Case Presentation

An unmistakable tumour of the tongue

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Description

A 37-year-old woman suffered from a painful and swelling lesion of the base of the tongue, with no history of smoking or medical history of tumours. On physical examination, she was found to have an ulcerated and painful induration of the base of the tongue, with no other localization. No cervical adenopathy was found. The CT scan was normal. A Biopsy was done. The exact histological diagnosis couldn't be done, and pathologists concluded a malignant proliferation. A glossectomy surgical resection was done. The specimen measured 4.5×3×2 cm. The Tumour size was 3×0.5 cm. It was poorly circumscribed and infiltrated the adjacent tissue. The cut surface was greyish-white. The tumour was composed of a monomorphic population of polygonal to round large cells that have optically clear or pale eosinophilic cytoplasm. The nuclei are round and often eccentric, basophilic and sometimes they contain a small nucleolus. Mitotic figures are scars. These tumour cells are arranged in sheets, cords, and nests in a fibrous stroma, with sometimes a scirrhous appearance due to hyalinized collagenous stroma (Figure 1).

On immunohistochemistry, tumour cells were positive for P40 and negative for PS100, CD10 and Vimentine.

Question

What is your diagnosis?

Replies

The diagnosis was hyalinized clear cell adenocarcinoma (HCCA) of the minor salivary gland origin.

HCCA is a rare tumor of the salivary gland characterized by myxoid or hyaline degeneration of the stroma arranged in trabeculae, nests, and cords [1]. It can appear in all salivary gland sites with squamous differentiation but mostly presents in the oral cavity at the base of the tongue, palate, or parotid glands [2]. It often metastasizes to locoregional lymph nodes [1,3]. HCCA accounts for only 1% to 2% of all primary salivary gland tumors [4]. Women and men are affected nearly equally [5]. This tumor can be misdiagnosed as

More Information

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mucoepidermoid carcinoma (MEC), epithelial-myoepithelial carcinoma (EMC), and metastases from the kidney of clear cell carcinoma, thyroid, or large bowel can be challenging. Hyalinizing clear cell carcinoma is immunoreactive to p63 and cytokeratins (CK5, CK7, CK8, CK14, and CK19) and negative for calponin, S100 protein, smooth muscle actin (SMA), and glial fibrillary acidic protein (GFAP) [1]. HCCA is most commonly associated with the EWSR1-ATF1 gene fusion. HCCA is typically low-grade and not associated with significant disease-related mortality. Treatment includes primary surgical resection followed by radiation. Data are limited for chemotherapy.

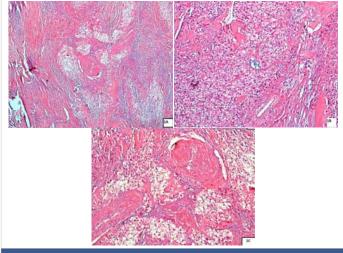


Figure 1: Mitotic figures are scars.



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