Malignant Peripheral Nerve Sheath Tumor Arising from Neurofibromatosis

- Electron Microscopic Study -

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A 65-year-old woman complained of a pedunculated, huge mass on the anterior chest associated with neurofibromatosis type 1. The clinical and histological data showed the possibility of a malignant peripheral nerve sheath tumor. On electron microscopy, we found the characteristic findings of a malignant peripheral nerve sheath tumor, such as the foci of entangled cytoplasmic processes, undifferentiated spindle cells having large pleomorphic nuclei with prominent nucleoli, distinct basal cell lamina, rudimentary cell junctions, intermediate filaments and clumps of glycogen in the tumor cells.

We, herein, report a case of malignant peripheral nerve sheath tumor arising from neurofibromatosis, studied by electron microscopy. (Ann Dermatol 8:2(153-157, 1996).

Key Words: Electron microscopic study, Malignant peripheral nerve sheath tumor, Neurofibromatosis

The malignant peripheral nerve sheath tumor (MPNST) is the neoplasm that most often arises in anatomically discernible peripheral nerves or neurofibromas, and shows a wide spectrum of histologic features. This rare tumor usually has a strong association with neurofibromatosis type 1 (NF-1), also known as von Recklinghausen's disease.

The diagnosis of MPNST may be difficult or impossible by light microscopy alone, if the neoplasm is poorly differentiated and there is no clinical evidence for its occurrence from a nerve trunk. Often electron microscopy will be necessary to confirm this tumor.

We report a case of MPNST developing from NF-1 ascertained by ultrastructural study, which will help confirm difficult cases.

REPORT OF A CASE

A 65-year-old woman presented with an asymptomatic, pedunculated, large mass which had abruptly enlarged on the anterior chest over the previous 4 months (Fig. 1). It was dark-brownish, 12.5 × 8.0 × 7.5 cm sized and ulcerated on its surface. On physical examination, multiple, variable-sized, skin-colored or brownish, soft nodules and several well-defined hyperpigmented patches were seen to be distributed on her whole body (Fig. 1). Her family history showed that her two daughters had the same lesions as hers. Neurological examination was unremarkable. Laboratory studies showed mild anemia and hypoproteinemia. Liver function test, renal function test, and other immunological studies, such as immunoglobulin G, A, M, E, and VDRL were within normal limits or negative. EKG and roentgenogram of chest and abdomen showed no definite abnormality. The abdominal CT showed multiple cutaneous nodular masses in the anterior and posterior aspects of

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the abdomen, and mild to moderate biliary ductal dilatations.

Under general anesthesia, the removal of the mass on the anterior chest was performed at the Department of General Surgery. Grossly, the tumor showed a firm, pale-yellow to pink, myxoid cut surface with some foci of hemorrhage, necrosis and ulceration.

The histopathologic findings of the anterior chest mass revealed a fascicular pattern of spindle cells, which had hyperchromatic and elongated nuclei, and a bizarre appearance. The tumor showed hypercellularity, significant nuclear atypia and increased mitotic figures (Fig. 2). In the peripheral area, the typical histologic patterns of benign neurofibroma, that is, interlacing loose fascicles of cytologically benign spindle cells, were also found (Fig. 3).

Immunohistochemical staining for S-100 was positive, but neuron specific enolase was negative.

An electron microscopic examination revealed the foci of entangled cytoplasmic processes (Fig. 4), undifferentiated spindle cells having large pleomorphic nuclei with prominent nucleoli (Fig. 5), distinct basal lamina, rudimentary cell junctions (Fig. 6), intermediate filaments, and clumps of glycogen in the tumor cells (Fig. 7). We failed to achieve a further evaluation of hepatic ductal dilatations because of her refusal.

After the surgical removal of the tumor, she remained without evidence of local recurrence or any new tumor development during the 3 months of the follow-up period.