

Case Report

## Adenoid Cystic carcinoma of nasal cavity- A Rare Case Report

Venkatraman J<sup>1</sup>, Roopa URS A.N<sup>2</sup>, Dhananjay S Kotasthane<sup>3</sup>

<sup>1,2</sup> Assistant Professor, Department of Pathology, Mahatma Gandhi Medical College and Research Institute, Pondicherry

<sup>3</sup> Professor and Head, Department of Pathology, Mahatma Gandhi Medical College and Research Institute, Pondicherry

**\*Corresponding author**

Venkatraman J

Email: [drvenkatraman1983@gmail.com](mailto:drvenkatraman1983@gmail.com)

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**Abstract:** Adenoid cystic carcinoma is most common tumor arising from major and minor salivary glands and rarely occurs in nasal cavity and paranasal sinuses. ACC was first described by French scientists—Robin, Lorain and Laboulbène in 1853 and 1854. Adenoid cystic carcinoma of the paranasal sinuses or nasal cavity is an aggressive neoplasm that results in a high incidence of both local recurrence and distant metastasis, regardless of treatment modality. Diagnosis and treatment of ACC pose numerous challenges, partly due to its biological behavior of slow growth, high tendency of local recurrence, and perineural spread. Here we report a case of ACC of Nasal cavity presented for its rarity.

**Keywords:** Adenoid cystic carcinoma, paranasal sinuses or nasal cavity

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**INTRODUCTION**

Adenoid cystic carcinoma is most common tumor arising from major and minor salivary glands and rarely occurs in nasal cavity and paranasal sinuses. Adenoid cystic carcinoma is a rare tumor corresponding to 1% of the head and neck region malignant tumors and 10% of all salivary gland tumors [1, 2]. ACC was first described by French scientists—Robin, Lorain and Laboulbène in 1853 and 1854 [3]. Adenoid cystic carcinoma of the paranasal sinuses or nasal cavity is an aggressive neoplasm that results in a high incidence of both local recurrence and distant metastasis, regardless of treatment modality [4]. Diagnosis and treatment of ACC pose numerous challenges, partly due to its biological behavior of slow growth, high tendency of local recurrence, and perineural spread [4, 5]. Three basic growth patterns have been identified, tubular, cribriform, and solid, with the cribriform pattern being the most frequent. Most patterns are often mixed, and the solid component of the tumor correlates with aggressive behavior [5]. In this paper, we describe a case of ACC of the nasal cavity and present the clinical, radiological and histopathological features.

**CASE REPORT**

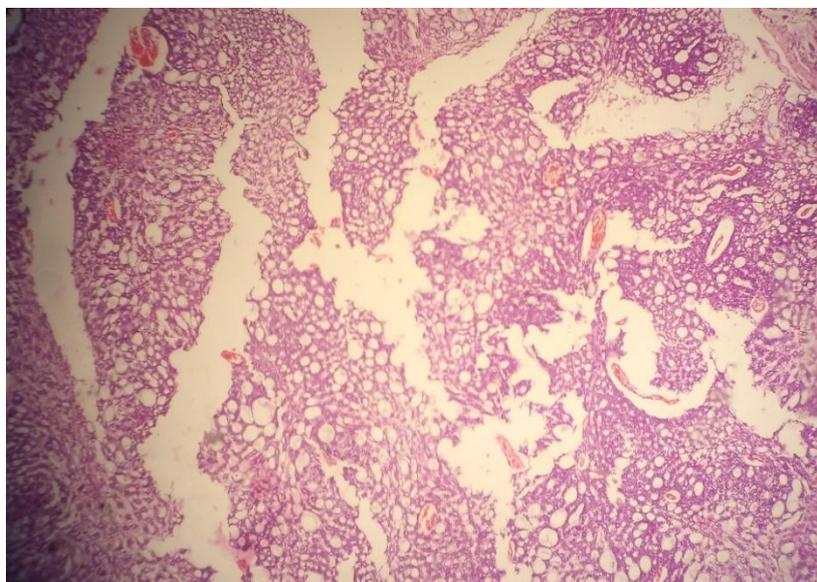
A 29 year-old female came to us with complaints of bilateral nasal obstruction since 9 months and swelling over left side of face since 4 months.

Initially nasal block started on the right side which was insidious in onset, gradually progressive and proceeded to complete nasal block on both sides from the last 6months. The patient also complains of swelling in the left side of face from the last 4 months. The swelling was initially smaller in size and was gradually progressive. On anterior rhinoscopy – mass was seen in right nasal cavity displacing the septum on the left side, occupying floor and lateral wall of the nose. Mass was smooth and relatively painful. Bilateral airway block was noted. Clinically differential diagnosis such as adenocarcinoma, squamous cell carcinoma and inverted papilloma were considered.

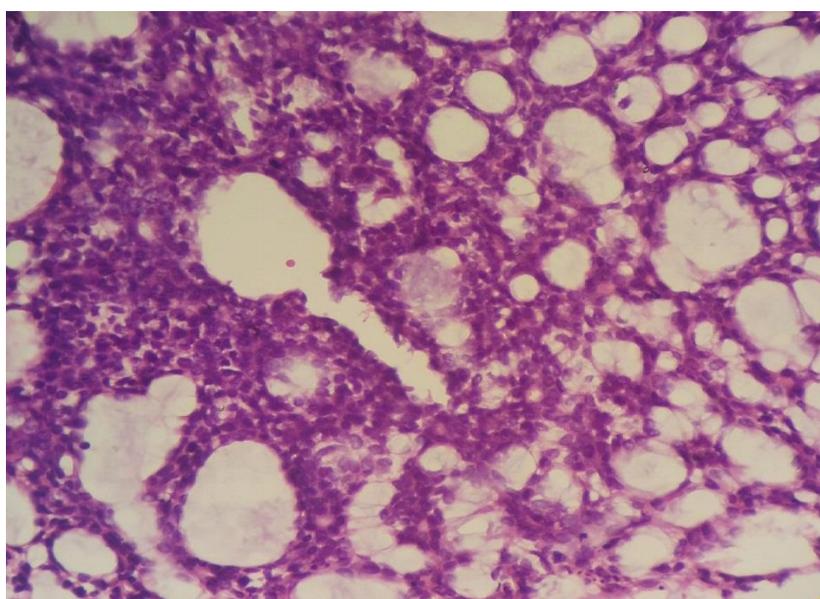
X-ray PNS showed DNS to the right and haziness of left maxillary sinus without any erosion of the wall. CT of Paranasal sinus showed erosion of the inferior sphenoid wall with extension into sphenoid sinus. It showed no vascular invasion and no intracranial extension. Sonography of the abdomen and X-ray chest revealed no metastasis. Excision biopsy was done and sent for histopathological examination. Grossly multiple grey white soft tissue fragments and a cartilaginous fragment were received. Histologically, the tumor cells were predominantly arranged in cribriform pattern and also in tubular and anastomosing trabecular pattern (Figure 1) Tumor cells were round to oval, having hyperchromatic nuclei, scant cytoplasm

and indistinct cell borders (Figure 2). The microcystic spaces of cribriform pattern were filled with eosinophilic hyaline material. Tumor cells were seen infiltrating the adjacent mucosal glands and interstitium.

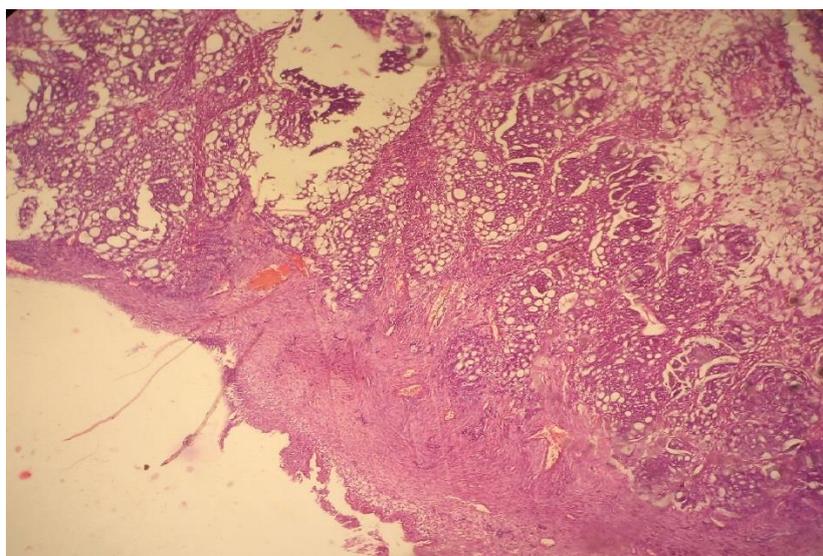
Perineural invasion, cartilaginous destruction and vascular emboli were also noted. (Figure 3). Finally the diagnosis was made as adenoid cystic carcinoma with perineural invasion.



**Fig 1: Tumor cells arranged in cribriform pattern (H&E, 4X)**



**Fig 2: Individual cells are round to oval, having hyper chromatic nuclei and forms mucin filled microcystic spaces. (H&E, 40X)**



**Fig 3: Perineural invasion by the tumor cells (H&E, 10X)**

#### **DISCUSSION**

Adenoid cystic carcinoma is most common tumor arising from major and minor salivary glands. The paranasal sinus is the most common site of minor salivary gland involvement [2, 4]. Of all malignant paranasal sinus tumors, 5 to 15% are adenoid cystic carcinomas which are considered to be very rare [4]. Lack of publications on endonasal removal of an ACC of paranasal sinuses and its prognosis matters a thorough discussion on this topic [5]. The peak incidence of ACC is found to be in fourth to sixth decades occurring slightly more in females [5]. When located outside the salivary glands, ACC can present as nasal congestion or obstruction, facial pain and swelling, and sinus pressure. This contrasts the presentation of ACC in the salivary glands, which presents most commonly as a painless mass or swelling of the parotid, submandibular, or sublingual glands with or without involvement of the facial nerve [6, 7].

It is an aggressive neoplasm with slow growth rate, resulting in high incidence of both local recurrence and distant metastasis [6]. Three histologic growth patterns have been identified and described: solid, cribriform, and tubular. Cribriform is the most common histologic subtype and similar predominant pattern was observed in our case. Most patterns are often mixed, and the solid component of the tumor correlates with aggressive behaviour. Assessment of the histologic grade is of significance in predicting the likelihood of tumor recurrence and survival [8]. In a series of studies, 5-year recurrence rates of 100%, 89%, and 59% were reported for tumors with solid, cribriform, or tubular growth patterns, respectively [8, 9]. Similarly, the

presence of greater than 30% solid growth has been reported to have a significantly poor survival (5%) when compared to tumors with a predominant cribriform (26%) or tubular (39%) growth pattern.<sup>9</sup> Also comparison with cribriform and tubular forms, predominant solid-type ACCs have been associated with higher rates of perineural invasion, higher S-phase fractions, and a higher incidence of aneuploidy [10]. Adenoid cystic carcinoma has the tendency to spread hematogenously and perineurally but not lymphatically. Metastasis rates vary considerably in the literature and the main metastatic sites are lung and bone [11].

Immunohistochemical staining for biomarkers in ACC can augment current information regarding prognosis. Since c-KIT mutation expression in ACC can range anywhere from 78% to 100%, there has recently been an increasing intrigue in determining the efficacy of c-KIT inhibitors in treatment of ACC [12]. Histological studies have shown that overexpression of c-KIT mutations in tubular and a solid ACC subtype confers a poorer prognosis [13].

ACC of the nasal cavity as a rare clinical entity continues to pose diagnostic and therapeutic challenges to physicians. This rare pathology presents in a nondescript manner, as facial pain and swelling or sinus congestion and pressure, and early diagnosis requires vigilance for suspicion of this disease and close follow-up [12]. Research on the possible role of chemotherapy and c-KIT inhibitors may help in improving treatment modalities of patients with ACC [13].

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