Primary Cutaneous Mucoepidermoid Carcinoma - Masquerading as a Benign Cyst

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ABSTRACT

Mucoepidermoid carcinoma (MEC) of the skin is a highly uncommon neoplasm, although it exhibits a higher prevalence in the main and minor salivary glands, constituting roughly 30% of all malignant tumours originating from these glands. In this report, we provide a clinical case involving a 52-year-old male patient who presented with swelling located on the posterior aspect of his neck. The patient's condition was classified as a benign sebaceous cyst based on clinical and radiological findings. The histopathological examination indicated the presence of primary cutaneous mucoepidermoid carcinoma (PCMEC). Histopathology is therefore crucial for early diagnosis as well as prompt treatment.

Keywords: Benign sebaceous cyst, primary cutaneous mucoepidermoid carcinoma.

INTRODUCTION

The tumor known as mucoepidermoid carcinoma (MEC) is relatively prevalent. About 30% of malignant tumors developing in the major and minor salivary glands are primary MEC.¹ The salivary glands, particularly the parotid but also the submandibular and small salivary glands, are most frequently affected. There is a 1.5–4% incidence in the lacrimal gland. Furthermore, apart from the breast, thymus, thyroid, pancreas, oesophagus, lung, bronchi, and trachea, there have been infrequent reports of its occurrence in the oesophagus, lung, bronchi, and female genital tract.¹,² The medical literature has described a total of 20 cases where cutaneous involvement was identified as the primary cause.³ As a result, it's critical to rule out metastases from a distant place when the skin is affected. In order to distinguish primary epidermal or adnexal neoplasms from metastatic tumours, several publications have suggested using p63, a member of the p53 gene family.⁴ High-risk Human Papilloma virus (HR-HPV)-mediated carcinogenesis is believed to be the primary cause of the dramatically higher incidence of oropharyngeal malignancies over the past three decades, particularly in young women. The SEER 9 data also shows a tendency towards an increase in the incidence of mucoepidermoid carcinoma (MEC) in females between the ages of 15 and 34. This raises the question of whether HR-HPV can also play a role in the development of MEC cancer.⁵

CASE REPORT

A 52-year male, came to the surgery OPD of GMERS hospital, Valsad with a complaint of swelling on back of his neck for six months. On examination, swelling was soft measuring 2.5 x 2.5 cm. Clinically and radiologically it was diagnosed as sebaceous cyst. The lesion was surgically
removed with intact capsule and sent to the histopathological department of GMERS college. The patient was then lost to follow up.

**Histopathological Examination**

On gross examination, single, irregular, skin covered, greyish brown, soft to firm in consistency measuring 2.5 x 2 x 0.5cm. On cut section, greyish brown, solid cystic areas seen (Figures 1 and 2).

Microscopic examination showed stratified squamous epithelial lining beneath which tumor cells were composed of a squamoid type with keratinization, intermediate cells and mucous secreting cells. Tumor cells are arranged in solid, trabecular, nests and in cystic forms. Squamoid type cells are polygonal, with intercellular bridging, mildly pleomorphic nuclei and moderate amount of keratinous cytoplasm. Intermediate cells are round to oval with bland nuclei and scant amount of cytoplasm. Mucous secreting cells are large, with mild pleomorphic nuclei and pale mucinous cytoplasm. Few mucous secreting cells are having clear cytoplasm. At places dense lymphoplasmacytic infiltrate, haemorrhage, congested blood vessels and few adnexal structures of skin is seen. Histopathological diagnosis of primary cutaneous mucoepidermoid carcinoma, Intermediate grade was given. (Figures 3-7)
DISCUSSION
Stewart et al. reported a series of 45 salivary gland tumors in 1945, establishing mucoepidermoid carcinoma as a distinct entity. Mucous-secreting cells, epidermoid cells and intermediate basaloid cells are present in various combinations in this tissue. All variants are currently treated as malignant, despite early attempts to discriminate between benign and malignant variants. The two most common subtype classifications in histology are low-grade (well-differentiated) and high-grade (poorly differentiated), while an intermediate grade has also been suggested. Additionally, three groups have been identified based on the percentage of mucin-secreting cells, including squamous, intermediate and clear cells, as well as the maturity level of these cells. Immunohistochemical studies are positive for CK7, PanCK, EMA and carcinoembryonic antigen while they show negativity for CK20 and anti-human gross cystic disease fluid protein.

The biologically significant, transcriptionally active HR-HPV was found in about one-third of MEC. Given that MEC1-MAML2 fusion transcripts are believed to be an early event in multistep carcinogenesis, it is possible that HR-HPV oncoproteins promote MEC as a later event. This may be caused by the HR-HPV E6 and E7 oncoproteins' cumulative effect on the overall loss of tumour suppression function. This work expands the range of relationships between HR-HPV and head and neck neoplasia and can provide the justification for subsequent investigations that deepen our understanding of the etiology.

Mucoepidermoid carcinomas (MECs), which are neoplasms originating from the skin, exhibit a comparatively low incidence rate. To date, a total of 20 cases have been reported.

The typical differential diagnosis for malignancies in this region mostly encompasses squamous cell carcinoma, adenocarcinoma, undifferentiated carcinoma, melanoma, lymphoma, and infrequently, mucoepidermoid carcinoma.
The site of MEC on back of neck is very rare, presenting with unusual symptoms of swelling.

The tumour is graded based on morphologic features such as:
1. Neural invasion
2. Proportion of cystic and solid elements
3. Necrosis
4. Mitotic rate
5. Anaplasia

Each of these characteristics is assigned a numerical value, and the total sum of these values is used to classify the tumour grade as either low, intermediate, or high. The tumour grade of the present instance was evaluated using the designated criteria and scoring system, yielding an intermediate grade. The study conducted by Spiro et al. shown that a significant proportion of low grade (90%) and high grade (42%) mucoepidermoid carcinomas located in the head and neck region achieved complete remission after a decade. Furthermore, it has been noted that in addition to high grade and bone invasion, variables such as age over 60, pain, positive cervical nodes, and facial nerve palsy are seen as additional unfavourable prognostic indicators.

CONCLUSION
We hence emphasize that while working upon any odd cyst-like lesions, malignancy should always be considered in the differential diagnosis to ensure an early diagnosis, complete removal and a better outcome.

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REFERENCES