Peripheral nerve sheath tumor classification

Peripheral nerve sheath tumor represent a heterogenous group with a wide spectrum of morphological features and biological potential. They range from benign and curable by complete excision (schwannoma and soft tissue perineurioma) to benign but potentially aggressive at the local level (plexiform neurofibroma) to the highly malignant (malignant peripheral nerve sheath tumors [MPNST]

The most common solitary nerve tumor is the neurilemmoma.

Neurofibroma. Most commonly found within the genetic disorder of neurofibromatosis. Neurofibromas may be solitary, multiple, or associated with von Recklinghausen's disease. They are usually centrally placed with nerve fibers traversing the tumor mass making it more difficult to remove the tumor without producing permanent neurological damage.

Schwannoma. These are nerve sheath tumors and can occur in isolation. Less commonly these tumors can occur in patients suffering from neurofibromatosis or schwannomatosis.

Malignant Peripheral Nerve Sheath Tumor. These very aggressive tumors are cancerous nerve sheath tumors and should be managed by a multi-disciplinary team. They can occur in association with neurofibromatosis Type 1.

Malignant tumors include neurofibrosarcomas which often are very aggressive, requiring wide excision or amputation, and the rare neuroepitheliomas. Reported nerve tumors, intraneural in location but nonneural in origin, include fibrofatty infiltration of the median and digital nerves, intraneural lipoma, hemangioma, and ganglion cysts. These lesions may be treated by decompression or excision, depending on the nature of the tumor. Four unusual cases are described [1)].

A nerve sheath tumor is a type of tumor of the nervous system (nervous system neoplasm) which is made up primarily of the myelin surrounding nerves.

A peripheral nerve sheath tumor (PNST) is a nerve sheath tumor in the peripheral nervous system.

Schwannoma is the most common nerve sheath tumor.

Benign peripheral nerve sheath tumor

Benign peripheral nerve sheath tumor.

Malignant peripheral nerve sheath tumor

Malignant peripheral nerve sheath tumor (MPNST) is a cancerous peripheral nerve sheath tumor.

Intraneural perineurioma

see Intraneural perineurioma.

Spinal neurofibroma

see Spinal neurofibroma.

Elsherif et al., noticed the coexistence of peripheral nerve sheath tumors (PNST) with lipomas within a subgroup of our patients. Given the prevalence of lipomas in the general population, we sought to investigate the extent of coexistence of the two entities aiming at uncovering any plausible association between both.

A retrospective review of all peripheral nerve sheath tumors (sporadic and syndromic forms) treated by a single surgeon between January 2009 and August 2015 was done. We recorded demographics (i.e., gender, age at diagnosis, imaging information, time to diagnosis) in addition to the method of diagnosis, subtype, number and location of lipomas, if present.

Over 6 years, 309 patients with PNST were operated/evaluated. These included 141 sporadic (schwannomas, neurofibromas) and 168 syndromic (neurofibromatosis type 1 and 2 and schwannomatosis). We found 32 patients [10.3%, 95% confidence interval (CI) = 7.43%-14.3%] with coexistent lipomas, some of whom also had a family member with lipoma (n = 3). Of these 26 had schwannomas, 3 had neurofibromas and 3 lacked definitive PNST histopathological diagnosis. Fourteen percent of patients with schwannomas and 2.9% of patients with neurofibromas had coexisting lipomas.

They believe there is an increased association of peripheral nerve tumors and lipomas overall [2].

1)
Strickland JW, Steichen JB. Nerve tumors of the hand and forearm. J Hand Surg Am. 1977 Jul;2(4):285-91. PubMed PMID: 197148.
2)
Elsherif MA, Babovic-Vuksanovic D, Spinner RJ. An association of peripheral nerve sheath tumors and lipomas. Acta Neurochir (Wien). 2017 Jan;159(1):185-190. doi: 10.1007/s00701-016-3038-3. PubMed PMID: 27900489.