CASE REPORT

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RECURRENCE OF SOLITARY FIBROUS TUMOR OF THE CERVICAL SPINAL CORD

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ABSTRACT

Solitary fibrous tumor (SFT) mostly originates from the pleura because of proliferation of fibroblast cells. It is extremely rare for the tumor to originate from the spinal cord. Here, we report a rare case of SFT in the spinal cord that recurred repeatedly and progressed from intramedullary to extramedullary. A 40-year-old man underwent C4-5 intramedullary and extramedullary tumor resection in another hospital. Eighteen years later, he experienced symptoms of myelopathy because of tumor recurrence; therefore, he consulted with our hospital and underwent tumor resection again. During surgery, we found that the tumor had an intramedullary and extramedullary location. Only partial resection was possible because of intraoperative deterioration in the compound motor action potential (CMAP). After resection, the pathological diagnosis was SFT. The postoperative course was good. However, two years later, a third tumor resection was required because of dysuria and tumor growth. In this surgery, total resection of the tumor was possible without intraoperative deterioration of the CMAP. The tumor has not subsequently recurred. However, SFT recurrence is relatively common and careful follow-up is required for early detection of recurrence, even after successful removal of the tumor.

Key Words: solitary fibrous tumor, intramedullary, extramedullary, cervical spinal cord, recurrence

INTRODUCTION

Spinal solitary fibrous tumor (SFT) is a rare disease that commonly arises from mesenchymal cells, but is often generated from the pleura.[1)] Carneiro reported the first spinal case of this disease in 1996.[2)] Thereafter, 34 cases of this type of spinal tumor have been documented.[2-31)] However, to the best of our knowledge, there has been no report of recurrent regrowth of spinal SFT that required repeated surgeries over a long period of time to treat disorders related to the spinal cord. Here, we present a case of SFT of the cervical spinal cord as a rare clinical entity that recurred after tumor resection and we review the literature on the occurrence and diagnosis of this tumor type.

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CASE REPORT

In January 1987, a 40-year-old male with gait disturbance and numbness in both hands underwent spinal tumor resection at another hospital. The surgery was a C3–6 laminectomy after dural incision. A tumor was found on the dorsal side of the spinal cord and contiguous from the extramedullary to intramedullary regions at the C4–5 level. Adhesion was mild. Subtotal tumor resection was performed and the tumor was benign pathologically. Thereafter, symptoms improved and there was no recurrence for a long period.

In September 2004, the patient presented with numbness in his upper extremities and progressive lower extremity weakness with a complaint of gait disturbance. In the initial consultation with our hospital, spinal magnetic resonance imaging (MRI) revealed recurrence of an intradural 1.9×1.7×1.9-cm nodular lesion at the C4–5 level, which was clearly circumscribed by the spinal cord (Fig. 1). The tumor appeared as a hypointense lesion on T1-weighted images with homogeneous enhancement by gadolinium. Computed tomography displayed no calcification around the tumor. Symptoms worsened and tumor resection was performed at our hospital in January 2005. During surgery, after C7 laminectomy, ultrasonic imaging showed that the tumor was located in the intramedullary and extramedullary regions. After debulking of about two-thirds of the tumor, an intraoperative deterioration in the compound motor action potential (CMAP) prevented further mass reduction of the tumor. After resting for 30 min, the CMAP decrease was reversed. However, complete tumor resection was not performed.

Although the tumor could not be resected completely after surgery (Figs. 2A, 2B), there was improvement of symptoms such as gait disturbance and numbness, which enabled the patient to return to work. There was no deterioration of symptoms for the following two years, but

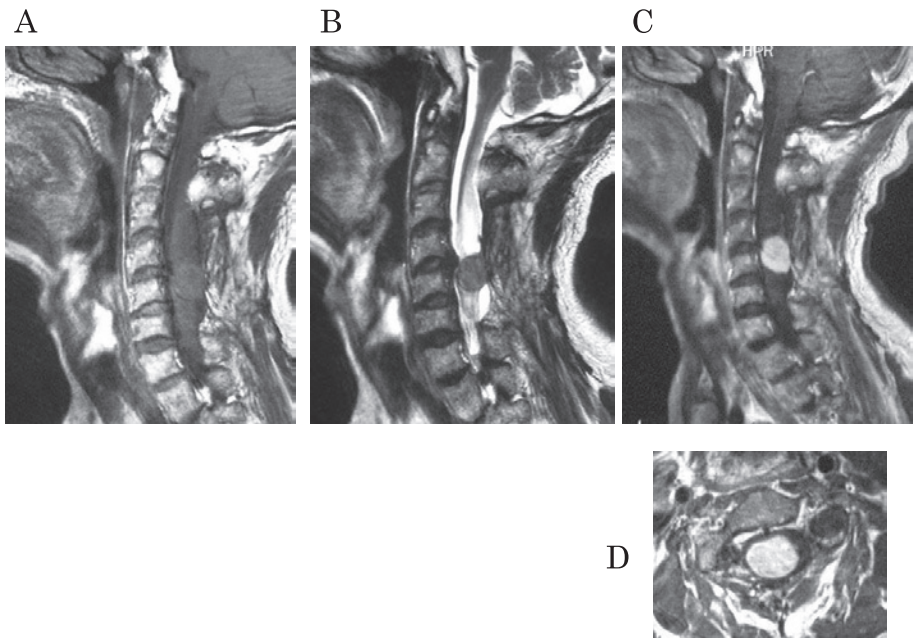

Fig. 1 Magnetic resonance imaging at the time of the initial consultation with our hospital. Sagittal T1-weighted (A) and T2-weighted (B) of the cervical cord gave isointense and iso- to hyperintense signals due to a intradural tumor at the C4–5 level. Sagittal T1 gadolinium-enhanced (C) and axial (D) images showed homogenous enhancement.

in November 2007 the patient experienced further neurological deterioration in the bilateral lower extremities and developed a neurogenic bladder. MRI revealed recurrence of an intradural 2.2×2.1×2.0-cm nodular lesion along the posterior aspect (Figs. 2C, 2D). Recurrence was accompanied by paralysis of the lower extremities and a revision surgery with the goal of total resection of the tumor mass was performed in January 2008. Total resection of the tumor without compromising the CMAP was achieved during this surgery.

Postoperative MRI performed 3 years after the revision surgery confirmed total resection and showed no evidence of tumor recurrence. Lower extremity strength had fully returned and the patient had recovered to his preoperative state (Figs. 3). Histological examination revealed a well circumscribed, partially encapsulated tumor with bland spindle cells with somewhat vesicular nuclei. Immunohistochemical staining was strongly positive for vimentin and CD34, but negative for S-100 and epithelial membrane antigen (EMA). The Ki-67 proliferation index was low, at approximately 1%. These findings are consistent with SFT (Fig. 4).

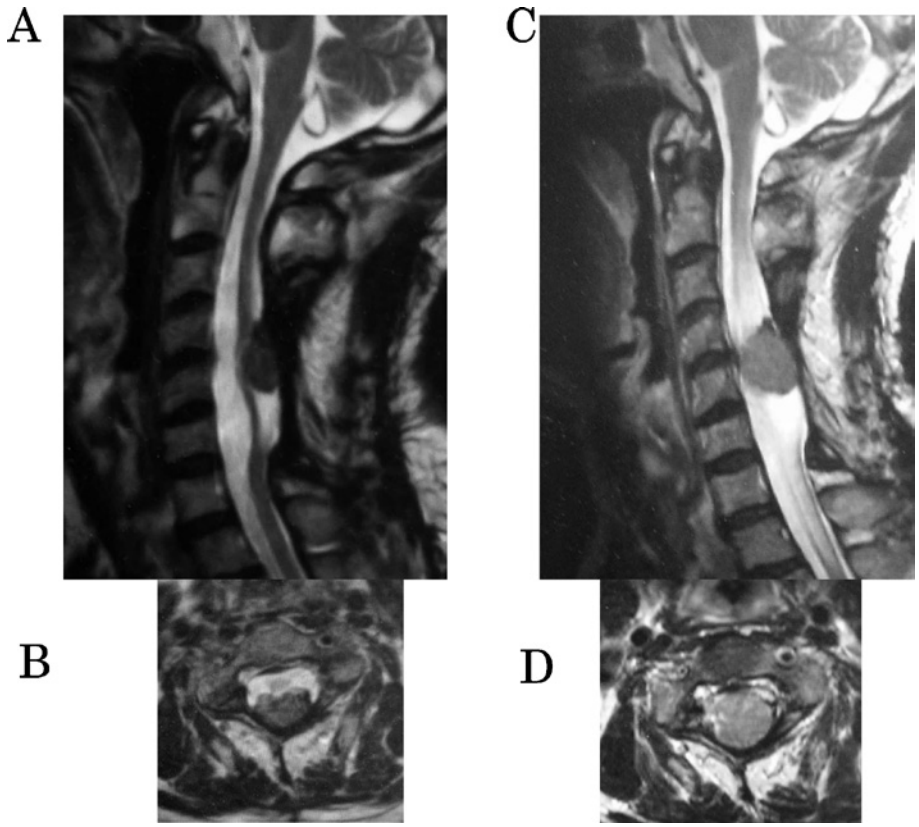

Fig. 2 Sagittal T2-weighted (A) and axial (B) imaging showed recurrence of the tumor six months after surgery at our hospital. Sagittal T2-weighted (C) and axial (D) imaging revealed tumor growth two years after surgery at our hospital.

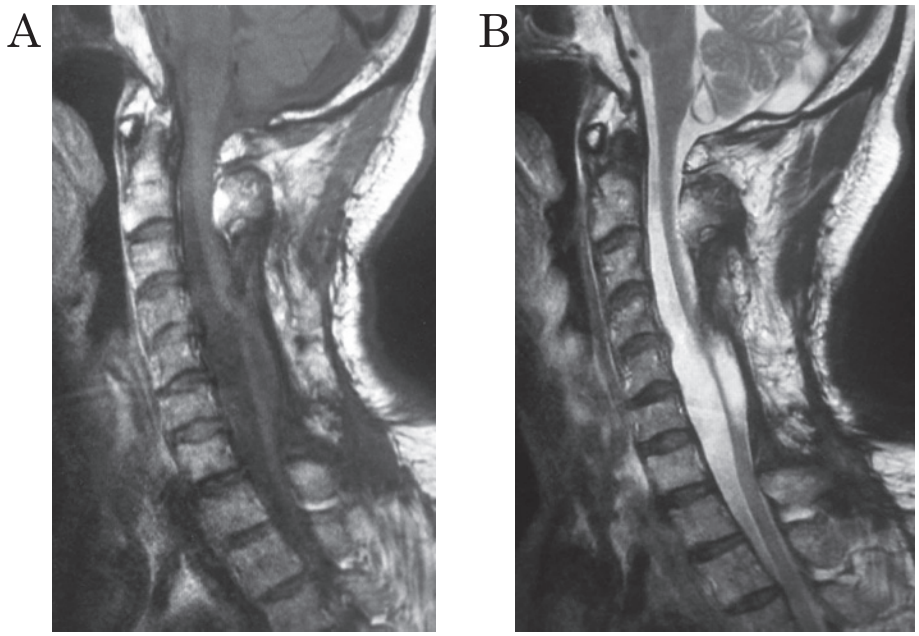

Fig. 3 Postoperative sagittal T1-weighted (A) and T2-weighted (B) images showing the absence of a residual tumor at the present time.

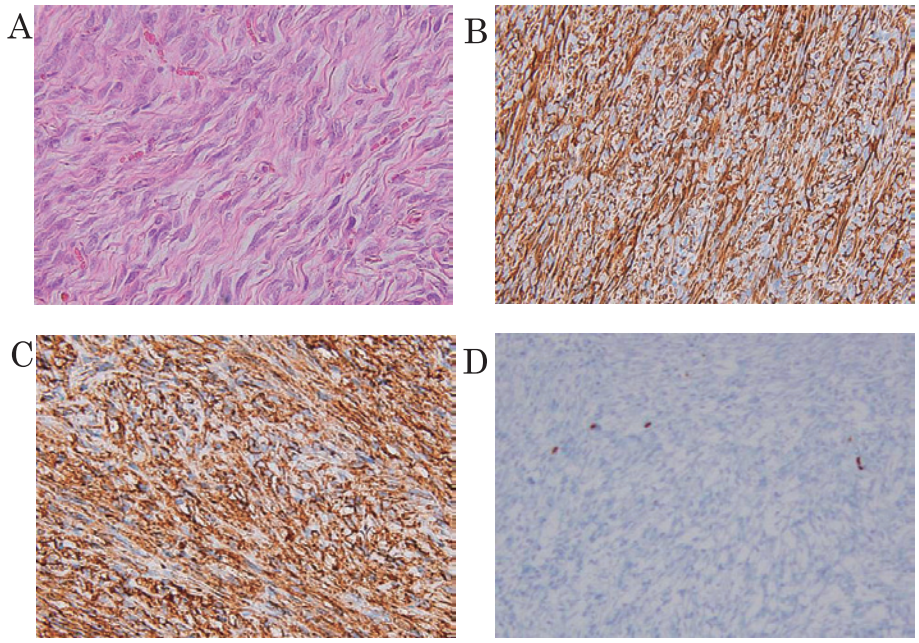

Fig. 4 Microscopic examination of the tumor specimen after hematoxylin and eosin staining showed a well circumscribed, partially encapsulated tumor along with bland spindle cells with somewhat vesicular nuclei (A). In immunocytochemical analysis, the tumor cells exhibited strong positivity for vimentin (B) and CD34 (C). The Ki-67 proliferation index was low (approximately 1%) (D). (A-D:×400).

DISCUSSION

SFT usually originates from mesenchymal cells such as those in the pleura and peritoneum.[1] Of all cases of these tumors, 40% are found in the subcutaneous tissue[2] whereas others are found in deep soft tissues of the extremities or extra-compartmentally in the head and neck region.[2,3,5] Histologically, SFT consists of spindle cells with a mixture of hypocellular and hypercellular areas, and the stroma is rich in collagenous connective tissue with a keloid-like pattern.[19]

Carneiro *et al.* described the first case of spinal SFT in 1996[2] and there are currently 34 cases described in the literature.[2-31] These tumors originated in the meninges, spinal cord parenchyma, nerve root, and dura[19-20,32] and occurred in the intramedullary (64%), intradural-extramedullary (12%), and extradural (24%) areas. The location was mainly in the thoracic region (56.3%), followed by the cervical region (31.2%) and the lumbar region (12.5%).[22] Among intramedullary SFTs, the origin has been reported to be the perivascular connective tissue, pia arachnoid and pia mater.[9,26,33] Most cases may be subpial in origin, similar to intramedullary exophytic tumors, because the pial surface of the spinal cord seems to be involved in the tumor during surgery. The tumor in our case had an intradural-extramedullary location and the origin was unclear. There have been four previous reported cases of intra-extramedullary spinal SFT,[2,5,6,14] all of which occurred in the cervical region, with attachment with the leptomeninges, nerve root, spinal cord film, and unknown in one case each. One of these cases recurred five years after surgery, but none of the cases had recurrence after 20 years, as found in our case.

In MRI, SFTs appear as hypointense or isointense lesions in T2-weighted images because they are rich in collagen fibers, and can be homogeneous or heterogeneous in Gd-enhanced images because they are rich in blood vessels.[3,8,16] These findings are useful for differential diagnosis with meningioma. In addition, reported cases of SFTs have not shown the “dural tail” sign, which is suggestive of meningioma.[9]

Immunostaining for pathological diagnosis in our case was not performed after the initial surgery. Thus, the patient was diagnosed with SFT based on immunostaining performed after the second surgery. Differential diagnoses of intradural tumor include meningioma, malignant peripheral nerve sheath tumors, and hemangiopericytoma (HPC).[22,32] SFT is characterized by positive immunostaining for vimentin and CD34.[26,34] In contrast, most fibrous meningiomas are positive for EMA, malignant peripheral nerve sheath tumors are generally positive for S-100, and HPCs are generally only focally and weakly positive for CD34 antigen.[22,32,34] Therefore, immunostaining is specifically required for SFT diagnosis.

Most cases of SFT are benign, but the behavior is unpredictable. Most histologically benign SFT prove to be non-recurring and non-metastasizing lesions and most histologically malignant tumors behave aggressively.[36-38] However, even a benign SFT may have multiple relapses and metastasis. Muñoz *et al.* reported a case of spinal cord SFT that was primarily diagnosed as benign, but showed hepatic and pulmonary dissemination after primary tumor resection.[22] Because the possibility of recurrence is as high as 20%,[8] SFTs should be treated as a low-grade malignancy.[35-37] In our case, total resection was not possible in the earlier surgery and recurrence was observed and required a reoperation. This illustrates that frequent examination of tumor regrowth is required after partial resection of SFT and early reoperation may be needed for total resection. In addition, even after successful removal of the tumor, careful follow-up is important for early detection of recurrence. Our experience in this case indicates that complete removal after an accurate diagnosis and careful follow-up is desirable for SFTs.

CONFLICT OF INTEREST

None of the authors have a conflict of interest regarding the contents of the manuscript.

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