Epithelial-myoepithelial carcinoma of the hard palate

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ABSTRACT
Epithelial-myoepithelial carcinoma (EMC) is one of the tumours of the salivary glands. It rarely occurs and accounts less than 1% of all salivary gland tumours. It commonly arises in the major salivary gland especially in the parotid gland, and less frequently in the minor salivary gland. We are reporting a case of a young man diagnosed with EMC in the right hard palate that extended into the maxillary sinus and pterygopalatine fossa. The incidence, clinical features, treatment of choice and the disease progression are discussed in this report.

Keywords: Epithelial-myoepithelial, salivary gland, hard palate, neoplasm

INTRODUCTION
In the major and minor salivary glands, certain neoplasms that arose from here are composed solely of myoepithelial cells. 1 Myoepithelial cells have contractile properties and display both epithelial and smooth muscle phenotype. 2 They are important elements in numerous salivary gland tumours, however, pure myoepitheliomas is unusual. Myoepithelial carcinoma or epithelial–myoepithelial carcinoma (EMC) is rare and its incidence is reported to be less than 1% of all salivary gland tumours. 1-5 Majority of the cases occur in major salivary gland, particularly the parotid gland 3, 4, 6 and uncommonly, it will involve maxillary antrum and palate. 3 We are reporting a case of a young man that was presented with EMC that arises from the minor salivary gland of the hard palate.

CASE REPORT
A 28-year-old man was referred to our centre from a private hospital with five-year history of right intermittent palatal pain. He had undergone two dental extraction procedures but the pain persisted. He also noted a swelling at the right palatal region that was progressively increased in size over that period of time but he did not seek treatment for it beforehand. Examination of the oral cavity revealed a bul-
A Contrast-Enhanced Computed Tomography (CECT) scan of the paranasal sinuses showed soft tissue mass arising from the right maxillary sinus, heterogeneously enhanced with area of hypodensity suggestive of cystic component within. Posterior, lateral and medial walls of the maxillary sinus were remodeled with associated erosion. Right pterygoid bone was eroded, and the plane between the mass and the medial and lateral pterygoid muscles was obliterated at its attachment. The mass was encroached into the right nasal cavity with obliterated plane with the inferior turbinate. The nasal septum was not deviated. Tissue biopsy from the right nasal cavity revealed myoepithelial carcinoma that was positive for S100.

Treatment consisted of a tracheostomy, submandibulectomy, subtotal maxillectomy and selective neck dissection. The patient was nursed overnight in the intensive care unit and was discharged four days later with a tracheostomy and on Ryle’s tube feeding. Histopathology of the hard palate mass revealed malignant cells in lobules, mostly in solids structures. There were bilayered duct-like structures lined by cuboidal cells with dense, fine granular cytoplasm. The outer cells were round to oval mildly pleomorphic cells with vesicular nucleous, inconspicuous nucleoli and ill-defined eosinophilic granular cytoplasmic border. The findings were consistent with EMC. There was evidence of vascular invasion and the tumour was located 2mm away from the lateral soft tissue margin. Tissue sample from the pterygoid muscle showed presence of malignant infiltration.

The patient was referred to the Oncology team for completion of the treatment. He completed a course radiotherapy and was doing well on follow up.

DISCUSSION
Myoepithelial cells are found in various tissues of man and animal. They can be found in the major and minor salivary glands, secretory portion of the sweat glands, ducts of mam-
mary glands and Bartholin’s glands. They also present in the mucus-producing glands of the trachea and oesophagus, prostate and lacrimal glands. 

Fonseca and Soares reported EMC according to WHO classification of salivary glands tumour, is an intercalated duct originated tumour of low-grade malignancy. In the previous literatures, the incidence of the tumour peak at seventh and eighth decades of life with female preponderance. Many reports on EMC indicated the tumour occurs more commonly in the major salivary gland, in particular, the parotid gland. Around 10-15% of cases are found in the minor salivary gland.

The clinical presentation of the tumour in each gland may differ. EMC in the major salivary gland may exhibit as a painless mass but in some occasion pain and facial nerve palsy were reported. However, the features for EMC in the minor salivary gland was not well-reported as the incidence of the tumour in this region is low. In our patient, he reported painful dentition with a slow growing palatal mass. He did not complain any symptoms in the maxillary region despite having a tumour mass presented in the right nasal cavity and evidence of the tumour seen in the right maxillary sinus as noted from the CT scan.

Due to the rarity of the EMC in the minor salivary gland, there is no standard treatment guideline for it. Many authors reported surgical treatment for their patients with EMC of the salivary gland, with or without radiotherapy. Simpson et al had discussed a case series that showed patients who received a surgical resection of the tumour followed by radiotherapy remained free from tumour recurrence for nine years. In our case, we have proceeded with the subtotal maxillectomy and selective neck dissection based on the clinical and radiological assessment. The histopathological report revealed the tumour margin is 2mm away from the lateral soft tissue margin and the malignant cells infiltrated the right pterygoid muscle. Hence, we have referred him to the oncologist for further management.

EMC is considered as a low grade neoplasm, gradually infiltrated the local region and there is a possibility of recurrences after the surgical excision. There was a number of cases in which it developed disease complications and act as intermediate-degree or aggressive tumour. In about 20% of cases there were cervical metastases and, infrequently, produced multiple metastases. Death can occur in 8% of cases. Hence, there is a question whether the term low grade neoplasm needs to be revised.

In conclusion, EMC of hard palate rare and can occur in the salivary glands. Not
many cases of the tumour in this region have been reported in the literature. We hope our case will increase the awareness among clinicians.

REFERENCES


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