

A Rare Case of Sinonasal Lymphoepithelial Carcinoma in an Adult Male

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Abstract: Lymphoepithelial carcinoma is a histologic variant of squamous cell carcinoma was first reported by Regaud and Schmincke independently. Lymphoepithelial carcinoma mainly occurs in the nasopharynx. Rarely, tumour with same histomorphologic features has been described in the sinonasal tract. Due to the paucity of reported cases, there is no precise detailing of its management. Therefore, we report a case of right sinonasal lymphoepithelial carcinoma in a 56 years old male farmer in Sub Saharan Africa.

Keywords: Lymphoepithelial, Carcinoma, Sinonasal, Africa

INTRODUCTION

Lymphoepithelial carcinoma (LEC), also called lymphoepithelioma, is a histologic variant of SCC that was first reported by Regaud and Reverchonand independently by Schmincke [1]. The World Health Organization (WHO) has defined it as “a poorly differentiated squamous cell carcinoma or histologically undifferentiated carcinoma accompanied by a prominent reactive lymphoplasmacytic infiltrate, morphologically similar to nasopharyngeal carcinoma”[18]. LEC mainly occurs in the nasopharynx. Rarely, tumours with the same histomorphologic features have been described in the Sinonasal tract, oropharynx, salivary glands, oral cavity, larynx, trachea, hypopharynx, lung, thymus, stomach, skin, breast, uterine cervix, vagina, and urinary bladder under a variety of terms: undifferentiated carcinoma of nasopharyngeal type, undifferentiated carcinoma with lymphoid stroma, lymphoepithelioma, lymphoepithelium-like carcinoma, and LEC [1]. In contrast to LEC of the nasopharynx, LEC arising at these other sites, with the exception of the major salivary glands, does not exhibit a close association with EBV except in Chinese patients [1].

LEC is differentiated from nasopharyngeal carcinoma (NPC) by its location and clinical outcome [2, 3]. Sinonasal LECs have been described in literatures either as single case reports or clinical series [4, 5]. Due to the paucity of reported cases, there is no precise detailing of its diagnostic characteristics and standardized treatment policy reported to date for LEC of sinonasal tract [3].

Therefore, we report a case of right Sinonasal LEC in a 56 years male farmer in sub-Saharan Africa.

CASE REPORT

A 56 years old male farmer presented to the outpatient unit of department of ENT surgery with 2 years history of right sided constant nasal blockage, occasional epistaxis, right cheek swelling and pain, right upper jaw tooth ache. There was no associated history of snoring, otologic symptoms, neck swelling, neuro-ophthalmic symptoms, headache, cough, or weight loss. He neither smoked cigarette nor ingested alcohol.

Examination revealed elderly looking, preserved with non tender right cheek mass, hard and fixed to the underlying structure, overlying skin appeared normal. A fungating mass arising from the lateral wall of the right nasal cavity, with contact bleeding was noted in Fig. 1a. A neck examination revealed no palpable lymph nodes and no eye signs.

Plain X-ray of the paranasal sinuses showed complete opacity of right maxillary sinus, medial wall was pushed into the right nasal cavity. The bony walls were intact. Other routine laboratory investigations including chest X-ray were normal.

Examination under anaesthesia of the nose, biopsy specimen was obtained from the right nasal cavity and maxillary sinus. Right intranasal mass was cleared. Histological examination revealed lymphoepithelial carcinoma arranged in sheets and intermingled with lymphocytic infiltrate as in Fig. 1b. He was referred for chemoradiation at different centre.



Fig. 1: (a) Showing right sinonasal fleshy mass with contact bleeding, (b) Photomicrograph showing lymphoepithelioma (H&E X 220)

DISCUSSION

LEC has its highest worldwide incidence in the people of Southeast China, Southeast Asia, the Arctic regions, and Malaysia [6, 8]. Regions of occurrence have been designated as high incidence (e.g., South China province of Kwantung and Hong Kong), intermediate incidence (e.g., North Africa), and low incidence (e.g., Europe and the United States) [1].

Moreover, LEC is the most common type of nasopharyngeal cancer in young people (>90%) [9].

Other features specific for LEC are occurrence at a younger age than other head and neck SCCs, absence of a strong alcohol or tobacco etiologic relationship as in our case, lack of substantial risk for a second primary tumour (1.3%), and a very high rate of systemic dissemination [10].

Sinonasal LEC is very rare with a ratio of 1:564 to that of nasopharynx [11]. Sinonasal LEC is commonly diagnosed in patients between 40 and 70 years and also has a male predilection with a male to female ratio of 3:1 [1]. Our patient falls into this group. Recurrent nasal bleeding and nasal obstruction are main presenting symptoms with other non specific Sinonasal complaints related to acute or chronic rhinosinusitis [4, 5], as in this index case. Sinonasal LEC spread locally and none had neck metastasis from the previous reports [4], which is similar to our case. However the LEC developing from the sinonasal tract can present as an aggressive tumour with signs of local invasion to nerves, orbit, or the nasopharynx [12]. All these features were absent in our case.

In general, the routine laboratory tests are normal. In terms of imaging, LEC of sinonasal tract appears as a diffuse opacity of soft tissue density on standard radiography [13], as in our case. Paranasal sinus CT scan highlights a solid homogenous mass occupying the maxillary sinus cavity that is not enhanced with an intravenous contrast injection [14] which was not available at our centre at the time of

diagnosing the case. There are no pathognomic radiological criteria that differentiate the LEC from other tumours of the maxillary sinus, especially squamous cell carcinomas or lymphomas [5].

In general, clinicoradiological tests are non-contributory to the final diagnosis of LEC, although assist in determining extent of the disease. Histology and immunohistochemical analysis establish the definitive diagnosis of a LEC. The microscopic features of this tumour show epithelial cells with eosinophilic cytoplasm. Their large oval nuclei have fine vesicular chromatin with one to three prominent red nucleoli. The fibrous stroma is heavily infiltrated by plasma cells and lymphocytes. Individual tumour cells may be surrounded by the mixed infiltrate, resembling Hodgkin's disease [15]. The presence of non-caseating granulomas negative for acid-fast bacilli, sarcoid-like granulomas, and localized amyloid has been reported in the adjacent stroma [16].

Immunohistochemical staining shows that epithelial cells of LEC stain positively for pancytokeratin marker (MNF 116) and MIB-1 and stain negatively for CK 5/6, CK 7, and CEA. There is no immune reactivity for melanin A marker and CK 20 [1].

Due to the frequently not noticeable epithelial nature of undifferentiated carcinoma and its most common presentation is metastases to cervical lymph nodes, the differential diagnosis is diverse, and include Sinonasal undifferentiated carcinoma, Hodgkin's disease, large cell lymphoma, lymphoid hyperplasia, and melanoma. Capsular fibrosis and dense bands of collagen entrapping discohesive tumour cells are histologic findings that may be common to Hodgkin's disease and lymphoepithelioma [15].

However, LECs were subdivided into two histologic types: Regaud type (clusters, nests, or aggregates of neoplastic epithelial cells with lymphoid elements) and Schmincke type (dispersed

tumour cells forming a syncytial net beneath an inflammatory infiltrate [17]. These two types were essentially descriptions of two growth patterns of an undifferentiated carcinoma. Familiarity with these variations in the histomorphology of undifferentiated carcinoma is useful, particularly when the examiner is confronted with a small biopsy of the primary tumour or is evaluating a metastatic deposit in a cervical lymph node with an occult primary tumour. Designation of LEC as a Regaud or a Schmincke type does not have prognostic significance.

The initial treatment for maxillary sinus tumours has always been surgery. Lymphoepithelial carcinoma is known to be radio-sensitive [13].

CONCLUSION

Sinonasal LEC is a very rare malignant tumour with few cases reported; these pose a greater challenge in its management. A strong suspicion and the systematic use of various immunological tests, histopathological examination and immunohistochemical studies will help at a definitive diagnosis by excluding other tumours. The treatment consists of surgery and radiotherapy with the possibility of achieving acceptable cure rates.

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