Lumbar intraspinal microcystic/reticular schwannoma

Case report and literature review

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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. 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). 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.

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

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

| 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 | 1.0 | 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 | Mesentery | 4.0 | Encapsulated | ANED at 10 | Tang et al\(^{[2]}\) |
| 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\(^{[30]}\) |
| 11 | 70/F | Sigmod colon | 0.7 | UA | Unencapsulated | Kienemund et al\(^{[29]}\) |
| 12 | 79/F | Sigmod colon | 0.3 | UA | Unencapsulated | Kienemund et al\(^{[29]}\) |
| 13 | 61/M | Sigmod colon | 0.7 | Unencapsulated | ANED at 24 | Trivedi et al\(^{[4]}\) |
| 14 | 73/F | Rectum | 0.85 | Unencapsulated | ANED at 36 | Liegl et al\(^{[2]}\) |

(died of metastatic colon carcinoma)

Subcutaneous and soft tissue

| Case No. | Age(yr)/ Sex | Site | Size(cm) | Growth pattern | Final follow-up status | References |
|----------|--------------|------|----------|----------------|------------------------|------------|
| 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\(^{[10]}\) |
| 17 | 30/F | Upper arm | 0.7 | Partially Encapsulated | ANED from 26 to 60 | Luzar et al\(^{[29]}\) |
| 18 | 55/M | Right upper arm | 1.0 | Partially Encapsulated | ANED from 26 to 60 | Luzar et al\(^{[29]}\) |
| 19 | 56/F | Back | 0.5 | Encapsulated | ANED at 60 | 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\(^{[29]}\) |
| 22 | 26/M | Left Masticator space | 6.6 | Unencapsulated | ANED at 19 | Lau et al\(^{[31]}\) |
| Glands | | | | | | |
| 23 | 62/M | Pancreas | 5.0 | Unencapsulated | UA | Liegl et al\(^{[2]}\) |
| 24 | 41/M | Pancreas | 2.5 | Unencapsulated | ANED at 7 | Shen et al\(^{[19]}\) |
| 25 | 53/M | Left adrenal gland | 23.0 | Focal infiltration | ANED at 3(than lost to follow-up) | Liegl et al\(^{[2]}\) |
| 26 | 31/F | Adrenal gland | 4.0 | Encapsulated | ANED at 4 | Zhou et al\(^{[9]}\) |
| 27 | 60/M | Adrenal gland | 7.0 | Unencapsulated | ANED at 4 | Xie et al\(^{[9]}\) |
| 28 | 59/F | Parotid gland | 2.8 | Unencapsulated | UA | Pang et al\(^{[4]}\) |
| 29 | 34/M | Submandibular gland | 4.5 | Unencapsulated | UA | Lau et al\(^{[31]}\) |

Others

| Case No. | Age(yr)/ Sex | Site | Size(cm) | Growth pattern | Final follow-up status | References |
|----------|--------------|------|----------|----------------|------------------------|------------|
| 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\(^{[14]}\) |
| 32 | 61/F | Right mandible | 5.0 | Focal infiltration | UA | Yiu et al\(^{[4]}\) |
| 33 | 28/M | Right neck | 13.0 | Encapsulated | ANED at 5 | Gomi et al\(^{[14]}\) |
| 34 | 22/F | Palate | 4.0 | Encapsulated | UA | Gu et al\(^{[2]}\) |
| 35 | 35/M | Cervical spine | 3.5 | Unencapsulated | UA | Li et al\(^{[31]}\) |
| 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 × 0.8 cm in the present case was circumscribed and well-encapsulated. 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.2,21

Tumors arising in profundus soft tissue usually presented with enlarged masses.2,24 Cutaneous cases presented as painless slightly raised nodules.2,25 Besides, individual patients manifested as epigastric pain, indigestion, obstructive sensation during swallowing, or recurrent upper lobe pneumonia.2,27 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.1,2,5,21 Spindle-shaped cells with obvious myxoid stroma may lack palisading structures.2,21 And there is also a lackage of hyalinized 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.1,15 Cell atypia can be found but nuclear atypia is rare.1,21 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 expressed.

Table 2

| 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 |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 4        | (+)   | (+)F |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 5        | (+)D,S| (+)P |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 6        | (+)D,S| (+)P |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 7        | (+)D,S| (+)P |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 8        | (+)D,S| (+)P |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 9        | (+)F  | (+)P |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 10       | (+)   | (+)F |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 11       | (+)   | (+)F |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 12       | (+)F  | (+)P |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 13       | (+)F  | (+)P |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 14       | (+)D,S| (+)P |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 15       | (+)D,S| (+)P |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 16       | (+)D  | (+)F |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 17       | (+)F  | (+)P |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 18       | (+)F  | (+)P |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 19       | (+)D,S| (+)P |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 20       | (+)D,S| (+)P |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 21       | (+)D  | (+)F |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 22       | (+)D  | (+)F |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 23       | (+)S  | (+)F |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 24       | (+)S  | (+)F |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 25       | (+)D  | (+)F |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 26       | (+)D  | (+)F |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 27       | (+)   | (+)F |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 28       | (+)D,S| (+)P |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 29       | (+)F  | (+)P |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 30       | (+)F  | (+)P |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 31       | (+)   | (+)F |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 32       | (+)D,S| (+)P |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 33       | (+)   | (+)F |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 34       | (+)   | (+)F |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 35       | (+)S  | (+)S |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |
| 36       | (+)D,S| (+)P |    |     |      |                         |     |     |        |     |       |      |    |     |     |      |     |

\(\text{+}\text{=}\text{positive, }\text{-}\text{=}\text{negative, }\text{=}\text{availability, }D\text{=}\text{diffusely, }F\text{=}\text{focally, }P\text{=}\text{partly, }Pa\text{=}\text{patchily, }S\text{=}\text{strongly, }W\text{=}\text{weakly.}\)
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 MB-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 SOX10 (Fig. 3B), cytoplasmic positivity for GFAP (Fig. 3C) and Vim. Cytokeratin (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 chordomal is specific. The presence of immunoactivity 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 eosinophilic, 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.[14] 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.[43] The lacelike and cordlike structure in EMC may cause confusion with the more reticular growth pattern in MRS.[11] 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.[16]

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 thoracoscopic, 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 misdiagnosed as malignant tumor which was based on preoperative fine needle aspiration.[17] 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.

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