PERIPHERAL NERVE SHEATH TUMORS

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KEYWORDS

• Nerve sheath tumors • Neoplasms • Schwann cells • Peripheral nerve

ABSTRACT

This article presents an overview of the diagnostic categories of benign and malignant nerve sheath tumors, including neuroma, neurofibroma, nerve sheath myxoma, perineurioma, schwannoma, and malignant peripheral nerve sheath tumor. The discussion emphasizes histologic patterns; ancillary studies, such as immunohistochemistry; and differential diagnoses. The information is of value to practicing pathologists in both community and academic settings.

OVERVIEW OF PERIPHERAL NERVE SHEATH TUMORS

Peripheral nerve sheath tumors consist of a heterogeneous group of neoplasms derived from one or more of the cell types that accompany peripheral nerve fibers. The predominant cell types include Schwann cells and perineurial cells, which are described in this article, as well as nerve sheath dendritic cells and fibroblasts. Schwann cells are neural crest-derived spindle cells that form the innermost layer of the endoneurium and are intimately associated with the nerve fibers.[1] Schwann cell nuclei are wavy, spindled, and characteristically pointed at both ends. Schwann cells are diffusely and strongly immunoreactive for the S-100 protein[2] as well as Leu7 and laminin while negative for epithelial membrane antigen (EMA), desmin, and muscle-specific actin. Schwann cells are typically negative for cytokeratin expression; however, schwannomas with focal cytokeratin immunoreactivity have been described.[3] Perineurial cells are slender spindled cells with similarity to arachnoid mater cells, and they form an external layer outside of the endoneurium.[4] Perineurial cells are immunoreactive for claudin-1,[5] EMA (focally),[6] and glucose transporter 1 (GLUT1)[6] but are negative for the S-100 protein.

Benign nerve sheath tumors include neuromas, neurofibromas, nerve sheath myxomas, perineuriomas, and schwannomas. Benign nerve sheath tumors arise in the differential of bland spindle cell lesions of the superficial and deep soft tissues, which includes leiomyoma, fibromatosis, solitary fibrous tumor and low-grade fibromyxoid sarcoma. Benign nerve sheath tumors, however, may also display diverse cytomorphologic features, such as epithelioid morphology, hypercellularity, myxoid matrix, and degenerative atypia. Malignant peripheral nerve sheath tumors (MPNSTs) are the malignant and aggressive counterparts to the benign neoplasms. MPNSTs arise in the differential of intermediate to high-grade soft tissue neoplasms, which includes synovial sarcoma, fibrosarcoma, leiomyosarcoma, sarcomatoid carcinoma, and melanoma. A more detailed discussion of each of these individual non-nerve sheath entities can be found in the accompanying articles elsewhere in this issue.

NEUROMA

Neuroma is a benign, disordered proliferation of nerves that include all the cellular components of the nerve sheath in otherwise normal relationships to each other. Each component can be highlighted by the appropriate immunohistochemical stain (eg, neurofilament for axons, the S-100 protein for Schwann cells, and EMA for perineurial cells).

Traumatic neuroma presents as a painful or tender nodule arising adjacent to a nerve after trauma or surgery. Microscopically, it consists of a non-neoplastic, nonencapsulated, disorganized

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Key Features
Peripheral Nerve Sheath Tumors

1. May be associated with syndromes, such as neurofibromatosis type 1 or 2, multiple endocrine neoplasia (MEN), type II, and Carney complex
2. Composed of varying proportions of perineurial cells, Schwann cells, and nerve axons
3. Benign tumors include neuroma, neurofibroma, nerve sheath myxoma, perineurioma, and schwannoma
4. Benign tumors typically composed of bland spindle cells characterized by wavy nuclei with pointed ends
5. Benign tumors may display degenerative atypia, epithelioid morphology, myxoid stroma, or hypercellular regions
6. Neurofibromas characteristically contain dense collagen fibers and prominent mast cells
7. Schwannomas commonly display regions of alternating cellularity (Antoni A and Antoni B areas), Verocay bodies, fibrous capsules, and peripheral lymphoid cuffs
8. Schwannomas are diffusely and strongly immunoreactive for the S-100 protein
9. MPNSTs typically have sweeping fascicles of atypical spindle cells with wavy nuclei, tumor cell condensation around large vessels, frequent mitoses, and necrosis
10. MPNSTs are usually only focally immunoreactive for the S-100 protein

mass of nerve bundles embedded in scar tissue (**Fig. 1**). Mucosal neuroma presents as a small nodule located beneath the mucosal epithelial surfaces of the eyelids, intestines, and oral cavity. Clinically, mucosal neuroma is associated with MEN, type IIb, which is characterized by thyroid medullary carcinoma, pheochromocytoma, and parathyroid hyperplasia. Microscopically, it consists of nonencapsulated, disorganized bundles of nerve fibers with distinct perineurium (**Fig. 2**). Palisaded

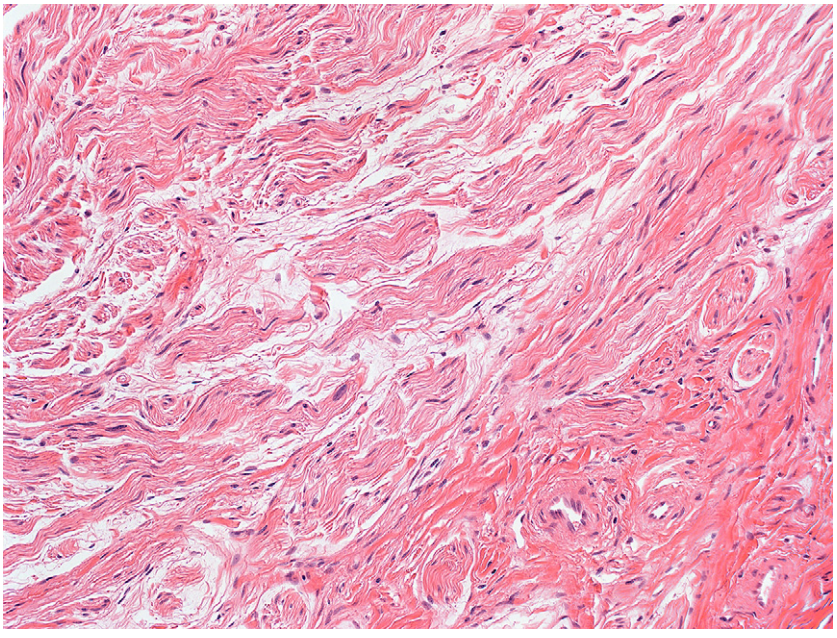

Fig. 1. Traumatic neuroma. Disorganized nerve bundles containing all normal cell types of the peripheral nerve are embedded in scar tissue and represent a non-neoplastic response to injury to a nerve (hematoxylin-eosin [H&E], original magnification ×64).

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