Solitary fibrous tumor of filum terminale

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Abstract
Solitary fibrous tumor (SFT) is a mesenchymal neoplasm that most commonly arises from the visceral or parietal pleura. Solitary fibrous tumor with a primary site in the filum terminale has not been reported previously in the literature. Here we report a case of SFT occurring in the filum terminale. The characteristic imaging feature of this tumor is hypointensity on T2-weighted images. Even though rare, SFT should be considered in the differential diagnosis of tumors occurring intraspinally, even in the filum terminale.

Keywords: Solitary fibrous tumor, spinal cord, MR imaging, CNS, adults, neoplasms primary

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Solitary fibrous tumor (SFT) is a mesenchymal neoplasm that most commonly arises from the visceral or parietal pleura. Many extrapleural sites of SFT origin have been reported in the literature since the first description of five pleural SFTs by Klemperer in 1931 (1).

Approximately 100 cases of central nervous system SFT have been reported including intracranial SFT and SFT in the spinal canal. Ciappetta et al. stated that intradural spinal SFT accounted for 73% of the reported cases, and the remaining 27% had intramedullary localization (2). However, to the best of our knowledge, SFT with a primary site in the filum terminale has not been reported yet. Here we report a case of SFT in the filum terminale. Both the rarity of the location and the characteristic magnetic resonance imaging (MRI) features are the highlights of this case.

Case report
A 50-year-old woman suffered from intermittent pain in the lumbosacral area for about 10 years. Her back pain became aggravated progressively and radiated to the bilateral lower extremities 1 year before she arrived at our hospital. A general physical examination was normal. Neurological examination, which included mental status, cranial nerves, cerebellar testing, and motor and sensory tests of the extremities, was also negative. Routine blood tests were within normal limits.

Magnetic resonance imaging was performed with a commercially available 3.0 Tesla MR scanner (Signa EXCITE HD, GE Healthcare, Milwaukie, WI, USA) and demonstrated a well-defined mass measuring 18 × 20 mm in transverse diameter located at the level of L2. The irregular intraspinal mass showed isointensity on T1-weighted images and hypointensity on T2-weighted images, and was avidly and homogeneously enhanced after intravenous injection of Gd-DTPA (Magnevist; Bayer Schering Pharma AG, Berlin, Germany) at a dose of 0.1 mmol/kg (Fig. 1).

A L2-L3 laminectomy was performed and an obvious tumor surrounded by nerve bundles was found. The dura mater was opened, as was the overlying arachnoid. Also, the nerve roots of the cauda equina were retracted. A well-defined tumor arising from the filum terminale was exposed. There was a dilated feeding artery and a draining vein running to and from the tumor. The tumor was completely removed. The microscopic appearance showed that the tumor was composed of cytologically scattered spindle cells arranged randomly. Immunohistochemically, the tumor cells had strong and diffuse positivity for vimentin, but were negative for S-100 and EMA antigen. With these findings, SFT of the filum terminale was considered to be the final diagnosis based on the World Health Organization’s classification of soft tissue tumors (2002).

Postoperatively, the patient became free of pain in the lumbosacral area, however, she had numbness of the buttocks and sacral region.

Discussion
SFT is a rare neoplasm of mesenchymal origin that arises most commonly in the visceral pleura (3, 4). Review of the literature also reveals extrathoracic involvement, such as liver (5), tunica vaginalis testis (6), the upper respiratory tract, epiglottis (7), thyroid (8), orbit (9), tibial periosteum (10), soft tissues (11), and the meninges (12). Recently, SFT
has been described in the central nervous system. The spinal canal is an extremely rare site of SFTs with only 19 cases presenting as an intradural extramedullary or intramedullary mass reported in the literature since 1996. To our knowledge, this is the first report of an SFT arising from the filum terminale.

The typical histological features of SFTs are spindle cells in interlacing fascicles or in a patternless manner with collagenous bands and branching vascular channels. Immunohistochemically, the tumors are usually positive for CD34, CD99, and vimentin as well as B-cell lymphoma 2 (bcl2), but negative for keratin, cytokeratin, S-100 protein, smooth muscle actin, factor VIII-related antigen, desmin, and epithelial membrane antigen. The Ki-67 labeling index is also positive in 4% of neoplastic cells (2).

The signal intensity of SFT reflects fibrous tissue with high collagen content. The appearance of a “black-and-white” mixed pattern or “yin-yang” pattern been reported as characteristic for the SFT. Areas of hyper-intense signal intensity have been seen within the lesion on T2-weighted images as a result of internal hemorrhage, cystic degeneration, or relatively fresh fibrosis. However, in this case, we observed the usual MRI findings, which refer to isointensity to hypointensity on T1-weighted imaging and hypointensity on T2 (13). Collagen content increases, the signal intensity of SFT on T2-weighted images decreases (14). Enhancement of a specific tumor is attributed to various factors such as vascularity, capillary permeability, and renal clearance, as well as composition of extracellular fluid. High vascularity due to prominent vascular channels within the tumor contributes to the marked enhancement (15).

According to Gold et al. (16), among tumors less than 10 cm in size, there have been only two of 15 SFTs with a histologically malignant component. Most SFTs have a favorable clinical course. Surgical treatment in time appears to be important. However, because of its rarity, SFT is often poorly recognized and remains a diagnostic challenge. The purpose of this article is to increase our understanding of SFT.

In conclusion, although its occurrence is uncommon, we suggest that SFT should be kept in mind in the differential diagnosis of a spinal canal mass, especially for lesions with hypointensity on T2-weighted images.

Conflict of interest: None.

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