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Case Report

A Rare Case of Low-Grade Mucoepidermoid Carcinoma Presenting as Chronic Facial Swelling and Nasal obstruction

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Abstract

Background: Mucoepidermoid carcinoma (MEC) of the sinonasal tract is an extremely rare malignancy that may present with nonspecific symptoms such as facial swelling, nasal obstruction, or trismus, leading to delayed diagnosis.

Case Presentation: A 46-year-old male presented with a 2-year history of right-sided cheek swelling and right sided nasal obstruction. Clinical examination revealed a nasal mass. Imaging and biopsy with debulking was conducted. Histopathology confirmed low-grade mucoepidermoid carcinoma. The patient was referred for chemoradiotherapy.

Conclusion: This case highlights the importance of including sinonasal MEC in the differential diagnosis of chronic cheek swelling and nasal obstruction. Timely diagnosis and tailored treatment are essential for better prognosis.

Keywords: Mucoepidermoid Carcinoma; Paranasal Sinus Tumor; Trismus; Sinonasal Malignancy; Nasal Mass

Introduction

Mucoepidermoid carcinoma (MEC) is a malignant tumor most commonly arising from salivary glands but may also occur in other locations such as the bronchial tree and sinonasal tract. Sinonasal MEC is exceedingly rare, accounting for less than 5% of all salivary gland malignancies and under 1% of all head and neck cancers [1,2]. The annual incidence of sinonasal malignancies is approximately 0.5–1 per 100,000 population, with MEC representing a small subset [3].

There is a slight female predominance for MEC overall, but sinonasal involvement does not consistently follow this trend [4]. The typical age of presentation ranges from the 4th to 6th decades. Risk factors include prior radiation exposure, smoking, and chronic inflammation [5].

Given the nonspecific and indolent presentation, diagnosis is often delayed. Histological grade and anatomical extent play a critical role in prognosis and treatment planning.

Case Report

A 46-year-old male presented to the ENT outpatient department with a 2-year history of progressive right cheek swelling, right nasal obstruction with intermittent history of epistaxis and loss of olfaction. Patient reported headache and facial pain increased in intensity since 1month.There were no other associated complaints.

Clinical examination revealed right facial fullness. Diagnostic nasal endoscopy revealed an ulcero-proliferative lesion in the right nasal cavity. No cervical lymphadenopathy was noted.

Imaging

CT scan of the paranasal sinuses showed a heterogeneously enhancing soft tissue mass of size 4 x 3.5 x 6 cm. It is seen in the right maxillary sinus extending into the right nasal cavity. Medially the lesion is seen causing mass effect on the septum and the laterally widening of the maxillary ostium was noted. Superiorly the mass is seen causing mass effect over the anterior skull base, causing erosion of adjacent bony walls. The lesion is also seen involving the right frontal, anterior and posterior ethmoidal cells. MRI revealed hyperintense lesion on T2-weighted images with moderate post-contrast enhancement and no perineural spread or intracranial extension. There was no evidence of intra-orbital or intra-cranial invasion.

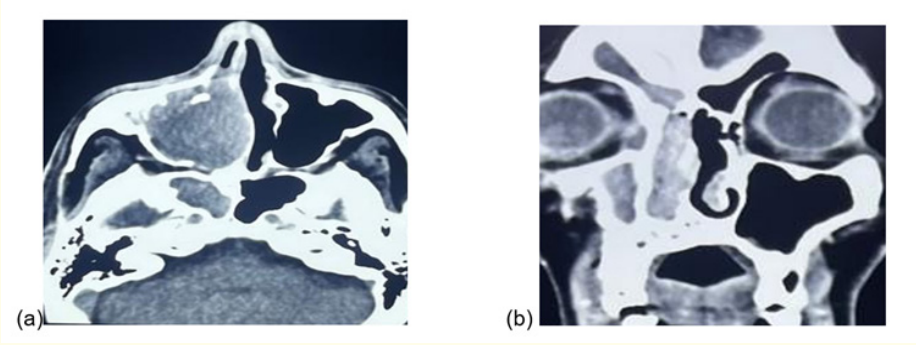

Image 1

Biopsy and histopathology

Biopsy was taken under general anesthesia from the nasal cavity. Debulking of the mass was done using microdebrider (Image 2). The mass was seen arising from the right maxillary sinus and having erosions of the lateral nasal wall and protruding into the nasal cavity. Medially the mass was seen having a mass effect on the septum causing deviation of the septum to the left. Superiorly the mass was extending upto the skull base. Sufficient biopsy was taken and remaining mass was debrided to clear the obstruction from the nasal cavity and the maxillary sinus and anterior and posterior ethmoidal region. Complete clearance of the mass from the floor of the maxillary sinus and the skull base was not possible. Histopathological examination revealed a neoplasm composed of mucous-producing cells, epidermoid cells, and intermediate cells arranged in cystic and solid nests, consistent with low-grade mucoepidermoid carcinoma There was minimal atypia, no necrosis, and no perineural or lymphovascular invasion, supporting the low-grade classification (Image 3) [6].

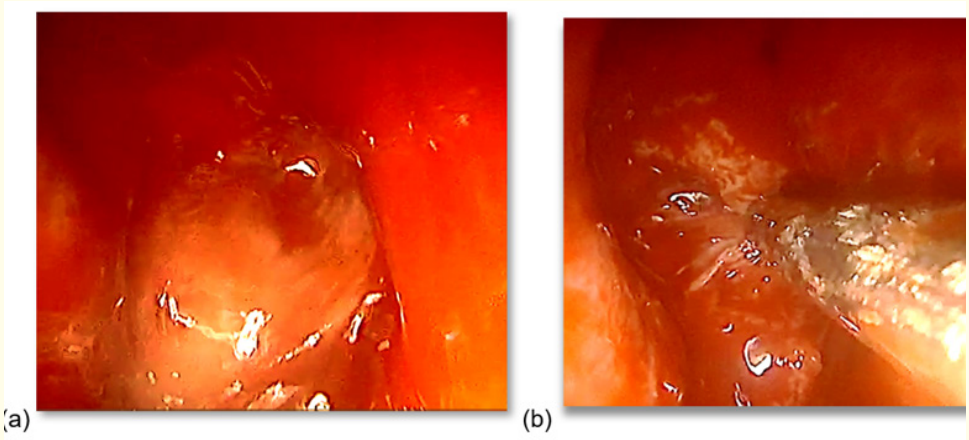

Image 2: Unilateral reddish mass, completely obstructing the nasal cavity. Surface is nodular and indurated. Consistency is firm. The mass is seen arising from the right maxillary sinus extending into the posterior choana and impinging on the right lateral nasal wall and medially over the septum.

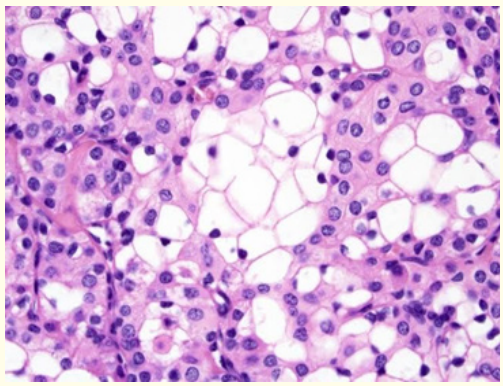

Image 3: Section shows features of low grade Mucoepidermoid carcinoma. Tumor shows solid growth pattern with three cell types- epidermoid cells, mucinous cells and basal cells. There is marked predominance of epidermoid cells arranged in solid pattern and in microcystic pattern. The cells show moderate nuclear pleomorphism and atypia. A few basal cells and mucinous cells are also seen. Mitosis is occasional. Necrosis is not seen.

As per TNM staging tumor is T3 (Disease having bony erosions, destruction and invading the ethmoid sinus, posterior wall). The patient was discussed in a multidisciplinary tumor board and referred for combined radiation and chemotherapy, given the location and potential morbidity of surgical resection.

PET-CT scan was done before initiating the Chemo – Radiotherapy. Definitive Chemotherapy with Cisplatin 100mg/m^2 3 weekly and IMRT with 60Gy in 35 fractions for 7 weeks was planned.

Follow up

Patient was kept on 6 montly follow up with DNE and PET CT scan. No recurrence was noted till 1 year.

Discussion

MEC of the sinonasal region is rare and often presents late due to vague symptoms. Symptoms may include nasal obstruction, epistaxis, facial swelling, trismus, or palatal ulceration—signs that often mimic chronic sinusitis or benign tumors [1,2].

Etiology of MEC includes genetic rearrangements such as the t(11;19)(q21;p13) translocation, which leads to CRTC1-MAML2 fusion, commonly found in low- and intermediate-grade tumors and associated with better prognosis [7].

Radiologic findings are vital for evaluating extent and surgical feasibility. CT scans delineate bony destruction, while MRI provides soft tissue detail. In this case, CT showed bony erosion, and MRI confirmed the absence of perineural or intracranial extension—critical for staging.

Histopathology remains the diagnostic gold standard. Low-grade MECs are defined by prominent cystic areas, minimal cellular atypia, and low mitotic activity. Absence of necrosis, perineural, and vascular invasion correlates with a favorable prognosis [6,7].

Yes, immunohistochemistry (IHC) can be helpful in confirming the diagnosis of mucoepidermoid carcinoma (MEC) and distinguishing it from other tumors, especially in small or poorly differentiated biopsies. Here are the key IHC markers commonly used for MEC:

- **CK7**: Positive in mucinous and glandular areas
- **CK5/6**: Positive in squamous components
- **p63**: Strong nuclear positivity in basal and intermediate cells
- **MUC5AC**: Positive, indicating mucinous differentiation
- **Ki-67**: Low proliferative index (<10%), consistent with low-grade tumor
- **DOG1 and SOX10**: Negative, helping rule out acinic cell carcinoma and myoepithelial neoplasms.
- In this case, IHC was instrumental in confirming the diagnosis:
- CK7 and MUC5AC highlighted the mucinous component.
- CK5/6 and p63 marked the squamous and basal/intermediate layers.
- A low Ki-67 index (<10%) correlated with the indolent behavior and low-grade status.
- Negative DOG1 and SOX10 helped exclude acinic cell carcinoma and other salivary gland neoplasms [6,7].

Molecular studies such as detection of CRTC1-MAML2 gene fusion (common in low-grade MEC) are useful for diagnosis and prognostication. Although molecular testing was not performed in this case, its presence would have further confirmed the diagnosis and predicted a better response to therapy [7].

Surgery is the mainstay of treatment when feasible, typically via endoscopic or open craniofacial approaches depending on extent. In unresectable cases, or when resection carries high morbidity (as in this case), definitive radiotherapy (RT) with concurrent chemotherapy is preferred.

Radiation regimens typically involve 60–66 Gy in 30–33 fractions over 6–6.5 weeks using intensity-modulated radiation therapy (IMRT) [8]. Chemotherapy: Cisplatin-based concurrent chemoradiotherapy is commonly employed for advanced cases or high-risk patients.Targeted therapy: For tumors harboring CRTC1-MAML2 fusions, targeted therapies like entrectinib or larotrectinib (in NTRK-fusion positive cases) are being explored but remain investigational [9].

Follow up is planned for 3 month – 6 months with DNE and PET – CT scans for first 2 years. Follow up monitoring for radiation induced osteonecrosis and other radiation induced side- effects is essential.

Prognosis

Prognosis depends on tumor grade, anatomical location, resectability, and presence of metastasis. Low-grade sinonasal MECs have a 5-year survival rate of 75–90%, whereas high-grade tumors fare significantly worse [6,10].

Conclusion

Sinonasal mucoepidermoid carcinoma, though rare, should be considered in the differential diagnosis of chronic facial swelling and trismus. This case emphasizes the need for thorough examination and timely biopsy in such presentations. Early diagnosis and appropriate multimodal treatment are critical for improved outcomes. In non-resectable cases, chemoradiotherapy remains a viable alternative to achieve disease control.

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