Case Report

Epithelial-Myoepithelial Carcinoma of the Base of Tongue with Possible Lung Metastases

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1. Introduction

Epithelial-myoepithelial carcinoma is a rare neoplasm that usually arises from the salivary glands, comprising approximately 1% of all salivary gland tumours [1, 2]. Since it was first described in 1972, there have only been 566 cases of epithelial-myoepithelial carcinoma described in the literature. Histologically, the tumour is biphasic with clear myoepithelial cells surrounding epithelial-lined ducts resembling intercalated ducts [3]. The largest series of cases to date was a recent multi-institution review of 246 cases which showed a disease specific survival at 180 months of 80.7% [1]. The case presented is an uncommon presentation of epithelial-myoepithelial carcinoma of the base of the tongue with only four other cases found in the literature.

2. Case

Our case is of a 52-year-old gentleman who presented with restricted tongue movement, dysarthria, and dysphagia that developed progressively over several months. He mentioned this at the Ear Nose and Throat clinic during follow-up of surgical drainage of a mucocele from the pterygopalatine fossa six months before. He also reported 15 kg of weight loss in the preceding four months. He is a nonsmoker with no medical history except for osteomyelitis following a meniscal knee repair four years before.

An MRI showed a 37 × 34 × 42 mm mass in the base of tongue extending to the geniohyoid muscle as well as the genioglossus muscle that was most concerning for squamous cell carcinoma as shown in Figure 1.

His PET/CT scan showed a large intensely FDG avid tongue mass arising in the posterior of the tongue within the body of the tongue and extrinsic muscles and extending posterolaterally into the region of the lingual tonsils and mid-line tongue superiorly. There was moderate FDG uptake in adjacent nodes consistent with metastases.

Multiple bilateral rounded pulmonary nodules were present within the lungs bilaterally, the largest of which measured 5 mm. These showed no significant FDG uptake and were below the resolution of the PET/CT to accurately characterise.

The lung lesions were too small for biopsy to confirm metastasis; however, it was judged to be metastatic given the rapidly progressive disease and bilateral neck metastases. The tumour was staged as T4N2cM1.

Two tongue biopsies performed were inconclusive; however, the third biopsy a month later showed an infiltrating...
biphasic tumour with solid nests composed of epithelial and myoepithelial cells as shown in Figure 2. The tumour cells also demonstrated perineural invasion as shown in Figure 3. There was nuclear atypia and prominent mitoses present. Tumour cells were positive for SMA, p40, p63, CK5/6, EMA (patchy and luminal), CEA (luminal), and CD43. Ki-67 reveals a high proliferation index (60%). The diagnosis was epithelial-myopithelial carcinoma with myoepithelial anaplasia.

Subsequently, the patient was offered radical local treatment with radiotherapy and chemotherapy. The patient declined these so as to maximise his quality of life. He was referred to the palliative care service for continued care and over a year later required a percutaneous endoscopic gastrostomy (PEG) tube for feeding. Approximately 18 months after the diagnosis, the patient passed away from obstruction of the airway.

3. Discussion

Epithelial-myoeptithelial carcinoma is a rare low-grade neoplasm that was first characterised in 1972 [7]. They most commonly arise in the salivary glands with approximately 60–80% arising in the parotid glands [1, 3]. The mean age is around 60 and affects females at a ratio of 1.5 : 1 with a mean tumour size of approximately 29 mm [3].

Epithelial-myoeptithelial carcinomas are primarily treated surgically, as shown in the largest study with all but 9 of the 207 patients undergoing surgery [1]. 85 of these patients (41.1%) received radiotherapy in addition to surgery; however, no survival benefit was noted for those who received radiotherapy compared to those who did not.

Epithelial-myoeptithelial carcinomas do not commonly have distant metastases and this case presents the possibility of lung metastasis, although unfortunately histological confirmation was not obtained. In a review of 58 patients with epithelial-myoeptithelial carcinomas, only 3 patients (5.2%) had evidence of metastatic disease with only one case being a distant metastasis to the iliac bone [3]. In the largest review of 246 cases of epithelial-myoeptithelial carcinoma, 11 patients (4.47%) had distant metastases; however, the locations were unspecified. There is only one case in the literature of pulmonary metastases which were discovered 14 years after a parotidectomy for epithelial-myoeptithelial carcinoma of the salivary gland [8].

The pathology in our case also showed a high proliferation index of 60% with Ki-67 staining. In contrast, a review of 61 epithelial-myoeptithelial carcinomas found a mean proliferative index of 16.9% with a range from 0 to 50% [3]. There have been cases of epithelial-myoeptithelial carcinoma dedifferentiating into a high-grade carcinoma (HGC) with a proliferation index of 67.1% in the HGC and 11.5% in the epithelial-myoeptithelial portion [9].

It is also rare for epithelial-myoeptithelial carcinoma to develop in the base of tongue with only 4 other cases in the literature to our knowledge [2, 4–6]. Two of these patients received surgery and two received chemoradiotherapy. Therefore, this is, to our knowledge, the first case of epithelial-myoeptithelial carcinoma in the base of tongue that has not been aggressively treated and instead given palliative support.

A comparison between the known cases in the literature of epithelial-myoeptithelial carcinoma in the tongue is summarised in Table 1 and is a continuation of the review done by Peters et al. [2].

Our case shows the potential malignancy of epithelial-myoeptithelial carcinomas with distant metastasis to the lungs and lymph nodes. It is the second case in the literature of
epithelial-myoepithelial carcinoma with metastases to the lungs and only the fifth case to occur in the tongue, to our knowledge. Although this is a rare presentation of the tumour, this case may help to guide patient decision-making and prognostic information in the future by demonstrating the potential severity of the disease.

**Disclosure**

No funding was received for this paper. The case here has not been presented nor published elsewhere. The patient’s information has been deidentified; however, unfortunately the patient has passed away before written consent could be obtained.

**Conflicts of Interest**

The authors wish to report no conflicts of interest.

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