

Adenoid Cystic Carcinoma – A Case Report

G. Lavanya, M. Keerthi, Aruna Kumari, P. Gouri

Department of Oral and Maxillofacial Pathology, Kamineni Institute of Dental Sciences, Narketpally, Telangana, India

Email for correspondences: dr.lavanya1606@gmail.com

ABSTRACT

Salivary gland neoplasms of the head and neck comprise of 1–5% of the total malignancies. Adenoid cystic carcinoma (ACC) is the second most common malignancy of the salivary gland. It is a slow-growing, rare malignant neoplasm of the salivary glands. It carries a poor prognosis due to aggressive nature of tumor and a high rate of recurrence. There is a female preponderance with peak incidence is in the fifth and sixth decades. Palate is the most common site to be involved in the oral cavity. They are usually asymptomatic and characteristically show infiltrative growth and perineural invasion. Perineural invasion along with cranial nerves is a pathognomonic feature of ACC. Surgery is the treatment of choice, but some cases may require radiation therapy after surgery. Rarely, chemotherapy may also be required to control the disease. Here, we present a case report of a 24-year-old female patient with a chief complaint of pain and swelling on the right side of palate for 4 months which was diagnosed histopathologically as a ACC.

Key words: Adenoid cystic carcinoma, malignant salivary gland tumor, palate.

INTRODUCTION

Adenoid cystic carcinoma (ACC) is a malignant salivary gland tumor that was first described by Billroth in 1859 and gave the term cylindroma attributing to its cribriform appearance formed by the tumor cells with cylindrical pseudolumina or pseudospaces.^[1] In 1954, Ewing introduced the term “ACC.” ACC is a relatively rare malignant salivary gland tumor comprising <1% of all malignancies of head and neck, represents 5–10% of all salivary gland neoplasms, and accounts for 17.7% of malignant palatal salivary gland tumors.^[2,3]


Around 55% of the cases are associated with parotid and submandibular glands, among which the parotid is the most common site of occurrence,^[1] whereas 50% of intraoral ACC occur on the palate. The other less common sites include the lower lip, retromolar tonsillar pillar area, sublingual gland, buccal mucosa, and floor of the mouth.^[3] The tumor, most often, exhibits a small size and slower

growth, which overlies its extensive subclinical invasion and early metastasis making the prognosis questionable.^[4] The three histological variants of ACC are cribriform, tubular, and solid, although cribriform is the most common and solid is the least common. Most ACCs do not occur in “pure” cribriform, tubular, or solid types, and they exhibit more than one histopathologic pattern in a single neoplasm. Tumors are classified generally according to the histologic pattern that predominates. Tubular pattern (well differentiated) has the best prognosis compared to cribriform pattern (moderately differentiated) and solid pattern (poorly differentiated). According to Szanto et al., ACC is graded as Grade I (cribriform or tubular), Grade II (<30% solid), and Grade III (>30% solid).^[5]

Here, we present a case report of a 24-year-old female patient with a chief complaint of pain and swelling on the right side of palate for 4 months which was diagnosed histopathologically as a ACC.

CASE REPORT

A 24-year-old female patient complains of pain and swelling on the right side of palate for 4 months. The patient gave a past history of extraction in relation to 18, 1 month back. The swelling was sudden in onset and was gradually progressing. On extraoral examination, no apparent abnormality was detected. Intraoral examination reveals a

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diffuse swelling measuring about 3 cm × 4 cm extending from mesial aspect of 15 to distal aspect of 17 [Figure 1]. Medially, the swelling extends from 0.2 cm below the gingiva to mid-palatal raphe. The overlying surface is smooth. Right submandibular lymph node enlargement is felt which is around 2 cm × 2 cm in size, oval in shape, and is non-tender. The computerized tomography showed the expansion of buccal cortical plates in relation to 16 and 17 [Figure 2]. The lesion was provisionally diagnosed as minor salivary gland tumor. An incisional biopsy was performed and was sent for histopathological examination [Figure 3]. H and E stained section shows the presence of cells arranged in the form of islands, nests, sheets, and cords. These malignant cells are basaloid in shape [Figures 4 and 5]. The

basaloid shape cells exhibited features of nuclear atypia such as nuclear and cellular pleomorphism and nuclear hyperchromatism. The islands show the features of duct-like structures with an inner layer of cuboidal or columnar cells [Figure 6]. There is also the presence of pseudocystic space enclosing of homogeneous eosinophilic material [Figure 7]. The connective tissue exhibited hyalinization and necrosis. Based on histopathological features, the final diagnosis was given as ACC. In the present case, cribriform and tubular types are identified.

DISCUSSION

The ACC constitutes approximately 29.6% of minor salivary gland tumor. Most commonly



Figure 1: Clinical picture showing a diffuse ill-defined swelling in the right side of the hard palate



Figure 3: The incisional biopsy specimen measuring about 0.5 cm × 0.5 cm in size that was sent for histopathological examination

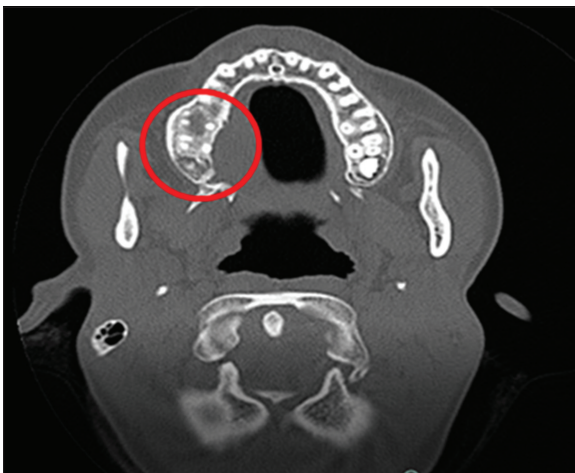


Figure 2: Computerized tomography scan reveals expansion of buccal cortical plates in relation to 16 and 17 tooth region

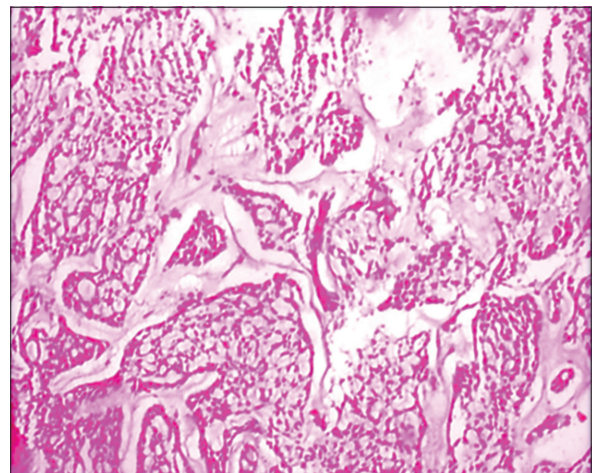


Figure 4: H and E stained section shows cells that are arranged in islands, cords, nests, and sheets (×4)

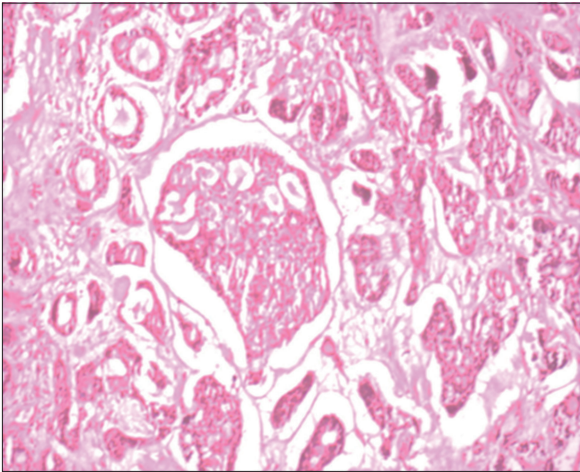


Figure 5: H and E stained section shows malignant glandular epithelium with basaloid cells ($\times 10$)

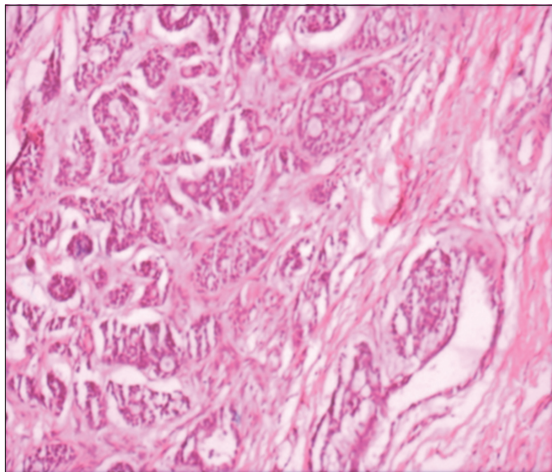


Figure 6: H and E stained section shows duct-like structures with an inner layer of cuboidal or columnar cells ($\times 10$)

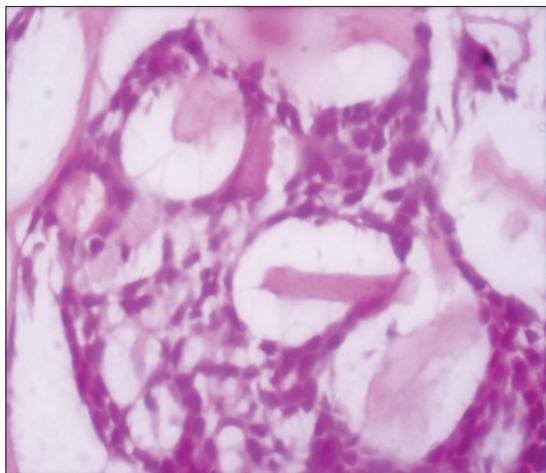


Figure 7: H and E stained section shows multiple pseudocystic spaces of variable sizes filled with eosinophilic material ($\times 40$)

involved site is palate followed by tongue, floor of mouth, and lip. Other rare locations include the aerodigestive tract, minor salivary glands, lacrimal glands, and adnexal skin glands. Rarely, it may also present as primary intraosseous tumors of the maxilla and mandible.^[6]

ACC 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.^[7]

ACC is a diagnostic and treatment challenge due to the rarity of the lesion. More studies and further information regarding its clinical behavior as well as treatment modalities and their results are required for ACC. Most cases of ACC are seen in the age of 24–78 years. The age of patients affected with major salivary gland tumors has been shown to be younger (mean 44 years) compared to the age of those who developed tumors of the minor glands (mean 54 years) and shows female predilection (female:male 1.2:1).^[8] In the present case, patient was 24-year-old female, which is a common age and gender for ACC. Pain is a common and important finding, occurring early in the course of the disease before there is a noticeable swelling.^[8] Among the malignant neoplasms of minor salivary glands, the most common was mucoepidermoid carcinoma (21.8%) followed by polymorphous low-grade adenocarcinoma (PLGA) (7.1%), and ACC was the third most common (6.3%).^[9,10] ACCs of the minor glands have been reported to have a worse prognosis than those of the major salivary glands.^[11,12] Since the present case is arising from a minor salivary gland, it is expected to have a poor prognosis. Tumors involving the nose, paranasal sinuses, and maxillary sinus have the worst prognosis as they are usually detected with higher stages at the time of diagnosis.^[13]

Tumors of minor salivary glands usually have the tendency to infiltrate extraglandular soft tissues and bone, thereby allowing increased dissemination of the tumor. Lymph node involvement is uncommon (<5% of cases) and is usually due to contiguous spread rather than lymphatic permeation or embolization.^[14]

Histologically, ACC of the salivary glands presents with variable combinations of the three main growth patterns, i.e., cribriform, tubular, and solid. In the present case, a combination of cribriform and tubular is seen. The cribriform is the most

characteristic in which the basaloid cells constitute the major cell population, showing mild nuclear pleomorphism and few or no mitoses; in the solid variant, these cells usually show a more pleomorphic appearance and mitoses are more commonly found.^[15]

Salivary gland ACC is graded using a specific three-tier grading system, originally proposed by Szanto et al. in 1984, which is solely based on the main type of growth pattern presenting the tumor. Grade 1 ACCs are well differentiated and composed of tubular and cribriform patterns without solid components, Grade 2 ACCs are characterized by a pure cribriform pattern or mixed with <30% of solid areas, and Grade 3 ACCs are tumors with a marked predominance of the solid pattern. The prognostic factor of ACC depends on the site of the tumor, stage of the tumor, the presence of perineural invasion, and grade of the tumor. Tubular and cribriform subtypes have a better prognosis than solid subtypes. Many authors described poor prognosis for tumors of the minor salivary glands, due to early local infiltration and invasion of surrounding tissue and bone.^[15]

Other special stains that are used to demonstrate the pseudocysts are periodic acid Schiff reagent and Alcian blue. On immunohistochemical analysis, epithelial cells are positive for carcinoembryonic antigen and epithelial membrane antigen. Duct lining cells are positive for C-kit (CD117) and myoepithelial cells are positive for S-100 protein, calponin, p63, smooth muscle actin, and myosin. Expression of S-100, glial fibrillary acidic protein, and neural cell adhesion molecule have been correlated with the presence of perineural invasion. P53 mutations appear to be involved with tumor progression and recurrence.^[16] Hypermethylation of the promoter region of the p16 gene was associated with higher histologic grades of malignancy. Microarrays and comparative genomic hybridization have been used to identify candidate genes for ACC.^[16]

CONCLUSION

ACC is an uncommon salivary gland malignancy. It is unique for its peculiar histopathological features and tendency for recurrence. Prognostic factors of ACC are the anatomic site, histologic subtypes, and metastasis. ACC is sometimes associated with a poor prognosis. Therefore, early clinical and histopathological diagnosis, prompt treatment, and long-term follow-up are essential in the management of this tumor.

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