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# A Long, Solitary, Rosary-Shaped Spinal Neurofibroma

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**Study Design:** Case report.

**Objectives:** We report the case of a long, solitary, rosary-shaped neurofibroma that was misdiagnosed as another disease due to the patient's surgical history involving repetitive procedures and its abnormal appearance.

**Summary of Literature Review:** Neurofibroma is an intradural-extramedullary spinal tumor. It is generally not difficult to diagnose due to its frequent occurrence and specific magnetic resonance imaging (MRI) findings. However, to date, neurofibromatosis stigmata and long, solitary, rosary-shaped neurofibromas have rarely been reported.

**Materials and Methods:** A 60-year-old woman was admitted to our hospital due to persistent pain, despite previous surgery and repetitive procedures. On physical examination, vision loss, hearing loss, skin discoloration, or subcutaneous nodules were not observed. A neurologic examination revealed normal motor and sensory function and voiding sensation. No pathologic reflexes such as the Babinski sign were observed. Previous sequential MRIs revealed intradural lesions that progressed from the thoracic vertebra 11 to the lumbar vertebra 3. She had no signs of neurofibromatosis stigmata, and the neurologic examination was unremarkable. The initial diagnosis was based on serial MRIs, which revealed a parasite infestation, a spinal cord tumor (myxopapillary-type ependymoma with hemorrhage), arachnoiditis, and vascular malformations. Total mass excision was performed, and the final diagnosis was neurofibroma.

**Results:** There were no signs of a tumor remnants or local recurrence in a 1-year follow-up MRI study.

**Conclusions:** Although intradural spinal tumors are very rare, their clinical features are nonspecific and resemble other degenerative spinal diseases, including spinal stenosis and disc herniation. These diseases may easily be overlooked by physicians.

**Key words:** Neurofibroma, Spinal cord neoplasms, Parasites, Diagnostic errors

Spinal neurofibromas are uncommon, accounting for 3% of all spinal cord tumors.<sup>1)</sup> They may occur as either sporadic lesions or multiple lesions. A diagnosis of the latter is relatively easy due to its association with neurofibromatosis.<sup>2)</sup> However, a solitary lesion is more difficult to differentiate from other solitary spinal cord tumors, especially schwannoma.<sup>3)</sup> Although a biopsy is necessary to make the final diagnosis of spinal tumor, early diagnosis of spinal tumor is not difficult due to the affected area having characteristic clinical features on magnetic resonance imaging (MRI). To date, there has only been one case describing spinal neurofibroma on a long segment in the literature;<sup>4)</sup> however, in that report the diagnosis was not difficult because it has characteristic features of nerve sheath tumor (i.e., adjacent bone remodeling and dumbbell-shape). Herein, we report a case of solitary neurofibroma that had

initially been suspected as parasite infestation, myxopapillary ependymoma hemorrhage, arachnoiditis, and vascular malformations, due to its long and grotesque shape.

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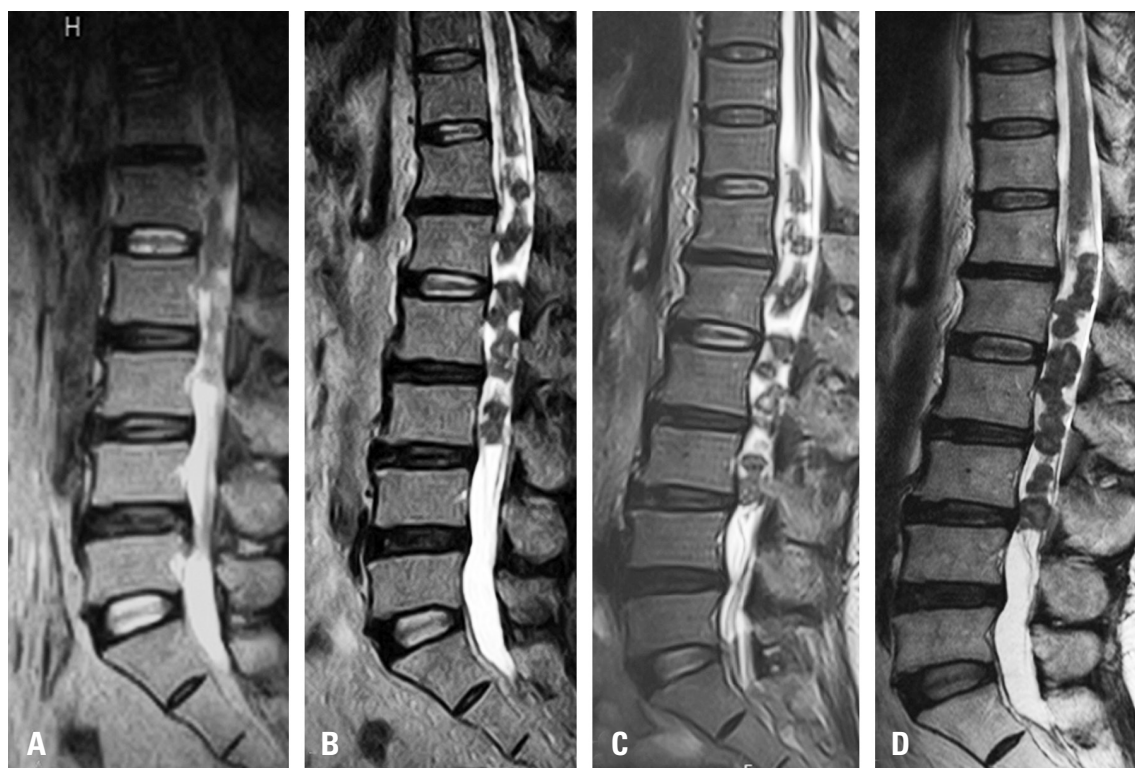
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## Case Report

A 60-year-old woman with a history of several years of lower back and bilateral lower extremities radiating pain presented with one month worsening pain, radiating to the posterior thigh, and posterior calf. She had no history of internal medicine problems. However she had received complicated therapies for her spinal pain including a discectomy of L4–5 level, a neuroplasty, and several times of epidural steroid injections. She had some improvement in the symptom, but it didn't clear. She lived in the countryside and often drank a natural mineral water from the mountains. She is a rural farmer who had occasionally been consuming raw meat and fish. On admission, her vital signs were stable and she complained only lower back and bilateral leg pain. She had checked several times MRIs from the other hospitals. At there, a mass-like lesion was detected from the 12th thoracic to the mid-body of the 3rd lumbar spine in both MRI examinations. Neurologic examination revealed normal motor and sensory function of lower extremity. Bilateral knee

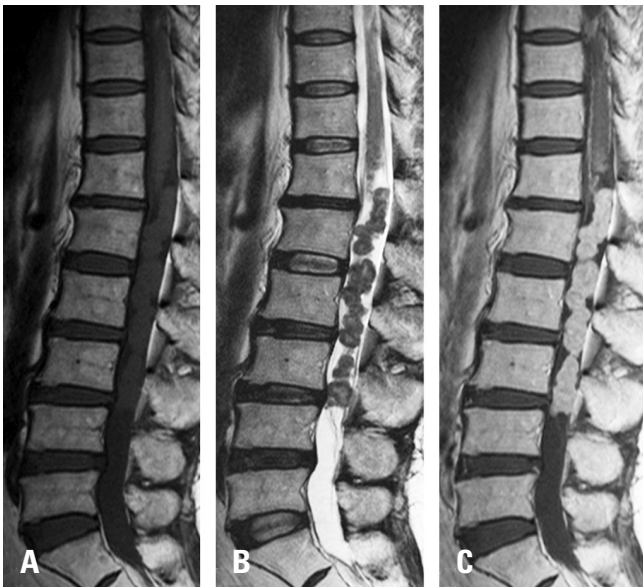
jerk and ankle jerk were +2/+2. No pathologic reflexes such as Babinski sign and ankle clonus were observed. The voiding sense and anal sphincter tone were intact. She had no signs of neurofibromatosis, such as café au lait spots, axillary or inguinal freckling, plexiform neurofibroma or neurofibromatosis, Lisch nodules, optic glioma, osseous change, or diagnosis of NF1 in a first-degree relative.<sup>1)</sup> The intraspinal lesion observed on previous sequential MRIs had grown or moved caudally (Fig. 1). Post-myelographic computed tomography demonstrated that the lesion had multiple, continuous, and well-defined boundary filling defects in the dural sac, from the T11 level to the L3–4 levels (Fig. 2). The findings prompted suspicion of intraspinal parasite. However, serum IgE and enzyme-linked immunosorbent assay (ELISA) for parasite antibodies were negative. Thereafter, an enhanced MRI revealed multiple, nodular and elongated well-defined heterogenous enhancing lesions in the dural sac (Fig. 3), consistent with a spinal cord tumor, especially myopapillary type ependymoma with hemorrhage, arachnoiditis, and vascular malformation. The patient underwent laminotomy and midline durotomy



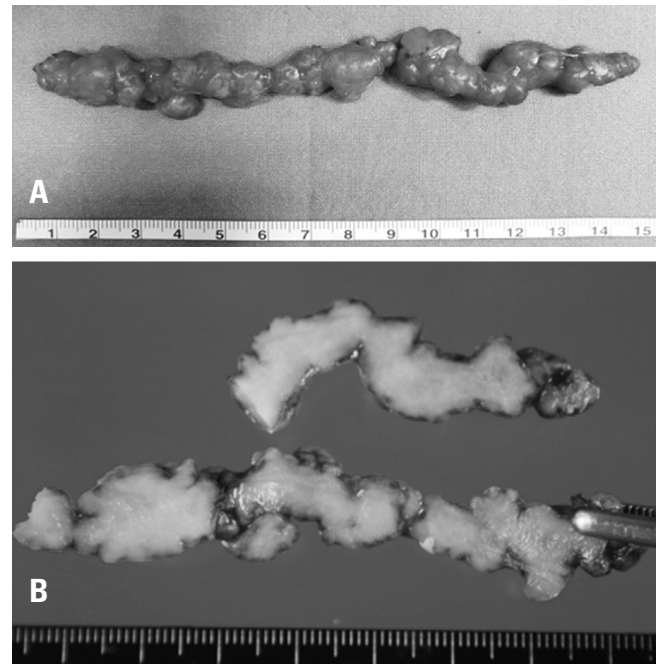
**Fig. 1.** Serial T2-weighted sagittal magnetic resonance imaging revealed that the mass-like lesion grew or moved caudally. (A-B) 27 months, (B-C) 12 months, (C-D) 5 months.



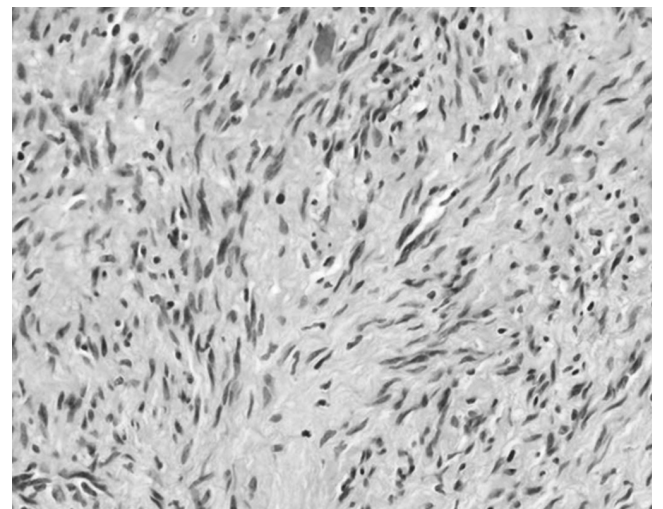
**Fig. 2.** A post-myelographic computed tomography scan revealed multiple filling defects in the dural sac, from the T11 to L3-4 disc levels. **(A, B)** Coronal and sagittal images show a tortuous lesion in the spinal canal with an elongated shape.



**Fig. 3.** Preoperative sagittal magnetic resonance imaging demonstrated that the mass-like lesion extended from the T11 level to the L3-4 disc levels. It was shaped like a rosary or pea in the dural sac. **(A)** Isointensity on T1; **(B)** mixed or slightly low intensity on T2; **(C)** a heterogeneous well-enhanced image post-contrast.



**Fig. 4.** Photographic findings. The mass had a serpentine shape with firm consistency. It measured 1.0×1.5×14 cm and weighed 10 g. **(A)** Gross surface, **(B)** longitudinal cross-section.



**Fig. 5.** Histological findings. A high-power section revealed a proliferation of spindle cells with wavy nuclei, consistent with a neurofibroma (hematoxylin and eosin staining, ×400 magnification).





**Fig. 6.** Postoperative 1-year follow-up magnetic resonance imaging. There was no evidence of tumor remnants or local recurrence of the tumor.

from T11 to L3. The mass-like lesion occupied most of the intradural space, displacing the spinal cord and cauda equina to the right. The elongated mass was multifocal fusiform or dumbbell shaped and the consistency was rubbery-hard. It had well encapsulated and clear boundary. Two feeder vessels were cauterized. Several nerve fascicles traversed a long coiled mass, and the mass was cut and removed to reduce nerve damage. Total mass excision was achieved, and the mass was measured to be 1.0×1.5×14 cm in size and 10 gm in weight (Fig. 4). Both ELISA for parasites and pathology for malignant cell in the cerebrospinal fluid were negative. Histological findings using Hematoxylin and eosin stain (Fig. 5), specimen are constructed with proliferation of spindle cells with wavy nuclei, and it has intra-tumoral infiltration of lymphocyte, and there are no signs of neurogenic sarcoma. Using S-100 stain the result shows possibility of cellular neurofibroma. Post-operatively, back pain and radiating pain were greatly improved. There were no signs of remnant or local recurrence of tumor on a 1-year follow-up MRI (Fig. 6).

## Discussion

According to the serial MRI data, the lesion was serpentine or rosary in shape and grew or moved caudally. We initially suspected parasite infestation, spinal cord tumor (especially myxopapillary type ependymoma with hemorrhage), arachnoiditis, or vascular malformations. All of the above diseases occur for a long time and manifest nonspecific symptoms and difficult to differentiate radiologically. Therefore we should effort to get more detailed medical history, family history, diet, physical examination, blood test, more specific radiological examination. Initially, the lesion looked like a parasite infestation morphologically, particularly sparganum. Sparganosis is detected more frequently in eastern Asia than in other areas.<sup>5)</sup> In this region, sparganosis infection in humans occurs by accidental consumption of water contaminated with infected copepods or by ingestion of raw or inadequately cooked snakes or frogs infected with sparganum.<sup>6)</sup> The radiologic findings of spinal sparganosis are hypointensity on post-contrast MRI. However, if it was surrounded by acute inflammation, hyperintensity is possible,<sup>5)</sup> which hinders the distinction from a spinal cord tumor. If viable, it may grow gradually and move slowly.

Preoperative diagnosis of spinal sparganosis, based on radiological findings, may be difficult. It is nonspecific to serum eosinophilia. The presence of anti-sparganum antibody in cerebrospinal fluid or serum, measured by ELISA, is highly sensitive and specific in the diagnosis of sparganosis.<sup>7)</sup> In this case, we initially considered it as sparganosis since the patient is a rural farmer who had occasionally been consuming raw meat and fish. However, sparganosis was excluded due to low eosinophil count, negative ELISA result, and hyperintensity in post-contrast MRI. As another possibility, the lesion looked like a primary spinal tumor, especially neurofibroma and myxopapillary-type ependymoma with hemorrhage. The slow growth over a long period and detection of a heterogenous hyperintense mass-like lesion in post-contrast MRI prompted an inclusion of neurofibroma in the differential diagnosis. However, there was no indication or family history of neurofibroma, and no erosion of adjacent bony structure had occurred during the period of growth. Radiologically, myxopapillary ependymoma with hemorrhage may also exhibit the shape observed. It is a variant type of spinal

ependymoma occurring most commonly in conus medullaris and filum terminale.<sup>3,8)</sup> It may often accompany hemorrhage, calcification, and cyst formation. If hemorrhage was present, a mixed T2 weighted MRI signal is necessary.<sup>3)</sup> The lesion needed to be differentiated from arachnoiditis, since the patient had received a discectomy and several injections in the spinal canal. The radiologic findings of arachnoiditis are lack of normal fanning, abnormal free-fall in the dependent position, and empty sac sign.<sup>9)</sup> However, these indications were absent. Although vascular malformations are rare, it must be identified in these radiologic findings. These are characterized by spinal cord enlargement on the MRI, and spinal angiography is generally useful in the differentiation.<sup>10)</sup>

Histologically, the present case was confirmed as a neurofibroma. A neurofibroma is a common benign tumor of the nerve sheath in company with schwannoma. These two tumors originate from the schwann cells of the nerve. They can be distinguished histologically, but it is difficult to differentiate radiologically. Most of the nerve sheath tumor grows slowly, symptoms happen very slow, rarely accompanied by neurologic abnormalities.

Simple radiographs and computed tomography may be accompanied by an enlargement of the intervertebral foramen, erosion of the pedicle, scalloping of the posterior vertebra, thinning of the lamina. On MRI findings, T1-weighted images show iso-low signal intensity compared to spinal cord, high signal intensity on T2-weighted images, cystic change (40%) and edema of adjacent spinal cord. Both tumors are virtually enhanced by contrast MRI, but heterogeneous enhancement with low signal is more characteristic of a neurofibroma.

Neurofibromas are sub-classified according to their growth patterns—localized, plexiform, and diffuse. This case presented the localized type. Although giant neurofibroma sub-classified as plexiform or diffuse type has been previously reported in many cases, to the best of our knowledge, the localized type has only been reported in one case to date.<sup>4)</sup> However, since erosion of adjacent bony structure occurred, diagnosis was not difficult. In the present case, erosion was absent, which delayed the final diagnosis.

Although parasite infestation, spinal cord tumor, arachnoiditis, and venous malformations are very rare conditions, clinical features were nonspecific and similar to degenerative spinal diseases, including spinal stenosis and

disc herniation. These diseases may be easily overlooked by physicians. In cases with a long duration of symptoms and a complex clinical history, these aforementioned diseases should be considered.

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## 염주알 모양의 긴 고립성 신경섬유종

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**연구 계획:** 증례 보고

**목적:** 수술 및 반복적인 시술의 복잡한 치료병력과 기괴하게 생긴 모습으로 인해 다른 질환으로 오인되었던, 염주알 모양의 긴 고립성 척추 신경섬유종의 증례를 보고하고자 한다.

**선행문헌 요약:** 신경섬유종은 경막내-척수와 종양으로 발생하는 위치와 특징적인 자기공명영상의 모습으로 초기 진단이 어렵지는 않다. 신경섬유종증의 징후가 없고, 장분절에 걸쳐 발생한 염주알 모양의 긴 신경섬유종은 보고가 드물다.

**대상 및 방법:** 지속적인 양하지 방사통으로 수술과 반복적인 시술의 복잡한 치료병력이 있는 60세 여자 환자가 내원하였다. 이학적 검사상 시력장애, 난청, 피부의 색소 침착이나 탈색, 피하 결절 등은 관찰되지 않았고, 신경학적 검사상 하지 감각, 근력, 심부건 반사는 정상이었으며, 배뇨 장애의 소견은 없었고, 족간대성 경련, 바빈스키 징후(Babinski sign) 등의 병적 반사는 관찰되지 않았다. 순차적인 MRI 소견은 흉추 11번에서 요추 3번까지 경막내 염주알 모양의 병변이 진행되는 모습이었다. 초기 감별질환은 기생충 감염, 출혈성 점액유두성 상의세포종, 지주막염, 혈관 기형이었다. 흉추 11번부터 요추3번까지 후궁 절개술 후 경막을 절개하여 종양 제거 수술을 시행하였고, 최종 진단은 신경섬유종으로 확진 되었다.

**결과:** 1년 후 추시 MRI에서 종양의 잔존 또는 재발된 징후는 없었다.

**결론:** 비록 경막내 종양은 매우 드문 질환이지만, 임상적 양상은 비특이적이며 척추관 협착증, 추간판탈출증과 같은 퇴행성 척추질환과 유사하다. 복잡한 치료병력이 있는 환자들에게서 이러한 질병은 쉽게 간과될 수 있다.

**색인 단어:** 신경섬유종, 척추종양, 기생충, 진단 오류

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