

Peripheral nerve sheath tumor

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A large majority of peripheral nerve tumors are benign. Some are caused by [neurofibromatosis](#), or [schwannomatosis](#).

Peripheral nerve tumors comprise less than 5% of all tumors of the hand.

Classification

[Peripheral nerve sheath tumor classification](#).

Pathology

Belakhoua and Rodriguez published pathologic features of common peripheral nerve sheath tumors, particularly those that may be encountered in the intracranial compartment or in the spine and paraspinal region.

They discussed [schwannoma](#), neurofibroma, atypical neurofibromatous neoplasms of uncertain biological potential, intraneural and soft tissue perineurioma, hybrid nerve sheath tumors, [MPNST](#), and the recently renamed enigmatic tumor, malignant melanotic nerve sheath tumor, formerly referred to as melanotic schwannoma. They also discuss the diagnostic relevance of these neoplasms to specific genetic and familial syndromes of nerve, including neurofibromatosis 1, neurofibromatosis 2, and schwannomatosis. In addition, they discuss updates in our understanding of the molecular alterations that represent key drivers of these neoplasms, including neurofibromatosis type 1 and type 2, [SMARCB1](#), [LZTR1](#), and [PRKAR1A](#) loss, as well as the acquisition of [CDKN2A/B](#) mutations and alterations in the polycomb repressor complex members ([SUZ12](#) and [EED](#)) in the malignant progression to MPNST. In summary, this review covers practical aspects of pathologic diagnosis with updates relevant to neurosurgical practice ¹⁾

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Belakhoua SM, Rodriguez FJ. Diagnostic Pathology of Tumors of Peripheral Nerve. Neurosurgery. 2021 Feb 16;88(3):443-456. doi: 10.1093/neuros/nyab021. PMID: 33588442.

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