SKIN CANCER (OTHER THAN MELANOMA)

NEUROFIBROMATOSIS TYPE I WITH MALIGNANT PERIPHERAL NERVE SHEATH TUMOR AND DESTRUCTION OF SKELETON: A CASE REPORT

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Background: Neurofibromatosis type 1(NF-1) is a genetic disease that commonly associated with benign or malignant tumors in both the central and peripheral nervous systems. We present a case of NF-1 with malignant peripheral nerve sheath tumor(MPNST) and destruction of skeleton.

Observation: A 62-year-old female presented with café-au-lait spots for 62 years, and increased nodules for 48 years, augmented for 2 year. Physical examination revealed scattered café-au-lait spots, and extensive skin-colored soft nodules, one huge nodule was on the left side of back with diameter about 15cm, and there was a subcutaneous nodule with diameter about 20cm on left hip. The positron emission tomography computed tomography(PET-CT) scan showed occupying lesion on the left Coxa, with destruction of coxal bone. The histopathologic examination showed normal epidermis, and extensive spindle cell in dermis and subdermis, with pale plasma and slender wave-shaped nucleus, there were areas with rare cells and areas with intensive cells, and atypical cells with trachychromatic nucleus could be seen in all areas. Immunohistochemistry showed that the tumor cells were stained positive for Vimentin, NSE, P53, and partly positive for S-100, and negative for EMA, SMA, Melan-A, CK and Myo, with Ki67 positvie for 15% of the tumor cells. The diagnosis was cutaneous neurofibiromatosis with malignant peripheral nerve sheath tumor (MPNST) and destruction of skeleton. The patient gave up treatment and died 10 months later.

Key message: Neurofibromatosis type I with malignant peripheral nerve sheath tumor and destruction of skeleton is rare clinically. Here we present such a case and so we could know it better.