

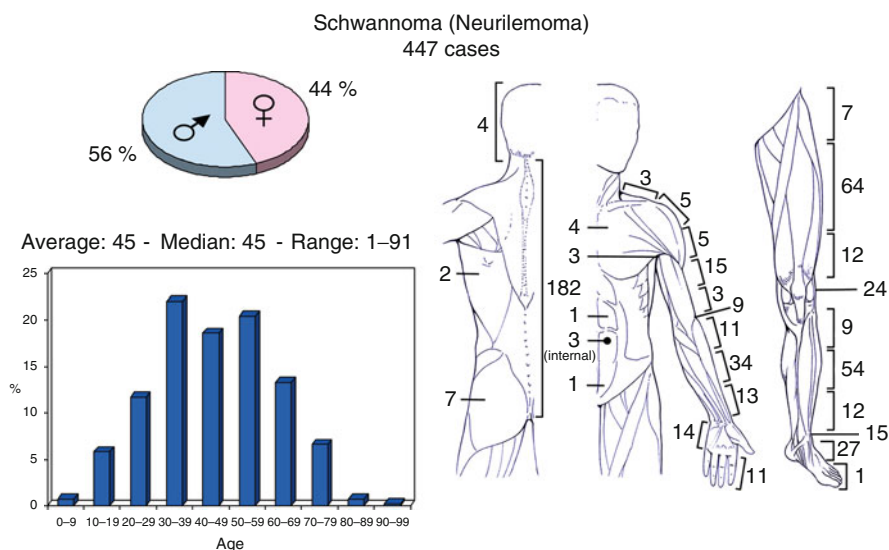
Chapter 71

Schwannoma (Neurilemoma, Neurinoma)

Marco Gambarotti

Definition: An encapsulated benign nerve sheath tumor composed of differentiated neoplastic Schwann cells.

Epidemiology: 20–50 years old. No sex predilection. Rarely associated with neurofibromatosis.



1900–2012 - Istituto Ortopedico Rizzoli - Laboratory of Experimental Oncology - Section of Epidemiology - Bologna - Italy

M. Gambarotti, MD
 Department of Anatomy and Pathological Histology, Istituto Ortopedico Rizzoli,
 Via del Barbiano 1/10, Bologna 40136, Italy
 e-mail: marco.gambarotti@ior.it

Location: Spinal roots, nerves of the mediastinum or of the retroperitoneum, peroneal, and ulnar nerve. Single nodule.

Clinical: Slow and progressive pain and neurological symptoms. Liquoral block, compression of the other roots or of the spinal cord, rachialgia, nocturnal increase in pain, stiffness, spinal contractures. Sharply painful, irradiated pain, paresthesia, algodystrophic syndrome when in peripheral nerve.

Imaging: On x-ray: scalloping of bone. On CT: well-circumscribed, homogeneous lesion, with muscle density. Frequent nonenhancing necrotic and cystic areas that cause an inhomogeneous hyperdense lesion after contrast administration. On MRI: nerve along the site of the mass. Capsulated. Homogeneous, isointense to muscle with frequent areas of low signal on T1, inhomogeneous, sometimes target appearance, higher than fat intensity on T2, diffused or peripheral enhancement with central necrotic unchanged zones on contrast T1.

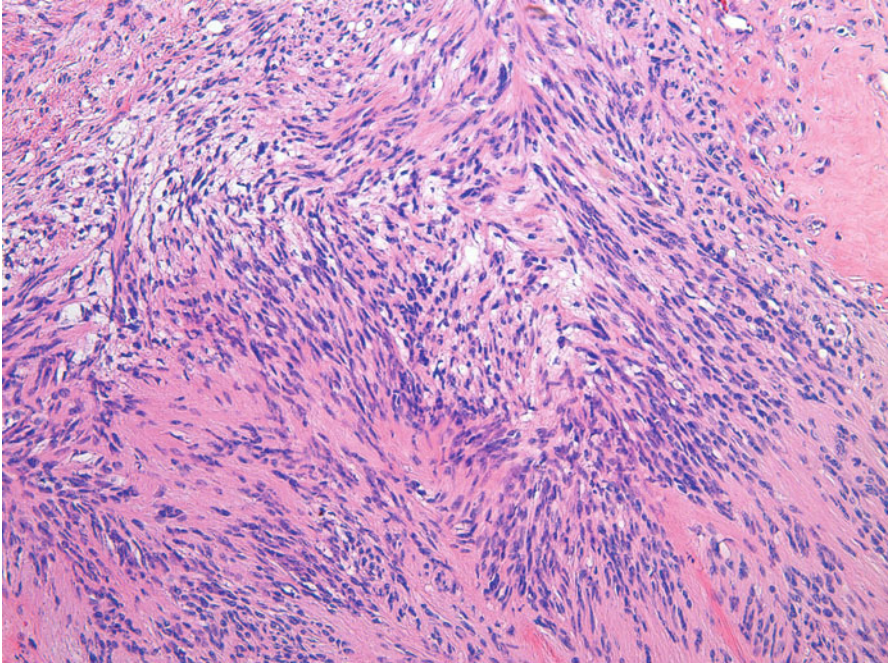
Histopathology: Globose “sausage-like” or “dumbbell mass,” elongated along the sac, well delimited by a capsule, soft, pink, white, or yellow, connected to a nerve root. Rarely, it expands like an hourglass inside and out of the intervertebral root foramen. S. of the peripheral nerves appears like an “onion bulb” or a piece of fruit attached to its stem. When very large, it may cause wide osteolyses. Usually encapsulated. Proliferation of spindle cells with ill-defined cytoplasm, in interlacing bundles, nuclear palisading, whorling of the cells, Verocay body formed by compact rows of well-aligned nuclei and cell processes that assume a roughly oval shape. Compact areas of spindle cells (Antoni A) alternating with loosely arranged areas (Antoni B). Rare if any mitotic figures. Large, irregularly spaced vessels with thickened wall and filled with thrombus material.

Course and Staging: Slow growth, rare recurrences after even incomplete surgery. Usually, stage 1 or 2.

Treatment: Marginal excision is curative. Malignant transformation exceptional.

Immunohistochemical Panel

| | |
|------|---|
| VIM | + |
| S100 | + |
| GFAP | + |



Elongated spindle cells with wavy nuclei organized in palisades (Antoni A areas) or loose sheets of cells (Antoni B areas)

Selected Bibliography

- Asthagiri AR, Helm GA, Sheehan JP (2007) Current concepts in management of meningiomas and schwannomas. *Neurol Clin* 25(4):1209–30. Review
- Benz MR, Czernin J, Dry SM, Tap WD, Allen-Auerbach MS, Elashoff D, Phelps ME, Weber WA, Eilber FC (2010) Quantitative F18-fluorodeoxyglucose positron emission tomography accurately characterizes peripheral nerve sheath tumors as malignant or benign. *Cancer* 116(2):451–8. Erratum in: *Cancer*. 2010 Feb 1;116(3):775
- Coulon A, Milin S, Laban E, Debiais C, Jamet C, Goujon JM (2009) Pathologic characteristics of the most frequent peripheral nerve tumors. *Neurochirurgie* 55(4–5):454–8. Review. French
- Levi AD, Ross AL, Cuartas E, Qadir R, Temple HT (2010) The surgical management of symptomatic peripheral nerve sheath tumors. *Neurosurgery* 66(4):833–40
- Woertler K (2010) Tumors and tumor-like lesions of peripheral nerves. *Semin Musculoskelet Radiol* 14(5):547–58. Review