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.

(Arch Pathol Lab Med. 2004;128:335–337)

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 parenchyma4–6; 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-

Accepted for publication November 6, 2003.

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The authors have no relevant financial interest in the products or companies described in this article.

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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 asymptomatic 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 breast15 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