Adenoid cystic carcinoma of palate: Report of a solid variant

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INTRODUCTION

Adenoid cystic carcinoma (ADCC) is a malignant salivary gland tumor. The credit for its initial description goes to Theodor Billroth who discovered the lesion (1859).[1] He suggested the nomenclature cylindroma attributed to its cribriform appearance formed by the tumor cells with cylindrical pseudolumina (pseudospaces). The term “ADCC” was introduced by Foote and Frazell in their fascicle of major salivary gland tumors in 1954. ADCC is a relatively rare malignant salivary gland tumor comprising <1% of all malignancies of the head and neck. It is the fifth most common malignancy of salivary gland origin, representing 20% of all salivary gland neoplasms.[2,3] The tumor is most often clinically deceptive by its small size and slow growth, which actually overlies its extensive subclinical invasion and marked ability for early metastasis making the prognosis questionable, aptly referred as “wolf in sheep’s clothing.” Cribriform, tubular and solid are the three recognized histopathological patterns. In ADCC, one of the important prognostic factors is the histological grade determined by the percentage of solid component in the tumor, which in the present study comprise more than 30% of the entire lesion rendering it an unfavorable prognosis.

CASE REPORT

A 68-year-old male patient reported with a chief complaint of pain on the right side of lower jaw radiating to ear for the past 15 days [Figure 1]. According to the patient, 2 months before our consultation, there was a nasal discharge from the right nostril which stopped after ayurvedic treatment. The patient revealed that a mild pain was persistent even after taking analgesics. Right submandibular lymph nodes were palpable but nontender. Intraoral examination revealed a solitary ulcer of size approximately 1 cm × 1.5 cm in posterolateral region of soft palate extending from the distal aspect of the second premolar till tuberosity region [Figure 2]. Computed tomography scan revealed the presence of a tumor mass obliterating right maxillary sinus and eroding bone in tuberosity region [Figures 3 and 4]. The lesion was provisionally diagnosed with ADCC. The differential diagnosis included basaloid variant of oral

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squamous cell carcinoma and polymorphous low-grade adenocarcinoma (PLGA).

An incisional biopsy of the lesion was performed. Histopathological examination revealed nests and sheets of small, darkly staining basaloid cells and identifiable small duct-like structures. Few cell groups revealed cribriform pattern composed of uniformly shaped basaloid tumor cells with scanty cytoplasm and angulated basophilic nucleus [Figures 5 and 6]. The overall clinicopathological correlation was suggestive of solid variant of ADCC.

DISCUSSION

ADCC is a malignant neoplasm arising from salivary glands of the head-and-neck region. ADCC is thought to arise from the mucous-secreting glands. It arises specifically from the intercalated ducts, and electron microscopy shows that it arises from cells that can differentiate into epithelial and myoepithelial cells. ADCC is confined to structures derived from the foregut (i.e., parotid, submandibular and sublingual glands, and the mucus glands throughout the upper respiratory tract).[4]

Among the major salivary glands, parotid is the most common site of occurrence followed by submandibular glands and together they account for 55% of the cases intraorally 50% of ADCCs occur on the palate. ADCC accounts for 8.3% of all palatal salivary gland tumors and 17.7% of malignant palatal salivary gland tumors (Armed Force Institute of Pathology [AFIP] series). The other less common sites are lower lip, retromolar area, sublingual gland, buccal mucosa and floor of the mouth.[5,6] The nose and paranasal sinuses represent the next most common sites for minor gland ADCCs.

**Figure 1:** A clinical image shows 68-year-old male c/o of pain on the right side of lower jaw radiating to ear for 15 days

**Figure 2:** Intraoral examination showing ulcer of size approximately 1 cm × 1.5 cm in posterolateral region of soft palate extending from distal of 15 to tuberosity region

**Figure 3:** Coronal computed tomography section showing obliteration of the right maxillary sinus and erosion of bone in tuberosity region by tumor mass

**Figure 4:** Axial computed tomography section showing erosion of the right maxillary tuberosity
The three recognized histological variants of ADCC are cribriform, tubular and solid, cribriform being the most common and solid the least common. It is recognized that most ADCCs do not occur in “pure” cribriform, tubular or solid types. It is common to have more than one histopathologic pattern in a single neoplasm and rather all three patterns can be observed in the majority of tumors. The most frequently observed pattern is the cribriform pattern, characterized by nests, sheets and clusters of tumor cells discretely separated from each other by uniform bands of the collagenized stroma. These cribriform nests are composed of small, darkly stained basal or myoepithelial cells, with inconspicuous duct-like structures containing the excretory product. Mitoses are usually minimal. round to oval uniformly-sized intercellular spaces filled with hyaline or basophilic mucoid material lends it a “Swiss cheese appearance.”

In the tubular pattern, well-formed ducts and tubules are seen with the lumen at the center and further lined by an inner layer of cuboidal to columnar ductal cells and an outer layer of smaller, darkly staining myoepithelial cells. In the solid pattern, isomorphic tumor cells characteristic of cribriform and tubular types are arranged in nests or sheets of varying size and shape. There is the minimal tendency of formation of cystic spaces as in cribriform type or duct-like structures as observed in tubular type. In the central portion of solid tumor islands, areas of necrosis may be found along with cellular pleomorphism and mitoses which are usually not found in other variants of ADCC.

Perineural and to a lesser extent, the intraneural invasion is a common and frequently conspicuous feature of ADCC. Tumors can extend along nerves for a considerable distance beyond the clinically apparent boundaries of the tumor. Perineural invasion (PNI) is grouped into two categories as follows: incidental and clinical. Incidental includes asymptomatic patients with evidence of microscopic PNI detected only by histopathology. Clinical PNI includes patients with evidence of a cranial neuropathy on physical examination and/or radiographic evidence of gross tumor involvement along the tract of the nerve. In addition, the tumor may invade bone extensively before there is radiographical evidence of osseous destruction.

The histopathological feature has a significant effect on grading and consequently the treatment modality. According to Szanto et al., ADCC can be graded into three categories; Grade I - cribriform or tubular pattern, Grade II - <30% solid pattern and Grade III - more than 30% solid pattern.

In most of the studies, a solid growth pattern has been associated with the worst prognosis due to advanced stage and development of distant metastasis. An increased solid component is also associated with increased risk of local recurrence.

The differential diagnosis includes PLGA, salivary duct carcinoma, basal cell adenoma (BCA) and basaloid squamous cell carcinoma (BSC).

Considering PLGA, a polymorphous architecture characterizes PLGA, whereas ADCC has a more limited range of histologic patterns with no more than three patterns. Foci of papillary growth and areas of single cell infiltration are characteristic of PLGA. Basophilic pools of glycosaminoglycans are seen in ADCC but not in PLGA. PLGA shows uniform cell population with cytologically bland, round or oval vesicular nuclei and pale eosinophilic cytoplasm whereas cells in ADCC have clear cytoplasm, angular, hyperchromatic nuclei and may show mitotic activity.

The differential diagnosis includes PLGA, salivary duct carcinoma, basal cell adenoma (BCA) and basaloid squamous cell carcinoma (BSC).
In salivary duct carcinoma, the central portion of nests of tumor cells contains necrotic material (comedonecrosis), whereas no such features are observed in ADCC. Cribriform structures may sometimes be observed in BCA and such cases can be differentiated from ADCC on the basis of gradual structural alteration from areas typical of BCA. The peripheral palisading and focal squamous differentiation with whirling pattern present in BCA are not usually encountered in ADCC.\[11\]

BSC should be differentiated from solid ADCC of minor salivary glands. The basement membrane material secreted by BSC tends to dissect between tumor cells rather than form crisp cribriform spaces seen in ADCC. Focal keratinization, attachment to rete pegs, and the presence of surface squamous dysplasia or carcinoma in situ help to distinguish it from ADCC.

Possible treatments of ADCC include four different modalities as follows: surgical therapy, radiotherapy, chemotherapy and combined therapy (surgery and radiotherapy, radiotherapy and chemotherapy) being the latter in most cases, the treatment of choice. Only surgical removal or radiotherapy in isolation may fail to eliminate the possibility of spread in surgical margins, as well as the occurrence of metastasis in cervical lymph nodes, lungs, bones, and brain.\[23\]

In general, in ADCC, it is observed that tubular pattern (well differentiated) is believed to have the best prognosis compared to the cribriform pattern (moderately differentiated) and solid pattern (poorly differentiated). It is generally agreed that solid type ADCC has a relatively poor prognosis. However, the amount of solid regions within this often mixed type tumor that predicts a poor prognosis is not firmly established. Some authors stipulate that the presence of a solid component regardless of the amount is a poor prognosticator where others argue that the amount should be taken into consideration. Two grading systems most commonly used are those described by Perzin et al., Szanto et al. and Spiro et al., respectively. They report that prognosis of ADCC is poor if >30% and >50% of the tumor volume has a solid growth pattern, respectively.\[6,7\] ADCCs have a well-known prognostic profile. The 5-year survival rate is 75%, but the 10-year survival rate is only 20% and survival rate at 15 years is about 10%.\[13\]

**CONCLUSION**

ADCC is rather an uncommon salivary gland malignancy. It is unique for its peculiar histopathological features and tendency for PNI. Prognostic factors of ADCC are the anatomic site, histologic subtypes and metastasis. ADCC with a solid histopathologic pattern is associated with a worse prognosis than those with a cribriform or tubular arrangement. Therefore, early detection, prompt treatment and long-term follow-up are essential in the clinical management of this tumor.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

**REFERENCES**

1. Lucas RB. Pathology of Tumors of the Oral Tissues. 4th ed. London: Churchill Livingstone; 1998. p. 330-5.
2. Schwarz S, Müller M, Ettl T, Stockmann P, Zenk J, Agaimy A, et al. Morphological heterogeneity of oral salivary gland carcinomas: A clinicopathologic study of 41 cases with long term follow-up emphasizing the overlapping spectrum of adenoid cystic carcinoma and polymorphous low-grade adenocarcinoma. Int J Clin Exp Pathol 2011;4:336-48.
3. Luksič I, Suton P, Macan D, Dinjar K. Intraoral adenoid cystic carcinoma: Is the presence of perineural invasion associated with the size of the primary tumour, local extension, surgical margins, distant metastases, and outcome? Br J Oral Maxillofac Surg 2014;52:214-8.
4. Marx RE, Stern D. Oral and Maxillofacial Pathology, a Rational for Diagnosis and Management. Chicago: Quintessence; 2002. p. 550-3.
5. Bradley PJ. Adenoid cystic carcinoma of the head and neck: A review. Curr Opin Otolaryngol Head Neck Surg 2004;12:127-32.
6. Balamucki CJ, Amanda RJ, Werning JW, Vaysberg M, Morris CG, Kirwan JM, et al. Adenoid cystic carcinoma of the head and neck. Am J Otolaryngol 2012;33:510-8.
7. Gneppe DR, Henley JD, Roderick HW, Eveson JS. Salivary and lacrimal glands. In: Gneppe DR, editors. Diagnostic Surgical Pathology of the Head and Neck. 2nd ed. Philadelphia: Saunders; 2009. p. 482-6.
8. Bianchi B, Copelli C, Cocchi R, Ferrari S, Pederneschi N, Sesenna E, et al. Adenoid cystic carcinoma of introral minor salivary glands. Oral Oncol 2008;44:1026-31.
9. Barrett AK, Speight PM. The controversial adenoid cystic carcinoma. The implications of histological grade and perineural invasion. In: McGurk M, Renahan A, editors. Controversies in the Management of Salivary Gland Disease. Oxford: Oxford University Press; 2002. p. 211-7.
10. Chandra NS, Amudala R, Thakapam P, Nagarajua C D. Adenoid cystic carcinoma of palate: A case report and review of literature. Dent Res J (Isfahan) 2013;10:274-8.
11. Nagao K, Matsuzaki O, Suga H, Sugano I, Shigematsu M, Kanelo T, et al. Histopathologic studies of basal cell adenoma of the parotid gland. Cancer 1982;56:736-45.
12. Manisha S, Aelu RR, Govind P, Amina R. Adenoid cystic carcinoma. J Ahmedabad Dent Coll Hosp 2013;4:41-50.
13. Pushpanjali M, Sujata DN, Subramanyam SB, Jyothsna M. Adenoid cystic carcinoma: An unusual presentation. J Oral Maxillofac Pathol 2014;18:286-90.