Reminder of important clinical lesson

Mucoepidermoid carcinoma of sublingual gland: a malignant neoplasm in an uncommon region

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Although mucoepidermoid carcinomas (MECs) are considered as the second most common malignant salivary gland neoplasm, only 0.5–1% of epithelial salivary gland tumours have been reported to arise from the sublingual salivary gland.¹ The authors report a case of a low-grade MEC of the sublingual salivary gland in a 35-year-old woman. Wide excision of the tumour along with continuous marginal mandibulectomy was performed. There has been no recurrence till date.

BACKGROUND

Sublingual salivary gland tumours are rare. They are hard to distinguish from submandibular gland extension. They may mimic minor salivary gland tumours in the floor of the mouth. Their prognosis correlates with clinical staging emphasising the importance of early diagnosis. Although mucoepidermoid carcinoma (MEC) is the most common primary salivary gland malignancy in both adults and children, only 1% of epithelial salivary tumours have been reported to arise from the sublingual salivary gland.²

CASE PRESENTATION

A 35-year-old woman of Indian origin reported to the Department of Oral Medicine and Radiology, with the chief complaint of a swelling in the floor of the mouth, for the past 2 months. She had noticed a small, painless swelling on the floor of the mouth, which had grown to the present size. She experienced dull, localised intermittent pain over the swelling, which aggravated on applying pressure; paracetamol gave temporary relief. She had difficulty in mastication, swallowing and talking, and no change in salivation or paraesthesia. Her medical history was unremarkable. She gave history of intermittent pain in the lower back tooth since 1 month.

Extraoral examination revealed asymmetry of the face due to a diffuse swelling present below the left parasymphysal region. Intraoral inspection revealed a solitary, well-defined, ovoid swelling, on the floor of the mouth (approximately 3 × 4 cm in size), obliterating the left lingual vestibule. The surface of the swelling appeared smooth, shiny and pink. Mesiodistally the lesion extended from 31 to 36 and buccolingually from the lingual border of the alveolar mucosa towards the ventral surface of the tongue. It was hard in consistency, with well-defined borders, movable and tender (figure 1). There was reduced salivary flow from the orifice of the left Wharton's duct. Deep cervical lymph node examination findings were positive for Lib - 2 cm and LIII - 1.5 cm both of which

were firm and mobile. A clinical diagnosis of a tumour of the left submandibular/sublingual salivary gland was arrived at.

Examination of teeth showed that there were root stumps of 15, 16, 17, 24, 26, 27, 28, 34, 36, 45, 46 and missing 38 and 47. Generalised periodontal pockets were present indicating generalised chronic periodontitis and periapical abscess in the 34.

INVESTIGATIONS

The orthopantomogram (figure 2) showed root stumps in relation to 15, 16, 17, 24, 26, 27, 28, 34, 36, 45, 46 and missing 38 and 47 with generalised horizontal bone loss indicating generalised chronic periodontitis. Occlusal radiograph (figure 3) and left lateral oblique view of the mandible (figure 4) did not show any evidence of salivary calculi or any other abnormality.

Ultrasonogram showed a large heterogenous hypoechoic mass on the floor of the mouth on the left side, compressing the left submandibular gland salivary gland and measuring 43.1 × 31.6 mm with enlarged left submandibular lymph nodes (figure 5).

CT plain revealed a large hypodense mass on the floor of the mouth on the left side with close approximation to the mandibular lingual cortex with enlarged left submandibular lymph nodes and a goitreous nodule with colloid cyst on the left lobe of thyroid (figure 6). CT with contrast enhancement (figure 7) revealed a poorly enhancing/non-enhancing hypodense mass in relation to floor of the oral cavity extending from the antero-lateral part of the base of the tongue along the mandible of approximately 48 mm length. This was suggestive of a neoplastic mass, though an inflammatory/infective collection could not be ruled out.

Laboratory investigations revealed a normal hemogram and thyroid hormone levels. The fine needle aspiration cytology from the lesion was reported as retention cyst. An incisional biopsy however, turned out to be low-grade

Figure 1 Intraoral figure showing a well-defined swelling on the floor of the mouth on the left side.

Figure 2 Panoramic radiograph – loss of lamina dura and horizontal bone loss seen with periapical radiolucency in 35 indicating generalised chronic periodontitis with root stumps in 34, 36 and periapical periodontitis in 35.

Figure 3 Left lateral occlusal view of mandible – no abnormality seen.

Figure 4 Left lateral oblique view – no radiopaque abnormalities seen indicating the absence of salivary calculi.

MEC. The patient was planned for ablative surgery with radical neck dissection of the left side. The lesion and the associated lymph nodes were surgically excised. Wide excision of the tumour along with continuous marginal

mandibulectomy and modified radical neck dissection was done and subjected to histopathology. The intraoral defect on the floor of the mouth was reconstructed using a left lateral tongue flap.

Figure 5 USG of left sublingual gland – a large heterogenous hypoechoic mass on the floor of the mouth on the left, compressing the left submandibular gland – measuring 43.1 × 31.6 mm. Enlarged left submandibular lymph nodes seen.

Microscopic picture of the tumour showed abundant cystic spaces surrounded by mucous cells and few epidermoid cells. Histopathologically as the cystic component was <20% and neural invasion, necrosis, mitosis and anaplasia were absent, it was given a point score of 2. Based on the histopathological findings (according to WHO staging of salivary gland tumours, 2005), a final diagnosis of a ‘low-grade mucoepidermoid carcinoma of the sublingual salivary gland’ was given (figures 8 and 9). The margins did not show any evidence of tumour infiltration. No metastasis was observed in the dissected lymph nodes and the margins of the defect were free of tumour residues. The TNM staging of the lesion was given as T3 N0 M0 and the tumour grouped as stage III.

DIFFERENTIAL DIAGNOSIS

- The differential diagnosis includes mucocele, neoplasm arising from submandibular/sublingual gland, sialolithiasis and Kuttner’s tumour.
- Mucocele, low-grade MEC and mucous cell adenocarcinoma produce mucus and thus resemble each other.
- Sialolithiasis is one of the commonest pathologies to affect the submandibular gland/duct and hence is considered here.
- Kuttner’s tumour is an inflammatory disorder of the submandibular gland presenting as an enlarged firm painful gland. Its proximity in location prompted its inclusion.

Figure 6 CT-plain of the mandible – a large hypo dense mass on the floor of the mouth on the left side. Enlarged left submandibular lymph nodes and goitreous nodule with colloid cyst of the left lobe of thyroid.

Figure 7 CT-scan: contrast of the mandible – a fairly large cystic mass seen in relation to floor of the oral cavity extending from the antero-lateral part of the base of the tongue along the mandible – approximately 48 mm in length.

- Mucinous adenocarcinoma is an entity frequently found in palate and sublingual gland and hence needs to be ruled out.

TREATMENT

The lesion and the associated lymph nodes were surgically excised. Wide excision of the tumour along with

Figure 8 Histopathological picture – abundant cystic spaces surrounded by mucous cells and few epidermoid cells seen.

Figure 9 Histopathological picture – 40× magnification of histopathological specimen.

continuous marginal mandibulectomy with modified radical neck dissection was performed. The decision to perform an radial neck dissection was based on the fact that Myers and Ferris in their book ‘Salivary Gland Disorders’ stated that ‘neck dissection is advocated in patients in

stage III or IV cancer of the sublingual salivary gland....’ Only this would ensure longer survival of the patient as patients usually report back only if they have a significant discomfort and that in such cases is usually too late. Marginal mandibulectomy was performed as the needle

of suspicion of the involvement of lingual periosteum by the tumour was high as seen in the CT. Moreover in this geographic region, attempting a conservative procedure could lead to disastrous results, as patients are usually lost to follow-up.

OUTCOME AND FOLLOW-UP

Patient is disease free and currently under regular follow-up.

DISCUSSION

Malignant tumours comprise 15–32% of parotid tumours, 41–45% of submandibular tumours, 70–90% of sublingual tumours, and 50% of minor gland tumours. Sublingual salivary carcinomas embrace only 1% of major salivary gland carcinomas, that is, only 0.5–1% of all epithelial salivary tumours origin.^{2–4} Eighty to 90% of sublingual salivary gland tumours are frankly malignant, with MEC being rated as the most common malignant salivary gland neoplasm (ie, it accounts for 30% of all salivary malignancies, 10% of all salivary tumours and 5% of head and neck salivary gland cancers).^{1 4} 50–60% of MECs arise from the major salivary glands of which only about 2–4% bracket the sublingual salivary glands, (the rest 80% arise from the parotid and 8–13% from the submandibular). It is also found to occur in minor salivary glands (most often in the palate) and the seromucous sites of the sinonasal tract, larynx and the trachea.¹

Smith, in a review article on the mixed tumours of the salivary glands, stated that in a study by Dr BR Shore at St. Luke's hospital, only one sublingual tumour was found during the past 40 years.⁴

Goode *et al* in an analysis of 234 cases of MEC of the major salivary glands found only 3% in the sublingual gland.⁵

Ma'aita *et al* evaluated the types and clinical outcomes of 221 salivary gland tumours in Jordan and found only one case (0.4%) of sublingual salivary gland tumour.⁶

Poomsawat *et al* in a retrospective study of 60 cases of salivary gland tumours in a Thai population found that 10% were major salivary gland tumours. Out of these six cases, only two cases had their origin from sublingual glands.⁷

The causative role in MECs hold no significant familial history nor are they associated with trauma, sialoliths, any abuse of alcohol or tobacco.

Clinical features

Primary tumours of the sublingual salivary gland are very rare and clinically present themselves as a slowly enlarging mass situated under the tongue. The mass, if benign, may be present as a painless lesion for many years whereas a malignant tumour may occur within duration ranging from a few months to a year and may be associated with pain when perineural, cartilage or bone invasion is suspected. Malignant tumours are usually concentrated around the 3rd to the 5th decade of life, with a slight inclination towards the female sex.¹

Goode *et al* found that 80% of patients with sublingual gland tumours were symptomatic.⁵

Imaging

Plane radiographs, sialography and nuclear scans add very little to the diagnostic information and are seldom indicated for evaluation of the sublingual gland masses. Sonographically, MECs, which are smaller than 2 cm, have a homogenous structure with smooth borders and may erroneously considered as benign. CT and MRI give a better visualisation of the masses within the salivary glands. CT scans are especially useful in assessment of the location and density of the lesion. They help to differentiate cystic from solid lesion and the evaluation of the relation to the adjacent structures including the bones and soft tissues.¹

Histopathological classification of salivary gland tumours

Histopathologically, MECs, by definition, contain three cellular elements in varying proportion: squamous cells, mucus-secreting cells and the intermediate cells. The gross appearance varies with the grade of the tumour. According to WHO classification (2005), the tumours of the salivary gland are graded as low-, intermediate- and high-grade tumours.² The low-grade tumour has a higher ratio of mucous cells and is a less aggressive lesion. While high-grade form is considered to be more malignant tumour and has a poorer prognosis,^{4 8} low-grade tumours produce well-circumscribed ovoid masses of 2–4 cm in diameter which contain solid gray-white or grey-pink areas which are mixed with mucus-filled macrocysts, infrequent solid nests of intermediated and epidermoid cells and no cellular pleomorphism.⁹

Treatment modalities

Salivary gland tumours are best treated by surgery, the extent of which depends on the location, size and the histopathological appearance. A malignant tumour may require an extensive resection which may involve the adjacent organs and dissection of the involved node.¹ A low-grade tumour may require a less aggressive approach when compared to a high-grade one and should be managed by wide local resection, en-bloc resection or hemimandibulectomy or hemimaxillectomy. Neck dissections are usually part of the treatment in cases where metastasis to the cervical nodes is suspected. Radiotherapy is recommended for high-grade MEC cases.⁴

Goode *et al* in an analysis of 234 cases of MEC of the major salivary glands stated that 208 received surgical therapy only, 22 received combined surgery and radiation therapy and only 2 received surgery with adjunctive radiation and chemotherapy.⁵

The prognosis of such tumours is a 5-year survival rate with 92–100% in low grade, 62–92% in intermediate grade and 0–43% in high grade. Despite low rate of occurrence of tumours in the sublingual salivary gland, the percentage of malignant tumours are reported to be very high. They are difficult to distinguish from an extension of the submandibular gland or a minor salivary gland tumour in the floor of the mouth and are usually misdiagnosed clinically and radiographically as a cyst or benign tumour. Hence, it is important to diagnose and treat the lesion in its early stages.^{10 11}

Learning points

- ▶ Despite low rate of occurrence of tumours in the sublingual salivary gland, the percentage of malignant tumours are reported to be very high.
- ▶ They are difficult to distinguish from an extension of the submandibular gland or a minor salivary gland tumour in the floor of the mouth and are usually misdiagnosed clinically and radiographically as a cyst or benign tumour.
- ▶ Radiographic features would suggest a need of suspicion which would have to be confirmed with a histopathological report.
- ▶ Early diagnosis of such lesions enable early treatment and reduce morbidity.

Competing interests None.

Patient consent Obtained.

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