

Cutaneous Superficial Malignant Peripheral Nerve Sheath Tumor

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Dear Editor,

Malignant peripheral nerve sheath tumors (MPNST) include a group of tumors that originate from the peripheral nerves or show differentiation along various elements of the nerve sheath, namely, Schwann cells, perineural fibroblasts, or fibroblasts (1). MPNST is rare soft tissue sarcoma with an incidence of 0.001% in the general population and it represents 5-10% of all such tumors with slight male predominance (2-5). One half to two thirds occur from neurofibromas or in the setting of neurofibromatosis type-1. It can increase to 5-42% in patients with neurofibromatosis type-1 (6). MPNSTs behave in an aggressive fashion, and carry a poorer prognosis than other soft tissue sarcomas (7). MPNSTs arise in deep soft tissues of trunk and extremities, but superficial primary MPNST with a cutaneous or subcutaneous origin have rarely been reported.

A 22-year-old male patient presented with a swelling in the right forearm that had been existing since childhood. There was no related trauma, local pain or infection at this localization and he had no significant history associated with neurofibromatosis. Physical examination revealed a painless, non-pulsatile, spherical subcutaneous mass that protruded about 1 cm above the skin surface. The tumor mass was completely resected with part of skin covering on it. The cut surface was greyish white in color and solid. Microscopically, multinodular tumor organization was observed which cover the whole dermis and extending subcutaneous adipose tissue. The tumor consisted of atypical epitheloid cells which some had spindled-shape, had ovaloid-round, hyperchromatic-vesicular nucleus and widely eosinophilic cytoplasm (Figure 1). Regressive changes like necrosis, hyalinization were seen in restricted areas. The mitotic activity was high (30/10 high-power fields) (Figure 2). Because there were no benign features, these findings indicated that the tumor didn't originate de novo or from malignant transformation of neurofibroma. Immunohistochemical studies played

a decisive role in diagnosis. Our case showed S-100, Vimentin, GFAP, CD68 positivity with over-expression of Ki-67 (mean 30%) (Figure 3). Metastatic malign melanoma and monophasic synovial sarcoma were included in differential diagnosis. Immunostaining EMA, CK7 and CD99 for synovial sarcoma; HMB45 and Melan-A for metastatic melanoma were used to distinguish from MPNST. They were all negative. Other markers, such as CK, CK19, Desmin, CA-125, CD31, CD34, Factor VIII didn't expressed in 'MPNST of the dermis' and were used exclude other tumors. By this findings, it was diagnosed as MPNST of cutaneous tissue.

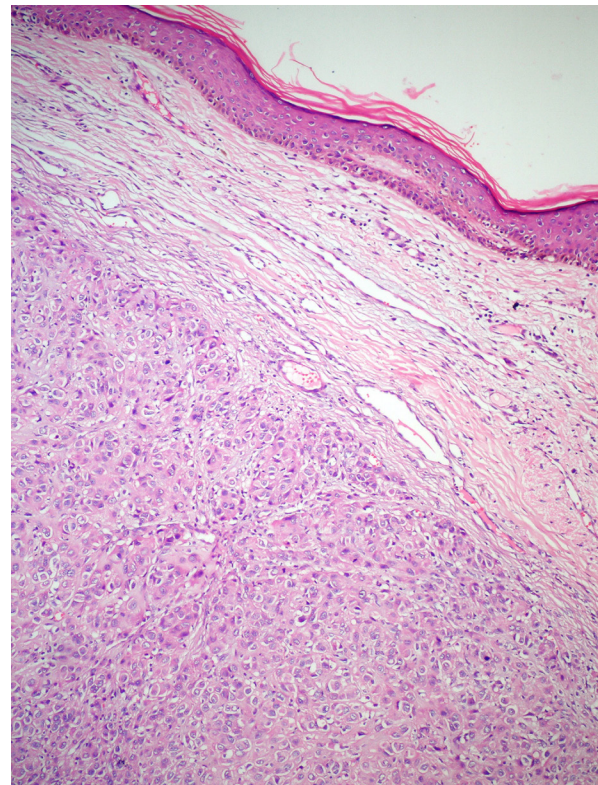


Figure 1. The tumor cells are pleomorphic obviously with ovaloid-round, hyperchromatic-vesicular nuclei and eosinophilic cytoplasm's (H&E, X100).



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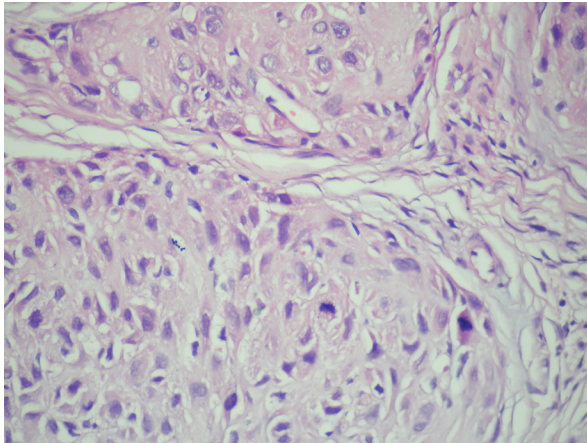


Figure 2. Atypical epithelioid cells are arranged in bundles, with nuclear pleomorphism and mitotic activity (30/10/HPF) (H&E, X400).

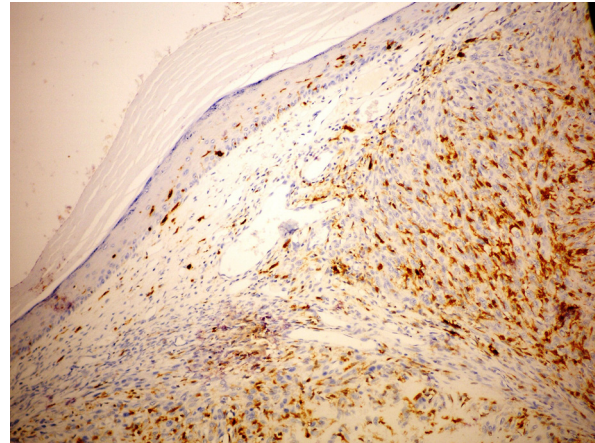


Figure 3. Immunohistochemical staining for S-100 was positive in most spindle shaped cells (X100).

The differential diagnosis between benign schwannoma and neurosarcoma may be challenging: One must look for necrotic foci, the number of atypical mitoses, and an absence of differentiated cells (6). Our case showed features that favor MPNST include hyperchromasia, large nucleoli, increased mitoses and necrosis.

Metastases arise in 39% of patients, lung being the most common metastatic site (6).

The treatment of option is surgery, but postoperative radio- and chemotherapy are part of integrated therapy. Gross total resection of the tumor is the most important therapeutic goal. When radical tumor removal is not possible, excision combined with radiation therapy seems to be the best alternative treatment. With the latest advances in molecular genetics, the target therapy for this tumor type is expected to be discovered (8-10).

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