Solitary Fibrous Tumor of the Thoracic Spinal Cord

—Case Report—

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Abstract

A 63-year-old woman presented with a rare case of primary solitary fibrous tumor (SFT) occurring in the extramedullary thoracic spinal cord. T₁-weighted magnetic resonance (MR) imaging showed the tumor as a mildly hypointense area with homogeneous enhancement by gadolinium. T₂-weighted MR imaging showed a hypointense mass with peritumoral edema. The tumor arose from one of the posterior spinal roots, with no attachment to the dura. The tumor was clearly circumscribed from the surrounding cord tissue and easily removed. Histological examination showed the tumor predominantly consisted of spindle cells separated by abundant collagen matrix fibers. Tumor cells were strongly positive for vimentin and CD34, but negative for glial fibrillary acidic protein, S-100 protein, epithelial membrane antigen, myelin basic protein, and keratin. SFT should be considered in the differential diagnosis of spindle cell central nervous system neoplasms, although SFT is extremely rare in the spinal cord.

Key words: solitary fibrous tumor, spinal cord

Introduction

Solitary fibrous tumor (SFT) was previously classified as a localized form of mesothelioma, which tends to occur in the pleura.^{8,13,14} However, the absence of epithelial markers and mesothelial features support a mesenchymal origin.^{4,13} SFT may occur in many locations throughout the body, not limited to the pleura or serosal surfaces, including the soft tissues, upper respiratory tract, peritoneum, retroperitoneum, and periosteum. Recently, SFT has been found in the central nervous system (CNS).¹⁻¹⁵

We report a case of intradural extramedullary SFT of the thoracic spine.

Case Report

A 63-year-old female was admitted to our hospital because of progressive sensory disturbance of the lower extremities. Her symptoms began 2 years ago as right toe numbness and paresthesia, and gradually extended to the right knee. She noticed numbness

of her left toes and gait disturbance 1 year after the onset. Neurological examination on admission revealed decreased pain, light touch, and vibration sensations below the L-3 level of her right lower extremity. Muscle weakness (grade 4/5) of all muscle groups was also demonstrated in the right lower extremity. Increased deep tendon reflexes were demonstrated in the bilateral knees and ankles.

Magnetic resonance (MR) imaging of the thoracic spine revealed an intraspinal mass measuring about 1 cm in diameter at the T-11 level, which was clearly circumscribed from the surrounding spinal cord. The mass appeared as a mildly hypointense area on the T₁-weighted images with homogeneous enhancement by gadolinium. The mass appeared as a clearly hypointense area accompanied by profound edema spreading longitudinally around the tumor on the T₂-weighted images (Fig. 1). Computed tomography myelography and cerebrospinal fluid study provided no additional information. Spinal angiography showed no tumor stain or feeding arteries.

The patient underwent tumor extirpation with

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Fig. 1 Sagittal (upper row) and axial (lower row) T₁-weighted magnetic resonance (MR) images revealing a round hypointense mass at the T-11 level (left column), with homogeneous enhancement by gadolinium (center column). T₂-weighted MR images showing a hypointense area accompanied by profound edema spreading longitudinally around the tumor (right column).

T8–12 laminoplastic laminectomies. Intradural exploration revealed a milky-white tumor dorsal to the spinal cord. The majority of the tumor tissue was buried in the cord. A definite plane was present between the tumor and the surrounding cord tissue, so total tumor removal was easily achieved. The tumor seemed to originate from a dorsal nerve root, which was sacrificed during the dissection. The histological diagnosis of the frozen specimen was fibrous meningioma.

The patient experienced transient worsening of the muscle weakness of the right lower extremity, but this gradually recovered to the preoperative state in a few months. Follow-up MR imaging taken 18 months after the surgery showed no tumor remnant or relapse.

Histological examination of the specimen revealed bland spindle cell proliferation with no pattern. Although Ki-67 staining was not performed, no mitotic figures were identified (Fig. 2A, B). Immunohistochemical study was performed using

formalin-fixed, paraffin-embedded tumor tissue and EnVision™ + System, HRP (DAKO, Glostrup, Denmark). Tumor cells were positive for CD34 (Fig. 2C) and vimentin (Fig. 2D), but negative for S-100 protein, keratin (cytokeratin AE1 + AE3, mouse monoclonal antibody; DAKO), c-kit, glial fibrillary acidic protein, neurofilament, and epithelial membrane antigens. These findings were considered to be compatible with SFT.

Discussion

The putative origins of CNS SFT include perivascular fibrous tissue or leptomeninges, as there are few sources of fibroblasts in the CNS.¹⁴⁾ SFT may be more common in the spinal compartment and posterior cranial fossa.⁸⁾ Intracranial and intraspinal SFTs seem to show considerable differences. The majority of intracranial SFTs had a dural origin, whereas one third of spinal SFTs had a dural attachment.^{2,8,10,13)} Since the present tumor was continuous with the spinal nerve root and not attached to the dura mater, the origin seemed to be the pial or vascular tissue of the nerve root.

SFT consists of spindle cells in a mixture of hypocellular and hypercellular areas. The stroma is rich in collagenous connective tissue, with a keloid-like pattern. The tumor cells usually proliferate haphazardly and may show storiform, herringbone, nuclear palisade, or hemangiopericytoma patterns.^{8,13,14)} The histological diagnosis primarily requires differentiation of SFT from other spindle cell tumors arising from the meninges, such as the fibroblastic variant of meningioma, as well as schwannoma, meningeal fibroma, myofibroblastoma, and hemangiopericytoma.^{1,8,13)}

Immunohistochemical staining shows the tumor cells are positive for vimentin and CD34, and negative for any epithelial marker, S-100 protein, alpha-smooth muscle actin, and desmin.^{1,8,13,14)} Meningioma shows a characteristic whorl formation, intranuclear cytoplasmic inclusions, and is positive for epithelial membrane antigen. Schwannoma includes bundles of tightly packed spindle cells, and is positive for S-100 protein. Histological and immunohistochemical examination of the present case was consistent with SFT and did not support the diagnosis of either meningioma or schwannoma.

The neuroimaging features of spinal SFTs including this case resemble those of meningioma.^{2,8,13,14)} Spinal SFT occurs as three types: intradural and dura-based, intradural extramedullary, and intramedullary tumors. MR imaging of this case was basically identical to that of previous cases. T₁-

Fig. 2 Photomicrographs of the specimen showing spindle cell proliferation with mild pleomorphism and dense intracellular collagen deposition (A: HE stain, $\times 100$), spindle cells demonstrating absence of pattern (B: HE stain, $\times 400$), and immunoperoxidase staining revealing strong, diffuse positive staining of tumor cells for CD34 (C: $\times 400$), and positive staining for vimentin (D: $\times 400$).

weighted MR imaging shows SFT as an isointense or mildly hypointense area with homogeneous enhancement by contrast medium. T_2 -weighted imaging shows SFT as a hypointense area. Reported cases of SFT have not shown the “dural tail” sign, which is suggestive of meningioma.¹⁴⁾

Early experience of reviewing process suggests that most extrathoracic SFTs tend to follow a benign clinical course. In addition, SFT in any location tends to have a clear cleavage plane, which allows gross total resection.^{1,14)} However, recurrence and metastasis may take place after apparently total removal of CNS SFTs.^{2,8)} Spinal SFTs including this case should be followed carefully even after successful removal.

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