Nerve Sheath Tumor

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43.1 Malignant Schwannoma (Malignant Peripheral Nerve Sheath Tumor—MPNST)

Definition: Malignant tumor that rises from a peripheral nerve, from a pre-existing benign nerve sheath tumor, in a patient with neurofibromatosis type 1, or with features of Schwann cells differentiation.

Epidemiology: 5% of all soft tissues sarcomas. Fifty percent in neurofibromatosis. Males prevail in patients with neurofibromatosis with a medium age of 30 years. Females prevail in sporadic cases in the adult age (>40 years).

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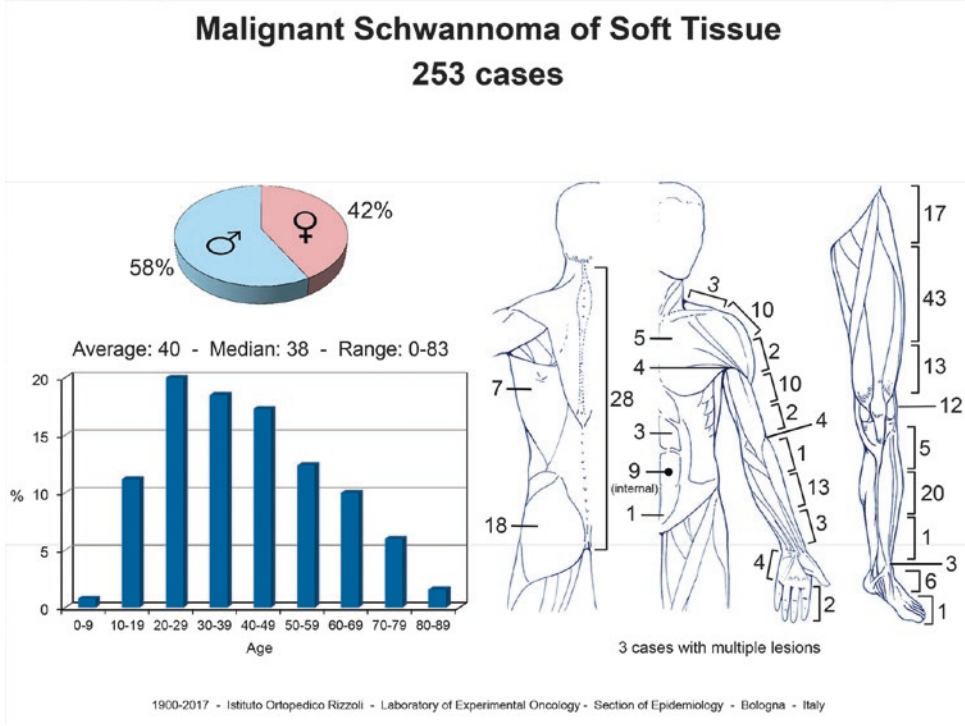
Malignant Schwannoma of Soft Tissue
253 cases
42%
58%
Average: 40 - Median: 38 - Range: 0-83
%
0-9 10-19 20-29 30-39 40-49 50-59 60-69 70-79 80-89
Age
3 cases with multiple lesions
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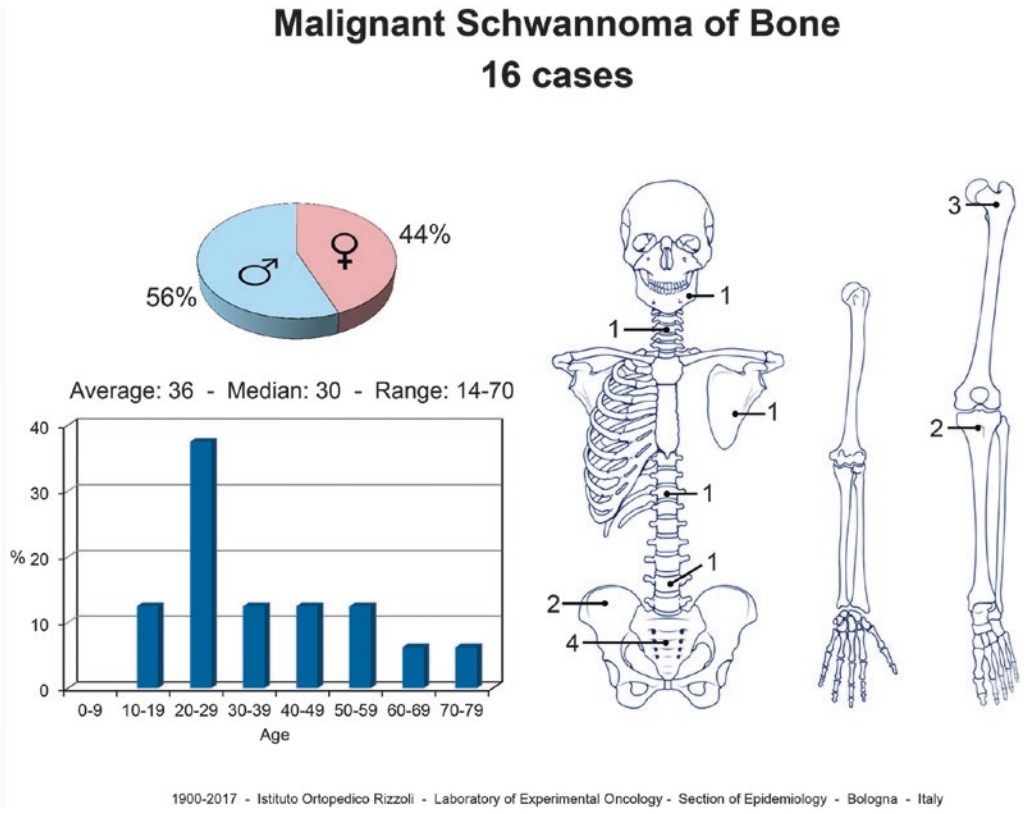
Malignant Schwannoma of Bone
16 cases
44%
56%
Average: 36 - Median: 30 - Range: 14-70
%
0-9 10-19 20-29 30-39 40-49 50-59 60-69 70-79
Age
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Location: In association with major nerve trunks: sciatic nerve, brachial plexus, sacral plexus. Gluteal region and pelvis, thigh region, shoulder region, and axilla are the most frequent sites.

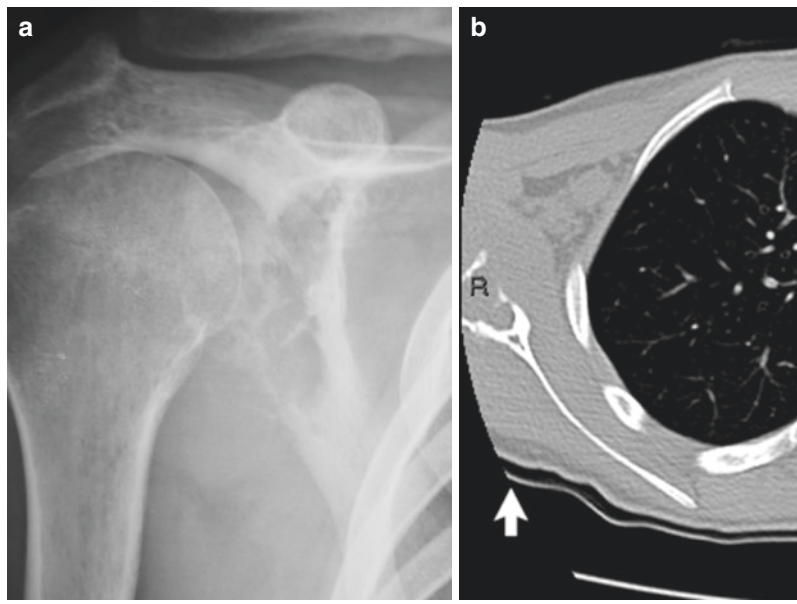
Clinical: Rapidly enlarging mass with pain. Sensory and motor symptoms including projected pain, paresthesias, weakness.

Imaging: Neither CT nor MRI can establish a definite diagnosis. On CT: mild enhancement with some nonenhancing areas of necrosis. On MRI: inhomogeneous and higher intensity than muscle on T1, markedly inhomogeneous and bright on T2, common necrosis in the center of the mass with peripheral capsule-like or irregular enhancement on contrast T1, a wide halo of edema unusual in benign lesions.

Histopathology: Large fusiform or eccentric mass within a major nerve. When the tumor spreads along the epi- and perineurium, it has a rosary bead aspect proximal or distal to the principal mass. Tumor is usually >5 cm, deep seated, but rarely superficial, fleshy, with hemorrhagic and necrotic diffused areas. Spindle cells with markedly irregular contours with wavy, buckled or comma shaped nuclei, lightly stained and usually indistinct cytoplasm. Densely cellular sweeping fascicules alternate with hypocellular, myxoid zones in the so-called "marble-like" pattern. A peculiar nodular or whorled arrangement of cells can be present. Focal nuclear palisading is present in only 10% of cases. Hemangiopericytoma-like vascular pattern can be present, with condensation of the neoplastic cells around blood vessels is characteristic. Heterologous elements such as glandular differentiation, skeletal muscle (malignant triton tumor), bone, cartilage, and angiosarcomatous areas are present in about 15% of cases. Epithelioid MPNST is a rare variant composed of epithelioid cells with abundant eosinophilic cytoplasm; it is not associated with NF1 and it is the most common type arising in schwannoma. On immunohistochemistry focal S-100 protein expression in classic MPNST. Epithelioid MPNST shows diffuse positivity for S100 protein and Sox10; it retains expression of H3K27me3 and mostly show loss of SMARCB1 expression.

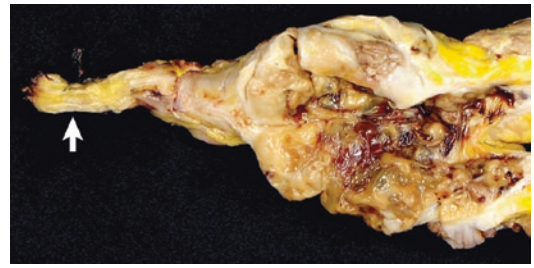
Course and Staging: Frequent local recurrence when surgery is inadequate. Metastases frequently develop in the lungs, liver, subcutis and bone, rarely in the lymph nodes. Usually, stage IIB.

Treatment: Very wide or radical local excision. Often, multicentric origin and diffusion along the nerve makes even radical surgery ineffective. Overall 5-year survival is 30% in patients with neurofibromatosis and 75% in sporadic cases. In neurofibromatosis, prognosis is worse because the tumor involves the trunk and proximal extremities with late diagnosis, it is larger and of higher grade, and multiple sarcomas may occur. Usefulness of radiotherapy and chemotherapy is uncertain.

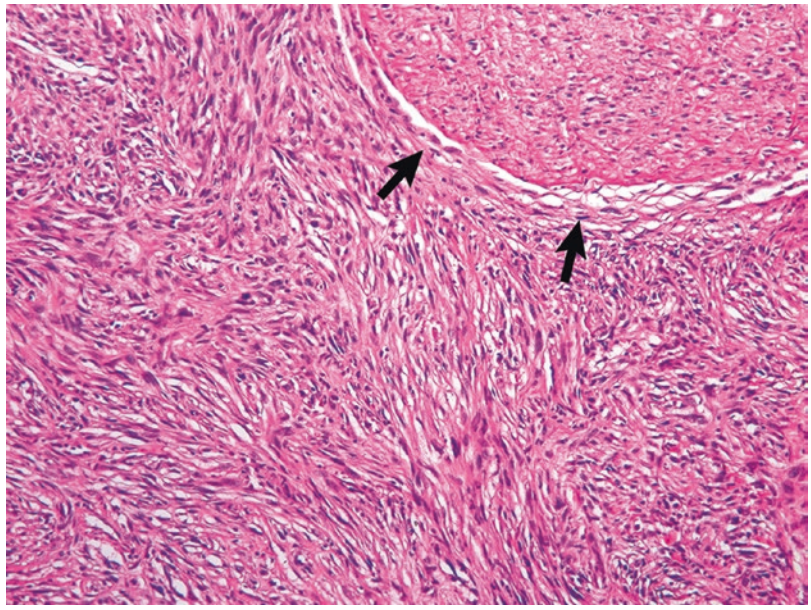


Radiograph (**a**), and CT (**b**) images. Poorly limited lytic and sclerotic lesion, destroying the cortex and invading the -soft tissues

Immunohistochemical panel	
• S100	+ (30% of cells in 30% of cases)
• GFAP	+ (30% of cells in 30% of cases)
• SOX10	+/–
• H3K23me3	–



The tumor arises from a peripheral nerve (*arrow*)



Highly atypical spindle cells with wavy or plump nuclei organized in storiform or fascicular pattern, frequently close to a peripheral nerve (*arrows*)

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