

Palatal Adenoid Cystic Carcinoma: a case report and Review of Literature

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ABSTRACT

Adenoid cystic carcinoma (ADCC) is a malignant salivary gland tumor that was first described by Billroth in 1859 under the name “cylindroma” characterizing to its cribriform appearance formed by the tumor cells with cylindrical pseudolumina or pseudospaces. The disease is seen most commonly in the fifth and sixth decades of life but may appear at almost any age. In a recent population study, ADCC occurred more commonly in women than men (60:40 ratio). The three recognized histological variants of ADCC are cribriform, tubular and solid although, cribriform is the most commonest and solid is the least common.

The article describes a case of a 46-year-old female patient reported to the college with a chief complaint of pain in his upper right back tooth region of the jaw since 2 months. According to the patient pain was persistent for 1 month before our consultation and was associated with a swelling. On intraoral examination, it was seen associated with a solitary erythematous swelling with clear borders involving the right posterior portion of hard palate. The swelling was small initially and gradually increased to the present size.

Keywords: Cribriform, Cylindroma, Salivary Gland Neoplasm

INTRODUCTION

Adenoid cystic carcinoma (ADCC) is a malignant salivary gland tumor that was first described by Billroth in 1859 under the name “cylindroma” characterizing to its cribriform appearance formed by the tumor cells with cylindrical pseudolumina or pseudospaces¹. The salivary glands are the most common site of origin for ADCC. As an uncommon tumor type, ADCC accounts for only 1% of all malignant tumors of the head and neck region and 10% of all salivary gland neoplasms^{2,3}. ADCC occurs more frequently in minor as compared with major salivary glands. Other sites of origin in the head and neck are the tongue, paranasal sinuses, palate, nasopharynx, lacrimal glands, and external auditory canal. ADCC may also arise in secretory glands located in other tissues, such as in the tracheobronchial tree, esophagus, breast, lungs, prostate, uterine cervix, Bartholin’s glands, and vulva^{4,5}. The disease is seen most commonly in the fifth and sixth decades of life but may appear at almost any age. In a recent population study, ADCC occurred more commonly in women than men (60:40 ratio)⁶. Due to the rarity of these tumors, there is little data on whether specific ethnic, geographic, exposure, or other factors predispose to the development of the disease. 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

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metastasis making the prognosis questionable. The three recognized histological variants of ADCC are cribriform, tubular and solid although, cribriform variant is the most common one. It is noticed that most of the ADCCs do not occur in “pure” cribriform, tubular or solid types. It is not uncommon to have more than one histopathologic pattern in a single neoplasm and rather all three patterns can be observed in the

majority of tumors⁷. Tumors are generally classified according to the histologic pattern that predominates. The main reason for histological typing is to assess the prognostic difference between histologic types. Tubular pattern (well differentiated) is believed to have the best prognosis compared to the cribriform pattern (moderately differentiated) and solid pattern (poorly differentiated). ADCC is graded according to Szanto, et al. as cribriform or tubular (grade I), less than 30% solid (grade II) or greater than 30% solid (grade III)⁸.

CASE REPORT

A 46-year-old female patient reported to the college with a chief complaint of pain in his upper right back tooth region of the jaw since two months. According to the patient pain was persistent for 1 month before our consultation and was associated with a swelling. The extraoral examination was within normal limits with no evidence of lymphadenopathy. Her medical history was noncontributory. On intraoral examination, it was seen associated with a solitary erythematous swelling with clear borders involving the right posterior portion of hard palate.(Figure 1) The swelling was small initially and gradually increased to the present size and was seen extending from mesial aspect of 16 to 2 cm anterior to 18, involving

the palatal area but not crossing the midline. The mucosa over the swelling was smooth and was continuous with the adjacent mucosa. On palpation, the inspector findings were confirmed. The swelling was nontender, nonfluctuant, firm in consistency and immobile. No bony lesion was observed in maxillary occlusal view. (Figure 2)

The clinical differential diagnosis included a benign or malignant neoplasm of minor salivary glands. Routine blood investigations were found to be normal. An incisional biopsy of palate was performed. On gross examination, multiple soft tissue specimen were received measuring approximately 1.5 cm x 1 cm x 1 cm. (Figure 3) Histopathological examination revealed fibrovascular connective tissue stroma with mild chronic inflammatory cell infiltrate consisting predominantly of lymphocytes. There was presence of numerous islands of small cuboidal to polygonal-shaped cells with little cytoplasm and hyperchromatic nuclei resembling basaloid epithelial cells along with multiple cylindrical cyst-like spaces filled with eosinophilic material.(Figure 4, 5) There was no evidence of perineural invasion even on serial sectioning. Based on histopathology, diagnosis of ADCC (cribriform pattern) was established. The patient was treated by wide surgical excision with clear



Fig. 1. Intraoral solitary erythematous swelling involving the right posterior portion of hard palate

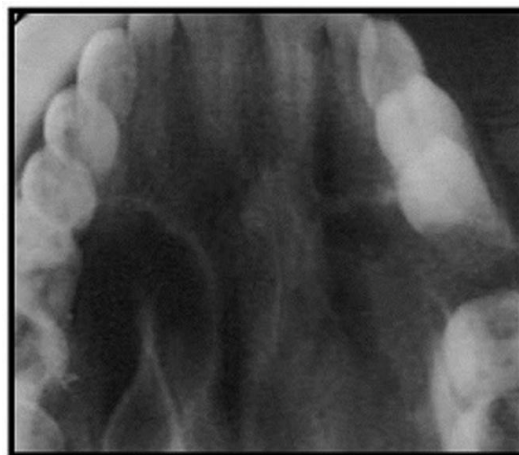
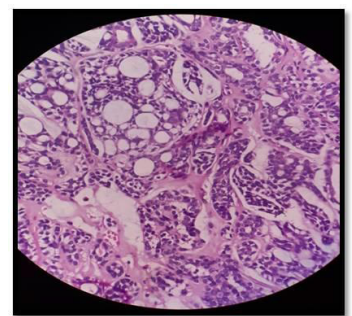
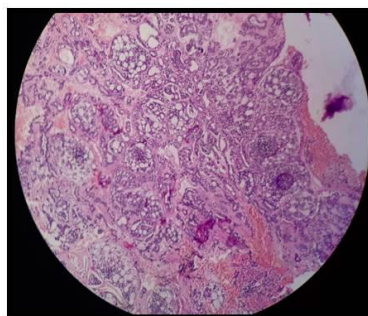


Fig. 2 Occlusal & Panoramic radiograph.



Figure 3. Multiple soft tissue specimens received after Incisional Biopsy



Figures 4, 5. Photomicrograph showing islands of small cuboidal to polygonal-shaped cells with little cytoplasm and hyperchromatic nuclei resembling Swiss-cheese (40X)

margins and hemi-maxillectomy of left maxillary region with postradiotherapy. The patient is under regular follow-up.

DISCUSSION

Adenoid cystic carcinoma most often presents a diagnostic and treatment challenge owing to the rarity of the lesion. Most findings regarding ADCC are actually based upon studies with a small number of patients and there is a need for further information regarding its clinical behaviour as well as treatment modalities and their results ADCC may occur at any age although in most cases the patients' age ranged from 24 to 78 years⁸. 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 (F:M – 1.2:1). Pain is a common and important finding, occurring early in the course of the disease before there is a noticeable swelling⁹. Among the minor salivary glands, palate is more commonly involved generally the area of greater palatine foramen.

Among the malignant neoplasms of minor salivary glands, the most common is mucoepidermoid carcinoma (21.8%) followed by polymorphous adenocarcinoma (PAC) (7.1%) and ADCC is the third most common (6.3%)^{10,11}. ADCCs of the minor glands have been reported to have worse prognosis than those of the major salivary glands. Tumors of minor salivary glands usually have the tendency to infiltrate extra glandular soft tissues and bone thereby allowing increased dissemination of the tumor¹². Lymph node involvement is uncommon (<5% of cases) and is usually due to contiguous spread rather than lymphatic permeation or embolization¹³. Microscopically, ADCC is composed of small basaloid epithelial, nonluminal, hematoxyphilic cells, with small or moderate cytoplasm which are arranged in three growth patterns: cribriform, tubular and solid as described earlier. Generally, nuclei are not so pleomorphic, with small or bland nucleoli. This type of tumor shows a predominant myoepithelial differentiation. Cribriform pattern is the most frequent and is composed of basaloid cells organized in oval/rounded masses of variable size, punched out by rigid, oval, cyst-like spaces (pseudolumina) that may contain "cylinders" (i.e., globules of hyaline material and/or myxoid glycosaminoglycans) and occasionally, small "true lumina" lined by luminal cells, composing a "swiss cheese pattern"¹³.

Diagnosis of salivary gland tumor by FNAC is difficult and is often compromised by false negative evaluations. Histopathological diagnosis remains the "gold standard" and especially necessary when planned therapeutic intervention involves radical surgery and possible sacrifice of facial nerve.¹⁴ Possible treatment modalities available for the treatment of ADCC are surgery, radiotherapy, chemotherapy and combined therapy. It requires excision with the widest surgical margins possible as tumor cells extend well beyond the clinical and radiographic margins. Neutron therapy, which involves larger particles of greater energy, can achieve reasonable local control as a primary therapeutic modality. Adoptive immunotherapy along

with chemoradiotherapy has recently shown promising results. E-cadherin expression can be used as prognostic marker. High argyrophilic nuclear organizing regions (AgNor) counts may be predictive of metastasis¹⁵. Main factors associated with patient survival are anatomic location, primary lesion size, presence or absence of metastasis at the time of diagnosis, perineural invasion and histological variant.

CONCLUSION

Salivary gland neoplasms should always be considered as differential diagnosis, especially when the lesion is involving palate. ADCC of head and neck is a common malignant tumor of minor salivary glands and has a tendency of perineural invasion and distant metastasis and therefore early diagnosis and long-term follow-up of patient is mandatory.

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