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Case Report

Central mucoepidermoid carcinoma of maxilla a case report and review of literature

Vivek Kumar Pathak[1,*], Anshul Chatrath[1], Riya Thakral[1], Abhinav Srivastava[1]

[1] *Dept. of ENT, School of Medical Sciences and Research (Sharda University), Uttar Pradesh, India*

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ABSTRACT

MEC is the most commonly occurring malignant salivary gland neoplasm, comprising 2.8–15% of all salivary gland tumors. Its behavior is like mucoepidermoid cancers elsewhere and maxillary cancers in general. These cancers typically impact the mandible and exhibit histological characteristics indicating low-grade malignancy. This paper reports the case of 50-year-old female with facial swelling, which on FNAC showed intermediate-grade epithelial malignancy. mucoepidermoid carcinoma. MRI showed lytic lesion in right maxilla. She underwent right subtotal maxillectomy and had good prognosis on one year follow up.

1. Introduction

MEC is the most commonly occurring malignant salivary gland neoplasm, comprising 2.8–15% of all salivary gland tumours.[1] Aberrant salivary gland neoplasms arising within jaws as primary central bony lesions are extremely rare, comprising 2–4.3% of all MECs reported.[2] Fewer than 100 cases have been reported to date. Like other maxillary cancers, it may first be noticed by its extension into adjacent structures, such as the jaw, palate, oral cavity, nose, orbit, and other paranasal sinuses. Its behaviour is like mucoepidermoid cancers elsewhere and maxillary cancers in general. These cancers typically impact the mandible and exhibit histological characteristics indicating low-grade malignancy. There are very few case reports available in the literature on central mucoepidermoid carcinoma of the maxilla. One such rare case is reported in our hospital.

2. Case Report

A 50-year female presented to ENT OPD of Sharda Hospital, Greater Noida with chief complaints of swelling on the right side of her face in the past 5 years. The swelling was insidious and progressed gradually. It started as pea-sized swelling and progressed to the current size of approx. 4 X 4 cm. There was no history of nasal bleeding, nasal blockage, skin changes, or change in vision. There was a history of loosening of teeth. There was no history of dental procedures or trauma to the face.

There was no significant past medical history, family history or personal history. On general examination she was poorly built and nourished, alert, responsive, and cooperative. All the vital signs fell within the normal range.

On local examination, a unilateral, solitary and diffuse swelling was present on the middle one-third of the right side of the face which caused gross asymmetry. The swelling was irregular, measuring about 3 cm in its greatest extension, located over the anterior wall of the maxilla extending from the infraorbital rim to the ala of the nose. The skin over the swelling was normal. On palpation,

* *Corresponding author*.
E-mail address: vivek.pathak@sharda.ac.in (V. K. Pathak).

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the swelling was tender, firm, non-mobile, non-fluctuant, non-compressible, and non-reducible. The skin above the swelling showed no abnormality.

On oral cavity examination, minimal swelling was noticed on the right side hard palate extending till midline. The associated palatal mucosa was erythematous. loosening of teeth was noticed. An ulcer was also noted on the buccal alveolar swelling with respect to 18. The ulcer was irregular, measuring 5 mm at its widest point, the floor was erythematous, the margins were sloppy and mildly tender and its base was indurated.

Lymph node examination, submandibular lymph node {level IB} was tender, mobile of approx 1*1 cm in greatest dimension.

On CEMRI of the nose and PNS showed soft tissue attenuation lytic mass lesion of size 3.95X3.76X3.55cm in the right maxilla extending superomedial up to the hard palate and laterally along gingivobuccal sulcus superiorly protruding through the anteroinferior wall of the maxillary sinus. Heterogenous enhancing lymph node of size 20*12mm in right level IB.

On Diagnostic nasal endoscopy, the middle meatus was normal, and no crusting or mass was seen. A biopsy of tissue from the middle meatus was taken and sent for HPE.

HPE showed a group and cluster of polygonal cells which showed a moderate degree of pleomorphism

FNAC from the cheek (right anterior wall of maxilla) showed a tight and loose cluster of basaloid epithelial cells showing mild to moderate pleomorphism and scanty cytoplasm. The cluster of cells with vacuolated and foamy cytoplasm. Few cells were polygonal with abundant cytoplasm. Cytology suggested intermediate-grade epithelial malignancy. mucoepidermoid carcinoma.

Considering that lesion was aggressively growing but not yet had involved other sinuses, muscles (pterygoid muscles) and nerves a surgical approach was planned immediately. All the maxillary teeth were extracted. Subtotal maxillectomy was performed under general anaesthesia along with a temporary obturator was given.

Post-operative specimen biopsy showed a large cellular tumour consisting of a squamoid area mixed with mucus-secreting cells. Tumor cells exhibited significant pleomorphism. Tumour is also seen infiltrating into the bony fragments (maxilla). Specimen suggested mucoepidermoid carcinoma.

3. Review of Literarture

Lepp discovered central mucoepidermoid cancer in the jaw of a 60-year-old female in 1939. There are only less than 200 cases reported in the literature. Various theories have been proposed on the origin of the tumour and is thought to be due to one of the following: 1. Metaplasia of the odontogenic cyst epithelium, 2. Entrapped salivary tissues or minor salivary glands during development, 3 from the

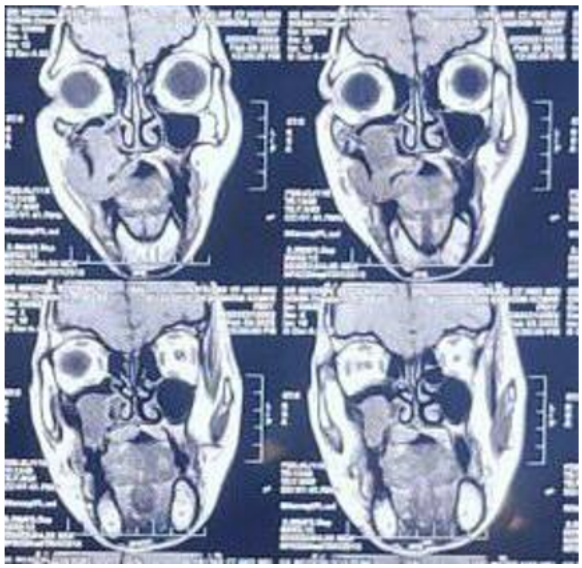

Fig. 1: Pre-operative MRI nose and paranasal sinuses

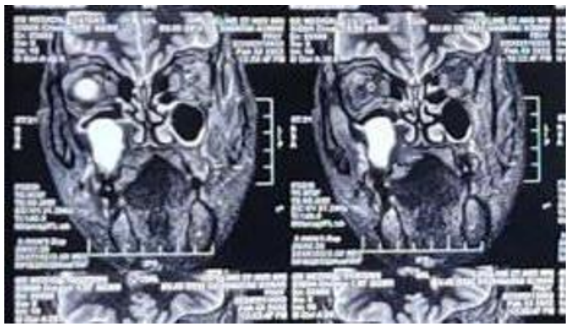

Fig. 2: Pre-operative CEMRI nose and paranasal sinues

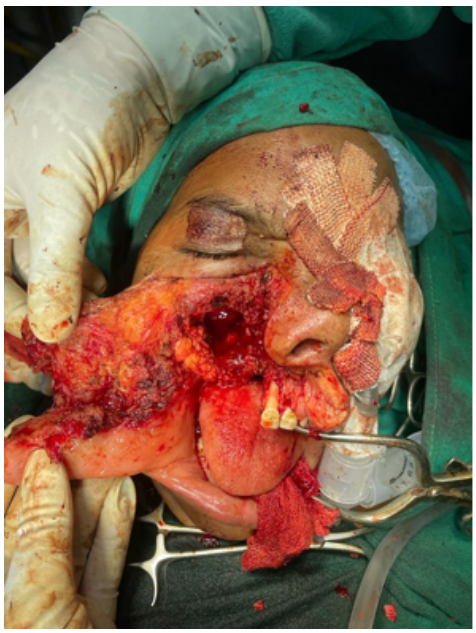

Fig. 3: Intra-operative subtotal maxillectomy

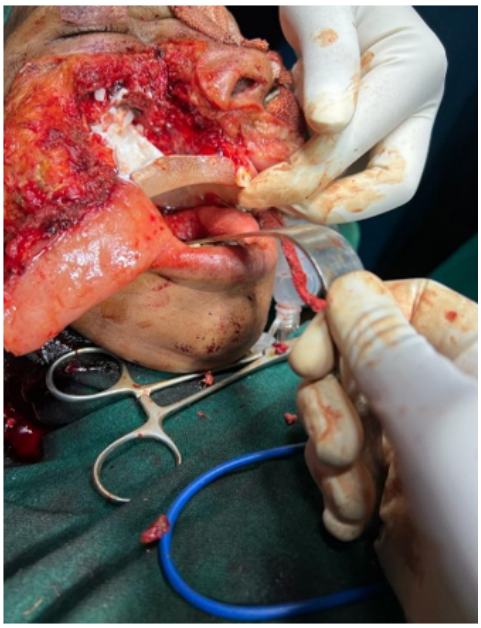

Fig. 4: Intra-opperative application of obturator

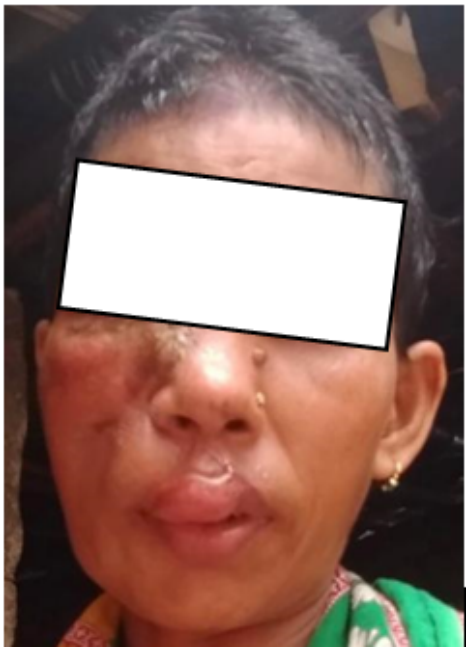

Fig. 5: Post-operative picture of the patients

Maxillary sinus epithelium, 4. Odontogenic remnants of the dental lamina. There are data regarding the incidence and management mucoepidermoid carina of the maxilla.

A study conducted at the University of Texas Health San Antonio in 2020 concluded Females were more affected (54.5%) by MEC and their average age was 48.8 years. MEC was most frequently seen in the parotid glands (56.8%), followed by the hard palate (18%). The most common clinical presentation was mass (65.2%), followed by ulcer (29.4%), with pressure as the primary symptom (64.4%). The most common histologic appearance was Low grade (46.7%), followed by Intermediate grade (27.3%), and surgery was the most commonly employed treatment (76.2%). The average follow-up was 138.5 months, and recurrence was reported at 8.5%.[3]

Another retrospective clinicopathologic study at Government Medical College and Hospital, Ambajogai, of 25 cases conducted in 2021 concluded that the relative frequency of MEC was 13.15%. The most prevalent MEC type was low-grade (44%) followed by intermediate-grade (36%), and high-grade (20%). The average age of occurrence of MEC was 44.28 13.29 years. MEC was more common in women (60%) than in men (40%). Thus, the overall female-male ratio was 1.5:1. Among minor salivary glands, the palate (48%) was the most common site, and among major salivary glands, the parotid gland (16%) was the common site.[4]

There is a systematic review article reporting molecular determinations of Mucoepidermoid carcinoma (MEC) and adenosquamous carcinoma (ASC) conducted in 2022. Adequate evidence for a distinctive molecular profile of either MEC or ASC was not found. Reported cases retrieved in this review point towards the relevance of MAML2 rearrangement in confirming the diagnosis of MEC when positive.[5]

Chuan-Xiang Zhou et al. conducted a clinicopathologic and immunohistochemical Study of 39 Chinese Patients on MEC. In their study radiographically, most cases showed a unilocular or multilocular radiolucency with bone destruction. Most cases were classified as low-grade MECs.All cases were found to be primary; local recurrence occurred in 8 cases, most (75.0%) of which were low-grade tumours. The immunohistochemistry of keratins helps in differential diagnosis. Radical surgery is the treatment of choice, whereas the role of radiotherapy or chemotherapy is still controversial, and careful long-term follow-up is necessary.[6]

A clinical study of 16 cases was conducted in 2006 at General Hospital, in Thessaloniki. They had a mean follow-up of 4-14 years. Out of 16 MECs patients, ten were alive and five (35.6%) died from the disease. Four patients were free of the disease for more than 5 years (range 8-14), five patients were free of the disease for 5 years and one of them was free of the disease for 4 years. One patient

lived more than 10 years and died from another disease. only one patient developed local recurrence after 10 years of the initial treatment. they compared the Ki-67 values in correlation with the histological grade of the tumours. The Ki-67 expression was only 1% in low-grade MECs, while in intermediate-grade tumours it was estimated between 3 and 4%. The high-grade tumours had increased expression (10%) of tumour cells. The preferred course of treatment for MECs is complete surgical excision. Adequate excision is important in all grades of tumours. The prognosis of MECs depends on the histological grade, adequacy of excision and clinical staging. The immunohistochemical study of Ki-67 expression may provide additional prognostic information for this tumour.[7]

There is still a review of lacuna regarding the MEC of the maxilla.

4. Discussion

Mucoepidermoid carcinoma is the most common malignant salivary gland tumour, accounting for about 2.8%-15% of all salivary gland tumours.[8] The most commonly affected major salivary gland is the parotid gland, and the most common intraoral site is the posterior palate.

Intraosseous carcinoma arising in the jaw bones was described as a central epidermoid carcinoma by Loos in 1913.11 Later, Pindborg coined the term ”primary intraosseous carcinoma” (PIOC) in the first edition of the World Health Organization classification for the histopathological typing of the odontogenic tumours12 Primary intraosseous MEC is an uncommon lesion which was first described and reported by Leep13 in 1939. Waldron and Mustoe14 proposed that intraosseous MEC be classified as type 4 primary intraosseous carcinomas of the jaws. Classification of PIOC14 N Type 1 PIOC ex odontogenic cyst N Type 2a Malignant ameloblastoma N Type 2b Ameloblastic carcinoma arising de novo, ex ameloblastoma or ex odontogenic cyst N Type 3 PIOC arising de novo: (a) keratinizing type; (b) non-keratinizing type N Type 4 Intraosseous MEC

Our case is of mucoepidermoid carcinoma of the anterior-inferior wall of the maxillary region and is a rare site of occurrence since salivary glands are absent in this site. Ectopic salivary glands in this area may be the cause of the development of mucoepidermoid cancer.[9] Adults in their second to eighth decades of life are more likely to develop mucoepidermoid carcinoma, with a minor female predisposition.[8] Similarly, our patient was a 50-year-old female.

low-grade MECs tend to be of long duration and asymptomatic. In the present case, the patient was asymptomatic for 5 years. As the lesion was asymptomatic, the patient failed to recognize the lesion, and there are possibilities of a much earlier occurrence of the tumour mass. In a few cases clinically the patient presents with pain, trismus and paresthesias. The pain sometimes radiates to the teeth, palate, face, and nose, misleading the clinician to focus on neural, dental or maxillary sinus problems. In our patients, there were nasal or oral symptoms or any problem related to the maxillary sinus

There have been reports of sclerosing, intraosseous, clear cell, goblet cell, and spindle cell types, among other histological varieties. Histological grades of MECs have been demonstrated far too frequently to exhibit a correlation with the tumour’s clinical symptoms.

Mucoepidermoid carcinoma is classified based on the degree of cyst formation, the proportion of cell types, and the presence or absence of cytomorphologic atypia as low-(15%-62% of patients), intermediate-(9%-48%), or high-grade (22%-38%) malignancy.

1. Low-grade: It is a highly differentiated neoplasm with a predominance of macro- and microcystins. Intermediate and mucin-producing cells with minimal cellular atypia are present.
2. Intermediate grade: It predominantly consists of intermediate cells with a few cysts. The presence of mucin-producing cells and islands of epidermoid cells are seen.
3. High-grade: It is a poorly differentiated neoplasm and consists of solid blocks of intermediate and epidermoid cells. Mucin-producing cells are present with nuclear pleomorphism and mitotic activity.[8]

In our case, the histopathological findings showed the predominance of mucous cells followed by a squamoid area mixed with mucus-secreting cells. Tumour cells showed a high degree of pleomorphism. Thus, a diagnosis of intermediate-grade mucoepidermoid carcinoma was made.

They commonly present as multilocular cyst-like radiolucent lesions, while few cases consist of radio opacities. Considering the location and radiological features, the differential diagnosis of ameloblastic carcinoma, and clear cell odontogenic carcinoma should be considered. In our case, MRI with contrast showed soft tissue attenuation lytic mass lesion of size 3.95X3.76X3.55cm in the right maxilla. Heterogeneous enhancing lymph node of size 20*12mm in right level IB.

Most of the central mucoepidermoid carcinomas reported in the literature are of low grade and carry a favourable prognosis. However, maxillary cases have a worse prognosis due to their extension into vital structures. Even though they are low-grade tumours, treatment includes wide local resection[10], en-bloc resection or hemi mandibulectomy, as a conservative line of management favour recurrences. When regional nodes are involved, neck dissection is done. Adjuvant radiotherapy is useful in high-grade tumours and in cases with positive surgical margins. Since our patient had intermediate grade MEC, subtotal maxillectomy was done. There was no radiotherapy given

postoperatively.

A prior study found a 12.7% local recurrence in a mean period of 41.7 months. The regional metastasis was 9.8% in 173 patients. They also mentioned a 92% to 100% survival rate for low-grade, 62% to 92% for intermediate grade, and 0% to 43% for high grade. Ellis et al[8] suggested that there is an overall recurrence of 25%, out of which 10% recurrence is for low-grade and 75% recurrence is for high-grade MECs. These reports confirm the good prognosis of low-grade carcinoma as reported in our case.

5. Conclusion

MEC is seldom found in the anterior maxillary region. However, their occurrence in the anterior maxillary region cannot be overlooked because they often manifest as a benign or inflammatory condition. The use of histopathological examination can greatly help in the diagnosis as well as identifying the involvement of the adjacent vital structures, which may change the treatment and the prognosis. To identify late local recurrence and regional metastasis, MEC should be followed up for a longer period of time.

6. Source of Funding

None.

7. Conflict of Interest

None.

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Author biography

Vivek Kumar Pathak, Associate Professor

Anshul Chatrath, Senior Resident

Riya Thakral, Junior Resident

Abhinav Srivastava, Assistant Professor

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