Case Report

Clear cell myoepithelial carcinoma involving vestibule and alveolus: A rare case report with review of literature

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Abstract

Myoepithelial carcinomas (MCs) are difficult to distinguish from their benign counterpart due to diverse morphology. This neoplasm was introduced by Stromeyer *et al*. in 1975. They comprise of <2% of all salivary gland carcinomas involving most commonly major salivary glands and are characterized by differentiation of tumor cells into myoepithelial cells. The cells may present as epithelioid, plasmacytoid, spindle, clear, stellate and mixed type predominantly. Literature search revealed very few cases reported as clear cell variant of MC. Here, we report a case of clear cell MC involving buccal vestibule extending up to alveolus. The diagnosis was confirmed, and the patient was surgically treated.

Keywords: Clear cell, immunohistochemistry, myoepithelial carcinoma, salivary gland

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INTRODUCTION

Myoepithelial carcinoma (MC) and myoepithelioma can be considered as doppelgangers, as they are difficult to distinguish due to diverse morphology and MC have infavorable outcome.[1] Published reports suggest that MC comprise of <2% of all salivary gland carcinomas involving parotid gland most commonly. Other sites involved are palate, maxilla, nasopharynx, liver, vulva and vagina.[2] However, this may be just a tip of the iceberg as due to increased recognition of this tumor, its incidence may be changed and may vary depending on demography. In fact, it is now believed that this tumor is the second-most common salivary gland malignancy arising from the benign adenomas.[3] It may also arise *de novo*. An accurate diagnosis for MC relies on exclusive myoepithelial differentiation (morphologic and immunohistochemical [IHC]) and clear-cut tumor infiltration into adjacent salivary gland or other tissues. We had earlier reported a rare case of clear cell variant of MC in an unusual location of upper lip.[4] We now report a case of yet again clear cell MC (CCMC) involving buccal vestibule extending up to alveolus.

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CASE REPORT

A 42-year-male patient reported with the chief complaint of swelling in his right buccal vestibule for 4 months. It involved lower alveolus and vestibule extending anteroposteriorly from the region of first premolar to first molar; and buccolingually, it involved buccal vestibule extending till the lingual vestibule, measuring approximately 3 cm × 2 cm in size. Overlying mucosa was smooth and had indentations of maxillary teeth. Surrounding mucosa appeared normal [Figure 1a]. There was no visible pulsation

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or discharge. On palpation, it was a firm, nontender swelling which was noncompressible and nonreducible. Single ipsilateral submandibular lymph node was palpable, hard, nontender and nonmobile.

His radiograph revealed a radiolucent lesion with ragged border causing displacement of adjacent teeth and thinning of lower border of mandible [Figure 1b]. The patient had the habit of tobacco and areca nut consumption since 20 years. Family history was noncontributory and there was no relevant systemic finding. His complete blood picture and random blood sugar investigations revealed no deviation of values from the normal range. A provisional diagnosis of malignancy of salivary gland or odontogenic origin was made.

The tissue was surgically excised. Macroscopic examination revealed a solid nodular mass measuring about 3 cm × 3 cm. Microscopically, plentiful polyhedral (epithelioid) cells with abundant cytoplasm (mostly clear) and round vesicular nuclei with slight pleomorphism were noted [Figure 2]. The stroma was scant and appeared hyalinized. Collagenous material was sparsely seen arranged focally into spherules. Necrosis was also noticed. Infiltration was observed in the surrounding bone. There were mitotic figures of more than 7 per 10 high power fields. Immunohistochemically, the tumor cells were positive for calponin [Figure 3], CD-10 [Figure 4], alpha SMA [Figure 5] and HMWCK [Figure 6]. We confirmed the diagnosis as CCMC. The patient was recalled after a year and showed no recurrence.

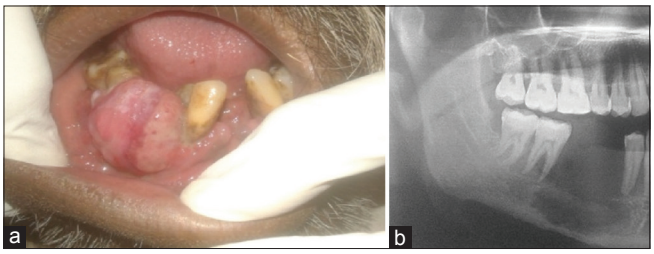

Figure 1: (a) Intraoral presentation. (b) Radiograph demonstrating radiolucent lesion with ragged border

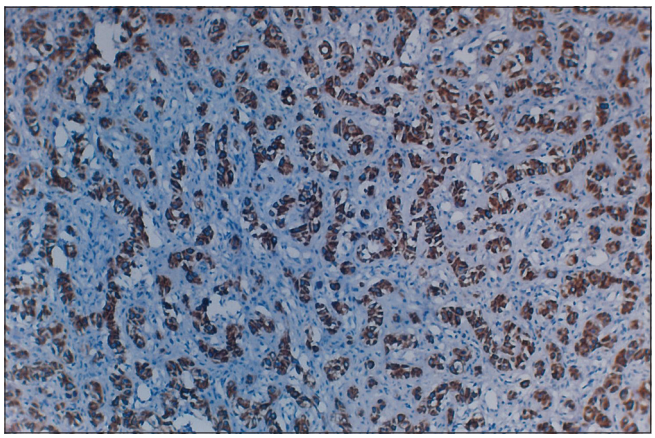

Figure 3: Immunohistochemical positive for Calponin (×100)

DISCUSSION

MC also known as malignant myoepithelioma (MM) affect most commonly major salivary glands and are characterized by differentiation of tumor cells into myoepithelial cells. Morphologic heterogeneity of tumor cells in MC include epithelioid, plasmacytoid, spindle, clear, stellate and mixed type.[5] Kane *et al*. in a series of 51 cases of MC, observed that minor gland involvement (almost 75%) exceeded major salivary gland involvement (<25%).[6] The CCMC affects parotid gland, submandibular gland, palate, retromolar area, maxillary sinus and even the base of the tongue, upper lip (only 1 case reported).[4]

Myoepithelial cells pose the ability to store glycogen as illustrated by Hamperl.[7] On the other hand, glycogen-rich, clear cells may be derivatives of different types of precursor

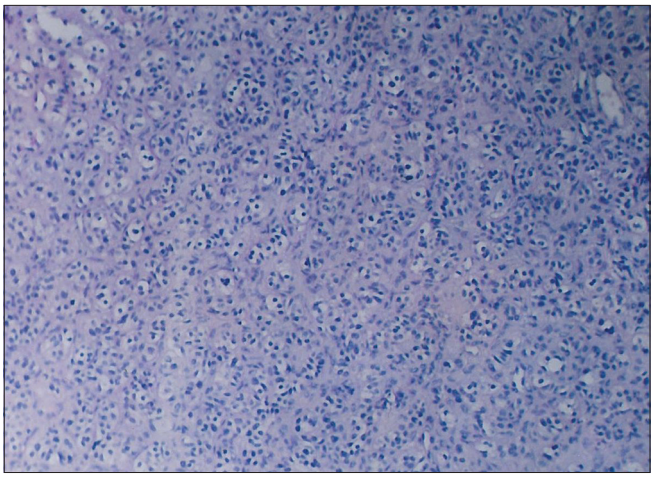

Figure 2: Clear cells with cellular and nuclear pleomorphism (H&E, ×100)

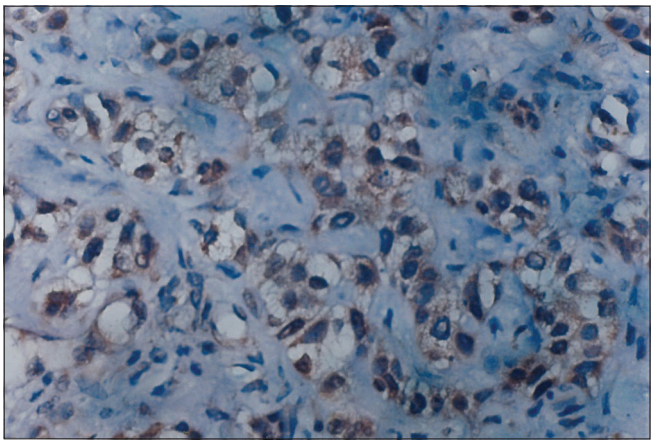

Figure 4: Immunohistochemical positive for CD-10 (×400)

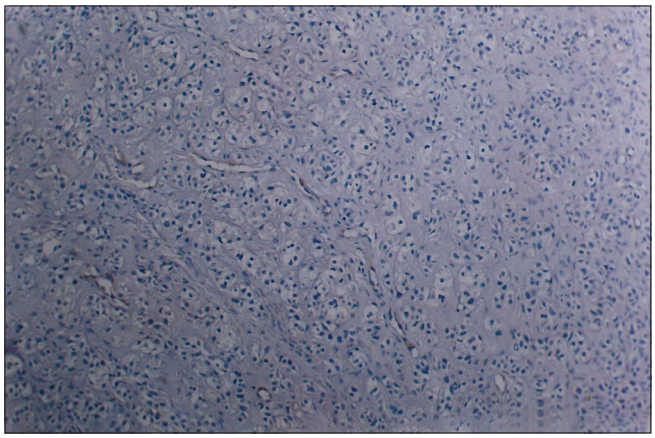

Figure 5: Immunohistochemical positive for alpha SMA (×100)

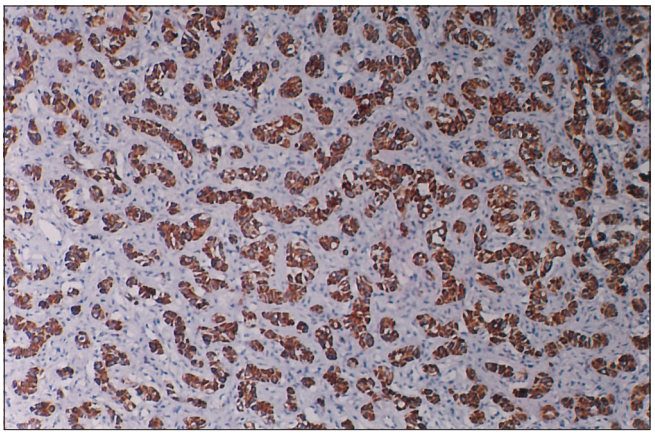

Figure 6: Immunohistochemical positive for HMWCK (×100)

cells, not confirming their origin. This implies that to label a myoepithelial tumor having predominant clear cell differentiation, it is necessary to confirm with IHC.[8]

We searched literature for clear cell variant of MC in the head and neck region. The search revealed the distribution of CCMC as depicted in Table 1. According to Losito *et al*. there were 16 cases (including 2 of his own) of CCMC reported till 2008.[12] The cases reported by Klijanienko *et al*. and Cassidy *et al*. have not been included by us as they are clear cell carcinomas and did not have characteristics of myoepithelial differentiation.[12] Kane *et al*. in 2010 found that 3 of their 51 cases showed clear cell differentiation.[6] They have not mentioned their particulars as to the site affected and other demographic details for those three cases. Apart from this, Liao *et al*. in 2005, studied 16 cases of MM and observed clear cell predominance in nine cases.[16] We found that the largest number of CCMC were studied by Skálová *et al*. (51 which arose de novo and 21 developed from preexisting pleomorphic adenoma).[17]

We report a case involving alveolus and vestibule. Our patient presented with a rapidly growing nodular mass. Ingle *et al*. have reported a similar case which clinically manifested as ulceroproliferative lesion of the alveolus. The ulceration proves a more aggressive variant histopathologically that is spindle cell variant.[5] Histopathologically, predominantly clear cells were present along with epithelioid cells in a hyalinized scant stroma. Benign and MMs may pose a dilemma in diagnosing as the features, and cellular type may be common. Malignancy may be considered by virtue of the tumor cells to show high proliferative index/increased mitotic activity and infiltration.[18] We observed high mitotic activity of more than 7 per 10 high power fields. Nagao *et al*. described 10 cases of MM and studied their IHC profile. They found epithelioid, spindle and plasmacytoid cell subtypes. Although few epithelioid cells had clear cytoplasm, there is no mention of predominance of clear cells in their 10 cases.[19] They also found the presence of osteoclast-like giant cells in 1 of their case and is regarded to have a very poor prognosis. We, however, did not observe any giant cell in our case.

Lata *et al*. have stated that MC arising from pleomorphic adenomas show a portion of ductal differentiation, whereas this may not be necessary for MC arising *de novo*.[20] We did not observe any ductal pattern.

Collagenous spherules (round, eosinophilic bodies) were appreciated and similar structures were confirmed by Michal *et al*. The spherules consists of a mixture of mucin, elastin, basement membrane proteins such as laminin and type IV collagen, admixed with collagen types I and III. This is a characteristic of tumors constituting myoepithelial cells.[9] Various IHC markers, as depicted in Table 1, aid in identification of myoepithelial cells. MCs have been considered to be low-grade malignancy. However, follow-ups have shown that MCs arising de novo tend to have a more unfavorable prognosis.[4,15,16] The patient was treated surgically and a follow-up of 1 year revealed no recurrence.

CONCLUSION

CCMC is a rare entity and may involve major as well as minor salivary glands. We report a case of CCMC in an unusual location involving vestibule and alveolus. The histopathologic findings and IHC profile corroborate a diagnosis of CCMC.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will

Table 1: Clear Cell Myoepithelial Carcinoma of Salivary Glands: Review of Literature

Authors	Number of cases	Age/sex	Year published	Site	Size (cm)	IHC markers positivity
Ogawa *et al*.[8]	1	64/male	1991	Palatal region	NA	S-100, vimentin, GFAP, actin
Michal et al.[9]	5	50/female	1996	Right submandibular gland	4.5×4×4	S-100, cytokeratin, actin
		70/female		Left parotid gland	4×4×3	
		66/male		Right parotid gland	4×3.5×3	
		37/female		Left parotid gland	3.8×2.5×2	
		78/female		Parotid	4×5×4.5	
Alós *et al*.[10]	2	49/male	1996	Parotid	4	GFAP
		85/male		Parotid	4	
Savera *et al*.[11]	4	74/female	2000	Parotid	3.5	AE1:AE3, 34ßE12, CK14, vimentin, S100, SMA,
		38/male		Maxillary sinus	5.5	calponin, MSA, GFAP
		46/female		Parotid	NA	
		51/male		Parotid	3.8	
Losito *et al*.[12]	2	67/female	2008	Left submandibular gland	12	CK-AE1/AE3, CK14, CK34bE12, Vimentin, S-100,
		64/female			6	calponin, p-63, alpha-SMA, GFAP, EMA
Yang *et al*.[13]	1	70/male	2010	Left retromolar region	6.5×5×4.5	Cytokeratin AE1/AE3, p63, S-100, vimentin, maspin, calponin
Park *et al*.[14]	1	52/female	2012	Left side of the tongue base	3×3	Cytokeratin 7, p63, HMWCK
Wang *et al*.[15]	5	48/female	2015	Larynx	4.1×3.5×3.1	CK 5, CK6, CK7, Pan-CK, p63, S-100, SMA, CEA,
		57/male		Floor of mouth	1×2	EMA, Calponin, Ki-67
		65/female		Submandibular gland	2.2×1.6	
		46/male		Parotid gland	2.5×3	
		66/female		Palate	2.5×2	
Gotmare *et al*.[4]	1	46/female	2017	Left upper lip	3×2	Alpha-SMA, HMWCK, CD-10, calponin

IHC: Immunohistochemical

not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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