

## Adenoid Cystic Carcinoma of the Maxilla – A Case Report and 5 Year Follow-up

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### Introduction

Adenoid cystic carcinoma is an uncommon tumour of the salivary and mucous glands which has been recognized for more than one hundred years. In 1359, Billroth [1] originated the word cylindroma in describing histologically four salivary gland tumours, one of which had recurred nine times after excision over a twenty year period. The term cylindroma was in common usage until Foote and Frazell [2] expressed their preference for the name adenoid cystic carcinoma in 1953. They credited the late James Ewing for having used that term for many years. The tumour has been variously referred to as “cylindroma,” “basiloma,” “adenocystic basiloid carcinoma,” and “adenoeplithelioma” [2].

It comprises of 5% to 10% of all salivary gland tumors, which account for 2% to 4% of all head and neck malignancies. It is most frequently located in minor salivary glands (31%), although it is found in the submandibular gland (14%) and the parotid (2%) [3-7]. Vokes et al. [6] have also reported frequent occurrence of Adenoid cystic carcinoma (ACC) in minor salivary glands of the nose and paranasal sinuses. Nearly half of all intraoral ACCs occur in the palate [8,9]. There is a wide age distribution but the peak incidence is in the fifth and sixth decade. There is also a predominance of cases in females [10,11].

The characteristic biological features of the ACC are local recurrences, perineural spread, and late distant metastases. These features make the local control of the disease difficult; even in cases with clinically clear surgical resection margins. Lymphatic spread occurs less commonly than with other malignant epithelial tumors. Blood spread to distant sites occurs particularly in the lung, and usually whenever the primary tumor has been inadequately treated [12-14].

### Case Report

A 72 year old male came to the department of oral and maxillofacial surgery at Sri Ramachandra University for the treatment of a swelling in the roof of the mouth, a dental cause for which could not be found. This swelling had gradually developed over the previous 4 months and had recently begun to cause the patient, an intermittent and dull pain. Patient also gave a history of nose block for the past 1 month and numbness over the right side of the upper lip. The patient had no significant medical history.

On general examination of the patient, the patient was moderately built and nourished, well oriented. There were no signs of pallor, icterus, cyanosis, clubbing, lymphadenopathy and pedal edema. Vitals were within normal limits (Figure 1).

On Local examination, Pupils were equally reactive to light, Extra ocular movement were intact, Face was Symmetrical, Mouth opening-39 mm. Intra oral examination revealed the swelling was present on the right side of the palate with a central area of ulceration. The swelling was Oval shape and measuring 2x1 cm in size. Anteriorly it extends from the right side Palatal Rughae and posteriorly to the soft palate. Medially it extends from the midline of palate and laterally to

the dentoalveolar process of tooth no's [15-17] region. On palpation, the swelling appears to be fixed, firm in consistency and non-tender (Figure 2). Teeth in relation to swelling are non-mobile/non-carious/non-tender. Paraesthesia was found on right side of upper lip. Regional lymph nodes were not clinically palpable.

Radiological investigations like: Computerized tomography scan of paranasal sinus, CHEST PA-X RAY, Ultra sound Abdomen, the latter two investigations was performed to identify distant organ involvement.

CT scan of PNS view suggestive of Hyper dense area in the right maxillary sinus extending medially to the inferior turbinate, Superiorly to the orbital floor, laterally contained within the right maxillary sinus and posteriorly extends to the pterygoid plates, perforation in the hard



Figure 1: Clinical Photograph -Patient.

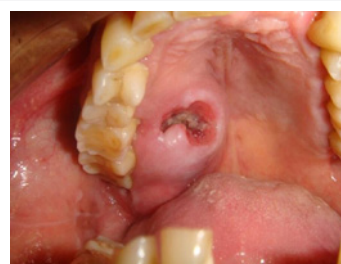


Figure 2: Swelling in the hard and soft palate with central area of necrosis.

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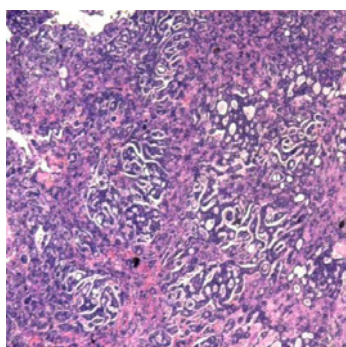
palate at the greater palatine foramen was evident (Figure 3). Chest X-ray and Ultra sound of the abdomen were normal for the patient.

Histopathological Examination Sections shows a tumor composed of monomorphic cells arranged in glandular and canalicular pattern. Focal areas show cribriforming with mucin-like material inside (Figure 4). Focal increase was observed in mitosis. CD-117 is strongly positive of immunohistochemistry which confirms cribriform type of Adenoid cystic carcinoma (Figure 5).

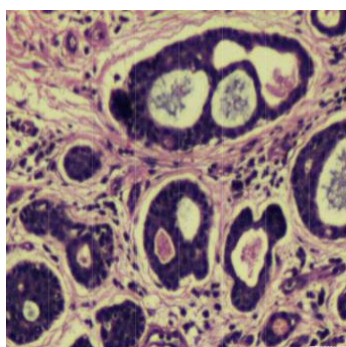
The patient was treated with right hemimaxillectomy (Figure 6) and adjuvant Radiotherapy. Postoperative review after 6 months revealed satisfactory wound healing and no nasal regurgitation, a final



**Figure 3:** CT-PNS -Tumor extending to the inferior turbinate with perforation of hard palate.



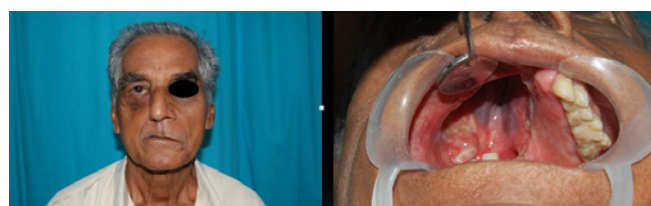
**Figure 4:** Histopathological section showing monomorphic cells arranged in glandular and canalicular pattern. Focal areas show cribriforming with mucin-like material inside.



**Figure 5:** Immunohistochemistry- Tumor cells strongly positive for CD-117.



**Figure 6:** Resected specimen after Right Maxillectomy.



**Figure 7:** 5 year disease free follow up of the patient.



**Figure 8:** Obturator in situ.

obturator was provided. The patient was well and clinically free of local or metastatic disease 5 year after surgery (Figures 7 and 8).

## Discussion

Adenoid cystic carcinoma (ACC) is a rare malignant tumour that accounts for less than 1% of all head and neck malignancies and approximately 10% of all salivary gland neoplasms [1]. Adenoid cystic carcinoma in the nasal cavity and paranasal sinuses origin often has a worse prognosis than in any other area of the head and neck region. It is reported to occur in any age group with a peak incidence in the fifth decade of life [12]. Presenting symptoms may be non-specific such as nasal obstruction, epistaxis and symptoms depending on the structure has invaded.

Three Histological growth patterns have been identified and described: Cribriform, Tubular, and Solid. Cribriform is the most common histologic subtype. Assessment of the histologic grade is of significance in predicting the likelihood of tumor recurrence and survival. In one series of studies, 5-year recurrence rates of 100%, 89%, and 59% were reported for tumors with solid, cribriform, or tubular growth patterns, respectively [17]. Perineural invasion along cranial nerves is a pathognomic factor of adenoid cystic carcinoma and is believed to be responsible for the high rate of local recurrence [16]. Lymphatic spread to local lymph nodes is rare. Lung is the

most common site for metastasis, liver brain, and kidneys are the less common sites of involvement. Fordice et al. considered that the use of appropriately aggressive surgery achieving negative margins was the linchpin of successful combined therapy [12]. A combination of radical surgery and postoperative radiotherapy was the main therapy for Sino nasal adenoid cystic carcinoma compared to either surgery or radiotherapy alone. But despite aggressive surgery, high incidence of positive margins was noted due to the anatomical complexity of the nose and paranasal sinuses. So adjuvant radiotherapy is necessary in such cases [2]. Chemotherapy appears to be ineffective in the treatment of adenoid cystic carcinoma. Long-term follow-up is necessary because of the high incidence of local recurrence and distal metastasis [16].

## Conclusion

Even though in our case the negative prognostic factors like tumor site within minor salivary glands, increased size and stage of the tumor and perineural invasion were present. Early diagnosis and adequate surgical resection with following adjuvant radiotherapy plays a crucial role in long term survival of the patient.

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