Solitary Fibrous Tumor of the Spinal Nerve Rootlet

Report of a Case Mimicking Schwannoma

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• We report a case of solitary fibrous tumor involving the spinal nerve root at the L1-L2 level in a 67-year-old man. The patient presented with lumbar pain and weakness in his right lower extremity. Histologically, the tumor was composed of a proliferation of monomorphous spindle cells in an abundant collagenous stroma; neither necrosis nor mitoses were evident. These cells were strongly immunoreactive with CD34, Bcl-2, CD99, and vimentin, but were negative with S100 protein, smooth muscle actin, and epithelial membrane antigen. Such an immunohistochemical profile was consistent with a solitary fibrous tumor of the spinal nerve rootlet and ruled out the main differential diagnoses, schwannoma and meningioma. The present case suggests that solitary fibrous tumor should be considered in differentiating spindle cell lesions of the spinal cord and nerve rootlet.

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Solitary fibrous tumors (SFTs) are ubiquitous mesenchymal neoplasms of putative fibroblastic origin.[1] They were originally described in the pleura, but subsequently have been reported in many extraserosal sites.[2,3] Solitary fibrous tumors may also occur in the meninges and central nervous system parenchyma[4–6]; in addition, spinal cord and spinal nerve involvement has been reported.[7–13]

We describe an SFT of the lumbar spine, which presented as a symptomatic mass and mimicked a schwannoma. We also discuss the differential diagnosis of intradural extramedullary masses, with emphasis on immunohistochemical findings.

REPORT OF A CASE

The patient was a 67-year-old man who complained of lumbar pain and weakness in his right lower extremity that had progressed during the preceding 3 months. Neurological examination showed no sensory loss and absence of tendon reflexes at the knees. Perianal sensation, bladder and bowel continence, and bulbocavernous reflex were normal.

Magnetic resonance imaging revealed an extramedullary, extradural, dumbbell-shaped mass that measured 5 cm at its great-

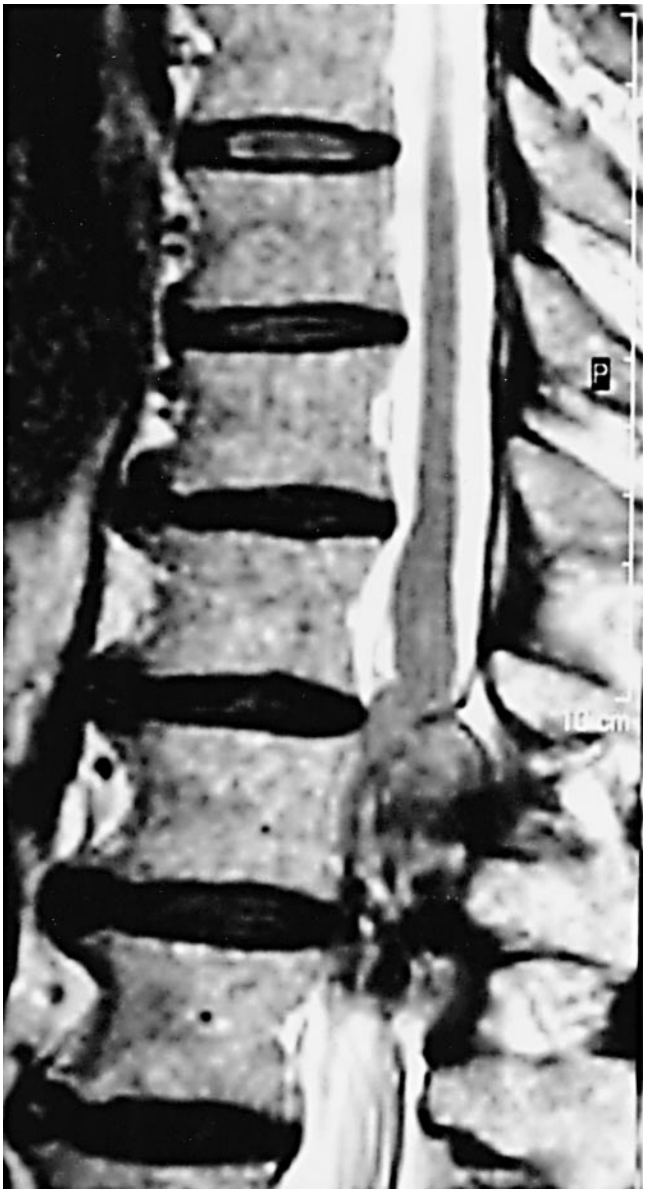

Figure 1. *A preoperative sagittal magnetic resonance image with gadolinium of the lumbar region shows an irregularly enhancing mass at the L1-L2 level.*

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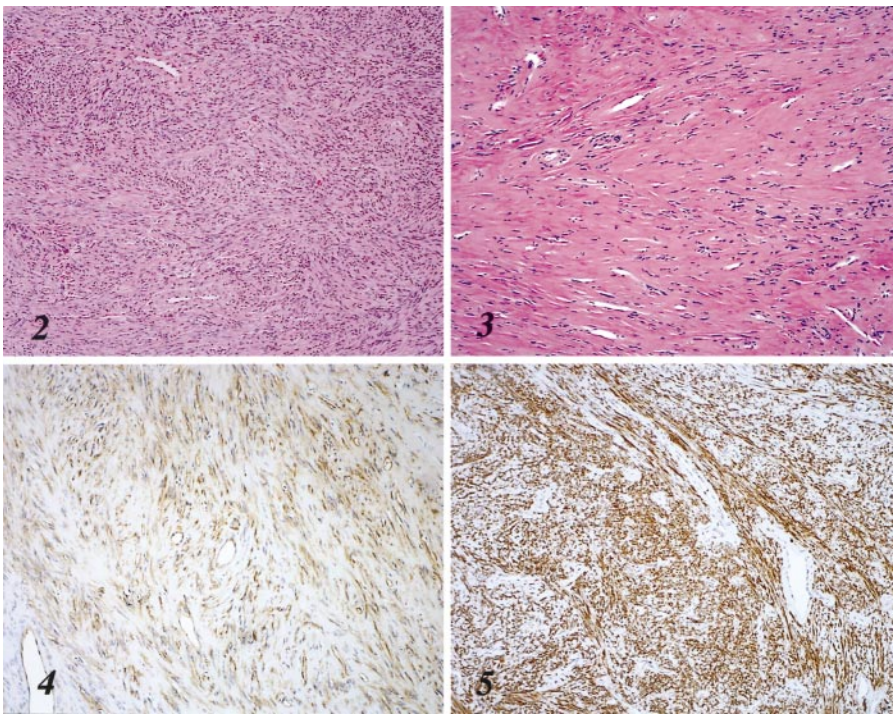
Figure 2. *Histologically, the tumor is composed of monomorphous spindle cells, arranged in short fascicles, with very low mitotic activity (hematoxylin-eosin, original magnification ×100).*

Figure 3. *Hypocellular areas with perivascular hyalinization are evident (hematoxylin-eosin, original magnification ×100).*

Figure 4. *CD34 is diffusely positive in neoplastic spindle cells and in endothelial cell (immunostaining, counterstained with hematoxylin, original magnification ×100).*

Figure 5. *Tumor cells are strongly immunoreactive with Bcl-2 (immunostaining, counterstained with hematoxylin, original magnification ×100).*



est axis. It was localized at the L1-L2 vertebral level and had heterogeneous contrast enhancement (Figure 1).

A presumptive clinical diagnosis of schwannoma was made, and the patient underwent surgical intervention. During surgery, the tumor appeared firmly attached to the posterior nerve rootlets, but gross total resection of the tumor was performed with sparing of the nerves. The postsurgical period was uneventful, and the patient was discharged with no neurological deficits.

MATERIALS AND METHODS

Tissue sections were routinely fixed, embedded in paraffin, and stained with hematoxylin-eosin. For immunohistochemistry, a standard avidin-biotin complex technique was used with the following antibodies: CD34 (monoclonal; BioGenex Laboratories, San Ramon, Calif), Bcl-2 (monoclonal; Dako, Glostrup, Denmark), S100 protein (monoclonal, BioGenex), smooth muscle actin (monoclonal, BioGenex), epithelial membrane antigen (monoclonal, Neomarkers, Fremont, Calif), and Ki-67 (monoclonal, Dako).

PATHOLOGIC FINDINGS

Grossly, the cut surfaces of the tumor were rubbery and tan with yellowish areas. Necrosis was not apparent. Microscopically, the tumor was composed of a proliferation of monomorphous spindle cells with scant cytoplasm, indistinct cytoplasmic borders, and inconspicuous nucleoli, embedded in a prominent collagenous stroma (Figure 2). Hypocellular areas alternated with hypercellular areas, and perivascular hyalinization was frequent (Figure 3). Neither necrosis nor mitoses were evident.

Immunohistochemically, neoplastic cells were diffusely positive for CD34 (Figure 4), Bcl-2 (Figure 5), CD99, and vimentin, but were negative for S100 protein, epithelial membrane antigen, and smooth muscle actin. The proliferation index (evaluated with Ki-67/MIB-1) was 2%.

COMMENT

Solitary fibrous tumors are peculiar fibroblastic proliferations that are now being described in an ever-increasing number of extrapleural sites. They can be asymptomatic and often display benign biological behavior, even though some authors would put them in an intermediate tumor category.[14]

The case described here presented as a symptomatic neoplasm, with clinical characteristics identical to those of a classic spinal schwannoma. Clinical discrimination from schwannoma or other spindle cell tumors arising from the meninges and nerves may not be possible; the differential diagnosis is based on histopathologic examination.

Typically, SFTs have distinctive, although nonspecific, histologic features, which include alternating hypercellular and hypocellular areas with perivascular hyalinization or myxoid degeneration. A hemangiopericytic vascular pattern is usually present.

Immunoreactivity with CD34, Bcl-2, and CD99 antibodies is quite typical. Combined expression of CD34 and Bcl-2 has been detected in other spindle cell neoplasms as well, for example, in breast[15] or soft tissue tumors.[16] Solitary fibrous tumor is just one of the vimentin$^+$/CD34$^+$/Bcl-2$^+$/CD99$^+$ family of tumors, as already emphasized by Magro et al.[15] However, immunohistochemical findings are often essential in the differential diagnosis.

Solitary fibrous tumors arising in spinal nerve roots must be differentiated from schwannoma, meningioma, and hemangiopericytoma. Even if morphologic features are helpful (schwannoma often shows alternating Antoni A and Antoni B areas, and in meningioma cellular whorls and psammoma bodies are frequent), again differential diagnosis depends on immunohistochemical results. Schwannomas are invariably S100 protein positive and meningiomas are epithelial membrane antigen positive, whereas both antibodies are absent in SFT.

About half of hemangiopericytomas are CD34 immunoreactive, at least focally. They also share many histologic features with SFTs, except for dense collagen bands, which are generally absent in hemangiopericytomas. They probably belong to the same tumor spectrum, and the same

features (necrosis, cellular atypia, and mitotic activity) are predictive of malignancy in both hemangiopericytomas and SFTs. According to some authors, distinguishing among questionable cases may not be important.[14]

In conclusion, SFTs should be included among the spindle cell lesions arising in spinal cord. Their recognition is important for an accurate nosologic classification, based on morphologic and immunohistochemical profile.

References

1. Fletcher CDM, Unni KK, Mertens F, eds. *Pathology and Genetics of Tumours of Soft Tissue and Bone.* Lyon, France: IARC Press; 2002. *World Health Organization Classification of Tumours;* vol 5.
2. Chan JKC. Solitary fibrous tumor: everywhere, and a diagnosis in vogue. *Histopathology.* 1997;31:568–576.
3. Hasegawa T, Matsuno Y, Shimoda T, Hasegawa F, Sano T, Hirohashi S. Extrathoracic solitary fibrous: their histologic variability and potentially aggressive behaviour. *Hum Pathol.* 1999;30:1464–1473.
4. Carneiro SS, Scheithauer BW, Nascimento AG, Hirose T, Davis DH. Solitary fibrous tumor of the meninges: a lesion distinct from fibrous meningioma: a clinicopathologic and immunohistochemical study. *Am J Clin Pathol.* 1996;106:217–224.
5. Brunori A, Cerasoli S, Donati R, Giangaspero F, Chiappetta F. Solitary fibrous tumor of the meninges: two new cases and review of the literature. *Surg Neurol.* 1999;51:636–640.
6. Tihan T, Viglione M, Rosenblum MK, Olivi A, Burger PC. Solitary fibrous tumor in the central nervous system. *Arch Pathol Lab Med.* 2003;127:432–439.
7. Alston SR, Francel PC, Jane JA Jr. Solitary fibrous tumor of the spinal cord. *Am J Surg Pathol.* 1997;21:477–483.
8. Malek AM, Weller SJ, Price DL Jr, Madsen JR. Solitary fibrous tumor presenting as symptomatic intraspinal mass: case report. *Neurosurgery.* 1997;40:844–847.
9. Kanahara T, Hirokawa M, Shimizu M, et al. Solitary fibrous tumor of the spinal cord: report of a case with scrape cytology. *Acta Cytol.* 1999;43:425–428.
10. Kataoka H, Akiyama Y, Kubo S, et al. Solitary fibrous tumor of the spinal nerve rootlet: case report and literature survey. *Pathol Int.* 1999;49:826–830.
11. Mordani JP, Haq IU, Singh J. Solitary fibrous tumour of the spinal cord. *Neuroradiology.* 2000;42:679–681.
12. Vorster SJ, Prayson RA, Lee JH. Solitary fibrous tumor of the thoracic spine: case report and review of the literature. *J Neurosurg.* 2000;92:217–220.
13. Kurtkaya O, Elmaci I, Sav A, Pamir MN. Spinal solitary fibrous tumor: seventh reported case and review of the literature. *Spinal Cord.* 2001;39:57–60.
14. Kempson RL, Fletcher CDM, Evans HL, Hendrickson MR, Sibley RK. *Tumors of the Soft Tissue.* Washington, DC: Armed Forces Institute of Pathology; 1999:52–55. *Atlas of Tumor Pathology*; 3rd series, fascicle 30.
15. Magro G, Bisceglia M, Michal M, Eusebi V. Spinal cell lipoma-like tumor, solitary fibrous tumor and myofibroblastoma of the breast: a clinico-pathological analysis of 13 cases in favor of a unifying histogenetic concept. *Virchows Arch.* 2002;440:249–260.
16. Suster S, Fisher C, Moran CA. Expression of bcl-2 oncoprotein in benign and malignant spindle cell tumors of soft tissue, skin, serosal surfaces, and gastrointestinal tract. *Am J Surg Pathol.* 1998;22:863–872.