American Journal of Cancer Case Reports

American Journal of Cancer Case Reports
http://ivyunion.org/index.php/ajccr
Vol.*, Article ID 201400372, 5 pages

Case Report

Epstein Barr Virus negative Lymphoepithelial Carcinoma Involving the Maxillary Sinus: A Rare Case Report

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Abstract:

Introduction: Lymphoepithelial carcinoma (LEC) is one of the rare malignancies involving the head and neck region. It is common in Eskimos and Chinese population with Epstein Barr virus association. Standard treatment protocol is not in place owing to the rarity of this tumor.

Case Presentation: The present case is a LEC involving the right maxillary sinus not associated with Epstein Barr Virus (EBV) and first of its kind presentation in the Indian population. Lack of awareness, nonaffordability and poor patient compliance resulted in poor response to radiotherapy. The patient succumbed within fourteen months of diagnosis.

Conclusion: Cancer treatment is still not at reach to the poor and the illiterate. Epidemiological studies targeted at genetic basis of origin will be beneficial in identifying rare, and unusual presentation of the disease. We believe that this case will highlight the importance of risk assessment and early diagnosis of cancer at mass level.

Keywords: Lymphoepithelial carcinoma; Nasopharyngeal carcinoma; Immunohistochemistr; Ohngren's line; Gene migration

Reviewers: Hosni Khairy Salem, MD, Urology Dept., Cairo University, Egypt; Sevim Ozdemir, MD, Department of Radiation Oncology, İstanbul University, Turkey

Received: April 21, 2014 **Accepted**: June 11, 2014 **Published:** July 4, 2014

Competing Interests: The authors have declared that no competing interests exist.

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Introduction

Among the major malignant tumors involving the paranasal sinuses, Lymphoepithelial Carcinoma (LEC) is rare [1]. First mention of malignant Lymphoepithelial lesion as a counterpart of the Benign Lymphoepithelial Lesion (BLEL) was made by Schminke in 1921 [2]. 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" [3]. Based on histopathology, Nasopharyngeal carcinomas (NPC) are divided into well-differentiated squamous carcinoma (WHO Type I), non-keratinizing squamous carcinoma (WHO Type II), and Lymphoepithelial carcinoma (WHO Type III), where non-keratinizing squamous carcinoma cells are mixed with numerous benign lymphocytes. Type III cancers respond well to treatment, but also have the greatest propensity for developing distant micro metastases [4].

Epistaxis develops as a clinical manifestation when the mucosa is ulcerated or tumor extends in to the sinus wall. Tumors involving the ethmoid, maxillary, or frontal sinuses may cause proptosis, restriction of eye mobility, diplopia or loss of vision. Epstein - Barr virus is strongly associated with NPCs as an etiological factor [5].

Case Report

A 45 years old female patient presented with a complaint of pain and swelling in the right middle third of the face since two months. Pain was severe, continuous and aggrevated during night. She also reported bleeding from the nose since 45 days.

On inspection extraorally, the swelling was 4x4 cms, diffuse; nonfluctuant, fixed, and tender to palpation. It was soft to firm in consistency, and the skin overlying the swelling was normal.

Intraoral examination revealed missing 14, 15, 16, and 17. Buccal vestibular obliteration was present in the premolar-molar region in the right maxilla. The swelling was firm to hard in consistency and tender. Aspiration was done which revealed dark tinge blood.

There was no significant medical history and no history of drug allergy. Neck examination was done, and no lymph nodes were palpable. Based on the history of epistaxis, long standing swelling and nonodontogenic origin of pain, a provisional diagnosis of malignancy involving the right maxillary sinus was given.

The patient was advised PNS radiograph followed by complete blood picture and CT-PNS view. Platelet count was elevated (415,000 cells/mm3), increase in total leucocyte count (11,800 cells/mm3), and elevated neutrophil count (79%). ESR was also elevated; 79 in the first hour and 140 in the second. Hb% was just below normal (11gm/dL). PNS radiograph revealed a hazy mass in the right maxillary sinus with evidence of irregular bone destruction. Plain CT scan showed a heterogeneously enhancing soft tissue mass of 3.5x3 cms size in the right maxillary sinus with evidence of irregular bone destruction of the anterior and posterior walls of the right maxillary sinus. Central necrosis of the tumor mass was evident with the hypodense area at the center and the medial side of the tumor mass [Fig. 1]

Incisional biopsy was performed. Histopathological examination revealed infiltrative tumor growth composed of pleomorphic round to large, as well as spindle shaped epithelial tumor cells. Areas of squamous differentiation, clear cell changes as well as foamy sebaceous metaplasia were also seen in the tumor nest. Numerous typical as well as atypical mitotic figures were evident. Areas of germinal centers in between tumor islands were seen.

Immunohistochemistry was performed; the cells were found to be diffusely positive for Pan CK [Fig 2], intensely for Vimentin, moderately for SMA, and negative for Myogenin and CD-34.

Chromogenic in-situ hybridization was done with negative result for EBV [EBER]. Based on immunohistochemistry, it was finally diagnosed as Lymphoepithelial carcinoma of right maxillary antrum.

The patient was referred to a local cancer hospital for management. She was subjected to external beam radiotherapy by antero-lateral portals on Cobalt 60 machine with a dose of 50Gy over 25 fractions at 2Gy per fraction from 22nd February 2013 to 1st April 2013. She discontinued the treatment as she couldn't bear the treatment expenses. She died on 5th March 2014. At the time of death she had stiffness of the neck multiple enlarged, matted lymph nodes on the right submandibular and the jugulo-digastric region and was also cachexic.

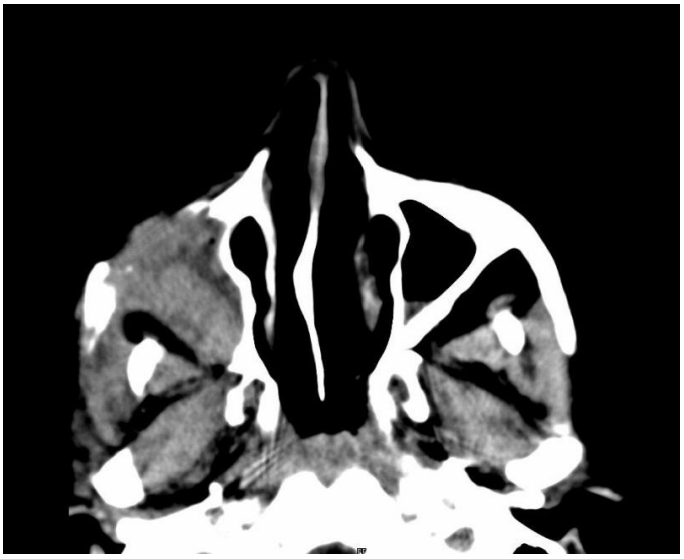

Figure 1 Axial CT Scan Image showing heterogeneous mass in the right maxillary sinus with necrosis

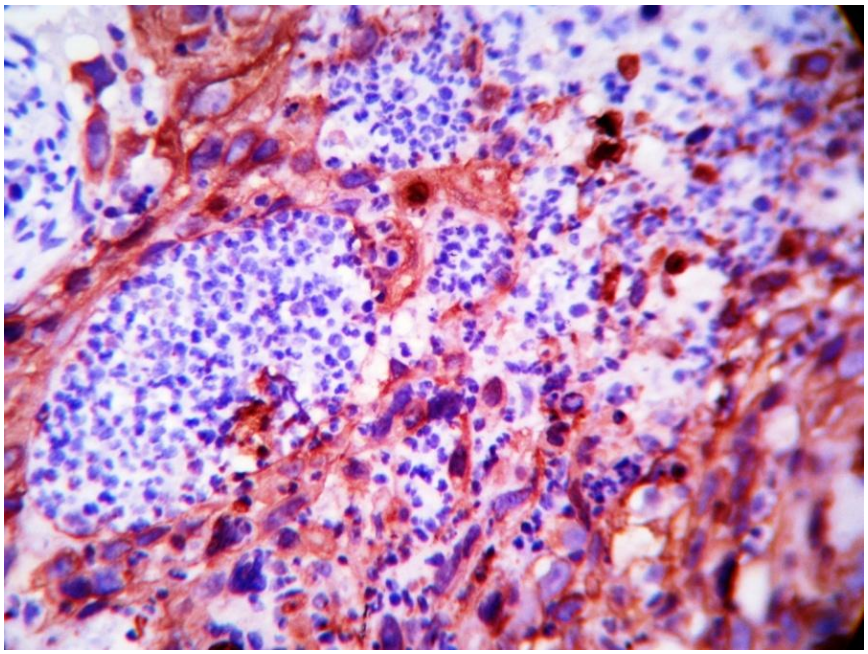

Figure 2 Pan CK positive Epithelial Island

Discussion

Incidence of LEC in Chinese population was reported to be 80 times higher than in the Western world. Certain HLA antigen types, diet, and EBV infection have been implicated as the etiological factors for the development of this tumor [6]. Extensive search through the medical literature has shown no case report from India. We therefore state that this is the first case report of LEC in Indian population. The probable etiological factor that might have contributed to this first appearance in Indian population might be Gene Migration [7]. Surgery and radiotherapy were the treatment modalities in all the previous case reports. Radiotherapy alone was opted in our case as the treatment modality owing to its radiosensitivity and N0 neck clinically and on contrast enhanced computed tomography (CECT).

One study has assessed the prognostic significance of maximum primary tumor diameter (MPTD) in nasopharyngeal carcinoma. A small MPTD of ≤3 cm was associated with a 5 year Local Relapse Free Survival (LRFS) ranging from 88.9% to 100% [8]. The location of the tumor in this patient was suprastructure or superoposterior to the Ohngren's line which is not favourable for a good prognosis [9].

Male patients with NPC are more at risk of distant metastases, have inferior overall survival and disease progression-free survival rates and a shorter interval before therapeutic failure than the female counterparts [10]. Size of the tumor mass being 3.5 cm, with central necrosis and involvement of the posterior wall of the maxillary sinus, prognosis was assessed to be poor. In general, malignancies involving the maxillary sinus extend along the planes with least resistance that is the nasal cavity and the orbits [11]. In this case the tumor must have involved the skull base which has led to stiffness of the neck at the time of death.

The survival data of patients with LEC as was reported in a study was 2yrs, 5yrs, and 10yrs with 91%, 66%, and 29% respectively [2]. Cancer treatment is still not at reach for the poor who also lack the awareness regarding the significance of early cancer detection and thereby timely intervention by the oncologic team. Educating the poor and the illiterate people in countries like India is the need of the hour to record a decline in the cancer morbidity and mortality.

We believe that this case report would definitely change the way cancer is studied at population based level. Many etiological factors are being studied correlated with the incidence of cancer, but little importance is given for the genetic basis of disease origin during epidemiological studies. If the genetic defects are studied, it is obvious that the disease process in rare incidences can be studied more aggressively, eventually may help plan preventive programs.

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