

Lymphoepithelial-like carcinoma of the submandibular gland

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Abstract

Lymphoepithelial-like carcinoma (LELC) of the salivary gland is uncommon, with about 80% of these occurring in the parotid gland. Its occurrence in the submandibular gland is very rare. It has a higher incidence in Eskimos and Orientals. Apart from a report about a North-African woman with LELC of the submandibular gland, to the best of the authors' knowledge, there are no other African reports in the English literature. We therefore report the case of a 3-year painless right submandibular swelling in a female Nigerian diagnosed as LELC. The patient was managed by submandibular salivary gland surgical excision with adjuvant chemotherapy and is currently disease free.

Key words: lymphoepithelial-like carcinoma, submandibular salivary gland

Introduction

Lymphoepithelial-like carcinoma (LELC) is a rare malignancy that has been categorised by WHO as an undifferentiated squamous cell carcinoma with a significant non-neoplastic lympho-plasmacytic infiltrate⁽¹⁻⁴⁾. It is histologically similar to nasopharyngeal carcinoma and is only differentiated from it by topography and clinical outcome^(2,5-7). LELC of the salivary gland is uncommon, constituting approximately 0.4% of salivary gland malignancies, with about 80% of these occurring in the parotid gland^(2,8). Its occurrence in the submandibular gland is very rare. In a recent report of 69 cases of LELC, 52 (75.4%) were in the parotid while 17 (24.6%) were in the submandibular gland⁽⁹⁾. Apart from a report about a North-African woman with LELC of the submandibular gland⁽¹⁰⁾, to the best of the authors' knowledge, there are no other African reports in the English literature. We therefore report a case of LELC in the submandibular salivary gland of a female Nigerian.

Case report

A 21-year-old female Nigerian reported at our clinic with a 3-year history of a painless right submandibular swelling. The swelling had been incompletely excised at another health facility some months before presentation via an intraoral approach.

Clinical examination revealed a dome-shaped swelling in the right submandibular region that was covered with apparently normal negroid skin (Figure 1a). The right submandibular lymph nodes were not palpable while those on the left were normal. The swelling was firm in consistency, non-tender, slightly mobile, well circumscribed and measured approximately 8.5 X 6.0 cm.

Intra-oral examination revealed a right-sided floor of the mouth ulcer which measured approximately 2.5 X 1.2 cm (Figure 1b). Incisional biopsy described an undifferentiated non-keratinizing squamous-cell carcinoma, presenting with syncytial cytoplasm, vesicular nuclei, and large central nucleoli with a remarkable lymphocytic inflammatory background. There was infiltration of adjacent fibrous connective tissues and the regional lymph nodes (Figure 2).

The patient was scheduled for surgical excision of the mass and associated gland with functional neck dissection and adjunctive chemoradiotherapy. An endoscopic

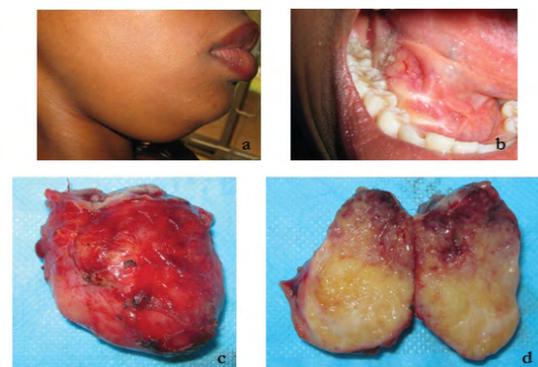


Figure 1. Clinical and Surgical profile showing a: extra-oral submandibular dome shaped swelling; b: ulceration and scarring in the floor of the mouth; c: circumscribed surgical specimen; d: cut section of gross specimen showing a yellowish fleshy mass.

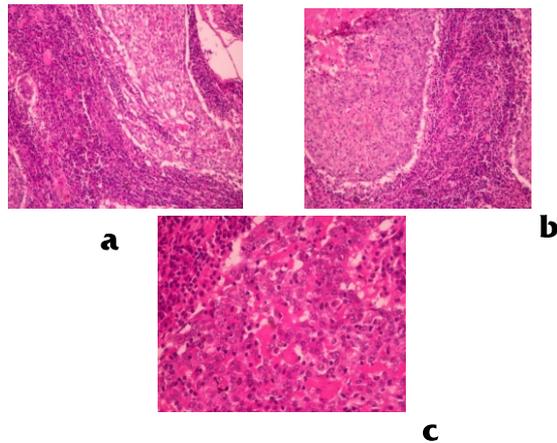


Figure 2. H&E section showing lympho-epithelial features including epithelial cells with syncytial cytoplasm, vesicular nuclei, and large central nucleoli dispersed in a chronic inflammatory background. (a & b X100) © X400

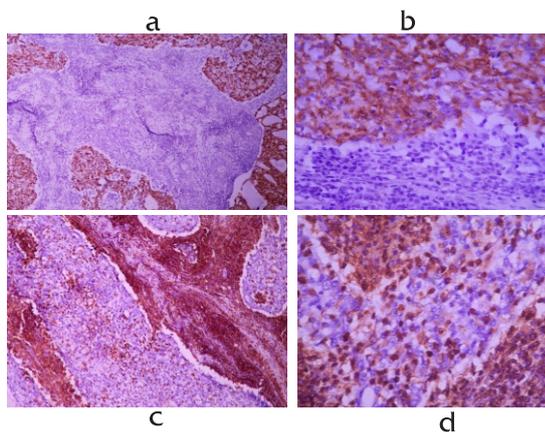


Figure 3: Immunohistochemistry showing a: AE1/AE3 antibody +++ staining intensity of the epithelial component. X100; b: AE1/AE3 antibody +++ staining intensity of the epithelial component. X400; c: LCA antibody +++ staining intensity of the lymphoid component. X100; d: LCA antibody +++ staining intensity of the lymphoid component. X400.

nasopharyngeal examination was also advised to rule out a primary nasopharyngeal carcinoma. The gross surgical specimen was fairly well circumscribed, lobulated, firm in consistency and measured about 7cm in its widest diameter (**Figure 1c**). The cut section surface was fleshy and yellowish in colour (**Figure 1d**). The excised tissue was fixed in 10% neutral buffered formalin and processed for haematoxylin/ eosin stain and antibodies to cytokeratin AE1/AE3. Positive and negative antibodies were employed for the antibodies tested.

Discussion

LELC is a poorly differentiated carcinoma that is composed of nests of neoplastic epithelial cells with indistinct cell membranes interspersed with benign lympho-plasmacytic infiltrate⁽³⁾. It can occur in other regions other than the orofacial tissues and the nasopharynx such as the larynx,

thymus, lungs, stomach, duodenum, breast, renal pelvis, urinary bladder, uterine cervix, ovary, vulva and vagina^(3-5,7). It has a higher incidence in the Eskimos and Orientals and it is usually associated with the Epstein Barr virus especially in Asians^(3,8,11). The viral proteins are found exclusively in malignant epithelial cells and never in the lymphocytes. Also, EBV is not usually identified in LELC of patients from Western Europe and USA⁽¹²⁾. Salivary glands LELC have a predilection for females, with a male to female ratio of 1:1.5⁽³⁾. Our patient is a 21-year-old female. Reports from Africa are extremely sparse in the English literature, thus, conclusive assertions regarding gender and age predominance on the African continent is difficult.

The clinical behaviour of LELC of the salivary gland typifies a benign neoplasm. It is usually painless and of long duration⁽²⁾, as in the present case. Grossly, LELCs are of varying sizes ranging from 1 to 10 cm⁽²⁾, often lobulated and either well circumscribed or locally infiltrative into the adjacent tissue^(2,3). The tumour is firm in consistency and the cut surfaces appear greyish with a mixture of yellowish and creamy colour^(2,3). Mostly all previously documented clinical and morphological estimates were comparable to our findings in this Nigerian case.

Following the diagnosis of a primary LELC in major salivary glands, it is pertinent to rule out a primary undifferentiated nasopharyngeal carcinoma^(2,5). Such assessment in our patient to support a primary LELC of the submandibular gland only without involvement of the nasopharynx could not be made, as she declined further evaluation. To determine metastatic nasopharyngeal carcinoma to the salivary glands thorough examination of the upper aero digestive tract with endoscopy and even random biopsy of the nasopharynx should be done⁽²⁾.

Adequate management of LELC entails surgical resection and radiotherapy^(2,5). Neck dissection is reserved for patients with clinically positive lymph nodes⁽²⁾. It has been suggested that LELC has better prognosis compared to other undifferentiated carcinomas of the salivary gland because the prominent lymphocytic infiltrate limits the aggressiveness of the lesion⁽²⁾. A 5-year survival rate of 50 to 87% has been reported⁽²⁾. Currently our patient is undergoing cycles of radiation therapy and has shown no signs of the disease, although follow-up is still in its early stages.

From existing English literature, we surmise that LELC is rare in African populations or most have gone un-reported or under-reported. Our report therefore is an important addition to literature, which will form part of the construct of a latter case series that should help characterise the neoplasm within the African context.

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