Case Report

Mucoepidermoid carcinoma: A rare case report

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Received - 18 February 2019 Initial Review - 04 March 2019 Accepted - 14 March 2019

ABSTRACT

Minor salivary gland carcinomas are rarely reported. Mucoepidermoid carcinoma (MEC) is one of the most common salivary gland malignancies. MEC accounts for 1% to 2% of major salivary gland neoplasms and 9% of minor gland tumors. It is most commonly seen in the parotid gland and usually appears as asymptomatic swelling. However, the palate is a frequent site when it occurs in the minor salivary glands. Follow-up of the patient is very important as local recurrence rate with conservative treatment is high. Here we are reporting a low-grade mucoepidermoid carcinoma involving palatal region in a 25-years-old female patient. The patient presented with a fast-growing swelling, invading the underlying anatomical structures and was treated successfully through the antrostomy procedure.

Key words: *Mucoepidermoid carcinoma, Palate, Salivary gland neoplasm, Surgery*

Mucoepidermoid carcinoma (MEC) is the epithelial salivary gland neoplasm of the oral cavity. In 1924, Masso and Berger first reported MEC. Stewart *et al* described it as a distinct pathology in 1945 [1]. As its name implies, it is composed of mucous producing, squamous, and intermediate type cells. About two-thirds MEC arises within the parotid gland, and one third arises within the minor salivary glands. Minor salivary glands are present in all areas of oral cavity except the gingival and the anterior portion of the hard palate. When MEC arises in minor salivary glands, it can be located on the palate, retromolar area, the floor of the mouth, buccal mucosa, lips and tongue [2]. Rarely, it can arise as a primary jaw tumor of the laryngeal, lacrimal, nasal, paranasal, tracheal, or pulmonary tumor.

MEC occurs most frequently in adults. Females are more commonly affected than males, in a ratio of 3:2. It occurs more commonly in the third to sixth decade of life [3]. Rarely, it is seen in the first decade of life. However, mucoepidermoid carcinoma is the most common malignant salivary gland tumor in children. Most patients are aware of the lesion for 1 year or less, although some report a mass of many years' duration. Histologically, MECs are classified into low, intermediate, and high-grade [4].

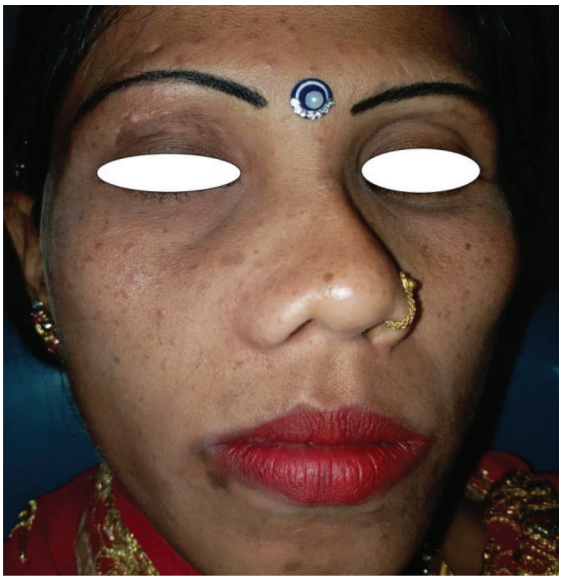

Figure 1: Extraoral image of the patient

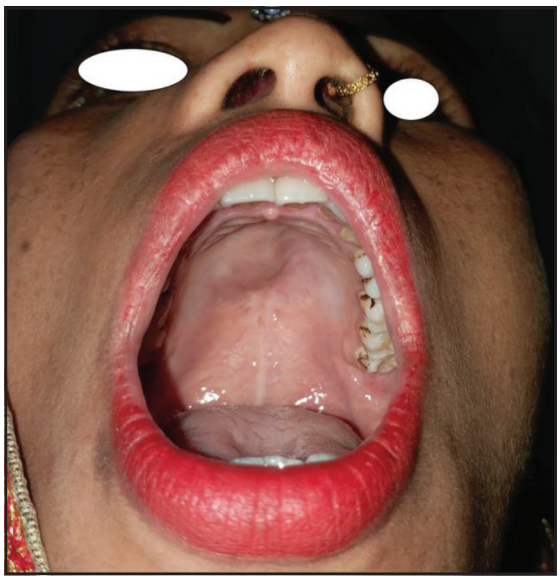

Figure 2: Intraoral image of the patient

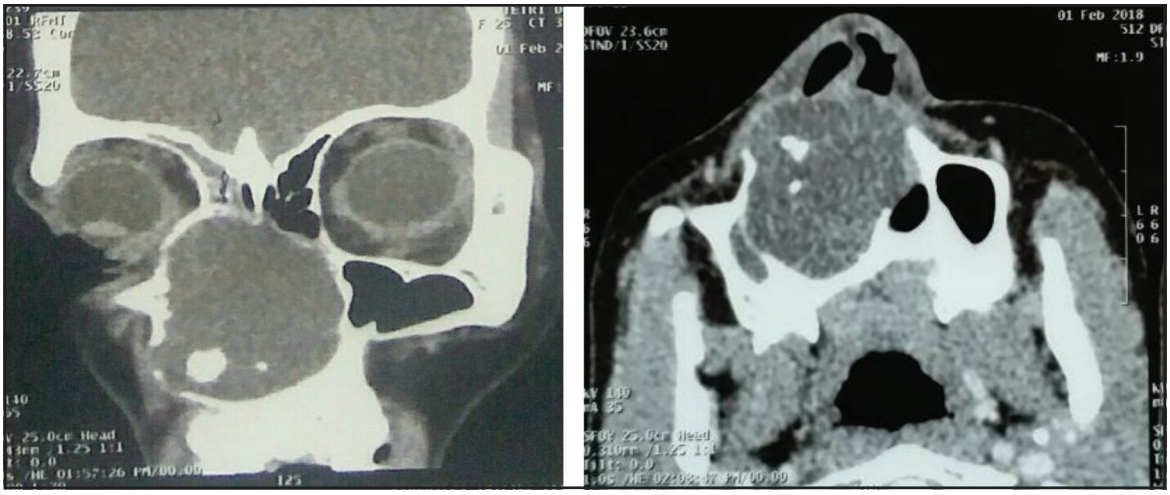

Figure 3: CT image of the patient showing a well-defined, lytic, expansible lesion

This article reports the case of mucoepidermoid carcinoma in a female patient involving the palatal region, presenting as a fast-growing swelling, invading the underlying anatomical structures and was treated successfully.

CASE REPORT

A 25-year-old female patient reported to the Department of Oral Medicine and Radiology with a chief complaint of swelling in the upper right front and back tooth region since 6 months [Fig. 1]. History of present illness revealed that the patient was apparently asymptomatic 6 months back before she experienced swelling in the maxillary right region. Initially, the swelling was smaller in size but gradually, it increased to the present size. There has been a constant increase in the size of swelling from the last 6 months. The swelling was also associated with pain which was sudden in onset, sharp, intermittent in nature and radiates to the whole of the face region on the right side. Her past medical, dental and family history was non-contributory.

On extraoral examination, a diffused swelling was present on the right facial region. Superior-inferiorly, the swelling extends from 0.5 cm from inner canthus of the eye till nasolabial fold and anterior-posteriorly, it extends from the midline of the nose involving the whole of the dorsum of the nose on the right side till 1.5 cm from the right zygomatic arch. There was a deviation in nasal septum and nasolabial angle. Nasolabial fold was raised. The color of the skin over swelling was of normal skin color. On palpation, the swelling was tender, firm and was not associated with any discharge.

On intraoral examination, a diffused swelling was seen in the palatal region, roughly oval in shape and 1.5 x 2 cm in diameter [Fig. 2]. Superior-inferiorly, it extends from rugae area till 2 cm posterior to the hard palate. Anterior-posteriorly, the swelling extends 1cm from the palatal surface of 16 till midline of the palatal region. Swelling is of normal color as of oral mucosa and surrounding mucosa appears to be normal. On palpation, all inspection findings were confirmed. The swelling was tender, soft to firm non-fluctuant, non-compressible and non-pulsatile. No mobility and displacement of teeth were present.

Based on the clinical findings, a provisional diagnosis of minor salivary gland tumor was given. In the investigation, computed tomography (CT) scan and biopsy were done. CT scan showed the presence of a well-defined, lytic, expansile lesion in the right maxillary bone causing bone resorption of the anterior nasal septum and the right nasal turbinate's causing complete nasal obstruction

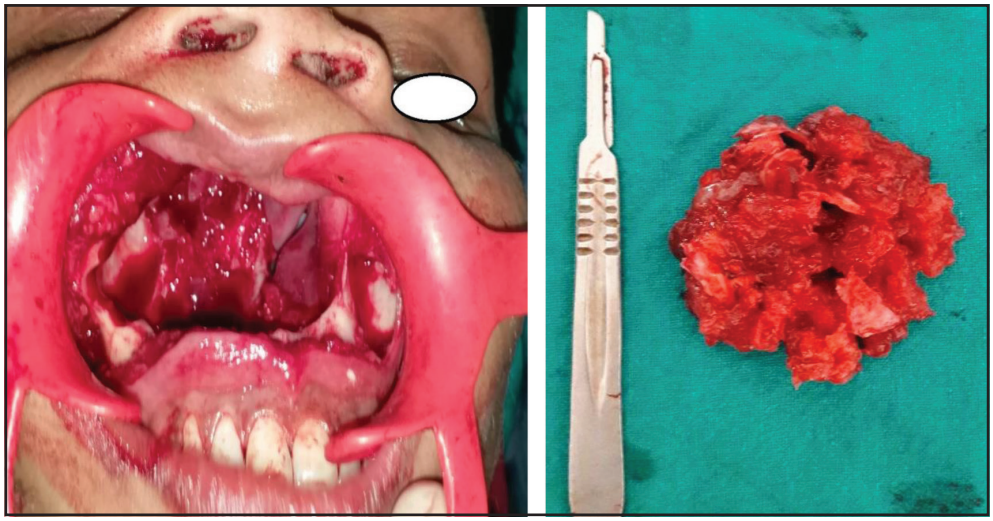

Figure 4: Image showing surgical excision of the lesion

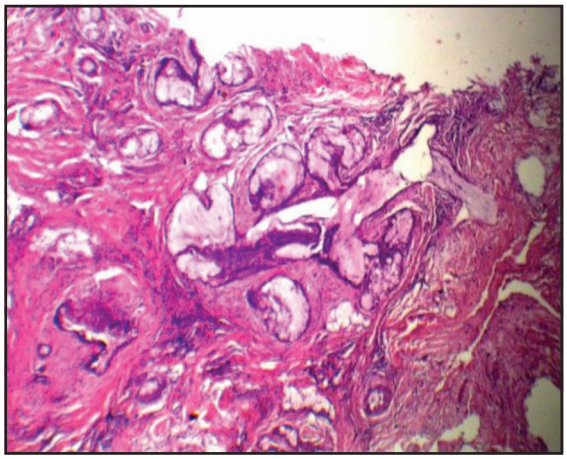

Figure 5: Histopathological image of the mucoepidermoid carcinoma

and significant mass effects leading to the lateral displacement of medial wall of the right maxillary sinus and the right cribriform plate bulging into the inferomedial aspects of orbit [Fig. 3]. Lesion extended to the right ethmoid and frontal sinus. Multiple destructive bones or calcifications within the lesion were seen. Mass lesion was measured approximately 39 mm x 44 mm x 50 mm.

As the lesion was aggressively growing, so immediate surgery was planned. After asepsis and anesthesia, a vestibular incision was given extending from the left canine to the right first molar region. A full-thickness mucoperiosteal flap was reflected to gain access to the maxillary sinus region [Fig. 4]. After proper osteotomy procedure, the lesion was resected and the specimen was sent for histopathological examination. Antrostomy was done and the pack was removed after 5 days. Histopathology report revealed the presence of malignant salivary gland tissue within connective tissue stroma. The section revealed the presence of ciliated columnar cell lining with predominantly located mucous pooled areas which appear to be surrounded by sheets of cells having squamoid features with polygonal shape and dysplastic features like increased nucleo-cytoplasmic ratio, nuclear and cellular pleomorphism, hyperchromatism and mitotic figures. Surrounding stroma revealed the infiltration of few chronic inflammatory cells, collagen fiber bundles with fibroblasts, endothelium-lined blood vessels with extravasated RBC's [Fig. 5]. This histopathology report was suggestive of low-grade mucoepidermoid carcinoma. After surgery, the patient was discharged and recalled after 3 months but unfortunately, the patient lost to follow-up.

DISCUSSION

Mucoepidermoid carcinoma is the most frequently diagnosed malignancy of the salivary gland. Pain or facial nerve palsy may develop, usually in association with high-grade tumors.

Table 1: Table showing differentiating features of the various form of MEC

S.No.	Lesion	Differentiating features
1.	Low-grade MEC	• Most common MEC. More benign in nature. • Low-grade tumors are soft and compressible. • Size of the lesion is generally less than 5 cm. • They appear as lobulated or irregularly sharply circumscribed cystic areas with intact underlying periosteum. • Low-grade MEC macroscopically is small and partially encapsulated. • Microscopically characterized by the presence of more mucous producing cells. • Prominent cystic structures lined by mature mucous, intermediate, or epidermoid cells are the hallmark of these tumors. • Solid areas are not evident and prominent fibrous stroma often is present. • Low-grade MEC grows in a well-circumscribed manner, without small infiltrative islands at the tumor border. • The treatment of low-grade MECs is complete, wide surgical resection of the tumor with free surgical margins. • Survival rates are 92% for low-grade MEC.
2.	Intermediate MEC	• Least common MEC • Clinically, they present as an ulcerated or solid mass. • Firm in consistency. Have normal colored overlying mucosa. • Can ulcerate in the early stages. • Bony invasion in the form of palatal bone erosion. • Intermediate comprises of solid than cystic architect with more intermediate cells. • These cells are capable of differentiating into mucous or epidermoid cells. • Survival rates are 83% for intermediate MEC.
3.	High-grade MEC	• Aggressive tumors. • High-grade lesions may be quite firm and accompanied with ulceration, resorption of bone, and numbness of adjacent teeth. • The high-grade tumors consist of epithelial cells, with very few mucinous cells. • They are less likely to demonstrate a capsule because of rapid growth and local tissue invasion. • Distant metastasis implicates an unfavorable prognosis but behavior of the metastatic deposits has a slow progression. • When distant metastases develop the average survival is 2.3 years for minor salivary gland tumors and 2.6 years is for tumors of the major salivary glands. • The lung is the most commonly involved site of metastasis. • High grade requires wide surgical excision, neck dissection and postoperative loco-regional disease control. • Survival rates are 24% for high-grade MEC.

Low-grade mucoepidermoid carcinoma is characterized by slow-growing painless swelling which rarely exceeds 5cm. They appear blue colored because of the mucin filled spaces. High-grade mucoepidermoid carcinoma shows a tendency for local tissue invasion in the early stages. In late stages, these invade underlying structures, perforate cortical plates and invade vital structures of maxillary antrum, nasal cavity and finally to infratemporal spaces and to the cranial base [4]. In our case also, local invasion to the maxillary sinus was seen. The minor glands constitute the second most common site, especially the palate. Retromolar area, the floor of the mouth, buccal mucosa and the lower lip are other common sites of MEC after palate [5]. However, pus discharge may be seen if the lesion is secondarily affected. Intraosseous tumors may also develop in the jaws and distant metastasis may be seen involving the lungs and the bones.

Traditionally, mucoepidermoid carcinoma has been classified histologically into three types based on the amount of cyst formation, the degree of cytologic atypia and relative numbers of mucous, epidermoid and intermediate cells [Table 1]. Low-grade MEC show prominent cyst formation, minimum cellular atypia and a relatively high proportion of mucous cells. High-grade MEC consists of solid islands of squamous and intermediate cells, which can demonstrate considerable pleomorphism and mitotic activity. Intermediate type shows feature that falls between those of low-grade and high-grade neoplasms. Cyst formation occurs but is less prominent than that observed in low- grade MEC [4,6]

Differential diagnosis of MEC involving palate should include minor salivary gland tumors, metastatic tumors, schwannoma etc. A low-grade MEC, bluish hue color, compressible may mimic underlying vascular lesions or melanoma if popular appearance is seen. Low and intermediate grade MEC with intact mucosa mimic pleomorphic adenoma and sometimes mucocele. Slow-growing intermediate-grade MEC having an ulcerated mucosa resembles adenoid cystic carcinoma or polymorphous low-grade adenocarcinoma. High-grade MEC with infiltration to underlying structures like sinus or maxillary sinuses gives an impression of nasal and sinus carcinoma.

Radiographically, MEC appears as a cystic or tumor-like lesion on radiographs. Low-grade mucoepidermoid carcinomas are typically not apparent on projection or plain images unless destructive changes to adjacent osseous structures have occurred. High-grade mucoepidermoid carcinoma shows irregular margins and ill-defined forms. Various radiographic techniques like sialography, multidetector computed tomographic imaging, cone beam computed tomographic imaging (CBCT), ultrasonography (USG), magnetic resonance imaging (MRI) and scintigraphy can be used for diagnosis of the lesion [7]. Immunohistochemistry also plays a vital role in the diagnosis of MEC. A study was conducted by AbdRabohNMto check the diagnostic role of discovered on gastrointestinal tumor-1 (DOG1) and p63 immunohistochemistry in salivary gland carcinomas. He along with his co-workers examined the expression of DOG1 and p63 immunohistochemistry in 33 MEC, 9 acinic cell carcinomas (ACC), 10 adenoid cystic carcinomas (AdCC) and 4 myoepithelial carcinomas. The result of a study showed that all 33 MEC cases expressed strongly to moderate positivity for p63 (P=0.001) while only 9.1% were weak to moderately positive for DOG1 whereas AdCC, ACC were positive for DOG1. He concluded that DOG1 is a sensitive marker for ACC whereas p63 is sensitive for MEC [8].

Treatment of MEC depends on aggressiveness and the extent of spread of the tumor. If there is any evidence of gross periosteal involvement or bone erosion, removal of a portion of the involving bone is mandatory and necessary. High-grade MEC requires a more aggressive surgical approach with or without postoperative radiotherapy and chemotherapy. Radical neck dissection is indicated in patients with clinical evidence of metastatic disease [9]. The survival rate of the patient with low-grade MEC is approximately 92%, whereas, in high-grade MEC, the survival rate is roughly 24 %.

CONCLUSION

Salivary gland malignancy is rare when compared to common lesions affecting the oral and maxillofacial region. So, it becomes very important for the dentist to take a proper history of the patient and get various investigations done to reach a proper diagnosis of salivary gland neoplasm. Proper surgery should be done by the clinician and a constant follow-up of the patient is advised for a prolonged period of time to check for any recurrence as the recurrence of such neoplasm is high.

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Funding: None; Conflict of Interest: None Stated.

How to cite this article: Alok A, Hasan K, Singh S, Jha A. Mucoepidermoid carcinoma: A rare case report. Indian J Case Reports. 2019;5(2):133-136.

Doi: 10.32677/IJCR.2019.v05.i02.013