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Extraglandular Submandibular Adenoid Cystic Carcinoma Involving the Lingual Nerve: A Case Report

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Abstract: *Malignant tumors of the submandibular salivary gland are uncommon and often difficult to differentiate clinically from benign lesions. Adenoid cystic carcinoma (ACC) is characterized by slow growth and a strong tendency for perineural invasion. We report a rare case of extraglandular ACC arising from accessory salivary glands in the submandibular region, independent of the submandibular gland. A 38-year-old woman presented with a painful left laterocervical mass associated with hypoesthesia in the distribution of the lingual nerve. Magnetic resonance imaging revealed an irregularly contoured mass in the left submandibular region. Surgical exploration identified a polylobulated tumor encasing the lingual nerve, necessitating tumor excision with nerve sacrifice and submandibulectomy. Histopathological examination confirmed adenoid cystic carcinoma with perineural and vascular invasion, while the submandibular gland was free of tumor. The patient received adjuvant radiotherapy. This case highlights an unusual extraglandular origin of ACC and emphasizes the importance of multidisciplinary management.*

Keywords : Adenoid cystic carcinoma; accessory salivary glands; submandibular region; lingual nerve.

1. INTRODUCTION

Adenoid cystic carcinoma accounts for approximately 5–10% of malignant salivary gland tumors and represents the second most common histological subtype. The submandibular region is involved in 12–30% of cases. ACC is considered an aggressive tumor because of its frequent perineural and vascular invasion. Although tumor progression is slow, there is a high risk of local, locoregional, and distant recurrence, sometimes occurring decades after initial treatment [2–3].

We report a rare case of extraglandular adenoid cystic carcinoma arising from accessory salivary glands in the submandibular region, encasing the lingual nerve and independent of the submandibular gland.

2. CASE REPORT :

A 38-year-old woman presented to the ENT department of Avicenne Military Hospital in Marrakech with a painful left laterocervical mass evolving for three months. Clinical examination revealed a hard, tender mass in the left submandibular region, fixed to the deep plane, with irregular margins and measuring approximately 4 cm in its largest diameter. The overlying skin was normal. Neurological examination showed hypoesthesia in the territory of the left lingual nerve. Examination of the floor of the mouth was normal.

Magnetic resonance imaging demonstrated a mass measuring 3.7 × 2.7 × 2.8 cm in the left submandibular region, with irregular contours, isointense signal on T1-weighted images, heterogeneous signal on T2-weighted images, and intense contrast enhancement. Diffusion-weighted imaging showed restricted diffusion with an apparent diffusion coefficient of 0.9, and perfusion analysis revealed a type C curve (Figure 1).

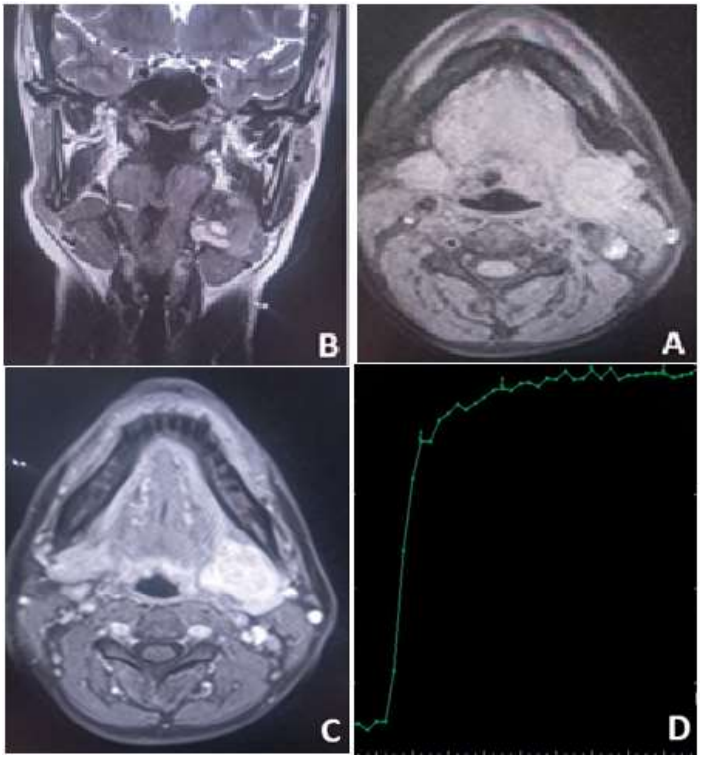

Figure 1: MRI of the submandibular region. A: Axial T1-weighted image without contrast; B: Coronal T2-weighted image; C: Axial T1-weighted image with contrast; D: Type C perfusion curve. A mass in the left submandibular gland measuring 3.7 x 2.7 x 2.8 cm, with irregular contours, isointense on T1-weighted imaging, heterogeneous on T2-weighted imaging, with intense enhancement, ADC = 0.9, and a type C perfusion curve.

Exploratory cervicotomy revealed a macroscopically normal submandibular gland and a hard, polylobulated tumor encasing the lingual nerve, making nerve dissection impossible (Figure 2). A submandibulectomy with complete tumor excision and sacrifice of the lingual nerve was performed, along with ipsilateral cervical lymph node dissection (levels Ib, II, III, and IV).

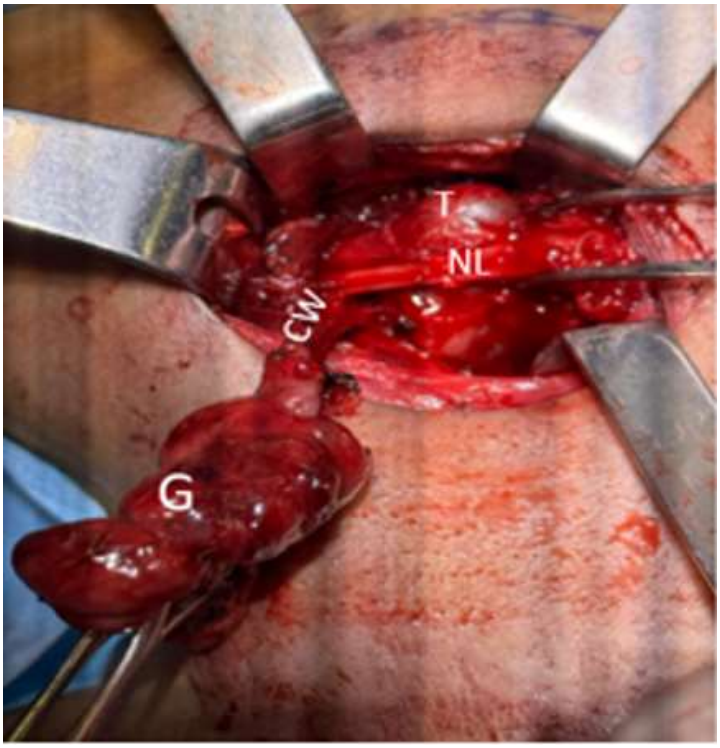

Figure 2 : Intraoperative view showing a normal-appearing submandibular gland and a hard, polylobulated tumor encasing the lingual nerve and fixed to the surrounding tissues. G: submandibular gland; CW: Wharton's duct; NL: lingual nerve; T: tumor

Histopathological examination confirmed adenoid cystic carcinoma with perineural and vascular invasion, including direct invasion of the lingual nerve. The submandibular gland showed no evidence of tumoral involvement (Figure 3).
After multidisciplinary discussion, the patient was referred for adjuvant radiotherapy with a total dose of 65 Gy.

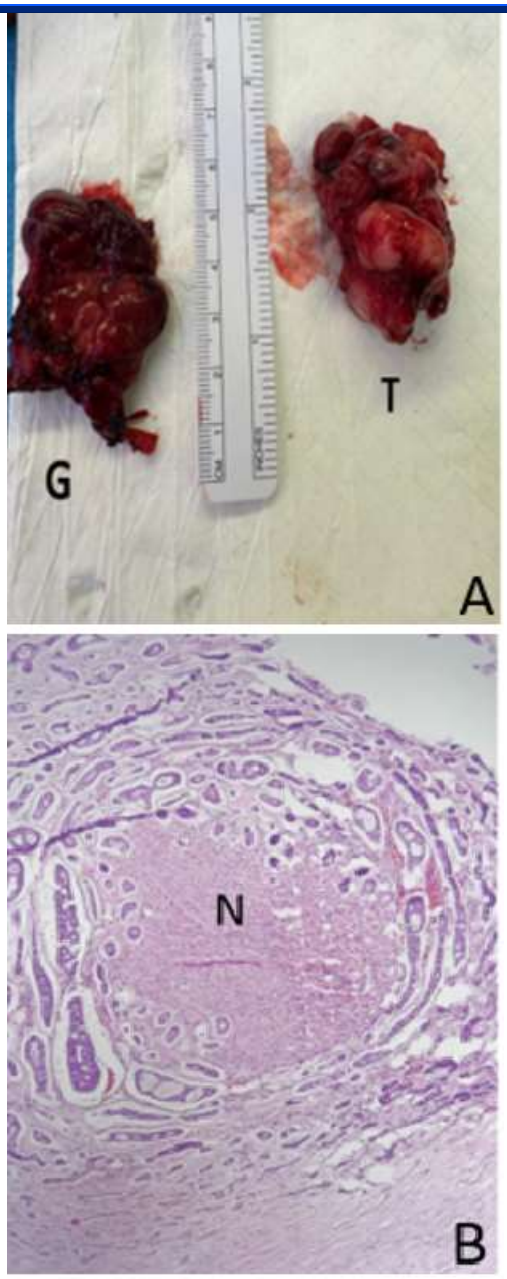

Figure 3: Examination of the specimen

A) Macroscopy: G, submandibular gland free of tumor, T: tumor mass

B) Microscopy: lingual nerve (N) entirely encased by cancer cells.

3. DISCUSSION

Adenoid cystic carcinoma was first described by Foote and Frazell in 1953 [1]. It involves the submandibular region in approximately 20–30% of cases and is characterized by marked perineural and perivascular invasion [2–3]. Three histological patterns have been described: tubular, cribriform, and solid. The solid pattern is associated with a poorer prognosis, whereas tubular and cribriform patterns are associated with better outcomes [4].

ACC affects both sexes, with a mean age at diagnosis between 40 and 60 years [3–4]. Clinically, it usually presents as a salivary mass without specific features; however, paresthesia and nerve paralysis may be present in up to 20% and 30% of cases, respectively [4]. Magnetic resonance imaging is the imaging modality of choice for evaluating tumor extension and perineural spread.

Surgical resection remains the primary treatment modality, with or without neck dissection. Adjuvant radiotherapy is commonly recommended to improve locoregional control, particularly in cases with high-grade histology, close or positive margins, lymph node metastases, advanced T stage, perineural invasion, or recurrence [5,8–10]. Untreated disease is associated with late local recurrence and distant metastases. Overall survival at 20 years ranges from 15% to 20%, with a median survival of approximately three years after the onset of metastases [6–7].

4. Conclusion

This case report describes an unusual presentation of adenoid cystic carcinoma arising from accessory salivary glands in the submandibular region, independent of the submandibular gland, with invasion of the lingual nerve. Recognition of this rare entity is essential for accurate diagnosis and appropriate management. Optimal treatment relies on complete surgical excision followed by adjuvant radiotherapy within a multidisciplinary framework.

5. References

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