Mucoepidermoid carcinoma of the tongue

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ABSTRACT

We report a 27-year-old Congolese man with mucoepidermoid carcinoma (MEC) of the tongue base, which presented as spontaneous intraoral bleeding. Optimal treatment of tongue base MEC is unknown. To our knowledge, this is the first reported case treated with transoral excision with carbon dioxide laser and selective neck dissection. Although immunohistochemical studies have revolutionised understanding of the disease, little else is known of the natural history of MEC. The majority of MEC is considered low-grade, with an indolent course without recurrence or metastasis. Nonetheless, MEC requires surgical management, postoperative radiotherapy and close long-term follow-up.

Keywords: carbon dioxide laser, minor salivary gland tumour, mucoepidermoid carcinoma, salivary gland, tongue tumour

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INTRODUCTION

Minor salivary glands are found in all areas of the oral mucosa with the exception of the gingiva and anterior portion of the hard palate. Tumours of the minor salivary gland constitute a small proportion of all head and neck malignancies, but are more frequently malignant, if present. Mucoepidermoid carcinoma (MEC) of the tongue base is rare, as MEC is usually found in minor salivary glands. Other unusual sites have also been reported in the literature, including the lacrimal duct, skin, lung and pleura.[1-5] We present a case of MEC of the tongue base and discuss its management. We review available literature and compare management of this rare carcinoma with previously-reported cases.

CASE REPORT

A 27-year-old Congolese man presented to the emergency department with heavy intraoral bleeding. There was no preceding trauma and bleeding had occurred spontaneously. He was a non-smoker and was otherwise healthy. There were no stigmata of bleeding disorders on direct questioning. He denied taking any illicit drugs. He had resided in the UK for the past three years and there was no recent history of foreign travel. Bedside flexible nasendoscopy was difficult due to the large amount of blood in the nasopharynx and oral cavity. The patient was fully conscious and was in control of his own breathing at all times. There was no concern of aspiration. He was instructed to sit leaning forward, and to spit out all blood and saliva. After appropriate resuscitation, he was transferred to the operating theatre for emergency pharyngoscopy. Haematenics and coagulation screen were within normal limits.

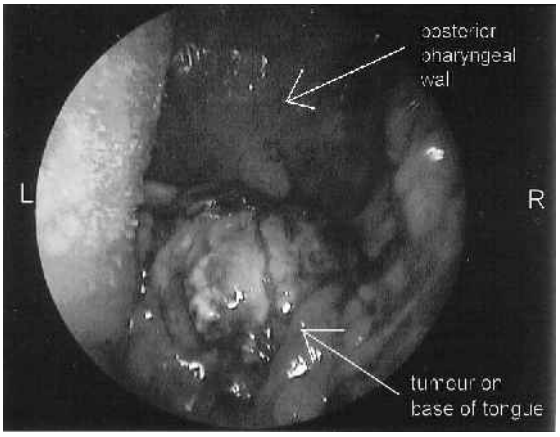

Fig. 1 Endoscopical photograph shows the tumour on the base of the tongue.

General anaesthesia was achieved with rapid sequence induction and naso-laryngeal intubation. A 3-cm diameter mass was found at the base of the tongue, on the left of the midline (Fig. 1). Haemostasis was accomplished with a combination of bipolar diathermy and adrenaline-soaked packing. On palpation, the mass was firm but mobile and did not involve the deeper substance of the tongue. There were no palpable neck nodes. Multiple biopsies of the tumour were taken. A tracheostomy was fashioned to protect the airway in the event of another intraoral bleed. Magnetic resonance (MR) imaging showed an irregular mushroom-shaped mass, 4 cm in diameter arising from the left tongue base and extending into the valleculae (Fig. 2). There was no lymph node involvement. Histology was reported as being

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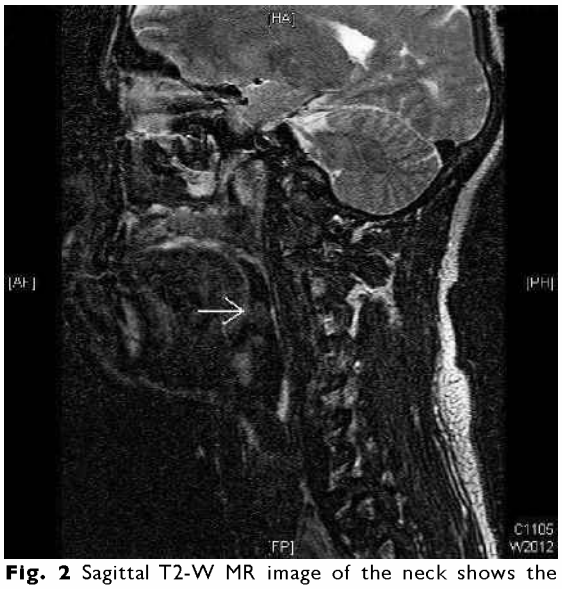

Fig. 2 Sagittal T2-W MR image of the neck shows the mushroom-shaped tumour (arrow) on the base of the tongue.

consistent with high-grade MEC. Ultrasonography of the kidneys was performed to rule out metastatic clear cell carcinoma. Chest radiograph was normal, as was his immune status.

He underwent transoral CO_2 laser excision of the tumour and selective left neck dissection. A nasogastric tube was inserted but was not used for enteral feeding. The tracheostomy was removed three days after surgery. He was assessed by the speech and language therapist on the second day, and was built up on soft diet, progressing to a full normal diet by the fourth day. Histology confirmed high-grade tumour with no cervical lymph node involvement. The tumour margins were clear. He was discharged from hospital five days after surgery. He received fractionated radiotherapy of the primary tumour site and neck, over a period of six weeks. The first course of radiotherapy (64 Gy, face and bilateral neck) was given over 32 fractions, the second course (50 Gy, face and bilateral neck) over 25 fractions and final course (10 Gy, bilateral neck). This was supplemented with cisplatin, which was given at the beginning, middle and end of radiotherapy. He had a percutaneous endoscopic gastrostomy (PEG) feeding tube inserted prior to commencing radiotherapy as a precaution against dysphagia. However, he only required PEG feeding for the last two weeks of radiotherapy and remained on PEG feeding two weeks after completion of the treatment. Swallowing improved progressively and normalised before the PEG was removed. He has been reviewed regularly since and remained disease-free at 12 months following the completion of treatment.

DISCUSSION

Although MEC is the most common malignancy of the tongue base, it remains a rarely-reported clinical entity. The first case was reported in 1973.[6] Since then, only 20 case reports or retrospective case-series have been reported in literature. In a retrospective study of tongue base tumours over a 30-year period, all 22 cases were malignant where 45% of these were diagnosed as MEC.[7] Goldblatt and Ellis found that 85% of their study cohort involving the tongue base was malignant in nature.[8] Of these, MEC accounted for 52%, followed by adenocarcinoma (20%). MEC of the tongue was more common in females and normally presented in the fifth decade.[8] While alcohol and nicotine are the main causes of intraoral carcinoma, the aetiology of MEC in unknown, as there are insufficient studies linking MEC to these causes. Cytogenic studies of MEC cell cultures have consistently shown the presence of trisomy 5, which the investigators have suggested as a possible precursor event in the pathogenesis of MEC.[9] In addition, mutation of the p53 oncogene has been demonstrated in in-vitro MEC cell lines.[10]

MEC has no distinctive cytological characteristic. The tumour is composed of epidermoid and mucin-producing cells, which take origin from the duct epithelial lining. The epidermoid cells proliferate in sheets or islands, and keratinising may occur. When the epidermoid constituent predominates, the histological appearance of the tumour may closely resemble that of squamous cell carcinoma, and it is thus classified as a high-grade MEC tumour. Conversely, the presence of mucin-producing cells within a predominately cystic architecture is regarded as low-grade MEC tumours. The morphological appearance of MEC share similarities to metastatic clear cell renal adenocarcinoma and clear cell bronchogenic squamous carcinoma.[11] Hence, appropriate work-up of the patient must also include imaging of the lungs and kidneys.

One of the most important criteria for measuring the biological behaviour and aggressiveness of MEC is cell proliferation. The proliferating cell nuclear antigen expression increases with the grade of malignancy.[12] Furthermore, mucin expression patterns can be useful for diagnostic and prognostic purposes. Membrane-bound mucins are expressed on the cell surfaces of MEC. Studies have shown that the presence of MUC-1 is related to aggressive tumour, while MUC-4 conferred greater cellular differentiation and better prognosis.[13,14] Cytokines have also been studied where over-stimulation of cell growth have been implicated in tumour growth. For example, transforming growth factor beta-1 (TGF-beta 1) affects

growth inhibition and stimulates extracellular matrix production and angiogenesis. The loss of TGF-beta receptor type II (TGF-beta RII) expression on the cell surface has been linked to resistance of TGF-beta1 mediated growth control and tumour progression, hence leading to uncontrolled cellular proliferation. Dillard et al showed that there was an inverse correlation between tumour grade and loss of expression of TGF-beta RII.[15] This membrane receptor was not present in high-grade MEC. Prognosis is not only adversely affected by high histologic grade, but is also compounded by rapid clinical presentation, old age and location of tumour in the tongue.[16]

The likelihood of developing distant metastasis is associated with high-grade tumour, where insufficient excision is the main factor to recurrence. Discussions at the local multi-disciplinary team revealed two distinct schools of thought. On one hand, the otolaryngologists favoured wide local excision and selective neck dissection. On the other hand, the maxillofacial surgeons advocated mandibulotomy, glossectomy, free flap reconstruction and bilateral neck dissections to ensure complete tumour excision and lymph node clearance. This rationale has been supported by several other authors who have advocated wide tumour margins as intraoral MEC tends to be more aggressive, with recurrence rate of up to 30%–40% with high-grade lesions.[6,17,18] Dequanter et al reconstructed the floor of the mouth with a sternocleidomastoid muscular pedicled flap for a similarly sited MEC tumour, while Deitmer and Stoll suggested lateral pharyngotomy to be included in the excision margins.[19,20] It is clear that due to the uncertain nature of MEC, most authors would favour extensive surgery to mitigate the likelihood of recurrence. This certainly brings to question the possibility of over-treating MECs.

Notwithstanding the significant morbidity from such an extensive procedure, the long-term functional disability of mouth malocclusion, compromised deglutition and altered speech was not sufficiently justified for an otherwise young and healthy individual in this case. Consensus was reached to attempt transoral CO_2 laser excision, accompanied by selective left neck dissection. Complete excision with the CO_2 laser was felt to be achievable as the tumour was mushroom-shaped, mobile and completely accessible with the Steiner laser endoscope. Furthermore, clear tumour margin was possible as there was no radiological evidence of deep invasion. This was supported by negative biopsies of the deep margins. There remains scant published information on the natural history and appropriate management of MEC of the tongue. MEC can present widely diverse biological behaviours based on the myriad of histological characteristics. MEC is a unique carcinoma as it demonstrates a broad spectrum of aggressiveness from indolent tumours that are cured by surgery alone to aggressive neoplasms that are prone to local invasion, recurrence, and metastasis. Whatever the treatment modality, MEC patients should be closely followed-up for life.[10]

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