MPNST ex. Diffuse Neurofibroma in a Patient with Unknown History of NF1, A Case Report.

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Abstract:

Malignant peripheral nerve sheath tumor (MPNST) is defined as spindle cell sarcomas arising from or differentiating towards the cells of the peripheral nerve sheath. MPNST is extremely rare in the parotid gland and, in most cases, the prognosis is grim. It usually develops in the setting of Neurofibromatosis type-1 (NF1), but can also develop sporadically. Herein, we report a rare case of MPNST of the parotid gland, in a patient with no previous history of NF1.

A 45 years old patient presented with a non-tender right parotid swelling of one-year duration. A sudden increase in the size of the parotid mass over the past two months alerted the surgeon to proceed to investigate the nature of the mass and the etiology of the sudden change in size. The patient's initial investigations, which included ultrasound imaging of the swelling, fine needle aspiration (FNA), Computed Tomography (CT) scan, and Magnetic Resonance Imaging (MRI) of the neck and swelling were consistent with a benign pleomorphic adenoma of the parotid gland. However, during the surgery the dissection was difficult and the tumor appeared adherent to the surrounding tissue, which lead to a total parotidectomy, in piecemeal!

The histopathology revealed a low-grade spindle cell neoplasm exhibiting a diffuse, infiltrative growth pattern, invading into and around salivary lobules, adipose tissue, skeletal muscle bundles and fascia. The morphology and positive s100 & CD34 stains suggested diffuse neurofibroma. Scattered foci of epithelioid cells with increased pleomorphism and mitotic activity were also evident.

The diagnosis rendered was thus MPNST of the parotid gland arising from a diffuse neurofibroma. The patient was also diagnosed with NF1 based on clinicopathologic correlation with a referral for confirmatory genetic testing. The patient underwent radiotherapy to continue his treatment following the resection of the tumor.

Images:

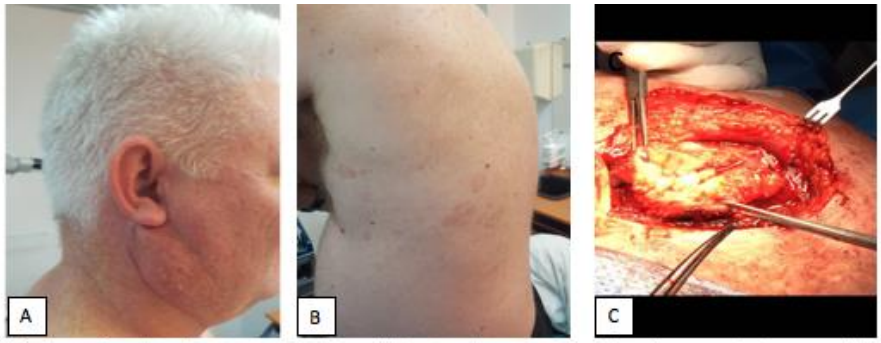

Figure 1. (A-C) Pre-operative clinical photographs showing a right parotid swelling (A) and café au lait macules on the patients torso (B); clinical photograph showing intra-operative view of the parotid swelling (C).

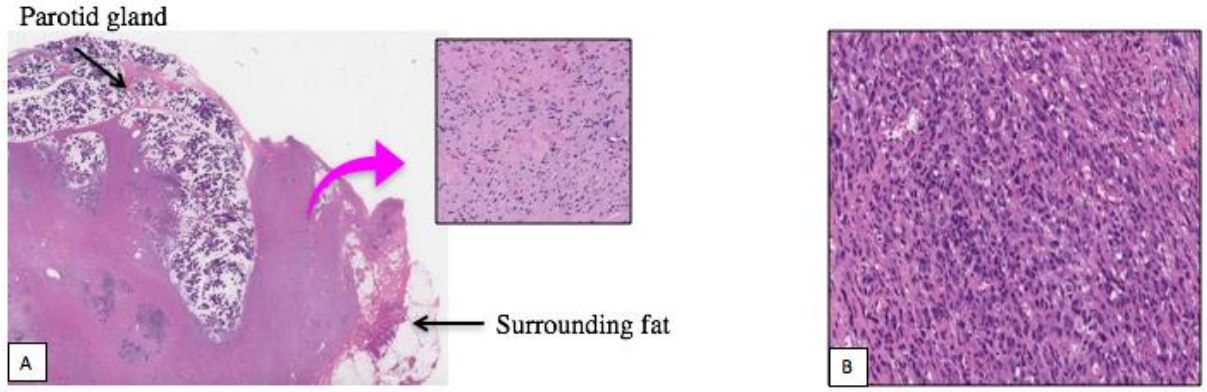

Figure 2. A, Histologically diffuse-type neurofibroma arising in the parotid region with diffuse infiltration between salivary gland lobules (arrow) and surrounding adipose tissue (arrow). Diffuse-type neurofibroma comprises tumor cells with round to fusiform nuclei in a fine fibrillary collagenous matrix (A, inset). B, Areas of increased cellularity with epithelioid cells showing nuclear atypia and mitotic activity indicate transition to MPNST.

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Biography

Mahmoud Ebrahim is a young doctor who just finished his internship year and has passion for otolaryngology and head and neck surgery. He is a hard working doctor at work who gives his maximum to the care of the patient's and there well-being. Outside of work he devotes his time in research and publications and attending courses. He is currently working as an assistant registrar in general surgery and attending ENT electives, as he is building up and strengthening his CV to apply for residency outside his home country.

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