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Case Report

Primary Salivary Gland Neoplasms (Malignant) Epithelial Myoepthial Carcinoma: A Case Report

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Abstract

Epithelial myoepthial carcinoma (EMEC) is rare, low grad tumor with epithelial and myoepithelial components and frequent local recurrence, also called glycogen rich adenoma (clear cell pattern), myoepithelioma (spindle and plamocytiod patterns), myoepithelial carcinoma (if only myoepithelial differentiation and marked cytologic atypia). Occurs most frequently in the major salivary glands and accounts for approximately 1% of all salivary gland neoplasms that usually occurs in parotid gland but can be occur in variety of sites such as the nasal cavity, paranasal sinus and base of the tong. In fact, due to the rarity of the epithelial myoepithelial carcinoma,there is no uniformity of data in the literature band vary different therapeutic strategies have been suggested.

Keywords: salivary gland; malignant epithelial; myoepthial carcinoma

Introduction

Epithelial-Myoepthial Carcinoma (EMC) is locally aggressive, low-grade tumor of duct -like structures composed of two cell type: small, inner duct lining cells and larger, peripheral myoepithelial cells.it represent 1% of all salivary gland neoplasm and over 75% occur in the parotid gland. Epithelial-myoepthial carcinoma EMC is histopathological term used to describe the biphasic pattern of clear staining of the myoepithelial cells surrounding variable proportions of the ducts with an epithelial lining [1]. EMC is a rare neoplasm, first described in 1972 by Donath et al [7]. Case Report: A 54-year old male patient, smoker and chawing Qat presented with a chief complain of flu (rhinorrhea) 4months follow by swelling and ulcer on the right side of his cheek, no trismus or respiratory distress was present. at 17/9/2019 PNS CT scan done showed malignant of right maxillary bone lesion mass destructive bone.

Biopsy taken showed epithelial myoepithelial carcinoma (spindle cell variant). Lab investigation with normal variant, tumor marker CEA: 3, CA19.9: 0.6. 19/9/2019 start chemotherapy with docetaxel +cisplatin receives one cycle than go for operation for wide excisional, no lymph node dissection.

Pathology

Mass is composed of multiple components with epithelial-myoepithelial carcinoma begin dominant 80% of tumor mass with minor component of variant adenoid cystic carcinoma and basal cell carcinoma, immunohistochemistry not done. than receive two cycles chemotherapy (docetaxel+ cisplatin) evaluation done CT scan neck +chest in 15/10/2019 showed residual mass at site of operation advised radiotherapy 60Gy/35 fraction, evaluation by CT scan in 4/3/2020 showed mild progression course irregular cicufertial soft tissue tumor thickening 16mm multifocal bony destructive and remolding including massive tumor infiltration Second line chemotherapy gemcitabine+ carboplatin AUC 5 received 6cycles with good response finish in 7/7/2020(figure1)

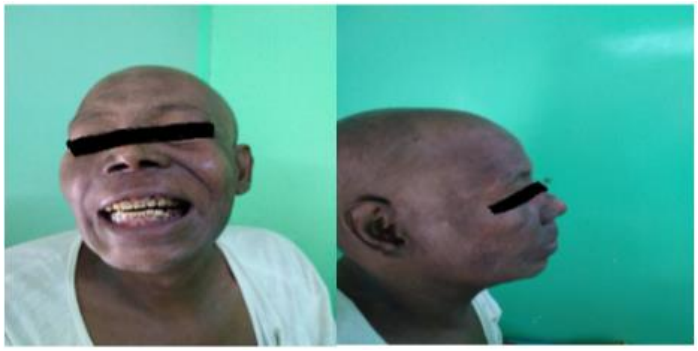

Figure 1: *post chemotherapy image of the patient*

After 3months CT scan 25/9/2020 show progression, patient not tolerated to chemotherapy developed sever neutropenia with mouth fungal infection (figure2).

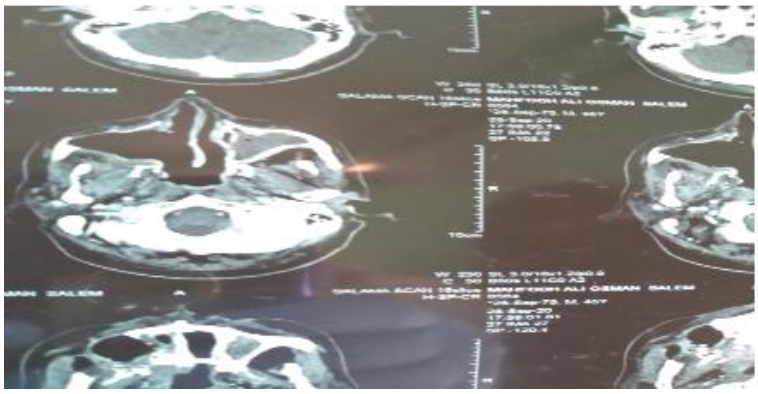

Figure 2: *CT scan: axial cut of upper neck with IV contrast showing destructive mass*

Start monotherapy with methotrexate weekly for 3weeks than progression with protrusion mass from nose with trismus with pus discharge from mass (figure 3).

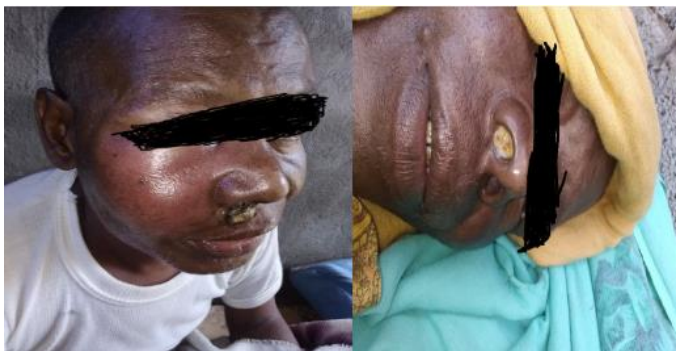

Figure 3: *image of the patient showing protrusion mass through nose.*

change to other line of chemotherapy with CAP (cyclophosphamide +doxorubicin +cisplatin) protocol receive one cycle but progression with massive mass protruded from nose and mouth with sever bleeding figure (4), patient receive one cycle paclitaxel 80mg/m2weekly than he refuses to continue chemotherapy, still complain bleeding from mouth and difficulty in swallowing and breathing, patient refuse tracheostomy escape to home, patient died due sever bleeding per mouth and difficulty breathing.

Figure 4: *Image of the patient with mass coming out nose and mouth with bleeding*

Discussion

There are three major salivary glands – parotid, submandibular, and sublingual-aswell as innumerable minor salivary glands distributed throughout the mucosa of the oral cavity and nasopharynx. All these glands are subject to inflammation or to the development of neoplasm [9]. These glands give rise of over 30 histologically distinct tumors [9]. Epithelial myoepithelial carcinoma is an extremely rare disease, first reported by Donath et al .1972 and officially classified by the world health Organization in 1991 [7,8]. The median age of occurrence is 60.4 years. [2, 3]. The prognosis of EMC is fairly good as the median disease survival rate is reported to be 11.34 years [2]. Long term follow-up is needed for detection of early signs of recurrence and proper management. Surgical excision with wide margins is the treatment of choice. Radiation therapy may be considered for tumors where resection involve significant cosmetic or functional deficit. Patients have 5- and 10-years survival rate of about 87.1 and 67.5% respectively [4]. Neck node dissection may be considered in patients with lymph node positivity in advanced or surgically non-respectable cases along with radio-Chemotherapy [5, 6].

With regarded to recurrence, in keeping with its low-grade malignant nature, a recurrence rate of EMC of 35-50% has been reported, with metastatic rate of 8.1-25% [10]. The present patient has shown sign of recurrences during 2months post -radiotherapy.

Conclusions

Epithelial-myoepithelial carcinoma is an uncommon biphasic malignancy most commonly seen in the parotid and submandibular glands. Although epithielial- myoepthelial carcinoma is rare in occurrence have immense destructive capabilities. Surgical treatment appears to be the mainstay of treatment, with radiation reserved for positive or close margins or patients who are not surgical candidates or who refuse surgery. The important of this case lie in rarity to see simultaneous different cancer of parotid gland and to know that treatment for such case is to words the tumor with the most aggressive type.

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