

Lymphoepithelial carcinoma of the maxillary sinus: A case report

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ABSTRACT

Lymphoepithelioma-like carcinoma or lymphoepithelial carcinoma (LEC) of the maxillary sinus is an extremely rare malignancy. It is a rare type of undifferentiated carcinoma, consisting of atypical epithelial cells surrounded by reactive lymphoplasmacytic infiltrate. The paper reports the case of a 76-year-old male patient, with complain of swelling on the hard palate and epistaxis. Biopsy from the hard palate mass showed features consistent with LEC, with contrast-enhanced computed tomography nasopharynx and neck showing soft-tissue lesion in the right maxillary sinus. The patient is currently under treatment with external beam radiotherapy.

Keywords: Lymphoepithelioma, Maxillary sinus, Nasopharynx, Undifferentiated

The term lymphoepithelioma was first used in 1921 by Regaud and Schmincke to describe certain nasopharyngeal tumors [1]. The incidence of sinonasal cancers is < 1% of all malignant neoplasms, and only 3% of all head and neck cancers. Lymphoepithelial carcinoma (LEC) consists of poorly differentiated squamous cell carcinoma or histologically undifferentiated carcinoma with prominent reactive lymphoplasmacytic infiltration [2]. Most common sites for LEC are the nasopharynx, oropharynx, hypopharynx, oral cavity, larynx, and salivary glands, with LEC occurring in the sinonasal tract being a rare entity, especially in the maxillary sinus [3]. It is generally asymptomatic or may present with non-specific obstructive nasal symptoms. It is strongly associated with Epstein Barr Virus (EBV) and Human Papilloma Virus [4]. Radiation is the treatment of choice for LEC, even when there is lymph node involvement [5].

In this report, we present a case of an Indian male with LEC of the left maxillary sinus with regional nodal involvement and review the radiological, macroscopic, and microscopic findings with the available treatment options.


CASE REPORT

A 76-year-old male patient attended the Department of Radiation Oncology, Regional Institute of Medical Sciences with complaints of swelling inside the oral cavity on the hard palate for the last 4 months and epistaxis for the past 20 days. The swelling was

initially small in size, but gradually increased in size over the course of 4 months to attain a size of 2.0 cm × 2.0 cm with development of multiple ulcers on the right side of the hard palate. The patient also complained of 2–3 episodes of epistaxis in the last 20 days. There was no history of cheek pain, chronic sinonasal infection, coughing, nasal congestion, cheek swelling, nasal obstruction, headache, visual disturbance, or trauma. There was no family history of malignancy or similar illness in the past. The patient gives a history of cigarette smoking (3–4 sticks/day) for the past 30 years (i.e., 6 smoking pack-years). The patient is on anti-hypertensive medication for the past 5 years.

On examination, the patient has an average built with the good general condition. His Body Surface Area was 1.7 m² and had a Karnofsky Performance Score of 90%. On extraoral examination, there was no cheek swelling, no facial asymmetry, or tenderness. Intraoral examination showed a single swelling of size 2.0 cm × 2.5 cm on the right side of the hard palate with irregular surface and margin, tender on palpation, along with multiple ulcerative spots on the hard palate and soft palate, with poor oral hygiene (Fig. 1). The oral hygiene of the patient was poor with staining of the teeth and gums and foul-smelling breath. There were no missing or sharp teeth.

Two neck nodes were clinically palpable, one on the right side of the neck, level 3 (1.0 cm × 1.0 cm in size, hard, mobile, non-tender), and other on the left side of the neck, level 2 (1.5 cm × 1.0 cm in size, hard, mobile and non-tender). No other lymph nodes or swelling were clinically palpable. Routine baseline investigations (Complete blood count, biochemistry, and electrocardiogram) were done and were found to be uneventful.

Access this article online	
Received - 11 April 2021 Initial Review - 29 April 2022 Accepted - 14 May 2022	Quick Response code 
DOI: 10.32677/ijcr.v8i5.3439	

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The metastatic workup (Chest X-ray and ultrasound whole abdomen) found no sites of metastasis or any organ involvement.

Contrast-enhanced computed tomography of the neck and nasopharynx showed a soft tissue lesion in the right maxillary sinus with an erosion of the bony wall of the maxillary sinus, the upper alveolar margin of the maxilla, pterygoid process, and hard palate of the right side. The nasal septum was deviated to the left side with an antero-choanae polyp in the right maxillary sinus, along with cervical lymphadenopathy (Fig. 2).

Biopsy of the hard palate swelling showed acanthotic, focally squamous keratinized epithelium with areas of ulceration covered with acute inflammatory exudate. The sub-epithelium showed a malignant tumor with lymphocytic stromal infiltration. All these features were consistent with the diagnosis of LEC of the maxillary sinus (Fig. 3a). Fine-needle aspiration cytology from the cervical lymph nodes showed metastatic poorly differentiated carcinoma. Immunohistochemistry (IHC) was positive for cytokeratin (Fig. 3b).

The patient was planned for External Beam Radiotherapy (EBRT) with Cobalt-60 teletherapy machine to the head and neck area, to a total tumor dose of 7000 cGy in 35#, 5 days a week for 7 weeks, to the right maxilla (primary site) by two fields

(antero-posterior and Right lateral) and a total dose of 6000 cGy in 30#, 5 days a week for 6 weeks, to the neck (secondary lymph nodes) by two parallel lateral opposed fields. The patient is currently under treatment with EBRT.

DISCUSSION

LEC of the maxillary sinus is a very rare malignant tumor [6]. It is more common in males, between the fifth and seventh decades of life, with a median age of 60 years [7]. LEC most commonly involves the nasopharynx, salivary glands, and larynx and it rarely affects the lungs, esophagus, skin, cervix, vulva, kidney, bladder, and central nervous system [1].

The clinical findings and symptoms vary according to the type, site, and stage of malignancy. They are generally asymptomatic and are discovered incidentally by imaging [8]. Patients may present with complaints of nasal obstruction, facial pain, or headache in the early stages, and in the advanced stage, the patients may complaints of epistaxis, postnasal discharge, or cranial nerve palsies. There is a strong association between LEC and EBV. The presence or absence of EBV has no importance in the prognosis of LEC of the maxillary sinus [9].

Differential diagnosis of LEC includes nasopharyngeal carcinoma (undifferentiated type), lymphoma, melanoma, and sinonasal undifferentiated carcinoma (SNUC). SNUC is a highly aggressive tumor and is a diagnosis of exclusion. Thereby, making it difficult to have a proper pre-operative diagnosis. On IHC, LEC stains positive for cytokeratin, CK 5/6, EMA, p40, p63, and negative for melanoma markers, hematolymphoid markers, and neuroendocrine markers [10].

There are no pathognomic radiological features for the diagnosis of LEC of the maxillary sinus, thereby making a clinico-radiological diagnosis difficult [11]. Definitive diagnosis of LEC is based on the histopathological and IHC analysis. Computed tomography and magnetic resonance imaging are important to assess the tumor extent and for proper treatment planning [12]. LEC generally spreads locally and even though the local spread is common at the time of diagnosis, the prognosis is very good [12]. On imaging, LEC of the maxillary sinus appears as a diffuse opacity of soft-tissue density [13].

Because of the rarity of these tumors, anatomic complexity of the location, and close proximity to critical structures, there is no standard treatment protocol for the management of LEC



Figure 1: Intraoral photo of the patient showing a mass on the right side of the palate with multiple ulcerative spots



Figure 2: Contrast-enhanced computed tomography of neck and nasal cavity (coronal view) showing a soft-tissue lesion in the right maxillary sinus with bone erosions

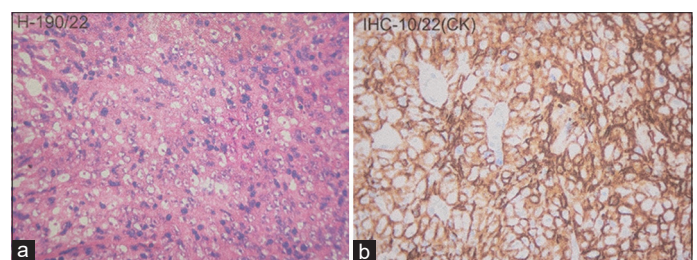


Figure 3: (a) Histopathological examination of the hard palate swelling showing features consistent with the diagnosis of lymphoepithelial carcinoma of the maxillary sinus; (b) immunohistochemistry report showing cytokeratin positive

of the maxillary sinus [11]. Surgery is the initial treatment of choice, but sometimes it is not possible due to tumor size, medical history, and age of the patient. As LEC is radiosensitive, radiotherapy is the cornerstone of treatment, even when there is lymph node involvement. Chemotherapy can be used as a neoadjuvant, concurrent, or adjuvant to radiotherapy. For patients with advanced and extensive disease, adjuvant chemotherapy and neck management can be considered [11].

The rate of recurrence of LEC of the maxillary sinus is around 25%, thus, regular follow-up and periodic CT scan are recommended [11]. The prognosis of LEC is very good for patients with the localized disease with a 5-year survival rate of 78% and progression-free survival rate of 69% [7].

CONCLUSION

LEC of the maxillary sinus is a rare type of malignant undifferentiated carcinoma. Histopathological analysis and IHC are required for proper diagnosis of LEC. Our patient was diagnosed with LEC of the maxillary sinus on the basis of HPE and confirmed with IHC analysis. The patient is currently under treatment with radiation therapy.

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Funding: None; Conflicts of Interest: None Stated.

How to cite this article: Mahawar R, Devi YS. Lymphoepithelial carcinoma of the maxillary sinus: A case report. *Indian J Case Reports*. 2022;8(5):154-156.