Mucoepidermoid Resembling Squamous Cell Carcinoma in the Premaxilla: A Diagnostic Challenge

Premaxillada Skuamöz Hücreli Karsinoma Benzeyen Mukoepidermoid: Tanısal Bir Zorluk

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ABSTRACT

Salivary gland malignancy represents a rare yet diverse entity that covers a heterogeneous spectrum of neoplasm of various clinical behavior, ranging from protracted, indolent to aggressive, life-threatening manner. Accurate histopathological diagnosis is imperative in terms of prognostication and further management planning. Herein, we report a case of an intermediate grade mucoepidermoid carcinoma of premaxilla masquerading as squamous cell carcinoma. Despite repeated biopsies, histological examinations failed to establish an accurate diagnosis. The patient eventually underwent wide surgical excision of the primary lesion with simultaneous bilateral supra-omohyoid neck dissection. Diagnosis dilemma encountered along with subsequent management are further discussed.

Key Words: Mucoepidermoid carcinoma, oral cavity cancer, minor salivary gland

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INTRODUCTION

Salivary gland malignancy is a rare entity which contributes around 5% of all head and neck tumours (1). Mucoepidermoid carcinoma (MEC) is the most common malignant neoplasm arising from salivary glands (1). It constitutes about 30% of all salivary gland tumours (2). Histologically, MEC can be categorized into mild, intermediate and high grade (1,3-5). Histological findings in high-grade MEC includes solid islands of intermediate and epidermoid cells, which resembles the histological finding in squamous cell carcinoma (SCC), attributing to the its diagnostic dilemma (1). Meticulous evaluation ought to be undertaken to avoid diagnosis inaccuracy.

CASE REPORT

A sixty-eight-year-old lady with no known comorbid presented with painless swelling over the alveolus for the past three months. The mass has progressively increased in size which eventually prompted her for the clinic visit. There were neither history of betel nut chewing nor alcoholic consumption. She also denied family history of head and neck malignancy or any prior radiation exposure. Clinical examination revealed an irregular mass measuring 3 cm x 3 cm located over the premaxilla region (Figure 1). The mass was non-tender, hard in consistency with irregular surface and border. Examination of other subunits of the oral cavity were otherwise unremarkable.
There was no palpable neck node. Contrast-enhanced computed tomography (CT) performed, demonstrated asymmetrical thickened mucosa over the premaxilla region, with intact underlying cortical bone. No regional and distant metastasis was noted.

An incisional biopsy under local anesthesia was performed. Histopathological examination (Hpe) revealed presence of small islands of stratified squamous epithelium, dyskeratotic cells with keratin pearl formation as well as microcystic formation (Figure 2). The Hpe was suggestive of MEC and SCC. Hence, a repeated incisional biopsy was performed, given the uncertainty in diagnosis.

The repeated sample showed presence of para-keratinised stratified squamous epithelium tumour composed of island, clusters, nests and strains of bland-looking neoplastic epithelial cells. They exhibited features of nuclear pleomorphism, hyperchromatic nuclei and microcystic formation. (Figure 3). No definite conclusion could be drawn from the second biopsy. The patient was counselled and underwent wide local excision of the primary lesion with bilateral supra-omohyoid neck dissection. Intraoperative frozen section analysis of the excised tumour demonstrated clear surgical margin. Final diagnosis confirmed intermediate grade MEC. She recovered well post-operatively and has been on active clinical surveillance with no evidence of recurrence till date (Figure 4).
First described by Massao and Berger back in 1924, the mucoepidermoid tumour was regarded as a benign lesion which composed of distinct group of neoplasm of salivary gland (2-4). It is reported to have slight female predominance with a ratio of 3:2 (2). MEC is more prevalent in the fifth and sixth decades of life (2). Approximately two-thirds of MEC arise within the parotid gland (1). The hard palate represents the most frequent site of a minor salivary gland MEC, followed by retromolar region, floor of the mouth, the buccal mucosa and the lower lip (2). Regardless of its location, the most typical clinical manifestation of a minor salivary gland MEC is an asymptomatic swelling that insidiously increases in size (5). Other presentations include pain, ulcer, regional nodal disease and resorption of the underlying bone, which often indicate disease advancement.

MEC is composed of three histological types of cells, namely (1) small basal and intermediate cells, (2) tall columnar or rounded mucus-forming cells and (3) epidermoid cell (3). Highly malignant MEC exhibited predominance of epidermoid and intermediate cells. Subsequently, Foote and Frazells reclassified all mucoepidermoid tumour as malignant, with different grading based on the degree of hyperchromatism in intermediate cells and the numbers of mucous-secreting cells (4). World Health Organization (WHO) eventually categorized it as malignancy in 1990, and coined the term mucoepidermoid carcinoma (4).

The aetiology of MEC remains elusive. History of radiation exposure has been suggested as a possible risk factor. Histologically, mucoepidermoid carcinoma displays a heterogeneity of behaviours. Based on the five histopathological characteristics (1) intracystic component, (2) neural invasion, (3) necrosis, (4) mitosis and (5) anaplasia, it can be categorized into low, intermediate or high grade (1). Low-grade tumours show a predominance of mucous – secreting cells with minimal cellular atypia. Intermediate grade tumours comprises of a mixture of the solid and cystic component with an increased number of intermediate cells. High-grade tumours displays aggressive features of perineural/vascular invasion and aphasia. Its predominant component being epidermoid cells with scanty mucous cell seen (1)(3).

The overlapping histological morphologies impose a significant challenge to differentiate between a moderate and high-grade MEC from SCC (1)(4)(6). Differentiating these two entities is deemed necessary as to prognosticate the survival outcome and to outline the possible treatment plan. The usage of Period acid-schiff (PAS) and mucicarmine stains were reported to be satisfactory in differentiating the two pathologies (6). This however, may not be applicable when squamous cell carcinoma invades the salivary gland structure, which may be easily mistaken as a MEC. Necrotic areas, which are weakly mucicarmine-positive, may also contribute to the diagnosis error. In our case overlapping features of keratin pearls with occasional microcystic formation led to the diagnosis of SCC.

Immunohistochemistry staining with various cytokeratins (CKs) can also help to differentiate the two entities due to their difference of tumour origin. SCC originates from epidermis and is positive for CK10, whereas MEC originates from the excretory duct of salivary gland and is CK13 positive (6). The other distinctive feature observed by Arojo et al. is that SCC is strikingly positive for CK14(6). Intraoperative frozen section provides insight into the adequacy of margin and to determine the involvement of perineural or lymph vascular involvement. Additionally, it has a respectable accuracy in differentiating benign lesions from malignancies, thereby avoiding under or overtreatment (7). Nevertheless, the employment of a frozen section in differentiating a high-grade MEC from squamous cell carcinoma may be of limited value due to the overlapping of histological morphology. Radiological modalities such as CT and Magnetic Resonance Imaging (MRI) are invaluable in delineating the extent of the pathology. Evidence of bony invasion is best seen with CT (2). MRI plays role in evaluating the soft tissues involvement of the surrounding oral cavity. Low-grade MEC behaves indolently with radiological features of lobulated or cystic lesion with underlying intact periosteum. Intermediate grade tumour may exhibit features of bony erosion, whereas high-grade MEC may show extensive cortical bony destruction and regional or distant metastasis (2).

The rationale of performing a selective neck dissection in a node-negative MEC remains contentious. Darling R. et al. reported an overall rate of nodal diseases as high as 40% in patients with high-grade MEC, whereas no regional metastasis was found in low-grade MEC (8). Sood S et al reported a 40 % incidence of lymph node metastasis in intermediate and high-grade tumours (9). An ipsilateral elective neck dissection is best performed when primary lesions are of a higher grade owing to their predilection of nodal metastasis.