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Case Report

Spinal solitary fibrous tumor: seventh reported case and review of the literature

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We present the clinical, radiological, and pathological features of a solitary fibrous tumor in the spinal cord. This case is the seventh spinal solitary fibrous tumor in the literature. The tumor caused clinical symptoms in a 70-year-old female, which indicated compression of the spinal cord. Magnetic resonance imaging showed an intradural extramedullary mass at T3 vertebral level. Surgically, the tumor was firm, in an intradural extramedullary location and attached to the dura. Histologically, the tumor was composed of spindle cells in a collagen-rich matrix but exhibited regional variations. CD34 and vimentin were diffusely positive during immunohistochemical stain testing. The tumor displayed no positive staining for epithelial membrane antigen, cytokeratin, S-100 protein, smooth muscle actin or desmin. The Ki-67 labeling index was low. Solitary fibrous tumors have been found in a variety of locations suggesting that a solitary fibrous tumor has a mesenchymal origin. This rare tumor should be considered in the differential diagnosis of spinal tumors.
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Introduction

A solitary fibrous tumor (SFT) is also known as a pleural neoplasm, formerly called solitary fibrous mesothelioma.[1] Although they were originally thought to arise from modified mesothelial cells, recently a fibrous tissue origin has been supported by the absence of epithelial markers and lack of mesothelial features in cell cultures. Additionally, ultrastructural studies further support fibroblast-like rather than mesothelial features of these cells. The occurrence of this tumor in extraserosal sites including orbit,[2] upper respiratory tract,[1] nasopharyngeal sinuses,[3] periosteum,[4] soft tissues,[5] skin,[6] prostate,[7] meninges,[8] spinal cord,[9–13] epiglottis,[14] liver,[15] and thyroid[16] suggest that a solitary fibrous tumor has a mesenchymal origin.

To the best of our knowledge, six spinal SFT cases have been reported in the literature till now. In this Case Report, we present the seventh case, analyse the spinal SFT literature and draw some conclusions regarding this rare spinal tumor.

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Case report

The patient was a 70-year-old woman who initially complained of a progressive right leg weakness over a 30-day period. Her neurological examination showed 1/5 global paresis of the right leg but there was no sensory loss and the patellar tendon, ankle tendon, and anal reflexes were normal.

Magnetic resonance imaging (MRI) revealed an intradural extramedullary mass at T3 vertebral level. The lesion showed an irregular isointensity in the T1-weighted image and irregular hypointensity and extensive oedema in the T2-weighted image (Figure 1). Gadolinium enhancement could be seen clearly (Figure 2).

The patient underwent T2–T4 laminectomies. The left paramedian durotomy revealed a gray-white colored rubbery mass, located at the left antero-lateral surface of the spinal cord being compressed and shifted to the right of the spinal canal. The tumor was attached to the dura-mater and removed totally by microsurgery. The intraoperative examination of the frozen sections resulted in a preliminary diagnosis of a benign tumor that was composed of fusiform cells suggestive of a neurofibroma. However, no actual attachment was determined from the spinal roots. After resection and hemostasis, the dura was sutured

primarily. The post-surgical period was uneventful; the patient experienced no strength deficits and was discharged on postoperative day 6. Twelve months after surgery her neurological exam showed no deficit and she continues to live a disease free life.

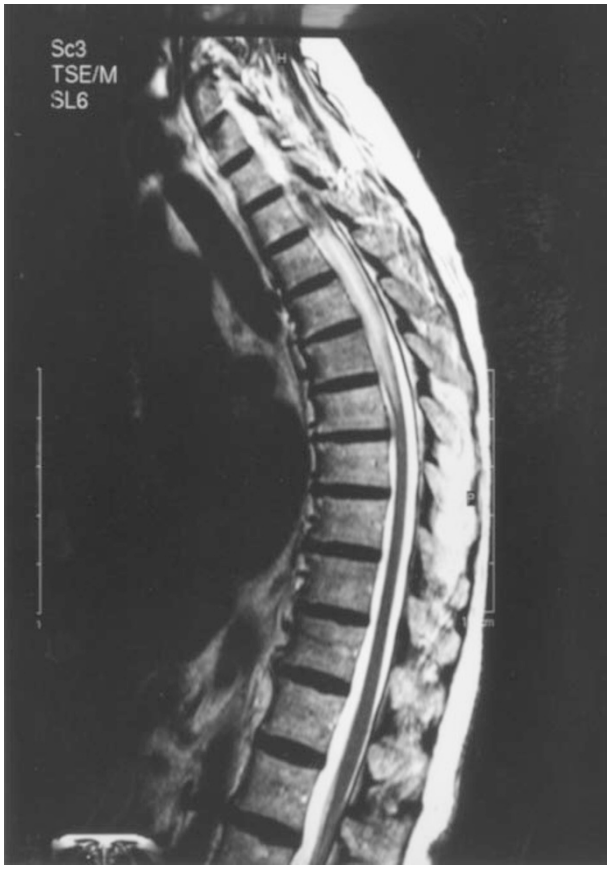

Figure 1 Magnetic resonance image shows an intradural extramedullary mass and extensive oedema at T3 vertebral level

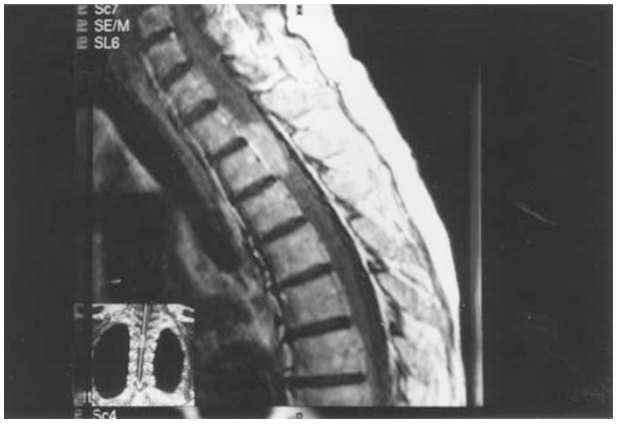

Figure 2 Post-contrast magnetic resonance image shows gadolinium enhancement

Pathology

Tumor tissue was fixed in 10% buffered formalin and embedded in paraffin. Tissue sections were stained routinely with hematoxylin and eosin. Selected sections of the tumor were stained with reticulin and trichrome stains. The tumor's immunohistochemical stains were tested for epithelial membrane antigen (EMA, BioGenex), vimentin (BioGenex), S-100 (BioGenex), cytokeratin (BioGenex), smooth muscle actin (SMA, BioGenex), desmin (BioGenex), CD34 (BioGenex) and Ki-67 (BioGenex).

The permanent histological examination of the tumor specimen revealed uniform oval to spindle cells arranged in interlacing fascicles (Figure 3). The tumor cells had elongated nuclei with slightly granular chromatin. In some areas, extensive collagen deposition was identified being intermingled with the

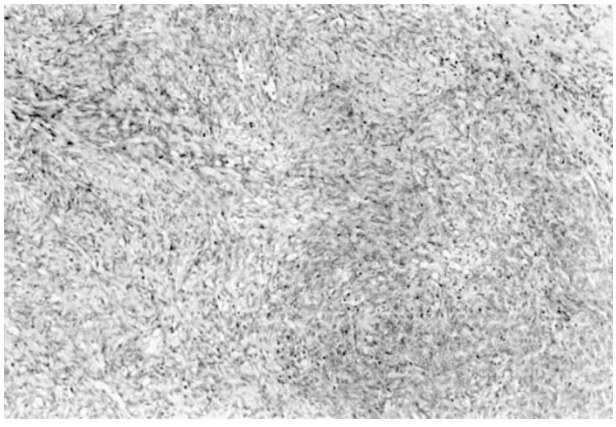

Figure 3 $\times 100$, H&E, highly cellular area of the tumor

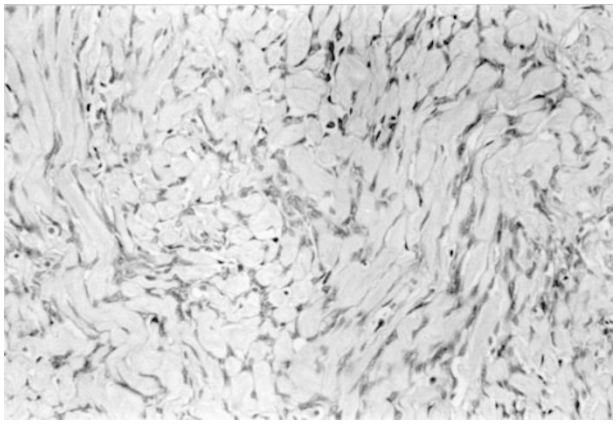

Figure 4 $\times 200$, H&E, neoplastic cells are crowded out by dense collagen deposition

neoplastic cells (Figure 4). A rich collagen network was demonstrated between many tumor cells with trichrome stain. Focal areas showed extensive fibrosis and hyalinization. Gaping and branching vascular channels, indistinguishable from those of hemangiopericytoma, were identified in some focal areas. Mitoses were present but the mitotic index was not high (<4 mitotic figures per 10 high power fields) suggestive of a benign phenotype. Pleomorphism and necrosis were absent. No psammoma bodies or whorls were identified.

Immunohistological findings revealed strong and diffuse immunopositivity with vimentin and CD34. Diffuse cytoplasmic immunopositivity with CD34 was a prominent and diagnostic finding of this tumor (Figure 5). The tumor cells did not exhibit positive staining for EMA, S-100, smooth muscle actin, desmin or cytokeratin. Ki-67 proliferation index was low, at 1.2%.

Discussion

The differential diagnosis of an intradural extramedullary lesion of the spinal cord includes: schwannoma, meningioma, hemangiopericytoma, non-neoplastic cysts, meningeal carcinomatosis, drop metastases and inflammatory lesions.

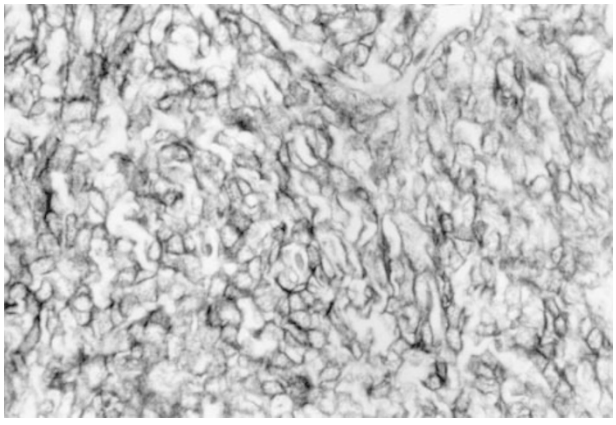

Figure 5 ×400, CD34 (BioGenex), diffuse cytoplasmic immunopositivity for CD34 in the tumor cells

To the best of our knowledge, six cases of primary solitary fibrous tumors occurring in the spinal canal have been reported in the literature,[9–13] our case being the seventh (Table 1). These seven cases suggest that this type of tumor is not only confined to the pleura and other serosal sites, but can be found in the spinal canal, in an intradural-extramedullary location. The histological features of a solitary fibrous tumor include spindle cells that are arranged as a storiform pattern in a densely collagenous matrix. The intercellular stroma has a distinctive fibrohyaline morphology with a keloid-like quality. Blood vessels may assume a branched appearance like those seen in hemangiopericytomas. Although mitotic activity is limited in solitary fibrous tumors, malignant cases have been reported.[17] Even though criteria for a diagnosis of malignant fibrous tumors has not yet been settled, increased mitotic rate (>4 mitotic figures per 10 high-power fields) and pleomorphism may suggest the malignant behavior.[9] Indicators of good prognosis are presence of a pedicle and good circumscription at the time of surgery, in addition to the absence of nuclear pleomorphism or mitotic activity.[18]

Immunohistology demonstrates somewhat diffused and strong reactivity to vimentin, but also occasional desmin immunoreactivity. The absence of cytokeratin, epithelial membrane antigen, S-100 protein and smooth muscle actin, and diffused immunopositivity for CD34 (also known as Q BEND 10) stains are consistent findings in solitary fibrous tumors. Ultrastructurally, these tumors consist of an abundance of collagen surrounding the fibroblast-like spindle cells with rough endoplasmic reticulum, cytoplasmic filaments and occasional primitive junctions but without microvilli, basal lamina, tonofilaments or well-formed tight junctions.[9]

The differential diagnosis for solitary fibrous tumors consists of meningiomas and neurofibromas. Solitary fibrous tumors do not exhibit immunopositivity for S-100 protein as seen in neurofibroma. Fibrous meningioma may show a storiform pattern but differs from solitary fibrous tumors only by positive immunostaining for epithelial membrane antigen.

Table 1 Spinal SFTs: Review of the clinical features of all cases

C	*Ref*	*Sex/age*	*Location*	*Extent of resection*	*Follow-up*
1	9	M/47	T4–T5 intradural/extramedullary	Total	NA
2	10	M/62	C6–C7 intradural/extramedullary	Total	NA
3	11	M/33	T7–T8 intradural/extramedullary	Total	NA
4	12	F/50	Spinal cord	Subtotal	After 5 year recurrence and DOD
5	12	F/54	L1–L3	Total	7 years after death with no recurrence
6	13	F/46	T12/L1 intradural/extramedullary	Total	4 months NED
7	▽	F/70	T3 intradural/extramedullary	Total	12 months NED

▽, presented case; DOD, died of disease; NA, not available; NED, no evidence of disease; C, Case number; M, male; F, female; Ref, references

This case contains hemangiopericytomatous areas as seen in many solitary fibrous tumors. Although CD34 immunopositivity of a solitary fibrous tumor is a consistent finding, CD34 positivity has also been reported in hemangiopericytomas.[8] Therefore the diagnosis relies on the distinct histopathologic features of a solitary fibrous tumor such as keloid-like collagen, extensive fibrosis and hyalinization.[9,11] Discrimination of spinal SFT from hemangiopericytomas is especially important. Leiomyoma that shows desmin and smooth muscle actin immunopositivity is another tumor that should be considered in the differential diagnosis of spinal solitary fibrous tumors. The most cellular types of spinal solitary fibrous tumors may be misdiagnosed as fibrosarcoma. However, the classical 'herring bone' pattern formed by tumor cells usually seen in the former and S-100 immunopositivity in the latter are helpful indicators to distinguish between these two spinal tumors.

Conclusion

These seven spinal SFT cases are not sufficient to reach firm conclusions, but three of them have appeared to follow a benign course, suggesting that no other treatment is needed after total surgical removal of spinal solitary fibrous tumors. Two of the five cases in the literature, which were totally removed and have follow-up have shared a benign, disease-free prognosis without any therapy. This approach is similar to the treatment principles in other locations of this tumor. We believe that awareness of this relatively rare spinal tumor may promote more frequent recognition. Solitary fibrous tumors must be differentiated histopathologically from other spindle cell neoplasms particularly the hemangiopericytoma. Hemangiopericytoma with high cellularity and rich in collagen can be mistaken for solitary fibrous tumors. Strong immune reactivity for CD34 is characteristic for SFT but not for hemangiopericytoma as well as other spindle cell tumors.

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