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Submucosal Globular Tongue Swelling: A Rare Presentation of Adenoid Cystic Carcinoma

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ABSTRACT

Introduction: Adenoid cystic is an infrequent malignancy of the salivary glands. Some of its unique features include slow growth, the tendency for perineural invasion, local recurrence, and the ability to metastasize. The mobile tongue's adenoid cystic carcinoma (ACC) is a rare reported case in the literature.

Report: We report a rare case of adenoid cystic carcinoma, in a 55-year-old female who presented with a swelling arising in the dorsal and lateral aspect of anterior tongue to Head and Neck surgery OPD in a teritary care cancer institute in North India. Histopathology after complete excision of the mass confirmed the diagnosis of adenoid cystic carcinoma. There has been no evidence of disease following excision with tumour-free margins.

Discussion: The pathophysiology of ACC is an understudied area due to the condition's rarity. Chromosomal abnormality and dysregulated MYB oncoprotein have to be thought to play a key role in the pathophysiology and proliferation of ACC.

Conclusions: Adenoid cystic carcinoma of the tongueis exceedingly rare but should be considered in the differential diagnosis in patients with tongue swelling. These tumours are malignant and treatment requires surgical excision and neck dissectionfollowed by adjuvant radiotherapy.

Keywords: Adenoid cystic carcinoma; Carcinoma tongue; Oral cavity; Perineural invasion; Lingual tumours.

INTRODUCTION

Adenoid cystic carcinoma (ACC) is an infrequent malignancy that affects secretory glands, most commonly the salivary glands. Although rare, it is an essential differential to consider for a painless swelling in the head and neck region because of its high likelihood to metastasis. ACC has a slight predominance in females, with a peak incidence in the fifth and sixth decades of life. It represents 1 to 2% of all malignant tumours of the head and neck with a reported incidence of 2.9% of all cases in the mobile tongue.[1] We present a case of a 55-year-old female with ACC of mobile tongue treated with surgery followed by adjuvant radiotherapy and a brief review of the literature.

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CASE REPORT

A 55-year-old female patient presented to the OPD with complaints of slow-growing swelling in the tongue for the past one year before the

consultation. She also complained of associated difficulty chewing and speaking for 2 months. There was no addiction history.

On intraoral clinical examination and palpation, an obvious smooth firm mass of about 2.5 cm in diameter on the left lateral side of the tongue with no surrounding induration and same colour as the surrounding mucosa was observed without other oral lesions. (Fig. 1) The cervical lymph nodes were not palpable.

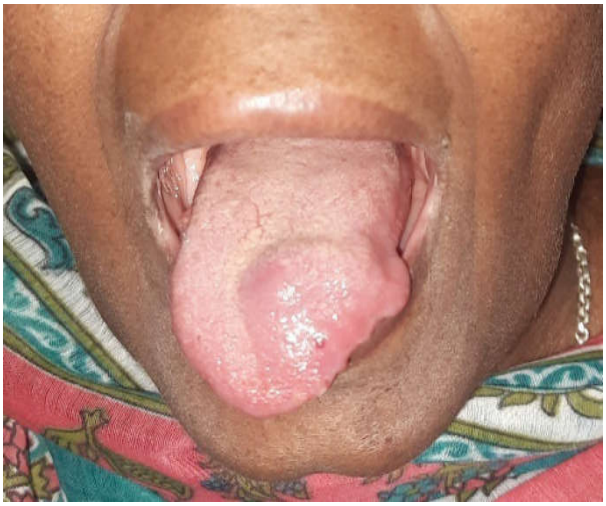

Fig. 1: Pre-operative image showing the swelling on the dorsal surface of left side of the tongue.

Contrast-enhanced magnetic resonance imaging was done for the patient which revealed a T2/STIR hyperintense enhancing tongue lesion of size 2.1 x 2.8 x 2.6 cm involving the left anterior and lateral border of the tongue, not involving the base and root of the tongue. (Fig. 2) The lesion appeared benign on clinic-radiological correlation.

She underwent an excisional biopsy under general anaesthesia. Intraoperatively a 2.5 x 2 x 1 cm well-encapsulated swelling was excised in toto (Fig. 3) and sent for histopathological examination.

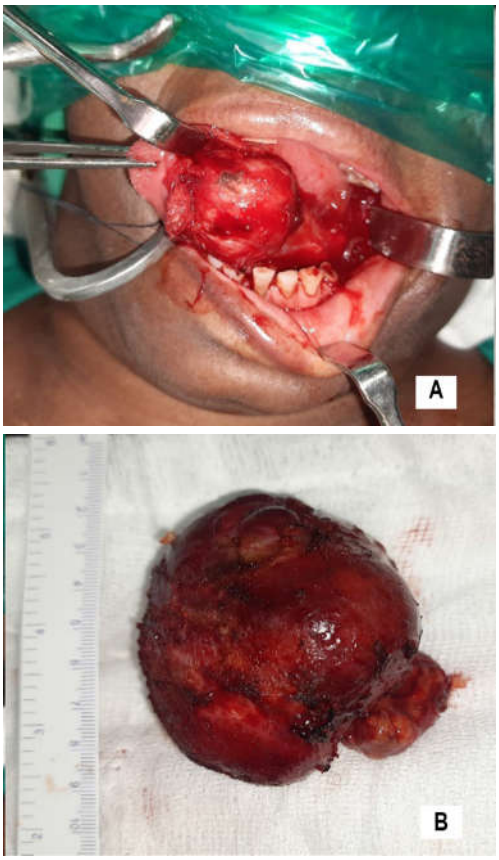

Fig. 3: (A) intraoperative picture showing the excision of the tongue mass. (B) Image showing the excised mass.

A detailed morphological examination revealed a basaloid tumour comprised of varied patterns,

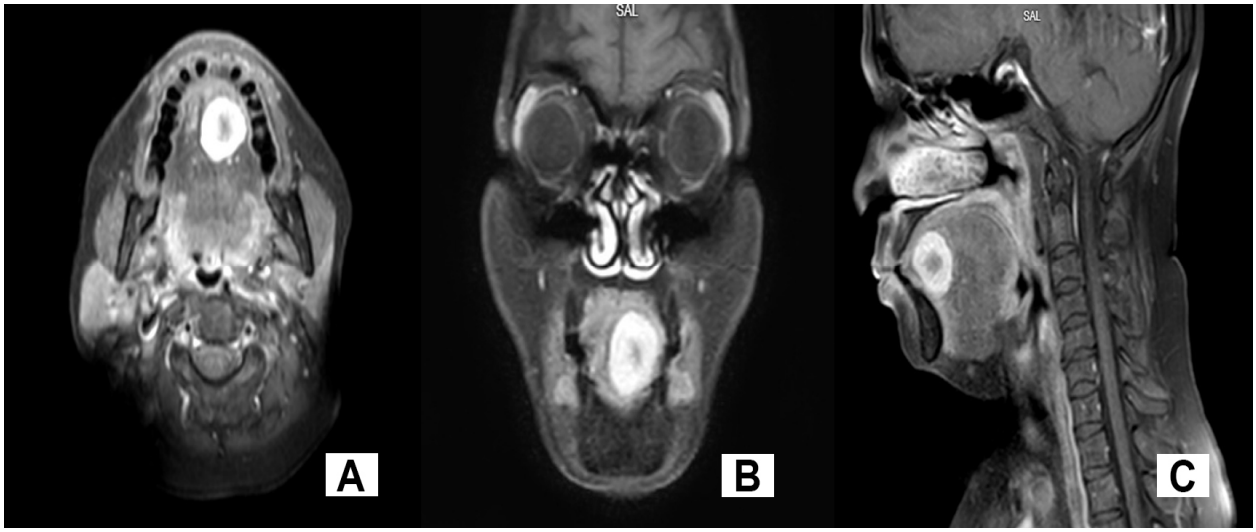

Fig. 2: MRI showing hyperintense enhancing lesion in the tongue (A) Axial view, (B) Coronal view & (C) Sagittal view.

amounting to 75% cribriform, 20% solid, and 5% tubular architecture. On immunohistochemistry, the ductal epithelial cells were positive for CK7 and c-kit, while the abluminal/myoepithelial cells were highlighted by p63 and focally by S100. Hence, a diagnosis of Adenoid Cystic Carcinoma, Grade 2 was rendered. No necrosis or features of high-grade transformation were noted. (Fig. 4)

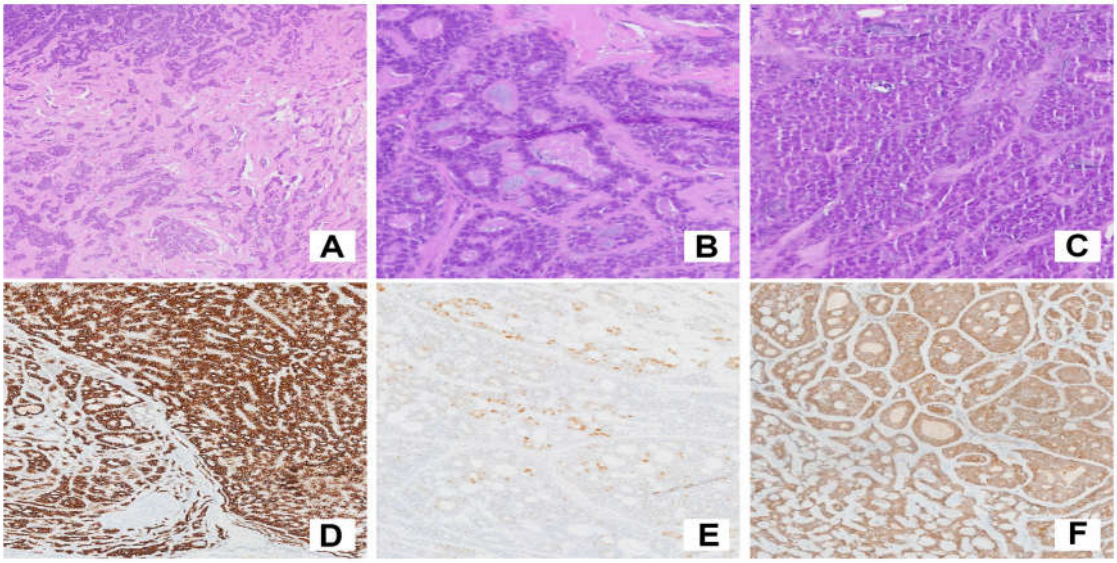

Fig. 4: A) Basaloid tumour with cribriform and tubular pattern (Haematoxylin and eosin,20x) **B)** Cribriform and tubular pattern (Haematoxylin and eosin, 400x) **C)** Solid area (Haematoxylin and eosin, 400x) **D)** Immunohistochemistry for CK 7, highlights the luminal and abluminal cells (400X) **E)** Immunohistochemistry for S-100 highlights few myoepithelial cells (400X). **F)** Immunohistochemistry for CD 117 highlights luminal cells (200X)

This unexpected diagnosis of malignancy, required a margin revision of the tongue lesion and left selective neck dissection (levels 1 to 4).The final histopathological report revealed scanty residual viable adenoid cystic carcinoma with all margins free of tumour. Total regional lymph nodes were sixty-six, negative for metastasis (0/66). No nodal metastasis was identified.

DISCUSSION

Billroth defined ACC as "cylindroma" and explained its recurring nature in 1859. In 1953, Foote and Frazell introduced the term "adenoid cystic carcinoma".[2]

Adenoid cystic carcinoma is a rare malignancy arising from the secretory glands. It most commonly affects the salivary glands. It accounts for about 1 % of the head and neck malignancies. It primarily affects the minor salivary glands, accounting for over 10% of salivary gland malignancy.[3] The tongue is the site of origin for ACCs in the head and neck region, accounting for 3.4% to 17.1% of instances, with the mobile tongue accounting for 2.9%. (1) The majority of cases are reported to be in the base of the tongue.[4] Nerve invasion is a hallmark of the tumour, which also implies a poor prognosis.

Histologically three distinct patterns are noted in ACC: tubular, cribriform, and solid, with cribriform pattern being the most common and associated with a better prognosis. Immunohistochemistry considerably contributes in the diagnosis of ACC since the tumor cells stain positive for smooth muscle actin, S100, vimentin, as well as for MYB and CD 117 (receptor tyrosine kinase c-KIT), which helps differentiate it from other malignancies.[5]

The treatment approach for adenoid cystic carcinoma of the tongue is surgical resection followed by radiotherapy. Adenoid cystic carcinoma is known to cause cervical lymph node metastases with an incidence of 17.6% in mobile tongue as reported by Carrasco et al.[6] Level 1b and II are the most commonly involved lymph nodes. Thus, selective neck dissection should be considered in patients with clinical N0 neck with ACC of the tongue. A combined approach in the management of ACC of the tongue includes wide local excision of the ACC of the tongue with adequate margins, with or without reconstruction, selective neck dissection, and adjuvant radiotherapy. Adjuvant radiotherapy is generally considered in advanced T-stage cases and the presence of positive tumour margins. Some authors have suggested radiotherapy in cases of advanced non resectable tumour.

Adenoid cystic carcinoma of the tongue has been rarely reported in the literature. Goldblat et al reported 5 cases of ACC of the tongue in 1987 with no mention of the specific site demarcation in the tongue.[8] In 2009, Luna Ortiz et al reported 2 cases of mobile tongue ACC in a retrospective study of 68 patients of head and neck ACC from 1986 to 2006.[7] Xi Tang et al reported a case of ACC of the mobile tongue treated with surgery, reconstruction with anterolateral thigh flap, and adjuvant radiotherapy in 2019.[9] Kumar S et al reported a case of ACC of the tongue in 2016, along with a review of literature demonstrating the rare presentation of adenoid cystic carcinoma in the mobile part of the tongue.[10] Thus, unusual presentations of adenoid cystic carcinoma of the mobile tongue have been recorded infrequently in the literature.

Postoperative management typically involves close surveillance to monitor for recurrence, as it carries a tendency for local recurrence and also distant metastasis.

CONCLUSION

We present a rare case of adenoid cystic carcinoma arising in the dorsal aspect of the tongue. Adenoid cystic carcinoma though rare, should be considered as a differential in growths identified in the tongue. Understanding the clinical presentation, diagnostic approach, histopathological characteristics, and treatment options for adenoid cystic carcinoma is crucial for achieving optimal patient outcomes and minimizing the risk of recurrence.

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Patient Consent: Verbal informed consent was obtained from the patient. The whole process of examination and the purpose of the article was explained to the patient and her relatives.

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