Malignant Triton Tumor

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168.1 Definition

Malignant Triton tumor is a variant of malignant peripheral nerve sheath tumor (→ see dedicated section) characterized by a rhabdomyoblastic cell component and a more aggressive behavior (worse prognosis) as compared to conventional MPNST.

This heterologous differentiation occurs in up to in 10% of cases and leads some investigators to consider Triton tumor as a mesenchymoma (→ see dedicated section).

The name "Triton" refers to experiments performed on salamanders of the genus *Triturus* (also known as Tritons), in which implantation of the cut end of a sciatic nerve into the soft tissue of the back resulted in the growth of a supernumerary limb.

The name includes the term "malignant" to differentiate this malignancy from the so-called benign Triton tumor (→ see dedicated section), a synonymous of neuromuscular choristoma (a rare benign peripheral nerve lesion in which well-differentiated skeletal muscle fibers are intimately associated or admixed with mature nerve fibers).

Suggested Readings

Bian (2019) A series of 10 malignant triton tumors in one institution. Medicine (Baltimore) 98(36):e16797

Fletcher (2013) WHO classification of tumours of soft tissue and bone (fourth edition)

Hornick (2019) Beyond "triton": malignant peripheral nerve sheath tumors with complete heterologous rhabdomyoblastic differentiation mimicking spindle cell rhabdomyosarcoma. Am J Surg Pathol 43(10):1323–1330

Thakrar (2014) Benign triton tumor: multidisciplinary approach to diagnosis and treatment. Pediatr Dev Pathol 17(5):400–405

Woodruff (1973) Peripheral nerve tumors with rhabdomyosarcomatous differentiation (malignant "Triton" tumors). Cancer 32:426–439

S. Mocellin, *Soft Tissue Tumors*, https://doi.org/10.1007/978-3-030-58710-9_168