

MUCOEPIDERMOID CARCINOMA OF HARD PALATE IN YOUNG FEMALE: A RARE ENTITY

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ABSTRACT

Mucoepidermoid carcinomas (MEC) of minor salivary gland are rare in origin in children and adolescents and also have been reported rarely. Literature regarding their clinical features and biological behaviour is scanty. The purpose of this case report is to discuss the clinical manifestation, diagnosis, and treatment plan of MEC of the palate. We are presenting a case of 21 year old girl visited to our department of ENT at tertiary health care centre complaining of painful swelling at left side of hard palate since 2 months. Several clinical, radiographic, and histopathological investigations were carried out to rule out the lesion. Incisional biopsy of the lesion confirmed the diagnosis of lesion as MEC of the palate. Following which a wide surgical excision with left horizontal Partial maxillectomy with adjacent free margins was carried out along with left supraomohyoid neck dissection done. Sample send for frozen section for confirmation of free margin and also for histopathological examination which was confirmative of MEC. This case report highlights the need for proper diagnosis and treatment plan in the cases of malignant tumours as it can help to reduce morbidity and mortality.

KEYWORDS: Minor salivary gland, Hard palate, Mucoepidermoid carcinoma.

INTRODUCTION

Mucoepidermoid carcinoma (MEC) is the epithelial salivary gland neoplasm of the oral cavity. It accounts for <3% of all head and neck tumours. About 5% of these tumours occur in patients younger than 18-year-old with women mostly affected.^[1,2,3]

MEC was first reported by Massao and Berger in 1942 and by Stewart et al. in 1945 as a distinct pathologic entity.^[1]

As its name implies, it is composed of mucus-producing, squamous, and intermediate type cells.^[1,4]

Histologically, it is classified into low, intermediate, and high grade.^[4,5]

Low grade tumours commonly develop a nesting pattern with multiple well-circumscribed squamous nests containing numerous clear cells. Intermediate-grade tumours are less cystic and show a greater tendency to form large sheets of

squamous cells and often have a more prominent intermediate cell population. High-grade tumours are predominantly solid, with greater degrees of atypia.^[1]

About two-third of MEC arise within the parotid gland, and one-third develops within the minor salivary glands. When it develops in minor salivary glands, it can be located on the palate, retromolar area, and floor of the mouth, buccal mucosa, lips, and tongue.

Treatment of MEC depends on its aggressiveness and the extent of its spread. When the tumour is confined to the palatal mucosa with intact periosteum, wide excision of lesion along with underlying mucoperiosteum is advised.^[6,7]

Prognosis of MEC is a function of the histological grade, adequacy of excision and clinical staging.^[8]

CASE REPORT

21 year old girl visited Department of ENT at tertiary health care centre complaining of swelling in oral cavity over left side palate since 2 months.

She was apparently alright 2 months back when she noticed swelling in oral cavity at left palate which was insidious in onset and painful in starting but over the period pain subsides with antibiotics analgesic. No history of change in size of a swelling. There was no any history of trauma or oral bleed. Not giving any positive history regarding nasal obstruction, recurrent rhinitis, facial swelling or parenthesis. Not giving any history related to odynophagia, dysphagia, change in voice or any sign related to aspiration. She did not have any co morbidity like Diabetes mellitus, Hypertension, Tuberculosis or Bronchial asthma. There is not any significant family and medical history.

General examination was normal

Ear and nose findings shows normal findings

Intraoral examination shows approximately 2×2 cm swelling with smooth surface similar to palatal mucosal colour with slight bluish tinge seen. On palpation swelling was hard in consistency, non tender, oval, smooth surface, regular margin involving left side of hard palate, alveolar ridge upto anterior margin of soft palate with no any pulsations (fig 1). Based on history and clinical examination initially, the differential diagnosis of the swelling was made as periodontal abscess, palatal region vascular malformations such as haemangioma, mucocele, sialometaplasia, pleomorphic adenoma, adenoid cystic carcinoma, MEC, and adenosquamouscarcinoma. CECT of Face done which shows heterogeneously enhancing well circumscribed lesion approximately 19×12 mm in size is arising from left side of hard palate. Mild mass effect and remodelling of left maxillary alveolar ridge was noted without cortical dehiscence (Fig 2). Few hypoechoic areas also noted within. Few enlarged left group lymph nodes group IA, IB, IIA, IIB noted.

Then incisional biopsy of swelling done which shows lobular growth pattern of tumour cells with minimal cytological atypia. The cystic spaces entrapped within the tumour islands are filled with mucin like substances with differential diagnosis like adenosquamous carcinoma and mucoepidermoid carcinoma of intermediate grade.

After all haematological work up and preanaesthetic evaluation and fitness for surgery patient posted for left hemimaxillectomy and left supraomohyoid neck dissection.

Patient taken under general anaesthesia

Gingival mucosal flap elevated to expose underlying maxillary bone and root of teeth from lateral canine to last molar. Canine tooth removed. Delineation for osteotomy done using drill from left canine socket till left last molar 2cm above the teeth .Osteotomy done with chisel and hammer.

1cm of normal tissue margin is kept around tumour on palatal side. Swelling along with maxilla (Horizontal Partial) removed in toto (fig3 a & b) and sent for histopathological reporting. Post excision cavity and maxillary sinus filled with medicated gelfoam .Haemostasis achieved. Mucosal flap sutured with vicryl 3.0 cutting Margins of specimen send for frozen section which showed no evidence of tumour.

After that neck dissection started with Transverse cervical incision along neck crease 2 finger breadths below mandible from mastoid tip till mentum taken.Subplatysmal flap elevated superiorly up to mandible inferiorly up to omohyoid superior belly. Dissection started in midline. Fibro fatty tissue dissected off from anterior belly of digastric upto superior belly of omohyoid and sternocleidomastoid. Submandibular gland identified and dissected. Facial artery identified and ligated and cut at two sites. Lingual and Hypoglossal nerve identified and preserved. Submandibular duct ligated and cut. (fig4 a & b)

Gland removed in toto and sends for histopathological reporting spinal accessory nerve identified and preserved. Level 1a, 1b, 2a, 2b and 3 groups of lymph nodes removed and sent for histopathological reporting separately (fig 5).

Fibrofatty tissue from carotid sheath removed and sent for histopathological reporting. Patient was kept on nasogastric tube feeding & hence Ryle's tube inserted .Histopathological report showed sheets and nest of tumour cells amidst mucin pool. Tumour is focally cystic and predominantly solid. Tumor cells are polygonal with distinct cell border and intercellular bridge with abundant eosinophilic cytoplasm. Interspersed round cell are seen with abundant blue mucin.Nuclei shows mild pleomorphism are vesicular with prominent nucleoli.Mitosis is inconspicuous. All this is confirmative of low grade mucoepidermoid carcinoma of palate (fig 6). Remaining lymph nodes were free of tumour.

Patient post operative finding were normal .Drain removal done on Post operative day 4 and suture removal done on day 10 (fig 6). Patient remains on Ryelstube feeding for 4 week .patient was advised palatal obturator for palatal defect.



Figure 1: Clinical and intraoral photo showing swelling over left hard palate.



Figure 2: CECT of face showing enhancing lesion within the mucosa overlying inferior surface of Maxilla.



Figure 3 (a & b): Intraoperative photo of left hemi maxillectomy.

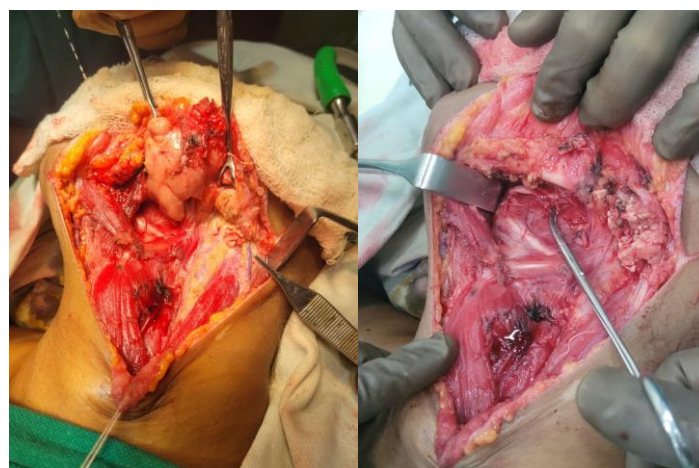


Figure 4: Intra operative photo after supraomohyoid neck dissection. (Removal of group Ia, Ib, IIa, IIb, III Lymph nodes and submandibular gland).



Figure 5: Specimen showing Lymph nodes with submandibular gland and left hemi maxilla with Teeth & Lymph nodes.

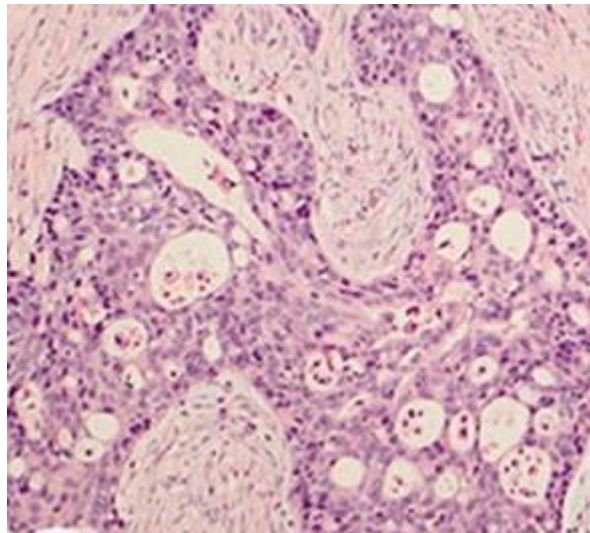


Figure 6: H and E staining of MEC.

DISCUSSION

It has been estimated that about 1–5% of all salivary gland tumors develops in children and adolescents.^[1,9]

The percentage of benign tumors occurring in the palate is higher than that of malignant tumors.^[10] Most of the malignant neoplasms of the salivary gland in children and adolescents occur in the parotid gland. In our case age of patient was 21 year old. Only a few cases are reported in minor salivary glands.^[5]

MEC is believed to arise from pluripotent reserve cells of excretory ducts that are capable of differentiating into squamous, columnar, and mucous cells.

MECs are generally found between 10 and 16 years. Clinically, the majority of palatal MEC appears as firm, painless swellings, it mimic mucoceles or vascular lesions.^[5]

Risk factors include female sex, history of radiation exposure, and a t (11; 19) (q21; p13) translocation that results in an MECT-MAML2 fusion gene.^[11]

Appearance on imaging depends strongly on tumor grade and guides preoperative planning. Treatment of low-grade tumors is conservative, usually necessitating solely wide local excision; treatment of high-grade tumors often requires complete parotidectomy with involvement of the facial nerve, neck dissection, and adjuvant radiotherapy. Low-grade tumors have a better prognosis when compared to high-grade tumours and have higher survival and lower recurrence rates.^[12]

Histologically Radical resection with wide safety margins of normal tissues.

Including neck dissection is the mainstay of treatment modality for all grades of minor salivary gland MEC of the palate. The patients with high grade disease should be treated with postoperative radiotherapy.^[13]

The patients with high grade disease should be treated with postoperative radiotherapy.

Intermediate grade fall between low and high grade. Cyst formation occurs but less prominent than low grade. All 3 major cell types will be present, but intermediate cells predominate. Cellular atypia may or may not be present.^[1]

Moraes et al. suggested that low to intermediate-grade MECs originating from intraoral minor salivary glands can be managed by wide local surgical excision that ensures tumour-free surgical margins.^[14]

Low- to intermediate-grade MECs originating from intraoral minor salivary glands has a very low recurrence rate (<10%) and a high survival rate (90%).^[15]

Low and intermediate grade MECs have an indolent clinical course and a rare chance for metastasis. Radical neck dissection is indicated if clinical evidence of metastasis. Prognosis depends on grade and stage of tumor.^[13] In our case as the tumour is of low grade and lymph nodes are free of tumour cells patient prognosis is good and managed without radiotherapy.

CONCLUSION

- Any palatal bulge on the hard palate even in young patient needs to be examined carefully & the possibility of carcinoma of maxilla needs to be evaluated.
- Early intervention & prompt diagnosis is needed to prevent the morbidity in such cases.
- Radical resection with wide safety margins of normal tissues including neck dissection is the mainstay of treatment modality for all grades of minor salivary gland MEC of the palate.
- A close clinical follow-up should be for lifetime because low and intermediategrade MEC in this age group can recur many years later.

Conflict of Interest- There is no conflict of Interest

REFERANCES

1. SJ Jarde, S Das, SA Narayanswami et al. Mucoepidermoid carcinoma of the palate: A rare case report, J Indian SOC periodontal, 2016 Mar-Apr; 20(2): 203–206.
2. Hicks J, Flaitz C. Mucoepidermoid carcinoma of salivary glands in children and adolescents: Assessment of proliferation markers. Oral Oncol, 2000; 36: 454–60.
3. Luna MA, Batsakis JG, El – Naggar AK. Salivary gland tumors in children. Ann Otol Rhinol Laryngol, 1991; 100: 869–71.
4. Neville BW, Damm D, Allen CM, Bouquot JE. Salivary gland pathology. In: Neville BW, editor. Oral and Maxillofacial Pathology. Missouri: Saunders, 2009; 495–7.
5. Aruna Laila Mathew, Biju Baby Joseph, Deepa Muraleedharan Sarojini, Preeja Premkumar, Sunil Sukumarannair. Mucoepidermoid carcinoma of palate - a rare entity, Clinpract, 2017 Sep 15; 7(4): 1009.
6. Bin Zhou, Zhuo-shan Huang, Wei-liangChen, Zi-xian Huang, Rui Chen, Kai-fang Yuan, Lei Hong, Yon-juChen. Outcomes of patients with mucoepidermoid carcinoma of minor salivary gland in palate undergoing radical resection followed by submental flap reconstruction. Asian Journal of Surgery, 2022; 45: 1225-1230.
7. Moore BA, Burkey BB, Nettekville JL, Butcher 2nd RB, Amedee RG. Surgical management of minor salivary gland neoplasms of the palate. Ochsner J., 2008; 8: 172-180.
8. K. Triantafillidou, J. Dimitrakopoulos, F. Iordanidis, D. Koufogiannis, Mucoepidermoid carcinoma of minor salivary glands: a clinical study of 16 cases and review of the literature Oral Dis, 2006; 12(4): 364-370.
9. Fonseca I, Martins AG, Soares J. Epithelial salivary gland tumors of children and adolescents in southern Portugal. A clinicopathologic study of twenty-four cases. Oral Surg Oral Med Oral Pathol, 1991; 72: 696–701.
10. Rajendran R. Tumors of the salivary gland. In: Rajendran R, Sivapathasundharam B, ed. Shafer's Textbook of Oral Pathology. India: Elsevier, 2009; 219-53.
11. Lee WH, Yoon JH. Mucoepidermoid carcinoma of the hard palate: a rare cause of hypervascular tumor. Yonsei Med J., 2003; 44: 723-6.
12. Bishop JA, Cowan ML, Shum CH, Westra WH. MAML2 Rearrangements in Variant Forms of Mucoepidermoid Carcinoma: Ancillary Diagnostic Testing for the Ciliated and Warthin-like Variants. Am J Surg Pathol, 2018; 42(1): 130-136.
13. Moraes P, Pereira C, Almeida O, et al. Paediatric intraoral mucoepidermoid carcinoma mimicking a bone lesion. Int J Paediatr Dent, 2007; 17: 151-4.
14. April MM, Brodsky LS, Cunningham MJ, et al. Mucoepidermoid carcinoma in a 10-year-old girl. Head Neck, 1997; 19: 431-5.
15. Ritwik P, Cordell KG, Brannon RB. Minor salivary gland mucoepidermoid carcinoma in children and adolescents: a case series and review of the literature. J Med Case Rep, 2012; 6: 182.