SKIN CANCER (OTHER THAN MELANOMA)

GIANT TUMOR ON NEUROFIBROMATOSIS TYPE 1, THINK OF A MALIGNANT DEGENERATION

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Introduction: Neurofibromatosis type 1 (NF1) is an autosomal dominant genetic multiorgan disease; it is de novo mutation in 50%. Different tumefaction complicates it. We report the case of suspicious infection on NF1.

Observation: This is a 50- years-old woman who had since the adolescence of cutaneous neurofibromas and plexiformneurofibromas, without any other particular antecedent. Plexiformneurofibroma of the left flank was rapidly increasing in volume, becoming ulcero-necrotic and inflammatory, malodorous. The examination showed on neurofibromas and cutaneous neurofibromas almost all over the body, neurofibromaplexiform in the occiput, vertex and vulva, and a budding swelling, whitish, ulcero-necrotic, firm, pedunculated, hemorrhagic, 210mmx170mm at the left flank. In fundus oculi, nodules of Lish were present. Biopsy of the giant tumor was a chronic suppurative inflammation. Chest radiography and abdominopelvic ultrasound were normal. The patient died before the surgery.

Discussion: The diagnosis of NF1 is based on NIH criteria. A Malign Peripheral Nerve Sheath Tumors (MPNST), a plexiform neuroma can be complicated. Our case is close to the MPNST. The diagnosis is confirmed by histopathologicalexamination with immunohistochemistry. The treatment of reference is exerese with surgical margins with radiotherapy at non-metastatic stage.

Conclusion: The risk of malignant transformation of neurofibromas should be systematically assessed during NF1.

Key words: Neurofibromatosis type 1-Malign Peripheral Nerve Sheath Tumors- Plexiform neuroma- Surgery-Immunohistochemistry