Giant Mucoepidermoid Carcinoma of the Parotid Gland: A Case Report and Review of Literature

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Abstract

Mucoepidermoid carcinoma is the most frequently encountered salivary gland carcinoma usually presenting as small, painless, asymptomatic mass. Presented is a 54-year-old male security personnel with a 5-year history of giant locally advanced, nodular, and ulcerated left sided facial tumour diagnosed as mucoepidermoid carcinoma on histology. Mucoepidermoid carcinoma can attain a grotesque dimension with management challenges. We present this case, as we have not encountered MEC as large as this and conclude that it should be considered as a differential diagnosis of facial tumours.

Introduction

Salivary gland carcinomas are a rare and clinically diverse group of neoplasms among which mucoepidermoid carcinoma (MEC) are the most frequently encountered [1]. The most common presentation is a painless, asymptomatic mass. Facial nerve weakness or paralysis occurs in 7-20% of patients with malignant parotid tumours with poor prognosis. Skin fixation, ulceration or fixation to adjacent structures may indicate malignancy [2]. The most important procedure for the treatment is a histological examination, which has statistical correlation with patients’ survival [1,3]. Traditionally MECs were histologically classified as low and high grade based on the relative proportion of cell types [1]. The main therapeutic method in the treatment of MEC is surgical resection [1]. Radiation therapy is considered the cornerstone of adjuvant therapy as no chemotherapy has been proven effective as single modality of therapy [2]. We present a giant size MEC of the left parotid gland in a 54-year-old male to highlight the fact that this lesion can present in grotesque dimensions with management challenges.

Case Report

A 54-year-old male security personnel presented as a referral with a 5-year history of growth on left side of his face, had a previous biopsy 3 years ago in another facility but did not state histopathology result. The growth progressively increased in size, initially smooth-surfaced but over the last 3 years ulcerated, discharging odourless fluid, and bled on slight contact. There was no history of fever, dysphagia, odynophagia, and dryness of mouth, difficulty in opening mouth, no speech defect, and no increase in size during mastication. There was also no history of trauma, irradiation, chronic cough, drenching night sweats, and weight loss. Defaulted from treatment from a hospital facility where he had a biopsy. However, he later presented at the maxillofacial unit of the University of Calabar Teaching Hospital from where he was referred to the surgical out patient department.

Examination revealed a middle-aged male in general good health, with a left-sided spherical facial multinodular exophytic growth measuring about 22 x18 cm with multiple ulcerations that discharged clear fluid. It was attached to the overlying skin and fixed to the underlying structures. Consistency varied, the superficial part was soft while the underlying area was hard. Mass was non-tender and bled on slight contact. There was no regional lymphadenopathy and evidence of facial nerve palsy (Figure 1). Examination by the Ear, Nose, and Throat surgeon revealed no abnormal finding. Chest and abdominal examination were unremarkable. A clinical diagnosis of malignant parotid tumour was made.

During hospitalization, he developed a cystic swelling at the most dependent part of the swelling; this was aspirated and sent for analysis and cytology.

Haemogram revealed a packed cell volume of 42%, white blood count of 9.5x10⁹/L, lymphocytes - 31%, neutrophils - 68%, eosinophils - 1%, and urinalysis was normal. The discharge from the lesion and aspirate from the cyst contained amylase on analysis. Cytology from the aspirate was negative for malignancy. Skull X-ray showed left sided soft tissue swelling, there was no evidence of bony involvement. Incision biopsy result revealed tumour composed of haphazardly dispersed cyst and irregular tumour nests composed of mucous, squamous (epidermoid) and intermediate cells in variable proportions. Immunohistochemistry showed the relative proportion of cell types [2]. The main therapeutic method in the treatment of MEC is surgical resection [1]. Radiation therapy is considered the cornerstone of adjuvant therapy as no chemotherapy has been proven effective as single modality of therapy [2]. We present a giant size MEC of the left parotid gland in a 54-year-old male to highlight the fact that this lesion can present in grotesque dimensions with management challenges.

Discussion

Mucoepidermoid carcinoma is one of the most frequent epithelial
Pathology.

A cystic collection would be a pointer to the affected organ and possible attendant challenges of management. The need for early presentation neglected, this lesion can attain very grotesque dimensions with the poor prognosis.

This tumour is well known to display a widely diverse biologic behaviour and variable clinical manifestation, which seems to correlate with tumour stage and grade. High grade MEC is highly aggressive tumour while its low-grade counterpart usually demonstrates a more benign nature though metastases have also been described in low grade MEC [7]. Metastasis may be lymphatic (cervical lymph node) and haematogenous. The most common site for the later is lungs [13]. Others include the liver, brain, skin [14], including other sites.

Rapidis et al. [1] in their series reported sizes of the lesions that ranged between 1.5 and 10cm. MEC is a rare disease of the salivary glands usually occurring as a small tumour [3]. It can secondarily involve the skin and delay in diagnosis result in unnecessarily large surgical procedures [11]. We present this healthy-looking patient with a locally advanced ulcerated MEC to highlight the fact that when neglected, this lesion can attain very grotesque dimensions with the attendant challenges of management. The need for early presentation and diagnosis and treatment is advocated for improved outcome.


Histology is the most important investigation for treatment [3]. The original description of MEC by Stewart et al. showed that salivary gland duct composed of several cell types (mucus secreting, basaloid, intermediate and epidermoid) and represents the histogenetical origin of MEC [12]. Traditionally MECs were histologically classified as low and high grade based on the relative proportion of cell types [13]. A three-tier grading scheme has replaced the previous classification, incorporating a third, intermediate grade that essentially resemble histologically with lower than with high-grade lesions. Results suggest that the intermediate grade tumours had a clinical behaviour closer to that of low-grade tumours [1]. In the three-tier grading, the low-grade tumour is characterised by the presence of mucus cells in greater number than epidermal cells including prominent cysts and mature cellular elements (Figure 2). In the intermediate group, the ratio of mucus to epidermal cells is equal, with fewer and smaller cysts, increased pleomorphism, and mitotic figures. The high grade is characterised by epidermoid cell greater than mucus with solid tumour cell proliferation [14]. Our experience was that of a low-grade tumour with locally advanced lesion without facial nerve palsy or regional lymphadenopathy on clinical evaluation.

Pathogenesis of this tumour based on genetic mechanisms is still poorly understood. Further studies of MECs both at molecular and clinical levels are needed to define biological groups with the same tumour entity and test the value of various therapeutic modalities [14]. Recent studies demonstrated that the epidermal growth factor receptor (EGFR) is expressed sharply in the cell membranes of parotid MEC and lymph node metastasis. EGFR targeting agents have potential to be used as therapy [15].

The large size of the tumour may be an obstacle in resection during an operation [3]. Surgical treatment is the main therapeutic method [11]. However, in giant lesions with skin involvement and fixicity to deeper structures, adjuvant therapy with radiation and chemotherapy remain possible options. Chemotherapy as single modality of therapy has not been proven to be effective [2]. Mendenhall et al. reporting on the role of radiotherapy in the treatment of salivary gland carcinomas stated that in advanced stages as was our experience, the efficacy of radiotherapy modality has a curability percentage approximately 20% [16]. Radiation therapy is considered the cornerstone of adjunctive therapy and plays an important role in those patients with advanced stage disease [2,17]. However, some studies report that MECs are radio resistant [18].

The biological behaviour is still hiding many secrets. Major determinants of survival are histology and clinical stage [1,14]. A trend towards better overall survival for patients less than 50 years has been suggested by Guzzo et al. [19]. The overall survival rate of patients with giant MECs has not been generally reported as most reports relate survival with histologic grade and metastasis. Patients with low grade tumours without nodal or distant metastasis 5-year survival is 75-95% while high grade tumours with lymph node metastasis at time of diagnosis have a 5-year survival of only 5%. Overall 10-year survival is 50% [2]. We present this case as we have not encountered a tumour as large as this and to highlight the challenges of management of locally advanced giant MEC lesion. It is concluded that MEC if neglected can attain a grotesque dimension with poor prognosis.

References


