

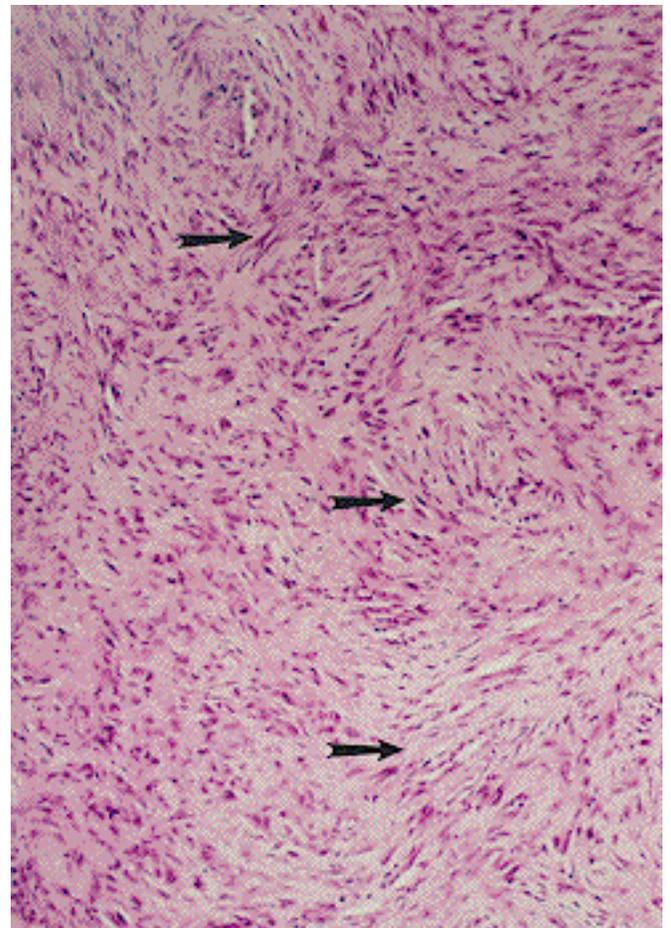
## MALIGNANT PERIPHERAL NERVE SHEATH TUMOR (MPNST)

MPNST arises from or shows differentiation toward peripheral nerve sheath cells. These tumors affect primarily the 20-50 year old age group, and about half develop in patients with neurofibromatosis—type 1. MPNST arise primarily in the extremities or the trunk. Cranial nerves are uncommonly involved.

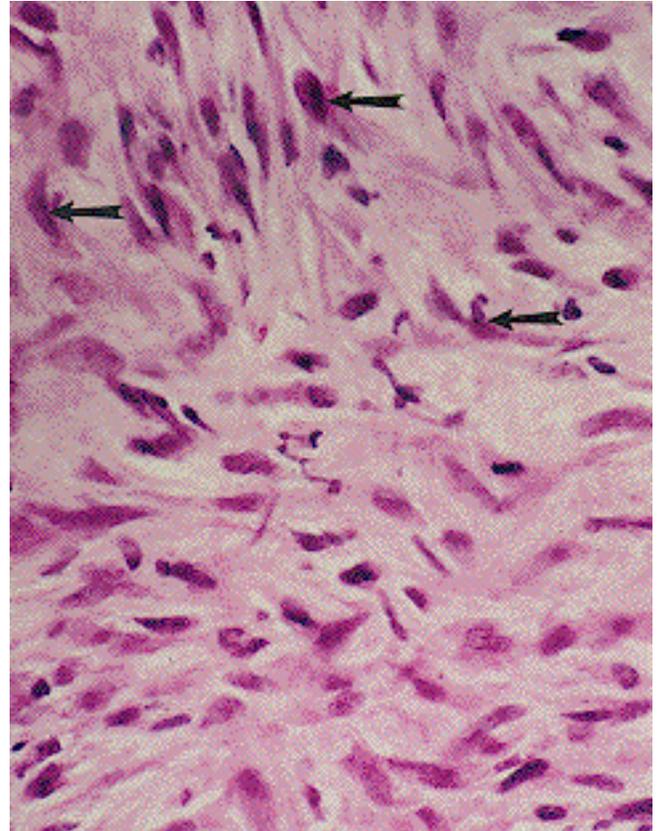
There is a wide histologic spectrum. The most common is that of a highly cellular, spindle cell tumor with a variable degree of nuclear pleomorphism.

Immunophenotyping shows focal staining for S-100 and CD57. MPNST is a high grade sarcoma in most instances with poor 5 year survival rates.

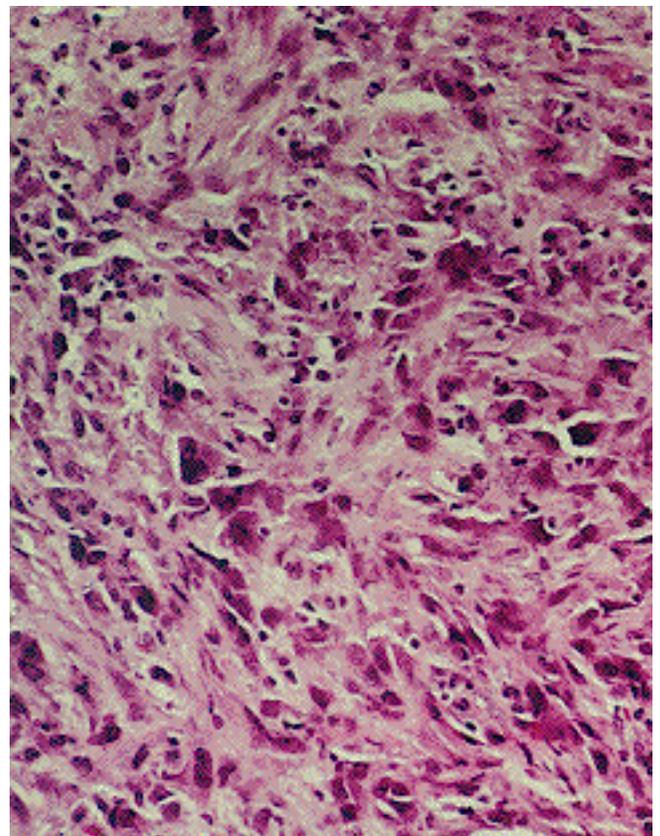
MPNST. 34 year old patient with neurofibromatosis and one lesion growing to the size of a grapefruit in only a few months. In this highly cellular tumor there is streaming or whorling (arrows) of spindle shaped cells pointing to a neurogenic origin. Nuclei are hyperchromatic and pleomorphic. Typically, there are, wavy or comma-shaped nuclei, mitoses, and myomatous ground substance. The distinction between this tumor and fibro-spindle cell sarcoma may be difficult.



MPNST, higher power of photo. Cytoplasm is eosinophilic, cell boundaries poorly defined (arrows), nuclei enlarged, hyperchromatic, pleomorphic, and curved or wavy.



MPNST, patient with neurofibromatosis. Note some suggestion of streaming and whorling. Pleomorphism and hyperchromatism prominent.



**CLINICAL ASPECTS**

The prognosis of MPNST, especially in patients with neurofibromatosis is poor. Some 2 to 15 percent of patients with neurofibromatosis may develop a malignant transformation in one of their tumors, chiefly those deep in the neck or extremities. Superficial lesions rarely become malignant.