Case Report

Mucoepidermoid carcinoma of the lower lip: A case report and review of literature

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Abstract

Mucoepidermoid carcinoma (MEC) is a rare salivary gland malignancy characterized by diverse cellular components. This case report presents a unique instance of low-grade MEC in a 28-year-old female, emphasizing the importance of accurate diagnosis and tailored management. The patient exhibited a painless, enlarging lower lip swelling over a year, with imaging suggestive of a fibrolipomatous lesion. Surgical excision was successfully performed, with subsequent histopathological analysis displaying mucinous cystic spaces, varied epithelial cell types and other characteristic features consistent with MEC. The case findings aligned with hallmark MEC features are seen in established literature, highlighting the significance of precise diagnosis and grading for appropriate management. This report contributes to the understanding of MEC's varied presentations and underscores the importance of thorough: histopathological examination, lower lip, mucoepidermoid carcinoma clinical evaluation, accurate histopathological analysis and interdisciplinary collaboration. The case highlights the value of considering MEC in younger patients, even with atypical presentations, and encourages ongoing exploration of diagnostic and therapeutic strategies to improve patient outcomes.

Keywords: Cystic spaces, epidermoid cells, mucicarmine stain, mucoepidermoid carcinoma, periodic acid–Schiff (PAS) stain

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INTRODUCTION

Salivary gland neoplasms, constituting a minority of head and neck tumours, encompass MEC as a significant subtype. While typically observed in adults, its manifestation in younger patients, especially within the lower lip region, remains exceedingly rare.[1] Mucoepidermoid was initially described by Masso and Berger in 1924. It was previously referred to as a 'mucoepidermoid tumour' and was considered a benign lesion. In 1990, the World Health Organization (WHO) reclassified it as a malignant neoplasm and renamed it as mucoepidermoid carcinoma (MEC).[2] Among malignant salivary gland tumours, MEC stands as the most frequent one. It constitutes around 35% of all salivary gland tumours and accounts for less than 3% of all head and neck tumours.[3] The onset of MEC is most commonly observed between the 3rd and 6th decades of life, with a higher incidence among women compared to

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men.[2,4,5] Predominantly, it occurs in the parotid gland, followed by involvement in minor salivary glands. Among the minor salivary glands, regions such as the buccal mucosa, floor of the mouth and labial mucosa are mostly affected. The palate emerges as the most frequent site for MEC, followed by the retromolar region, floor of the mouth, buccal mucosa and lower lip.[6] While the precise aetiology remains unclear, factors contributing to the carcinogenesis of these tumours are thought to include radiation exposure, endogenous hormones, viruses, lifestyle factors, and specific occupations.[2,5] Variations in the frequency of specific histologic types and the involvement of major and minor salivary glands have been noted in literature from different parts of the world.[3,7-11]

Low-grade mucoepidermoid carcinoma (MEC) is an infrequent salivary gland malignancy characterized by a diverse array of cellular components, ranging from mucin-secreting to intermediate and epidermoid-like cells. Although most commonly arising within major and minor salivary glands, MEC can also manifest in atypical sites, including the vermilion border of the upper and lower lips. This distinct presentation poses diagnostic challenges and therapeutic complexities, particularly when encountered in young individuals.[6] Existing literature has predominantly focused on the histopathology and management of MEC in conventional anatomical locales, often overlooking its variant presentations in unusual sites. This underscores the importance of investigating the clinicopathological attributes of low-grade MEC in the lower lip of young individuals, not only expanding the understanding of this malignancy but also underscoring the necessity of tailored diagnostic and therapeutic strategies.[12] The pathogenesis of mucoepidermoid carcinoma involves intricate genetic and molecular mechanisms. Somatic mutations, such as the MECT1-MAML2 gene fusion, play a pivotal role in the initiation of mucoepidermoid carcinoma. These genetic alterations disrupt critical signalling pathways, including NOTCH and MAPK, leading to the transformation of salivary gland cells into a malignant phenotype. Moreover, the interplay between tumour cells and the microenvironment, involving stromal and immune components, further contributes to tumour growth and progression.[13] To comprehensively understand the clinical presentation, histological attributes and treatment options associated with low-grade mucoepidermoid carcinoma in the lower lip of young patients, an integrative approach is crucial. By amalgamating clinical observations with detailed histopathological analyses, clinicians and pathologists can enhance their diagnostic accuracy and tailor-effective treatment strategies to address the unique needs of individuals affected by this variant of MEC.[14,15]

The rarity of this presentation underscores the need for broader consideration of diverse malignancy manifestations, highlighting adaptable diagnostic and therapeutic approaches. Through an in-depth exploration of this unique case, the study aims to enrich existing low-grade mucoepidermoid carcinoma literature. By revealing its occurrence in an unconventional site and emphasizing multidisciplinary diagnosis and management, these insights could aid healthcare professionals facing similar cases, potentially improving patient care and outcomes.

CASE REPORT

A 28-year-old female presented with a swelling in her lower lip that had been progressively enlarging over the past year. She described the initial presentation as a small, soft and spherical mass. However, she noticed a rapid increase in size, accompanied by changes in the swelling's characteristics. The swelling became irregular in shape and firm in consistency. Despite the growth, she did not experience pain, tenderness or associated symptoms. Notably, there were no palpable lymph nodes in the surrounding area [Figure 1].

High-resolution ultrasonography (USG) was performed to assess the nature of the swelling. Lower lip region was ultrasonographically scanned by high-resolution, high-frequency and near-focus linear probe with magnification for better tissue details. Post-op case of lower lip lesion histopathologically shows mucoepidermoid carcinoma (low grade). There is a presence of an oval hypoechoic lesion measuring 10.2 × 5.1 × 10.4 mm in the lower lip midline region. On colour Doppler application, mild peripheral vascularity was seen [Figure 2].

Microscopic examination of the excised specimen revealed a well-circumscribed, un-encapsulated and lobulated

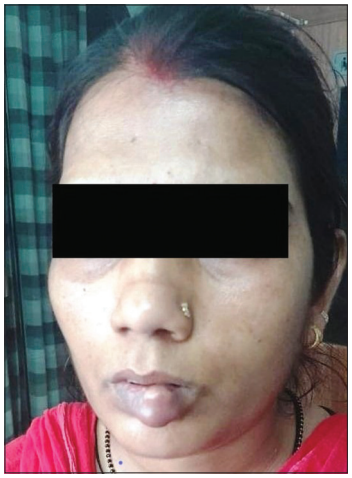

Figure 1: Photomicrograph showing dome-shaped swelling on lower lip

neoplastic lesion within a loose connective tissue stroma. Notably, large cystic spaces were observed within the oedematous connective tissue stroma, containing mucinous content. These cysts, along with small intra-cystic papillae, were lined by sheets of atypical large polygonal epidermoid cells and nests of non-specific intermediate epithelial cells. Islands of cells were observed floating within the mucinous pools. Focal areas exhibited squamoid cells with intercellular bridging. Importantly, the tumour cell nuclei were generally bland in appearance, with mild pleomorphism primarily seen in squamoid areas. Although there was minimal mitotic activity, no necrosis was observed. The stroma displayed slight chronic inflammatory cell infiltration [Figure 3]. Based on the comprehensive histopathological evaluation, the final diagnosis was established as low-grade mucoepidermoid carcinoma. The tumour's distinct characteristics, including the presence of cystic spaces, varying cell types, and infiltration of stroma, were consistent with this diagnosis. The application of the periodic acid–Schiff (PAS) stain in mucoepidermoid carcinoma (MEC) histopathology reveals distinctive features. This stain highlights the presence of mucin, which is a key component in MEC's composition, thus aiding in the identification of mucinous cystic spaces and pools within the tumour. The PAS stain enables the visualization of mucin-producing cells, a hallmark of MEC, offering valuable diagnostic insights by distinguishing this carcinoma from other salivary gland neoplasms. This stain contributes to a comprehensive histopathological analysis, facilitating accurate classification and characterization of MEC [Figure 4]. The utilization of mucicarmine stain, a specific diagnostic stain for MEC, facilitates the identification of mucin within tumour cells. Microphotographs stained with mucicarmine reveal mucicarmine-positive mucous cells and the contents of microcystic spaces, indicated by their magenta pink cytoplasm. The staining pattern demonstrates the presence of large mucous cells with abundant mucin, intermediate cells featuring basaloid nuclei and epidermoid cells characterized by ample cytoplasm [Figure 5]. The distinct staining characteristics align with the histopathological features indicative of low-grade MEC, offering valuable diagnostic insights.

The patient's post-operative recovery was uneventful. The surgical excision of the tumour had been successful in removing the neoplastic lesion completely. Regular follow-up appointments were scheduled to monitor the patient for any signs of recurrence or complications. This case highlights the importance of considering a wide range of diagnostic possibilities when encountering lip swellings, even in young individuals. The identification of low-grade mucoepidermoid carcinoma through thorough clinical evaluation, imaging and histopathological analysis

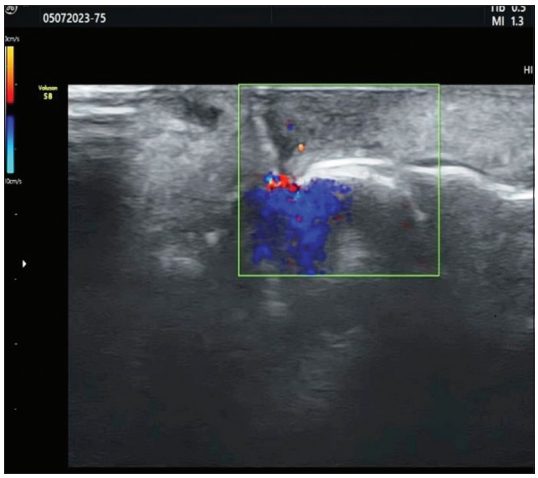

Figure 2: Oval hypo-echoic lesion measuring 10.2 × 5.1 × 10.4 mm in the lower lip midline region

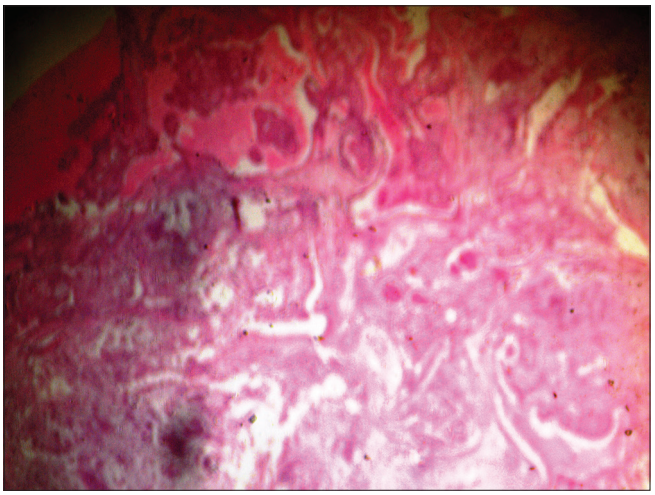

Figure 3: Photomicrographs demonstrating a sheet of tumour epidermoid, intermediate, and mucin-producing cells intermixed with some microcytic spaces 20X

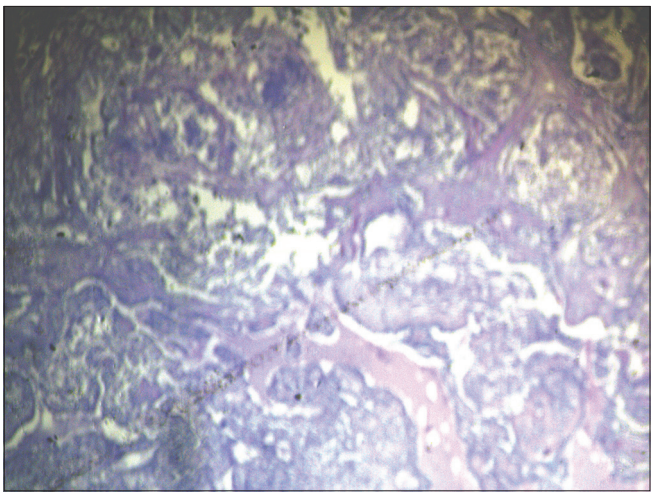

Figure 4: Photomicrograph showing PAS-stained mucinous cystic spaces within the tumour cells 20X

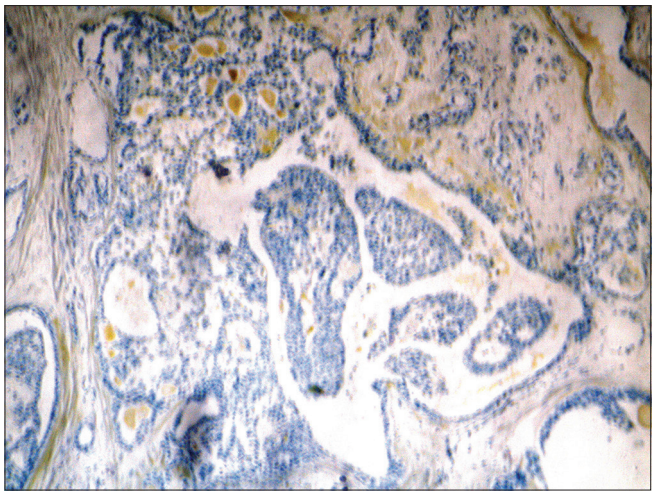

Figure 5: Photomicrograph showing mucicarmine-positive mucous cells and the contents of microcystic spaces, 20X

underscores the significance of a multidisciplinary approach in managing such cases.

DISCUSSION

The case of a 28-year-old female with a rapidly enlarging lower lip swelling, subsequently identified as low-grade mucoepidermoid carcinoma (MEC), provides insights into diverse clinical presentations, diagnostic challenges and management strategies for this rare tumour. This report delves into the case's contextual relevance vis-à-vis existing literature, MEC's histopathological features, molecular mechanisms and the broader landscape of minor salivary gland tumours.[15,16] The painless, enlarging lower lip swelling's clinical presentation mirrors a hallmark of MEC and aligns with prior reports, underscoring MEC's consideration even in younger patients, as evidenced by Sidhu *et al.* (2016)[15] in a comparable case. Analogous clinical scenarios have been documented across different anatomical sites, underscoring the tumour's varied impact.[1,6]

Histopathological analysis of the excised specimen reveals well-defined features consistent with established characteristics of low-grade MEC. The presence of distinct, lobulated neoplastic lesions containing mucinous cystic spaces and varied epithelial cell types aligns with recognized MEC histopathological patterns, as documented by other studies.[14,15,17] Similar observations were made by Chan *et al.* in 2007[6] and Manenzhe *et al.* in 2017,[1] highlighting MEC's heterogeneous cellular makeup. Identification of atypical polygonal epidermoid cells, intermediate cells and mucinous pools in the current case further supports alignment with prior case reports.[17,18] Focal squamoid regions and mild pleomorphism, typical of lower-grade MEC, further affirm the diagnosis.[14]

Molecular mechanisms play a pivotal role in MEC development, including genetic aberrations such as the MECT1-MAML2 gene fusion, which contribute to tumourigenesis.[13] Although not explored here, comprehending the molecular profiles of similar cases could unveil underlying pathogenic mechanisms, potentially guiding targeted therapies and prognostic insights. The surgical strategy applied aligns with established MEC management. Complete excision with clear margins is essential for preventing recurrence and ensuring favourable outcomes. The histopathological assessment's lack of necrosis and high mitotic activity align with lower-grade mucoepidermoid carcinomas' behaviour, affirming the selected surgical intervention's validity.[19]

Within the broader landscape of minor salivary gland tumours, differential diagnosis of intraoral neoplasms is intricate due to their varied presentations. Yih *et al.* (2005)[20] carried out a study on 213 cases of intraoral minor salivary gland neoplasms which underscores the spectrum within this category. Copelli *et al.* (2008)[21] discuss malignant tumours of intraoral minor salivary glands, accentuating precise diagnosis, especially for malignancies. This case, marked by an unusual site and atypical presentation, augments the knowledge in this domain, stressing meticulous clinical assessment and accurate diagnostics.[22] Ramesh *et al.* (2014)[19] performed a comprehensive study that delves into intraoral minor salivary gland tumours, offering insights into prevalence and characteristics within a specific region, enriching the comprehension of these neoplasms' occurrences. Stewart *et al.* (1945)[23] conducted a study on salivary gland mucoepidermoid tumours that contributed to the field's knowledge evolution. Namboodiripad in 2014[22] compiled a review work on the immunological markers for malignant salivary gland tumours and suggested the potential diagnostic aids of the same.

Some other noteworthy studies have advanced our understanding of minor salivary gland tumours. Spiro *et al.* (1978)[24] undertook a thorough clinicopathological analysis of salivary gland-originated mucoepidermoid carcinoma, expanding our clinicopathological comprehension. Auclair *et al.* (1992)[25] assessed grading criteria's significance in mucoepidermoid carcinoma's aggressiveness assessment. García-Cruz *et al.* (2010)[26] contributed insights about mucoepidermoid carcinoma of the lip. Mishra and Mishra (2014)[27] studied Indian cases over a decade, revealing minor salivary gland tumour

prevalence and epidemiology. Chandrasekar *et al.* (2016)[28] explored immunohistochemical markers for diagnosing salivary duct carcinoma and guiding diagnostics. Suvarna *et al.* (2018)[29] highlighted diverse tumour sites, like an unusual mucoepidermoid carcinoma case.

This case, along with existing research, adds to the ongoing discourse on the diagnosis and management of salivary gland tumours. To summarize, the presented case of a low-grade mucoepidermoid carcinoma in a young female patient underscores the importance of comprehensive clinical evaluation, accurate histopathological analysis, and consideration of molecular mechanisms in diagnosing and managing salivary gland neoplasms. The histopathological alignment with established patterns, the potential contribution of molecular insights, and the broader context of minor salivary gland tumours enrich our understanding of this rare tumour's intricacies. By correlating these findings with existing literature, clinicians can enhance their diagnostic acumen and contribute to improved patient care.

CONCLUSION

The correlation with existing literature emphasizes the importance of accurate diagnosis, comprehensive assessment and tailored management strategies for MECs. This case highlights the need for vigilance in recognizing unusual presentations and the significance of histological grading for appropriate treatment protocol. Further research and collaboration among clinicians can contribute to a deeper understanding of mucoepidermoid carcinomas and improve patient outcomes. The present case report stands as a vital contribution, shedding light on a rare manifestation and encouraging further exploration into diagnostic and therapeutic avenues for mucoepidermoid carcinomas in diverse anatomical locales.

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 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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