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Primary Cutaneous Mucoepidermoid Carcinoma of Parotideomasseteric Region: A Rare Case Report

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ABSTRACT

Mucoepidermoid carcinoma is a distinctive salivary gland malignancy and its presentation as a primary cutaneous carcinoma is a very rare occurrence. It presents grossly as an ill-defined mass with firm to hard consistency and can have cystic areas filled with mucin. Microscopically, they are characteristically composed of mucinous, intermediate and squamoid tumour cells forming cystic and solid patterns. Here we present a case of a 79-year-old female who presented with an ulcerative nodular exophytic growth over right parotideomasseteric region. CECT neck revealed it to be a heterogeneously enhancing lesion arising from the skin with the underlying subcutaneous fat, soft tissues and parotid gland to be normal. On histological and immunohistochemical examination, it was diagnosed as primary cutaneous intermediate grade mucoepidermoid carcinoma.

Key words: Mucoepidermoid carcinoma, cutaneous, rare

INTRODUCTION

Mucoepidermoid carcinoma is the most common glandular epithelial malignancy of the major salivary gland[1] which affects adults as well as children[2,3]. It predominantly involves parotid and intraoral minor salivary glands[4] but are also found to develop in lacrimal passages, sinonasal tract, oesophagus, bronchi, thymus, thyroid, breast, pancreas, uterine cervix and prostate[2]. Primary cutaneous mucoepidermoid carcinoma is a rare entity that often poses diagnostic challenges due to its uncommon presentation and overlapping features with metastasis from salivary neoplasm and other skin tumours.

CASE PRESENTATION

A 79-year-old female presented with a 7 months history of a progressively enlarging, painless, soft to firm nodule on the right parotideomasseteric region. The lesion was initially small but had grown to approximately 4x4x1.5cm in measurement by the time of presentation. There was no history of any trauma and the patient had no significant prior medical history. On examination, the nodule was well-circumscribed, firm, and non-tender, with no overlying ulceration or pigmentation. Regional lymphadenopathy was absent and there were no signs of systemic involvement. The routine haematological and biochemical parameters were found to be normal.

CECT neck revealed it to be a well-defined broad based polypoidal exophytic enhancing lesion seen arising from the skin in right parotideomasseteric region. The underlying subcutaneous fat and soft tissue were normal. Few subcentimetric homogenously enhancing discrete lymph nodes in station II, III and V were seen with largest measuring 5.8mm in right mid jugular node station. Both the parotids, submandibular glands, thyroid, nasopharynx, oropharynx and hypopharynx were found to be normal.

The microscopic examination showed an exoendophytic tumour lined by fibrinopurulent material. The underlying dermal tumour is arranged in cords and nests of epidermoid cells showing moderate nuclear pleomorphism (H&E, 20x; Figure 1). Minor component shows cystic spaces filled with mucin and large mucus secreting cells having eccentric nucleus and abundant fluffy cytoplasm. Intervening areas showing foreign body giant cell reaction. Mitosis was found to be 5-6/hpf. (H&E, 40x; Figure 2).

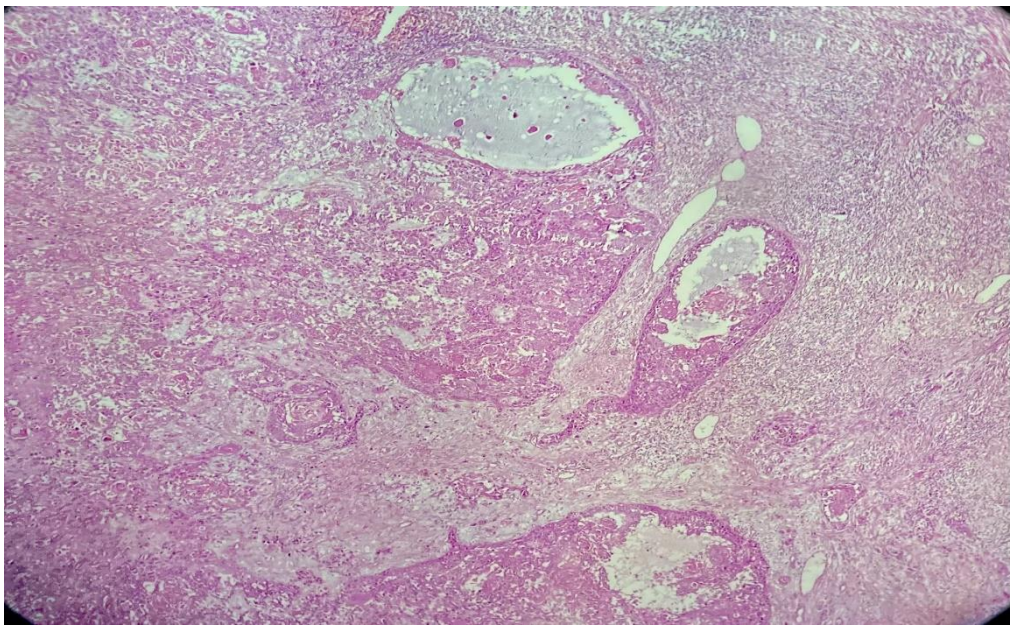

Figure 1: Microscopic examination shows cords and nests of epidermoid cells with cystic spaces filled with mucin and large mucus secreting cells. (H&E, 20x)

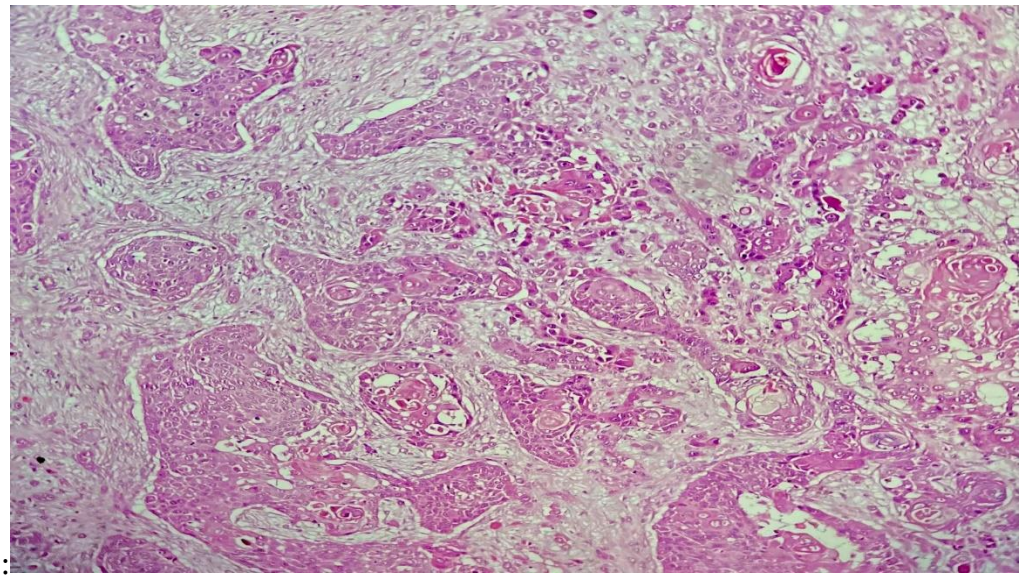

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Figure 2: Microscopic examination shows nests of epidermoid cells showing moderate nuclear pleomorphism. Mucus secreting cells having eccentric nucleus and abundant fluffy cytoplasm noted. Occasional mitotic activity seen. (H&E, 40x).

Positive staining for periodic acid-Schiff (PAS) in intracytoplasmic granules and extracellular mucin was seen (100x) (Figure 3). On immunohistochemistry, immunoreactivity for p63 was noted, thereby excluding metastasis. (40x) (Figure 4). CK7 and CEA were found to be negative. Overall features confirmed the diagnosis of cutaneous intermediate-grade mucoepidermoid carcinoma.

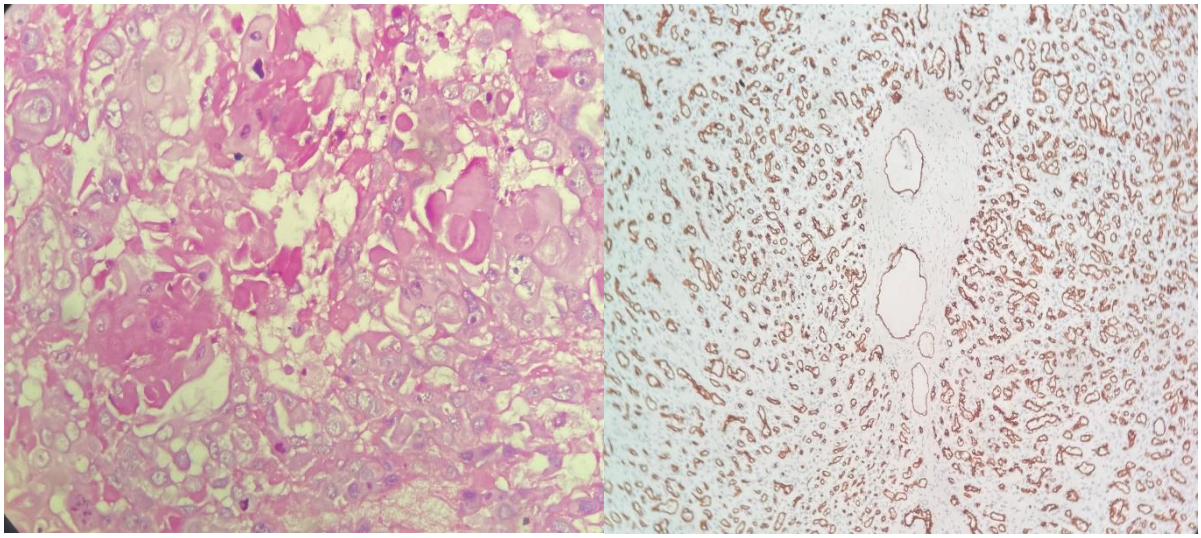

Figure 3 shows immunoreactivity for PAS stain for intracytoplasmic and extracytoplasmic mucin. (100x)
Figure 4 shows immunoreactivity for p63 (40x).

The patient was followed up for a period of six months and no signs of local recurrence or metastasis were noted.

DISCUSSION

Mucoepidermoid carcinoma is the most common primary salivary gland malignancy and was first described in 1945[5]. It is predominantly seen in parotid gland (45%) and intraoral minor salivary glands of palate (21%)[1,4]. It may also involve lacrimal passages, sinonasal tract, oesophagus, bronchi, thymus, thyroid, breast, pancreas, uterine cervix and prostate[2]. It is seen in adults as well as children [2] with a peak incidence in the second decade of life[3]. These are also found to develop secondary to radiation or chemotherapy. Clinically, they present as soft to firm nodular, circumscribed, erythematous to ulcerative infiltrative mass with cystic areas. It's presentation as a primary cutaneous carcinoma is very rare with few cases documented and first described by Galleger in 1959[6]. The primary differential diagnosis includes other cutaneous malignancies such as basal cell carcinoma, squamous cell carcinoma, and metastatic tumours. The literature suggests that primary cutaneous mucoepidermoid carcinomas are thought to arise from an ectopic salivary gland which are derived from sweat glands[7,8]. Primary cutaneous involvement include head and neck as the usual sites, though the axilla, vulva, finger and foot may also be involved.
Adenosquamous carcinoma and primary cutaneous mucoepidermoid carcinomas were initially thought to be identical conditions but Riedlinger *et al* [9]concluded that the two conditions have a different clinical course and histopathology. On microscopic examination, cutaneous mucoepidermoid carcinomas are predominantly dermal tumours characterized by variable squamoid cells showing atypia, intermediate cells with solid/ cystic growth patterns and peritumorous fibrosis. Abundant mucin producing cells are noted which show PAS positivity. The adenosquamous carcinoma on the other hand are predominantly intraepidermal and show less mucin-producing cells and more atypical carcinomatous cells. In addition, they have a more aggressive course. On

immunohistochemistry, p63 positivity is seen in primary cutaneous mucoepidermoid carcinoma and helps in differentiating it from metastatic carcinoma[10].

These tumours are generally less aggressive and are treated with complete surgical excision[11]. Adjuvant therapies such as radiotherapy or chemotherapy are generally reserved for high-grade tumours. The 10-year survival rate of intermediate grade mucoepidermoid carcinoma is approximately 70%[12,13].

CONCLUSION

Primary cutaneous intermediate-grade mucoepidermoid carcinoma is a rare and potentially aggressive tumour which requires early detection and appropriate management. The prognosis is generally favourable with complete surgical excision and clear margins. Clinicians should consider cutaneous mucoepidermoid carcinoma as a differential for tumours presenting in parotideomasseteric region and rule out other primary salivary gland or metastatic neoplasms.

Declaration by Authors

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