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Adenoid Cystic Carcinoma of Nasal Cavity; A Rare Case Report.

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Abstract: *Adenoid cystic carcinoma (ACC) is usually a slow growing but highly malignant neoplasm with a remarkable capacity for recurrence. It mainly affects salivary glands, palate and is rare in the nose and paranasal sinuses. A case report of 38 years old male with ACC of left nasal cavity who presented with gradually progressing left sided nasal obstruction is presented here. The CT scan showed mass in left nasal cavity eroding anterior part of medial wall of maxillary sinus. The tumor was excised with wide margin through a lateral rhinotomy approach under general anaesthesia. Histopathological report confirmed it to be a case of adenoid cystic carcinoma. The sinonasal ACC has an overall poor prognosis due to perineural invasion and post-operative radiotherapy may improve the prognosis.*

Keywords: *adenoid, carcinoma, cavity, cystic, nasal*

I. Introduction

Adenoid cystic carcinoma (ACC) is one of the commonest malignant tumors involving salivary glands. It may also arise in lacrimal glands, nose and paranasal sinuses, lung, digestive tract, breast etc. Approximately 20% of all ACCs occur in sinonasal tract. [1] In sinonasal cavity, maxillary sinus is the most common site for ACC followed by nasal cavity. [1]ACC most commonly presents in 4^{th} to 6^{th} decades. This tumor has a marked propensity for perineural spread and bony invasion. Distant metastasis to lung is more common than regional lymph node metastasis.

Histopathologically there are three characteristic growth patterns of ACC i.e. cribriform, tubular and solid. These patterns may be present in variable combinations in an individual case. As the cribriform pattern of the tumor forms cylindrical accumulations of basal lumina, glycosaminoglycan and stroma, the term cylindroma had been applied to ACC in the past.

Here we have reviewed the clinical features, histopathological & immunohistochemical findings, treatment and prognostic factors in a patient of adenoid cystic carcinoma of nasal cavity presented in our institution.

II. Case Presentation

A 38years old male patient came in the ENT department with complaints of gradually progressing left sided nasal obstruction for last 5 years. There was no other significant comorbidities. There were no palpable neck lymph nodes. Anterior rhinoscopy showed a mass in the left nasal cavity. The CT scan of paranasal sinus showed mass in left nasal cavity eroding anterior part of medial wall of maxillary sinus. USG abdomen and X-ray chest revealed no metastasis. An elective radical excision via left lateral rhinotomy approach was performed under general anaesthesia and the mass was sent to pathology department of our institution for further evaluation.

Grossly the mass was measuring $4\text{cm} \times 3\text{cm} \times 1.5\text{cm}$, greyish white, fleshy & soft to firm in consistency.Histopathological report revealed it to be a case of adenoid cystic carcinoma of nasal cavity. Sections showed tumor cells arranged in cribriform and focal solid pattern (Fig1). Individual tumor cells are small bland deeply basophilic cells with hyperchromatic ovoid nuclei and scanty cytoplasm. Tumor cells formed pseudoglandular or pseudocystic spaces containing basophilic mucinous material (Fig 2)& focally showed perineural invasion (Fig 3). Mitotic figures were very few. Immunohistochemical staining for S 100 showed positivity in tumor cells (Fig 4).

Presently the patient is undergoing radiation therapy and is asymptomatic now.

III. Figures

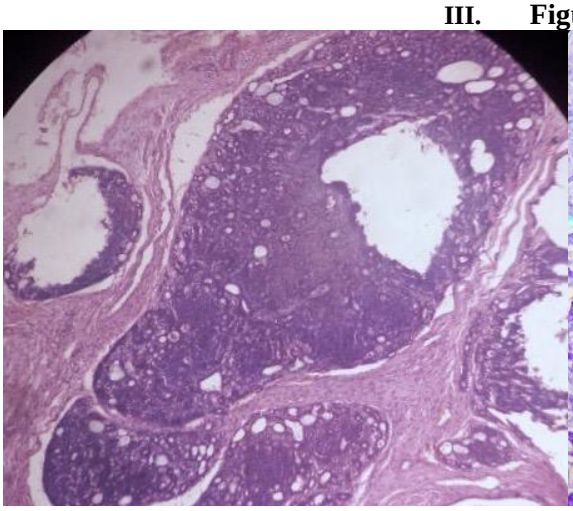

Fig 1: Focal solid and cribriform pattern showing pseudocysts surrounded by basaloid cells. (H&E; 10X).

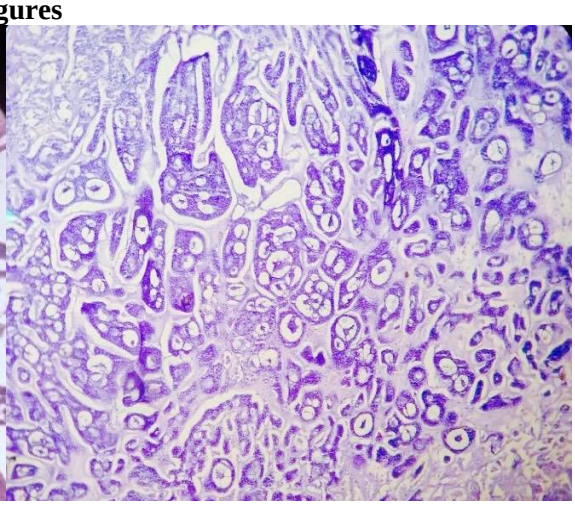

Fig 2: Cribriform pattern showing pseudocysts, surrounded by basaloid cells with hyperchromaticangulated nuclei. (H&E; 20X).

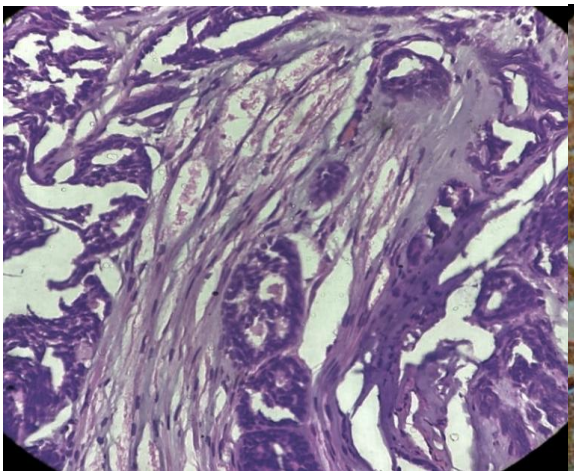

Fig 3: Tumor cells showing focal solid pattern and focal perineural invasion. (H&E; 40X).

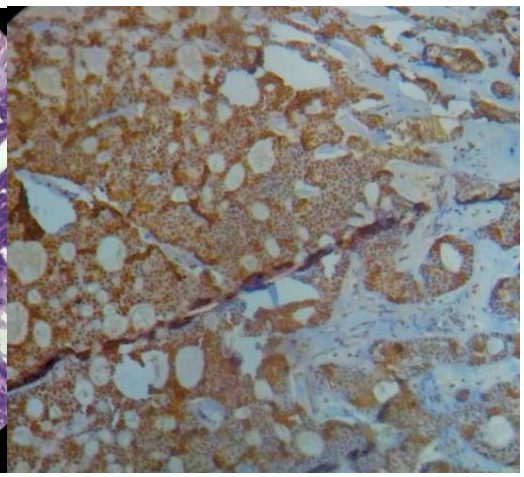

Fig 4: IHC staining for S100 showing positivity as a marker for myoepithelial differentiation. (40X).

IV. Discussion

Adenoid cystic carcinoma (ACC) accounts for 3 to 5% of all head and neck malignancies.[2] This is an invasive neoplasm composed predominantly of basaloid cells with myoepithelial/basal cell differentiation, accompanied by interspersed ductal structures. A distinctive chromosomal translocation t(6;9)(q22-23;p23-24) , which results in MYB-NFIB gene fusion , is found in adenoid cystic carcinomas.[3] They most commonly present as a slow growing mass in the sinonasal tract.

Immunohistochemical staining shows that tumor cells with differentiation express S100 and p63.[4] The interspersed ductal epithelial cells express CK7, CD117. The prognosis mostly depends on grading of tumor, which is based on proportions of the three histological growth patterns in the tumor.[5] The solid pattern have been associated with large tumor size, frequent early recurrence rates, low long time survival rate and higher distant metastasis rates.[6] Also in sinonasal tract ACCs of maxillary region have the worst prognosis.

Radical excision of the tumor followed by radiotherapy is most practiced treatment for sinonasal ACCs. [7] Irrespective of surgery and radiotherapy, the overall recurrence rate of sinonasal ACC is high. Although the tumor is generally indolent, the long term prognosis is poor. The 5 years survival is about 60% to 75%, but the 10 year survival drops dismally to 30% to 54%[8]The recurrencesand decrease in survival rates are due to intraneural, perineural infiltration and positive infiltration of margins while surgery.

V. Conclusion

Adenoid cystic carcinomas are slowly growing, rare malignant tumors, which are easily amenable to surgical management. Identification of the predominant histopathological growth pattern is very important for grading of the tumor. ACCs must be differentiated from basal cell adenoma/adenocarcinoma and epithelial/ myoepithelial carcinoma. Despite its rarity in the nasal cavity, adenoid cystic carcinoma should be taken into consideration in the differential diagnosis when a patient presents with slowly growing painful swelling in and around the nose. As they can mimic inflammatory process in sinonasal cavity, early diagnosis and management requires high index of suspicion.

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