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Case Report

Unveiling the enigma: A case report of adenoid cystic carcinoma in the hard palate

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Abstract

Adenoid cystic carcinoma (ADCC) was initially documented in 1853 and later termed by Spies in 1930. Previously perceived as benign, its malignant characteristics were elucidated in 1943. ADCC commonly originates from salivary glands, particularly from the ductal cells and myoepithelial cells, with key molecular abnormalities involving MYB and NFIB gene translocations. Clinically, ADCC predominantly affects adults aged 40-60 years with a slight female predominance and commonly arises in minor salivary glands. It presents as a painless, gradually enlarging mass. Histopathologically, ADCC exhibits cribriform, tubular, and solid patterns, with the solid subtype often indicating a poorer prognosis, and perineural invasion being a hallmark. Diagnostic challenges include distinguishing ADCC from similar tumors like polymorphous adenocarcinoma and pleomorphic adenoma. Management involves surgical excision, radiation, and chemotherapy. Here we present a case report of adenoid cystic carcinoma involving the hard palate in a 70-year-old male patient with a review of relevant literature.

Keywords: Adenoid cystic carcinoma, Hard palate, Perineural invasion.

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1. Introduction

Adenoid cystic carcinoma (ADCC) is a rare malignancy originating from the secretory glands, commonly found in the salivary glands. It comprises about 1% of Head and Neck malignancies.[1] It can arise in any salivary gland, with 50–60% occurring in minor salivary glands and a rare occurrence in the parotid gland (2–3% of all tumors).[2] ADCC is typically a slow-growing but aggressive tumor.[2] It has a high recurrence rate of 31.9%.[3] The peak incidence is among women in their 5th and 6th decades.[2] Histopathologically, ADCC presents three patterns as tubular, cribriform, and solid. The solid pattern is associated with higher recurrence, metastasis, and mortality rates.[4] Treatment primarily involves surgery, often supplemented with X-ray radiation, although recurrence and metastasis remain significant challenges, even with radical excision.[2] Due to its rarity, limited data exist on predisposing risk factors and advanced disease management.[1]

The case report emphasizes the histological variability observed in a salivary gland neoplasm and highlights the importance of utilizing immunohistochemical (IHC) markers to differentiate adenoid cystic carcinoma from other types of salivary gland neoplasms.

2. Case Description

A 70-year-old male patient presented with swelling in the upper right side on the roof mouth since 6 months with a history of blood discharge from the site without any pain. The patient is also a known case of hypertension, hyperthyroidism, and under medication for the same. Personal history revealed consumption of alcohol and beedi smoking since 30 years.

On clinical examination severe attrition was noted with 14, 16, grossly decayed with 27, cervical abrasion with 16, 26, 44, 45 and clinically missing 34, 35, 36, 38, 46, 48.

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Also, a diffused swelling of size 1x1cm was noted on the right side of the hard palate, soft in consistency, tender on palpation with a central ulceration, and active bleeding.

A provisional diagnosis of palatal abscess was made with a differential diagnosis of minor salivary gland tumour and necrotizing sialometaplasia.

The radiograph revealed no significant changes. An incisional biopsy was taken and sent for histopathological examination.

The gross specimen of 6 soft tissues bits were received, the largest measuring approximately 0.7x0.8x0.4 cms, irregular in shape with a rough surface, brown in color and fragile.

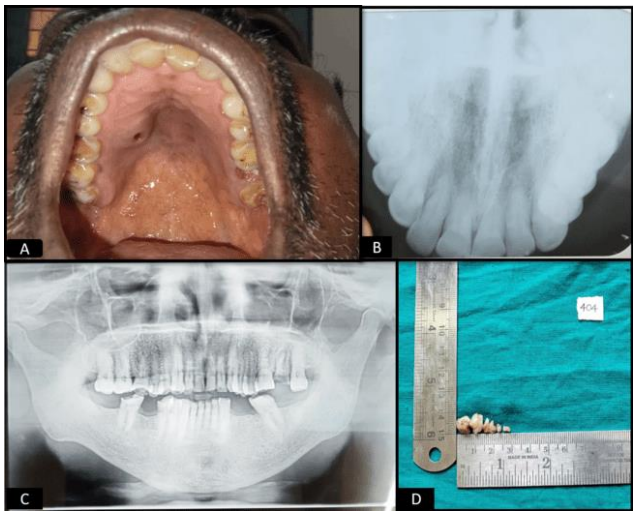

Figure 1: **A**): Palatal swelling with ulceration seen on the left posterior hard palate. **B&C)**: Occlusal radiograph & Orthopantomogram showed no significant changes. **D)**: Incisional biopsy consists of fragile soft tissue bits

On microscopic examination of H & E stained sections showed glandular tissue with tumour cell proliferation in the form of duct-like, cribriform and tubular patterns. Duct-like structures were lined by a single layer of cuboidal cells with hyperchromatic nuclei and scant cytoplasm with eosinophilic material inside the lumen, wedged between the cells with small, darkly stained nuclei and clear cytoplasm forming round to oval pseudocystic spaces containing pale blue fibrillar material. The tubular arrangement of basaloid cells with inner cuboidal and outer smaller cells, interconnecting at areas was evident. Hyalinized stroma was seen surrounding tumor cells with extravasated RBCs in the connective tissue. There was no evidence of perineural invasion.

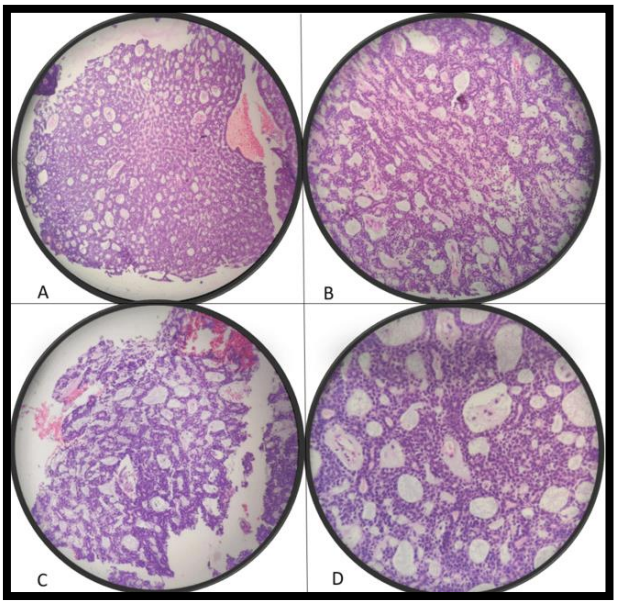

Figure 2: H & E stained sections showing (**A**): Cribriform pattern in 4X (**B**): Tubular pattern made up of double layer of cells in 20X (**C**): Duct like pattern with mucin in luminal spaces in 20X (**D**): Duct like structures lined by cuboidal cells with hyperchromatic nuclei in 40X

Histological differential diagnosis of adenoid cystic carcinoma, pleomorphic adenoma, polymorphous adenocarcinoma, epithelial myoepithelial carcinoma, and basal cell adenoma were considered.

Further immunohistochemical analysis was done. Ki67 index >10% indicating malignancy, Calponin showed strong positivity favouring myoepithelial origin, p63/p40 was strongly positive, and c-KIT also showed diffused strong positivity.

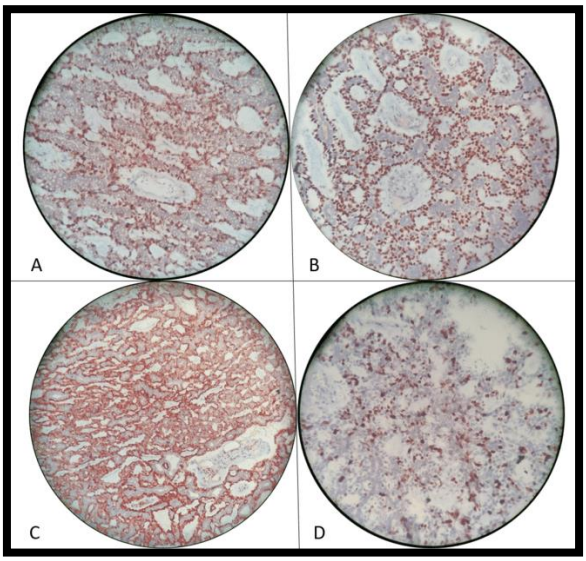

Figure 3: Immunohistochemical marker positivity seen with **A):** P40 (40X), **B):** P63 (40X), **C**): Calponin (20X), **D):** Ki67(40X)

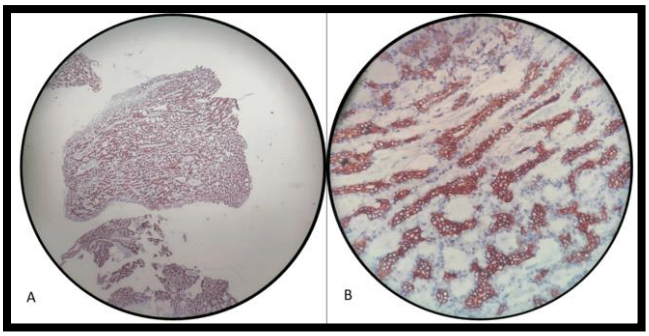

Figure 4: IHC positivity seen with c-kit in **A**): (4X) & **B**): (40X)

The final diagnosis of Adenoid Cystic Carcinoma was given based on histopathological and immunohistochemical findings.

The patient was treated with surgical excision.

3. Discussion

In 1853, Robin, Lorain, and Laboulbene initially documented two cases of an unusual epithelial tumor affecting the nose and the parotid gland, later it was termed "Cylindroma" by Billroth in 1856. It wasn't until 1930 that Spies coined the term "Adenoid cystic carcinoma (ADCC)". Before the 1940s, ADCC was perceived as a benign form of mixed salivary gland tumors. The malignant characteristics of ADCC were elucidated by Dockerty and Mayo in 1943.[5] ADCC typically exhibits slow progression, and it is remarkable for its tendencies towards local recurrence as well as regional and systemic metastasis.[6]

ADCC is thought to arise from the secreting glands, most commomly involving salivary glands. It arises specifically from the ductal cells, and electron microscopy shows that it arises from cells that can differentiate into epithelial and myoepithelial cells.[7]

Recent studies have identified key molecular abnormalities associated with ADCC development and progression. The most prevalent genetic anomalies involve translocations affecting the MYB gene on chromosome 6q and the NFIB gene on chromosome 9p. These genetic alterations result in increased expression of MYB and NFIB oncoprotein, which are crucial in regulating cellular processes such as growth, differentiation, and survival.[6] Adenoid Cystic Carcinomas demonstrate elevated expression of genes linked to myoepithelial differentiation, along with heightened levels of the transcription factor SOX4. SOX4 plays a crucial role in embryonic development and is considered a potential oncogene in humans. Additionally, they exhibit overexpression of other genes such as Casein Kinase 1-Epsilon and Frizzled-7, which are involved in the Wnt/beta-catenin signaling pathway and tumorigenesis. These tumors frequently display increased levels of receptor tyrosine kinase c-KIT and variable overexpression of other growth factor receptors, including Fibroblast Growth Factor Receptor 1 (FGFR1), Epidermal Growth Factor Receptor, and/or Human Epidermal Receptor-2 (HER2).[8]

ADCC typically occurs in adults aged 40 to 60 years but can also affect children and adolescents. While both genders are susceptible, there is a slight female predominance. ADCC commonly arises in the palate, tongue, and floor of the mouth, and can occur on the lips, buccal mucosa, retromolar trigone, and tonsillar area.[6]

ADCC arising from minor salivary glands usually presents as a painless, gradually enlarging mass. Patients might also report sensations of numbness or tingling in the affected region, attributed to perineural invasion, which is a hallmark of ADCC. Given that clinical manifestations can mimic other salivary gland tumors like pleomorphic adenoma and mucoepidermoid carcinoma, performing a biopsy is crucial for precise diagnosis.[6]

The gross pathology of ADCC typically presents as a firm, unencapsulated mass. Upon sagittal sectioning, the tumor appears gray-white in color. The presence of hemorrhage and necrosis within the mass suggests a high-grade transformation.[5]

Fine-needle aspiration cytology serves as a diagnostic tool, with characteristic findings such as large extracellular matrix globules, partially enveloped by basaloid cells, indicating adenoid cystic carcinoma (ADCC). However, efficacy of FNAC in diagnosing salivary gland cancers is hindered by frequent false-negative results.[9]

ADCC presents with three main histopathological patterns: cribriform, tubular, and solid. These subtypes are characterized by distinct arrangements of epithelial, luminal cells, myoepithelial cells, and the extracellular matrix. This tumor primarily presents with a cribriform pattern, which is the most prevalent, while the solid form is the rarest.[6]

ADCC can be categorized into high and low grades based on histological patterns. Low-grade includes Grade I: Predominantly tubular, with no solid areas or occasionally solid areas, and Grade II: Predominantly cribriform, with less than 30% solid component. High-grade includes Grade III: With more than 30% solid component.[10]

The histopathological patterns of ADCC, including cribriform, tubular, and solid subtypes, serve as crucial indicators for assessing clinicopathological and prognostic characteristics. Studies have shown that the solid subtype often exhibits less differentiation, a denser extracellular stroma, and heightened local aggressiveness, frequently associated with perineural invasion (PNI) and a poorer prognosis. PNI, a notable histological feature in ADCC, correlates closely with distant metastasis and adverse disease outcomes, often requires more aggressive treatment strategies.[6]

Pseudocysts exhibit positivity for Periodic Acid Schiff reagent and Alcian blue, containing basement membrane constituents like Type IV collagen, heparin sulfate, and laminin isoforms. Immunohistochemical investigations reveal that CEA and EMA show positivity in epithelial cells, while c-KIT (CD117) is expressed in duct lining cells. Myoepithelial cells demonstrate positivity for S-100 protein, Calponin, p63, SMA, and Myosin. Moreover, p53 mutations are implicated in tumor progression and recurrence.[5]

While the 5-year survival rate for these patients is relatively high, the long-term survival rates, particularly at 10 to 20 years post-diagnosis, are notably low.[5]

The differential diagnosis of ADCC includes tumors that also exhibit tubular and cribriform structures such as Polymorphous Adenocarcinoma (PAC) but it shows cuboidal or columnar cells with pale and ovoid nuclei with eosinophilic cytoplasm in contrast to hyperchromatic and angulated cells of ADCC. Also high c-KIT expression is seen in ADCC compared to PAC. Tumors with basaloid cellular morphology such as basal cell adenoma that has a capsule and lacks stromal and perineural invasion and basal cell adenocarcinoma which lacks clear cytoplasm and hyperchromatic, angulated nuclei but consists of peripheral palisaded nuclei, can be differentiated from ADCC. Because of tumors with a dual population of ductal and myoepithelial cells Pleomorphic Adenoma (PA) is considered as DD but PA consists of mesenchymal differentiation in the stroma and shows positivity with GFAP and CD57, but ADCC does not react with these markers.[11]

Management of ADCC is diverse, involving surgery, radiation, and chemotherapy, customized based on histological subtypes and the presence of perineural invasion (PNI). Despite advancements, surgery remains the primary treatment approach for ADCC arising from major and minor salivary glands in the head and neck area.[6]

4. Conclusion

Adenoid cystic carcinoma (ADCC) is a malignant tumor affecting the palate, buccal mucosa, and major salivary glands. The case report underscores the clinical significance of ADCC, highlighting its aggressive nature and diagnostic intricacy. Despite its rarity, ADCC necessitates prompt diagnosis and multidisciplinary management due to its propensity for perineural invasion and distant metastasis. Immunohistochemical studies play a crucial role in confirming the diagnosis and understanding of the molecular biology underlying ADCC. Continued research is essential to enhance our understanding of this neoplasm and improve treatment outcomes.

5. Source of Funding

None.

6. Conflict of Interest

None.

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