypia is rare.[5] In the tumor of present case, mucous network, cystic structure, and hyalinized blood vessels were detected. In addition, cytologic atypia, mitosis or necrosis was absent.

Immunohistochemistry plays an important role in differential diagnosis of MRS (Table 2). The majority of tumors were positive for S-100 protein, strongly and diffusely. Among the 30 cases performing GFAP staining, 23 were positive and 7 were negative. Staining for epithelial markers such as CK pan, AE1/AE3, or Cam 5.2 was performed in 23 cases and was negative in all of them. NF protein was focally positive in 1 case and negative in 10 cases. NSE and CD99 staining was performed in 2 cases, respectively, and was positive in all of these cases. In 12 out of 29 cases, EMA was expressed or focally expressed. CD34 was expressed or focally expressed in 11 out of 23 cases. CD117 was focally

Table 2

Immunohistochemical features of 36 cases.

Case No.	S-100	GFAP	NF	NSE	CD99	CK pan/ AE1/AE3/ Cam 5.2	EMA	SMA	Desmin	CD34	CD117	DOG1	P63	Syn	CgA	CD56	MIB-1
1	(+)D	(−)	/	/	/	(−)	/	(−)	/	(−)	(−)	/	/	(−)	(−)	/	/
2	(+)D,S	(+)P	/	/	/	/	(−)	(−)	(−)	(−)	(−)	/	/	/	/	/	/
3	(+)S	(+)F	(−)	/	/	(−)	(−)	(−)	(−)	(+)F	(−)	(−)	/	/	/	/	/
4	(+)	(+)F	/	/	/	/	/	/	/	(−)	(−)	(−)	/	/	/	(+)F	/
5	(+)D,S	(+)P	(−)	/	/	(−)	(+)F	(−)	(−)	(−)	(−)	/	/	/	/	/	/
6	(+)D,S	(+)P	(+)F	/	/	(−)	(+)F	/	/	/	(−)	/	/	/	/	/	/
7	(+)D,S	(+)	/	/	/	(−)	(−)	(−)	(−)	(+)a few	(+)F	(−)	/	(−)	/	/	<1%
8	(+)D,S	(+)P	(−)	/	/	/	(−)	(−)	/	/	(−)	/	/	/	/	/	/
9	(+)	(+)F	/	/	/	/	/	/	/	(−)	(−)	(−)	/	/	/	(+)F	/
10	(+)S	/	/	/	/	(−)	/	(−)	(−)	/	/	/	/	(−)	(−)	/	/
11	(+)S	/	/	/	/	/	/	(−)	(−)	(−)	(−)	/	/	/	/	/	/
12	(+)S	/	/	/	/	/	/	(−)	(−)	(−)	(−)	/	/	/	/	/	/
13	(+)D	/	/	/	/	(−)	/	(−)	/	(+)F	(−)	/	/	(−)	(−)	(−)	<2%
14	(+)D,S	(+)P	(−)	/	/	(−)	(+)F	(−)	(−)	/	/	/	/	/	/	/	/
15	(+)D,S	(+)P	(−)	/	/	/	(+)	(−)	(−)	/	/	/	/	/	/	/	/
16	(+)D,S	/	/	/	/	(−)	(−)	(−)	(−)	(−)	(−)	/	/	(−)	/	/	/
17	(+)	(+)P	/	/	/	/	(+)F	/	/	(+)F	/	/	/	/	/	/	/
18	(+)	(+)P	/	/	/	/	(+)F	/	/	(+)F	/	/	/	/	/	/	/
19	(+)D,S	(+)P	/	/	/	/	(+)	/	/	/	/	/	/	/	/	/	/
20	(+)D,S	(+)P	/	/	/	(−)	(−)	(−)	(−)	/	/	/	(−)	/	/	/	/
21	(+)	(+)P	/	/	/	/	(+)F	/	/	(+)F	/	/	/	/	/	/	/
22	(+)D,S	(−)	/	/	/	(−)	(+)	(−)	(−)	/	/	/	(−)	/	/	/	/
23	(+)S	(+)F	(−)	/	/	(−)	(+)F	(−)	(−)	/	(−)	/	(−)	/	/	/	<1%
24	(+)	(−)	/	/	/	(−)	(−)	/	/	(−)	(−)	/	(−)	/	(−)	(+)	/
25	(+)D,S	(+)P	(−)	/	/	(−)	(−)	(−)	(−)	(+)F,W	(−)	/	(−)	/	/	/	/
26	(+)D	(+)F	/	/	/	(−)	(+)F	/	/	(−)	/	/	/	/	(−)	(+)D	/
27	(+)	/	/	/	/	(−)	(−)	(−)	(−)	/	/	/	/	/	/	(+)	/
28	(+)D,S	(+)	/	/	/	(−)	(−)	(−)	(−)	(+)	/	/	(−)	/	/	/	/
29	(+)D	(−)	/	/	/	(−)	(+)F	(−)	(−)	(+)D	/	/	(−)	/	/	/	/
30	(+)D,S	(+)P	(−)	/	/	(−)	(−)	(−)	(−)	/	(+)F	/	/	/	/	/	/
31	(+)	(−)	/	/	/	/	(−)	/	/	(+)Pa	/	/	/	/	/	/	/
32	(+)D,S	(−)	(−)	(+)	(+)	(−)	(−)	(−)	(−)	(+)	/	/	(−)	(−)	(−)	/	<1%
33	(+)S	(−)	/	/	/	(−)	/	(−)	(−)	(−)	(−)	/	(−)	(−)	(−)	/	/
34	(+)S	(+)	/	/	/	(−)	(−)	/	/	/	(+)W	/	(−)	/	/	/	/
35	(+)S	(+)S	(−)	(+)S	(+)S	(−)	(−)	(−)	/	/	/	/	/	/	/	/	/
36	(+)D,S	(+)	/	/	/	(−)	(−)	/	/	(−)	/	/	/	(−)	(−)	/	/

n(−) = negative, (+) = positive, / = unavailable, D = diffusely, F = focally, P = partly, Pa = patchily, S = strongly, W = weakly.

expressed in 3 out of 19 cases. CD56 was positive in 5 cases in which 2 were focally positive while 1 was diffusely positive. Synaptophysin (Syn), SMA, Desmin, DOG1, P63, and CgA staining were performed in partial cases and were negative in all of them. Staining for the proliferation marker MIB-1 was performed in 5 cases and showed low nuclear reaction of $<1\%$ in 3 cases, $<2\%$ in 1 case. In the present case, tumor cells showed strong and diffuse nuclear and cytoplasmic positivity for S-100 (Fig. 3A), nuclear positivity for SOX 10 (Fig. 3B), cytoplasmic positivity for GFAP (Fig. 3C) and Vim. Cytokeratins (Fig. 3D), CD31, CD34, EMA, Syn, CgA, and CEA staining were performed with negative results.

Differential diagnosis of MRS which located in the spinal canal includes chordoma, spinal meningioma, neurofibroma and external mucosal chondrosarcoma (EMC).[1,38] Chordoma is a rare malignant primary bone tumor arising from fetal notochord. It usually occurs at the spheno-occipital region of the skull base (approximately 35%) or sacrococcygeal region (approximately 50%).[39] And chordomas account for about 20% of primary spinal tumors. Microscopically, cords and lobules of physaliferous tumor cells are separated by fibrous septa, with extensive myxoid stroma.[40] Cytoplasm of tumor cells is vacuolated and nucleus is prominent vesicular. And mitotic figures are rarely found.[40] Immunohistochemical staining of chordoma is specific. The presence of immunoreactivity for CK pan, EMA, and Vimentin helps in the differential diagnosis from MRS.

Another consideration of differential diagnosis is spinal meningioma. Meningiomas occurred below the C2 vertebral level is named spinal meningiomas. These tumors represent a minority of all meningiomas (approximately 12%) but are the second most common intradural extramedullary spinal tumors which account for 25% of all spinal neoplasms.[26] Spinal meningiomas are associated with dura mater and rarely enlarge vertebral foramen.[41] Histologically, cytoplasm of tumor cells are eosinophili, and nucleus is round and uniform, with commonly found intranuclear pseudoinclusions.[42] Besides, psammoma bodies are frequently detected. Immunohistochemically, it was recognized that spinal meningiomas only show infrequent and patchy positivity for S-100, whereas MRS mostly show strong and diffuse positivity. Furthermore, cytoplasmic EMA immunoreactivity is also useful to separate these entities.[43]

Neurofibromas are composed of mixed proliferation of all components of peripheral nerves, including axons, Schwann cells, perineers, and fibroblasts.[38] Schwann cells often play as the major cellular components, and mesenchymal myxoid degeneration can be significant. In the present case, stroma of the tumor showed abundant mucus and therefore histologically mimic neurofibromas with myxoid degeneration. However, lack of capsule, uniform arrangement of cellular components, absence of typical Antoni A and Antoni B regions and mast cell infiltration in stroma are useful to distinguish these entities.[44] In addition, neurofibromas show positivity for NSE and NF as well as negativity for EMA and D2-40.

EMC was first recognized in 1953 and was defined as a distinct clinicopathological entity 19 years later. EMC mostly arises at intramuscular or deep subfascial locations of the extremities with multinodular growth pattern.[45] The lacelike and cordlike structure in EMC may cause confusion with the more reticular growth pattern in MRS.[1] Nevertheless, the tumor cells of EMC are larger with more obvious eosinophilic cytoplasm. Besides, microcystic changes are generally absent. Furthermore, only 20% of EMC show sporadic S-100 positivity whereas MRS show diffusely S-100 positivity in nearly 100% of tumor cells.[46]

Complete surgical resection is considered as the preferred treatment for MRS according to the previous reports.[1–6,8–24] Tumors which arose in the gastrointestinal tract were thoroughly resected via thoracoscopy, laparoscopy or colonoscopy.[2,4,9] MRS which affected the glands or subcutaneous tissue were totally excised through open surgery.[6,8] Tumor that was located in the frontal lobe was completely removed through microsurgery.[13] Unfortunately, a specific patient received palliative chemotherapy followed by a Whipple surgery since misdiagnosis as malignant tumor which was based on preoperative fine needle aspiration.[7] A total of 25 patients gained follow-up of 2 to 60 months, respectively, with no recurrences or metastasis reported. The favorable prognosis indicated a similar biological behavior as conventional schwannomas. Therefore, adjuvant therapy for MRS was hardly needed.

In summary, the authors describe the first case of MRS which arose in the L spinal canal. MRS is a unique variant of schwannoma with benign biological behaviour. Raising awareness of MRS is essential for both pathologists as well as surgeons to prevent diagnostic pitfalls and make correct treatment strategies.

Author contributions

Conceptualization: Jingcheng Wang, Youwen Deng.
Data curation: Congcong Liu, Jing Li.
Formal analysis: Qing Liu.
Investigation: Lianqi Yan.
Methodology: Congcong Liu, Hongtao Jin, Jingcheng Wang.
Project administration: Jingcheng Wang.
Resources: Qing Liu, Hongtao Jin.
Software: Qing Liu.
Supervision: Youwen Deng.
Validation: Lianqi Yan.
Visualization: Lianqi Yan, Jingcheng Wang.
Writing – original draft: Congcong Liu.
Writing – review & editing: Congcong Liu, Youwen Deng.

References

[1] Scheithauer BW, Woodruff JM, Erlandson RA. Atlas of tumor pathology:tumors of the peripheral nervous system. Washington. DC: AFIP 1999.
[2] Liegl B, Bennett MW, Fletcher CD. Microcystic/reticular schwannoma: a distinct variant with predilection for visceral locations. Am J Surg Pathol 2008;32:1080–7.
[3] Chetty R. Reticular and microcystic schwannoma: a distinctive tumor of the gastrointestinal tract. Ann Diagn Pathol 2011;15:198–201.
[4] Zhou J, Zhang D, Wang G, et al. Primary adrenal microcystic/reticular schwannoma: clinicopathological and immunohistochemical studies of an extremely rare case. Int J Clin Exp Pathol 2015;8:5808–11.
[5] Xie J, Wang H, Deng N. Arenal microcystic/reticular schwannoma: a case report. J Diag Pathol 2016;23:626–8.
[6] Trivedi A, Ligato S. Microcystic/reticular schwannoma of the proximal sigmoid colon: case report with review of literature. Arch Pathol Lab Med 2013;137:284–8.
[7] Gu MJ, Choi JH. Microcystic/reticular schwannoma of the esophagus: the first case report and a diagnostic pitfall. BMC Gastroenterol 2014;14:193–6.
[8] Woodruff JM, Godwin TA, Erlandson RA, et al. Cellular schwannoma: a variety of schwannoma sometimes mistaken for a malignant tumor. Am J Surg Pathol 1981;5:733–44.
[9] Fletcher CD, Davies SE, McKee PH. Cellular schwannoma: a distinct pseudosarcomatous entity. Histopathology 1987;11:21–35.
[10] Dahl I. Ancient neurilemmoma (schwannoma). Acta Pathol Microbiol Scand A 1977;85:812–8.
[11] Killeen RM, Davy CL, Bauserman SC. Melanocytic schwannoma. Cancer 1988;62:174–83.

[12] Kao GF, Laskin WB, Olsen TG. Solitary cutaneous plexiform neurilemmoma (schwannoma): a clinicopathologic, immunohistochemical, and ultrastructural study of 11 cases. Mod Pathol 1989;2:20–6.
[13] Kindblom LG, Meis-Kindblom JM, Havel G, et al. Benign epithelioid schwannoma. Am J Surg Pathol 1998;22:762–70.
[14] Laskin WB, Fetsch JF, Lasota J, et al. Benign epithelioid peripheral nerve sheath tumors of the soft tissues: clinicopathologic spectrum of 33 cases. Am J Surg Pathol 2005;29:39–51.
[15] Woodruff JM, Christensen WN. Glandular peripheral nerve sheath tumors. Cancer 1993;72:3618–28.
[16] Font RL, Truong LD. Melanotic schwannoma of soft tissues. Electron-microscopic observations and review of literature. Am J Surg Pathol 1984;8:129–38.
[17] Michal M, Kazakov DV, Belousova I, et al. A benign neoplasm with histopathological features of both schwannoma and retiform perineurioma (benign schwannoma-perineurioma): a report of six cases of a distinctive soft tissue tumor with a predilection for the fingers. Virchows Arch 2004;445:347–53.
[18] Feany MB, Anthony DC, Fletcher CD. Nerve sheath tumours with hybrid features of neurofibroma and schwannoma: a conceptual challenge. Histopathology 1998;32:405–10.
[19] Weiss SW, Langloss JM, Enzinger FM. Value of S-100 protein in the diagnosis of soft tissue tumors with particular reference to benign and malignant schwann cell tumors. Lab Invest 1983;49:299–308.
[20] Wippold FJ2nd, Lubner M, Perrin RJ, et al. Neuropathology for the neuroradiologist: Antoni A and Antoni B tissue patterns. AJNR Am J Neuroradiol 2007;28:1633–8.
[21] Kurtkaya-Yapicier O, Scheithauer B, Woodruff JM. The pathobiologic spectrum of schwannomas. Histol Histopathol 2003;18:925–34.
[22] Nakamura Y, Becker LE, Marks A. Distribution of immunoreactive S-100 protein in pediatric brain tumors. J Neuropathol Exp Neurol 1983;42:136–45.
[23] Tang SX, Sun YH, Zhou XR, et al. Bowel mesentery (meso-appendix) microcystic/reticular schwannoma: case report and literature review. World J Gastroenterol 2014;20:1371–6.
[24] Gong S, Hafez-Khayyata S, Xin W. Microcystic/reticular schwannoma: morphological features causing diagnostic dilemma on fine-needle aspiration cytology. Am J Case Rep 2014;15:538–42.
[25] Luzar B, Tanaka M, Schneider J, et al. Cutaneous microcystic/reticular schwannoma: a poorly recognized entity. J Cutan Pathol 2016;43: 93–100.
[26] Solero CL, Fornari M, Giombini S, et al. Spinal meningiomas: review of 174 operated cases. Neurosurgery 1989;25:153–60.
[27] Agaimy A, Markl B, Kitz J, et al. Peripheral nerve sheath tumors of the gastrointestinal tract: a multicenter study of 58 patients including NF1-associated gastric schwannoma and unusual morphologic variants. Virchows Arch 2010;456:411–22.
[28] Lee SM, Goldblum J, Kim KM. Microcystic/reticular schwannoma in the colon. Pathology 2009;41:595–6.
[29] Kienemund J, Liegl B, Siebert F, et al. Microcystic reticular schwannoma of the colon. Endoscopy 2010;42(suppl 2):E247.
[30] Chaurasia JK, Afroz N, Sahoo B, et al. Reticular schwannoma mimicking myxoid sarcoma. BMJ Case Rep 2014;2014.
[31] Lau RP, Melamed J, Yee-Chang M, et al. Microcystic/reticular schwannoma arising in the submandibular gland: a rare benign entity that mimics more common salivary gland carcinomas. Head Neck Pathol 2016;10:374–8.
[32] Liegl B, Bodo K, Martin D, et al. Microcystic/reticular schwannoma of the pancreas: a potential diagnostic pitfall. Pathol Int 2011;61:88–92.
[33] Shen Q, Wang YF, Yu B. Clinicopathologic features of microcystic /reticular schwannoma in pancreas. Chin J Diagn Pathol 2014;21: 689–92.
[34] Pang JM, Mahar A, Shannon K, et al. Reticular and microcystic schwannoma of the parotid gland. Pathology 2013;45:96–8.
[35] Pearson L, Akture E, Wonderlick J, et al. Microcystic/reticular schwannoma of the frontal lobe: an unusual occurrence. Case Rep Pathol 2017;2017:1–5.
[36] Yin Y, Wang T, Cai YP, et al. Microcystic/reticular schwannoma of the mandible first case report and review of the literature. Medicine 2015;94:1–5.
[37] Guo JZ, Zhang XW, Yang SJ. Microcystic/reticular schwannoma of hard palate mimicking salivary grand tumor: report of a case. Zhonghua bing li xue za zhi Chin J Pathol 2017;46:431–2.
[38] Li BZ, Wang JW, Wei HQ. Microcystic/reticular schwannoma occurring in cervical spine: report of a case with literature review. Zhonghua bing li xue za zhi Chin J Pathol 2010;39:396–9.
[39] Gagliardi F, Boari N, Riva P, et al. Current therapeutic options and novel molecular markers in skull base chordomas. Neurosurg Rev 2012;35: 1–3.
[40] Barth TFE, von Witzleben A, Moller P, et al. [otochordal tumors: benign notochordal tumors and chordomas. Der Pathologe 2018;39:117–24.
[41] Bydon M, Gokaslan ZL. Spinal meningioma resection. World Neurosurg 2015;83:1032–3.
[42] Yamamuro K, Seichi A, Kimura A, et al. Histological investigation of resected dura mater attached to spinal meningioma. Spine 2012;37: E1398–401.
[43] Baxter DS, Orrego A, Rosenfeld JV, et al. An audit of immunohistochemical marker patterns in meningioma. J Clin Neurosci Off J Neurosurg Soc Australas 2014;21:421–6.
[44] Min H, Kim K. Differential diagnosis between nasal septal schwannoma and nasal septal neurofibroma. J Craniofac Surg 2017;28:1780–3.
[45] Meis-Kindblom JM, Bergh P, Gunterberg B, et al. Extraskeletal myxoid chondrosarcoma: a reappraisal of its morphologic spectrum and prognostic factors based on 117 cases. Am J Surg Pathol 1999;23: 636–50.
[46] Oliveira AM, Sebo TJ, McGrory JE, et al. Extraskeletal myxoid chondrosarcoma: a clinicopathologic, immunohistochemical, and ploidy analysis of 23 cases. Mod Pathol 2000;13:900–8.