

CASE REPORT

**Adenoid Cystic Carcinoma of Nasal Cavity in an Adolescent Male-
A Rare Presentation**

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Abstract

Adenoid Cystic Carcinoma (ACC) is uncommonly found outside the major or minor salivary glands and is especially rare when located in nasal cavity. A case report of 13 year old boy who presented with mass in nasal cavity and epistaxis is presented here. Endoscopic removal of mass was done and histopathology revealed ACC. ACCs are slow growing tumours with a propensity for frequent local recurrence and early perineural and haematogenous spread. Early diagnosis requires a high index of suspicion for this rare pathology.

Keywords: Adenoid Cystic Carcinoma, Nasal Cavity, Histopathology, Perineural

Introduction

Adenoid Cystic Carcinoma (ACC) accounts for approximately 10% of salivary gland tumours. It is the most common malignant tumour of the submandibular gland and minor salivary glands however, rare in nasal cavity and paranasal sinuses accounting for less than <5% cases [1, 2]. The age of the patients ranges from 20 to 84 years with a median age of 52 years [1]. It is common in females compared to males with a ratio of 2:1 [3]. The most common site affected is the maxillary sinus followed by nasal cavity [4]. Tumours of the sinonasal tract commonly present with symptoms that are identical to those caused by inflammatory sinus disease, such as nasal obstruction, nasal

discharge, epistaxis, headache, facial pain and cheek swelling [3]. Histologically, growth patterns are characterized as cribriform, tubular and solid. As the cribriform pattern of the tumour forms cylindrical accumulations of basal lumina, glycosaminoglycans and stroma, the term cylindroma had been applied in the past [5].

Although slow growing, these tumours have a propensity for frequent local recurrence and early perineural and haematogenous spread to lungs [6]. Tumours showing perineural invasion, cervical lymph node metastasis, solid histological features and distant metastasis are associated with increased treatment failures and recurrences [3]. Until today, most sources agree that aggressive treatment of these tumours is necessary. The mainstay of treatment of ACC is surgery; adjuvant radiation therapy is reserved in case of positive margins or advanced stage [3,4].

Case Report:

Here we present a case of 13 year old Indian boy with mass in the nasal cavity and epistaxis. The naso-endoscope examinations revealed a mass in the posterior right nasal cavity which was friable and bleeding easily. Clinically, possibility of juvenile nasopharyngeal angiofibroma was given. Computed Tomography (CT) scan revealed

heterogeneously enhancing lobulated mass lesion in the nasopharynx/posterior nasal cavity (Fig. 1). Thin bony septae were seen within it. Anteriorly, the mass was extending into bilateral posterior part of nasal cavity mainly into the right side, through posterior choanae and causing lateral bowing of medial wall of the right maxillary sinus and left deviation of nasal septum. Destruction of floor of sphenoid sinus was noted superiorly. The mass measured 5x4x3.5cm. There were no neck nodes or distant metastases. Neoplastic lesion in nasopharyngeal region was suggested.

The patient then underwent endoscopic removal of mass and sent for histopathological examination. On gross examination, multiple grey brown to grey tan soft to firm tissue pieces were noted altogether measuring 5x4x2 cm with two turbinates measuring 3x0.6 cm each. Microscopy revealed presence of a malignant tumour predominantly arranged in cribriform pattern comprising of islands of small uniform basaloid tumour cells with presence of microcystic spaces filled with Basophilic mucoid material (Fig. 2,3). Periodic Acid Schiff (PAS) stain was positive in mucoid luminal material. Focally, tubular pattern was also seen showing smaller duct like arrays of basaloid epithelial cells surrounding a central lumen like space. Surrounding stroma was hyalinised and showed mild chronic inflammatory cell infiltrate. Perineural invasion noted (Fig. 4). Rare mitosis was seen. Bony spicules and turbinates were free of tumour. The final diagnosis of Adenoid cystic carcinoma was made. The patient was referred to higher centre for postoperative radiation therapy.



Fig.1: Computed Tomography (CT) Scan Image shows Heterogeneously Enhancing Lobulated Mass Lesion in the Nasopharynx/ Posterior Nasal Cavity Extending Superiorly into Sphenoid Sinus

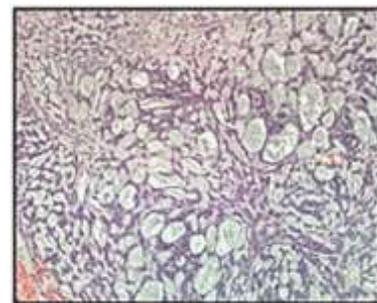


Fig.2: Photomicrograph in Low Power showing Cribriform Pattern of Adenoid Cystic Carcinoma (H & E X 100)

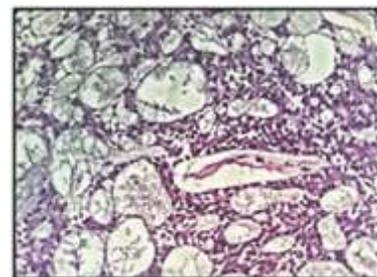


Fig.3: Photomicrograph in High Power showing Cribriform Pattern Comprising of Islands of Small Uniform Basaloid Tumour Cells with Presence of Microcystic Spaces (H & E X 400)

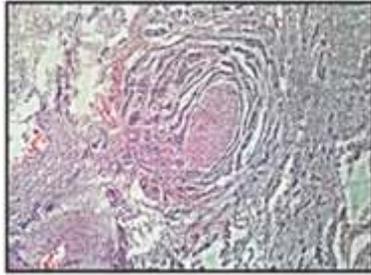


Fig. 4: Photomicrograph showing Perineural Invasion – A Characteristic Feature of ACC(H & E X 100)

Discussion:

ACC was first described by Billroth in 1856 [5]. ACC is a rare tumour of epithelial cell origin comprising 3 to 5% of all head and neck malignancies [7]. They usually arise in the major and minor salivary glands, but can occur in all sites comprising secretory glands (breast, cervix, colon, prostate) [2]. The peak incidence is from the fourth decade to the sixth decade, although cases have been reported in between ages of 11 to 90 years [3,6]. The maxillary sinus is the most common site of these tumours followed by the nasal cavity, as reported previously [8]. Common symptoms of sinonasal ACC include nasal obstruction, facial pain, epistaxis, nasal discharge, headache and loss of smell, the same symptoms observed in patients with sinusitis and inflammatory nasal conditions. This can cause a delay in diagnosis and treatment [3,8].

ACC can be composed of ≥ 1 histopathologic subtype(s), including tubular, cribriform, and

solid. Some studies have emphasized that histologic pattern correlates with prognosis (tubular = best, cribriform = intermediate and solid = worst) [1]. Cribriform is known as the most common type, [8] which also was demonstrated in the current study.

ACC of the head and neck, and specifically of the nasal cavity and paranasal sinuses, poses numerous treatment challenges for many reasons: it has a high propensity for local invasion to adjacent structures, making resection more difficult; it is commonly diagnosed late due to its insidious growth; and in 50% of cases it has already exhibited perineural spread at the time of diagnosis [7] as noted in our case also.

There are studies which indicate that surgery followed by radiotherapy is the most common treatment for patients with sinonasal ACC, [5] which was the treatment given in our case. Overall 5-year survival rates for patients with sinonasal ACC from 50% to 86% have been reported [9,10]. ACC of the nasal cavity is a rare clinical entity posing diagnostic and therapeutic challenges. Therefore, any patient with suspicious malignancy needs to be investigated thoroughly by histological diagnosis. It exhibits extensive local tissue infiltration and perineural spread, which results in a high rate of recurrence despite aggressive surgical resection. High index of suspicion is required for early diagnosis and optimal management.

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