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Case Report

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Clear Cell Variant of Mucoepidermoid Carcinoma Involving Palate

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ABSTRACT

Salivary gland carcinomas have clinical divergence with complicated overlapping histological patterns; Mucoepidermoid Carcinoma (MEC) being the most common among them. It accounts for about 5-10 % of all salivary gland tumors and 35.9% of all minor salivary gland neoplasms. Rarely these tumors occur in children and young adolescents. Clear Cells predominant MEC (cMEC) is a unique variant which is often difficult to distinguish from other clear cell tumors. We hereby report a rare case of Clear Cell variant of MEC involving palate in a 13-years-old female.

Key Words: *Mucoepidermoid carcinoma, Clear cell, Palate*

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INTRODUCTION

Salivary gland malignancy is a rare entity comprising less than 3% of head & neck cancers [1]. Out of which, Mucoepidermoid Carcinoma (MEC) is the most common type [2] affecting both major (60%) and minor salivary glands (35%) [3]. Intra-orally it has a higher predilection for palate [4] followed by buccal mucosa, antrum, tongue, gingiva, floor of the mouth & nasal cavity [3]. MECs most commonly occur in third to fifth decades of life with slight male predominance i.e.3:2 (male: female) excluding tongue, retrolomar and intraosseous lesions where female predilection is noted [3]. It was first described by Masson and Berger in 1924 & in 1945, Stewart at al. defined it as a separate entity [5]. The term MEC is an acronym of the original designation "mixed epidermoid and mucous-secreting carcinoma" [3]. Based on various clinical and histopathological characteristics in 2005 & 2017 WHO recognised MEC as a malignant glandular epithelial neoplasm consisting of epidermoid, intermediate and mucus producing cells, originating from pluripotent reserve cells of salivary gland ductal excretory system [5, 6]. Occassionally, some other types of cells could be noted such as clear cell, spindle cell, sclerosing cell, oncocyte etc. Calcification within a MEC was also reported in few cases [2]. Clear cells may be found scattered in the tumour, however, large areas predominantly composed of clear cells may also be noted [3].

CASE REPORT

A 13 years old female patient reported to the Department of Oral & Maxillofacial Pathology and Microbiology, Guru Nanak Institute of Dental Science & Research, Panihati, Kolkata, with the chief complaint of gradually increasing painless swelling on right side of palate since 1 year. Extraorally there was no swelling noted.

Intra-oral examination revealed the presence of a dome shaped soft to firm swelling (approximately 2cm in diameter) at the right side of palate extending from first molar to posterior palatal seal region and medially upto midpalatal raphe. Overlying mucosa was ulcerated in posterior aspect of the swelling. Nothing significant was found in Orthopantomogram (OPG) of the patient. Based on clinical examination, the provisional diagnosis of Salivary gland neoplasm was made.

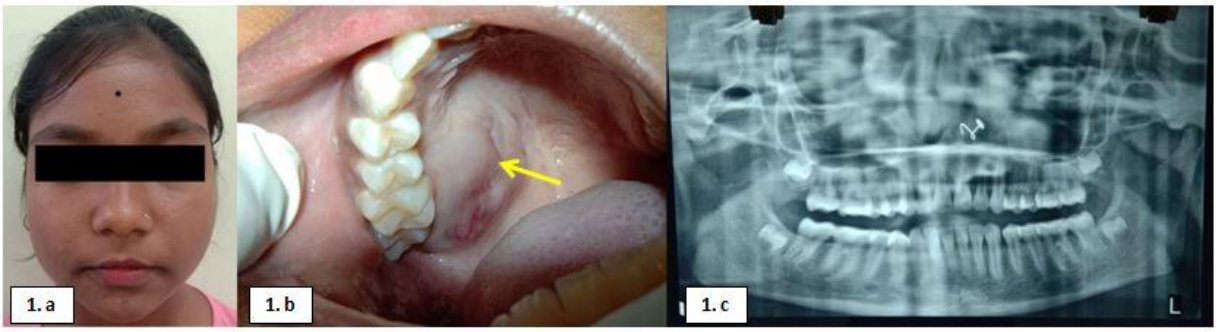

Fig: (1.a) Extra-oral photograph of the patient, (1.b) intra-oral photograph showing a dome shaped swelling over right palatal region (yellow arrow). (1.c) OPG of the patient.

Incisional biopsy was done from the representative area of the swelling. Microscopic examination of H&E stained sections revealed the presence of islands consisting of intermediate cells & mucous cells along with sheets of clear cells. These were large polygonal cells with centrally placed nuclei, clear cytoplasm & sharply defined cytoplasmic border. Intervening collagenous connective tissue was very scanty. Mucin filled microcysts were also present lined by mucous cells. No abnormal mitotic figure, necrotic area, haemorrhages and perineural invasion was noted. Thus, the microscopic features, along with the clinical findings confirmed the diagnosis of Clear Cell Variant of Low Grade Mucoepidermoid Carcinoma. The patient was then referred to the department of oral surgery for necessary surgical intervention.

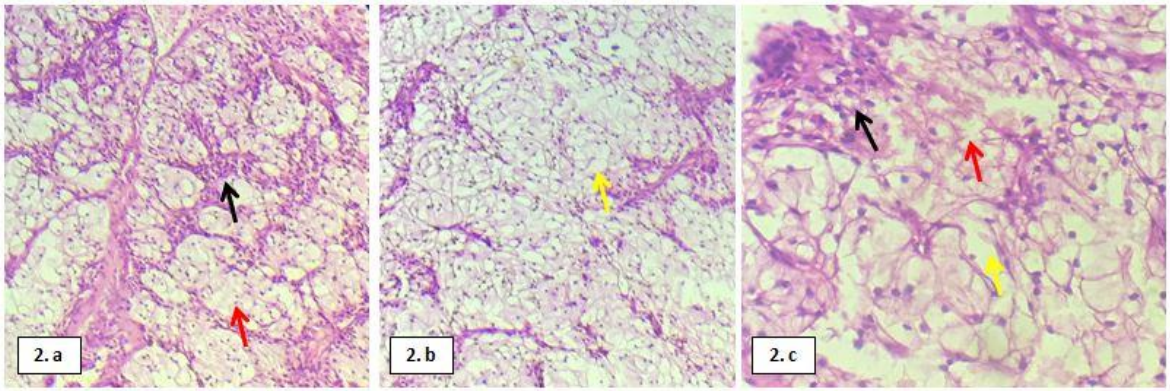

Fig: (2a & b) H & E stained image (10 X) showing zones of clear cells [yellow arrow] with intermingled hypercellular areas containing mucous cells [red arrow] and epidermoid cells [black arrow]. (2c) Amorphous eosinophilic deposits with close association with clear cells [yellow arrow].

DISCUSSION

The usual site for MEC is parotid gland (89.6%) followed by submandibular gland (8.4%). In case of minor salivary glands, it shows a strong predilection for palate [4]. MEC occurs most commonly in 50 to 60 years of age group with female predominance [5], Children are affected in about 1-5% of the cases. Most of the lesions found in children and adolescents involves parotid gland & rarely minor salivary glands [7].

Etiology of MEC is still unknown though some risk factors have been assorted i.e. radiation exposure, use of tobacco, genetic predisposition, virus and environmental chemicals etc [1]. An association was noted with a specific genetic translocation between chromosomes 11 and 19, t(11;19)(q14-21; p12-13) that leads to the formation of CRTC1-MAML2 fusion oncogene [5][6].

MEC clinically presents as solitary, painless, sessile, fixed, rubbery to soft mass measuring from 1cm to 3cm & as they are superficially located, they may appear as a blue-red tinged swelling intraorally mimicking mucocele or vascular tumors [6].

MECs can be studied for the extension of tumor and bone invasion with various radiographs i.e. OPG along with other advanced imaging modalities like MRI (Magnetic Resonance Tomography), MDCT(Multi Detector Computed Tomography),CBCT(Cone Beam Computed Tomography) etc [5].

Microscopically, MECs are predominantly composed of mucous cells and epidermoid cells in varying proportions along with intermediate cells, which is thought to be a progenitor cells of both mucous and epidermoid cells [8]. Intermediate cells are smaller than the polyhedral epidermoid cells with very little cytoplasm. The intermediate cells can be identified with mucicarmine staining or PAS positive, diastase resistance while the mucous cells are appreciated by

their mucicarminophilic nature [6]. Histologically it is classified as high-grade MEC with epidermoid cells predominency, intermediate grade with predominant intermediate cells and low grade MEC with the presence of mucin producing cells within a cystic framework [9]. This grading is associated with the degree of cystic architecture, mitotic rate, perineural invasion, necrosis and degree of cytologic atypia (anaplasia) [6]. The lesions of low and intermediate grade of MEC grow slowly, while the lesions having high histological grade are rapidly enlarging [5]. Low grade MEC most often affects minor salivary glands whereas high grade variant mostly involve major salivary glands, especially the parotid gland [6]. Epidermoid cells occasionally show dyskeratosis & produce keratin pearl. The epidermoid cells very rarely changeover to clear cell as cytoplasmic clearing [6]. Clear cells resemble as large, polyhedral cells with distinct outline and a hydropic, clear cytoplasm along with centrally placed piknotic nuclei. Clearing of cytoplasm can be due to three basic reasons- firstly, due to intracellular accumulation of nonstaining components like glycogen and rarely lipid or mucin secondly, due to paucity of cytoplasmic organelles and thirdly, due to fixation artifact [4]. Considering its histologic variations and degree of differentiation WHO classification in 1991 suggested that the term to be changed from "mucoepidermoid tumour" to "mucoepidermoid carcinoma" [8]. The confirm diagnosis of mucoepidermoid carcinoma can be made on histo-morphological features alone, without immunohistochemistry (IHC) or molecular confirmation [6].

Treatment of choice for MEC is surgical resection with disease-free margins. Postoperative radiotherapy is needed in advanced tumor stage, high-grade variant, perineural or lymphovascular invasion, close or positive resection margins, extra-parotid extension or lymph node involvement [6].

Histologic grade, tumor location, patient age, surgical margin, and lymph node involvement can act as prognostic factors in patients with mucoepidermoid carcinoma of minor salivary glands. 5-year survival rate of MEC is approximately 98.8% in low grade, 97.4% in the intermediate grade, and about 67% for high-grade tumors.

CONCLUSION

Mucoepidermoid carcinoma is most common salivary gland malignancy, though, clear cell variant of MEC in a young patient is very rare. Proper histopathological evaluation of this tumor is very important for proper diagnosis and prompt treatment for better prognosis.

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CONFLICT OF INTEREST: NIL

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