Adenoid Cystic Carcinoma of Nasal Cavity: A Rare Presentation Case Report

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Abstract- Adenoid cystic carcinoma (ACC) is one of the commonest malignant salivary gland tumors, affecting minor salivary glands and rare in the nose and paranasal sinuses. ACC in maxillary sinus usually mimic inflammatory disease and has a poor prognosis. A case report of 37 yr old male with ACC of left nasal cavity who presented with mass in left nasal cavity and swelling in left cheek is presented here. The CT scan showed mass in left nasal cavity and over frontal process of maxilla with bone resorption. Surgical excision was done by Lateral Rhinotomy approach and biopsy revealed ACC. Post op radiotherapy given and regular follow up done. The sinonasal ACC has an overall poor prognosis due to perineural invasion and post-operative radiotherapy is important to address the microscopic disease.

Index Terms- Adenoid cystic carcinoma, Nasal cavity, Paranasal sinuses

I. INTRODUCTION

Adenoid cystic carcinoma (ACC) is a malignant tumor of head and neck region, including trachea, bronchus, lung and mammary gland. It is an epithelial non epidermoid tumor and occurrences in the head and neck region are relatively rare. They arise most commonly in the major salivary gland, oral cavity, and pharynx and paranasal sinus. Clinical features of ACC are different from those of squamous cell carcinoma and thus we must be aware of its characteristics for the diagnosis and treatment.

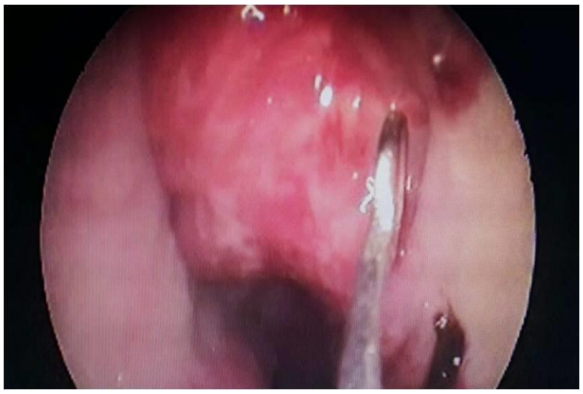
Adenoid cystic carcinoma most commonly involves the salivary gland tissue and is the second most common tumor of nasal cavity and paranasal sinuses. Sinonasal ACC accounts for 10-25% of all head and neck ACC. Sinonasal tumors are often asymptomatic and mimic inflammatory disorders leading to delay in diagnosis. Many patients present with advanced-stage disease and extensive involvement of surrounding structures including the dura, brain, orbits, carotids, and cranial nerves. In addition, ACC has a propensity for perineural spread and bony invasion, which can lead to significant skull base involvement and intracranial extension. These findings make treating sinonasal ACC challenging and potentially morbid.

ACC was first described by Billroth in 1856. Growth patterns are characterized as cribriform, tubular and solid. As the cribriform pattern of the tumor forms cylindrical accumulations of basal lumina, glycosaminoglycans and stroma, the term cylindroma had been applied in the past. We have reviewed the clinical features, treatments and prognostic factors in a patient who had undergone the primary treatment for ACC in our institution.

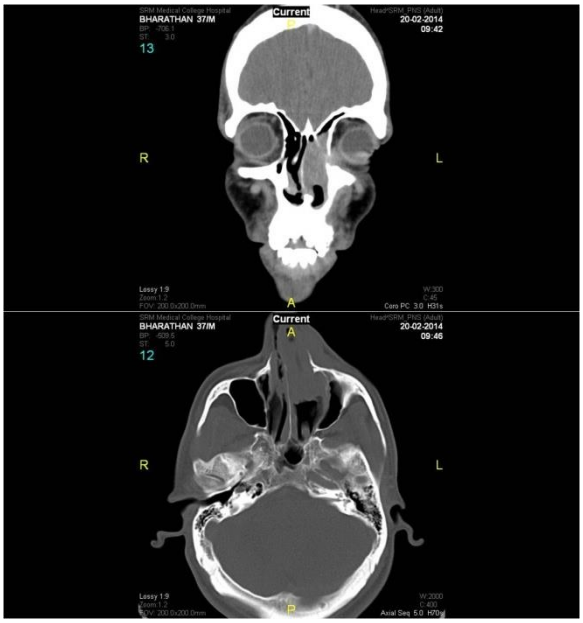
II. CASE PRESENTATION

A 37 year old male patient presented with complaints of left sided nasal block and left sided blood stained nasal discharge for the past 6 months. He also complained of swelling over left side of cheek along with headache and left sided facial pain for 6 months. Patient had similar complaints in the past for which he was twice operated 2 years ago. There were no associated co-morbidities. On local examination, a mass over left maxilla was palpated which was hard and tender measuring about 4x3cms with superior extension up to infraorbital margin. Anterior rhinoscopy showed a mass in the left nasal cavity, which appeared congested. No other extension was noted and no neck nodes were palpable.

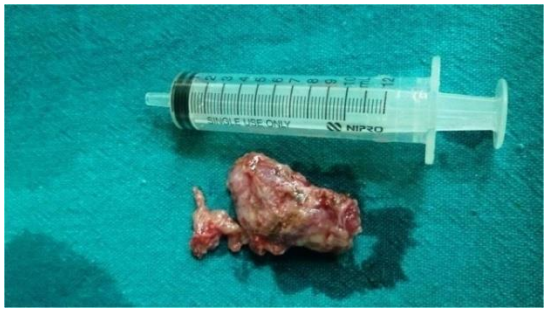
On Diagnostic Nasal Endoscopy (DNE), mass was seen between left inferior turbinate and the septum involving the lateral wall of nose. The mass was firm, appeared congested and was bleeding minimally on touch.



CT Paranasal sinus revealed a unilateral soft tissue mass involving left nasal cavity causing expansion of left nasal cavity and thinning of neighboring bone, with extension of mass into medial part of anterior wall of left maxilla with associated bone resorption.



Patient was taken up for elective surgery under general anaesthesia. Tumour was approached by left Lateral Rhinotomy (Moure's incision). The mass was delineated completely. The mass was found involving the lamina papyracea, frontal process of maxilla, medial part of anterior wall of left maxilla below the infraorbital rim. The orbit was lifted and the anterior ethmoid artery was ligated. Periorbita was found intact. The whole mass was removed in toto along with lamina papyracea, eroded frontal process of maxilla, eroded anterior wall of maxilla and sent for histopathological examination (HPE). After removal of the mass, the bony walls were drilled thoroughly.



HPE reported the specimen as Adenoid Cystic Carcinoma. The histological examination revealed malignant neoplasm in cribriform, tubular and focally solid pattern in myxoid and hyalinised stroma. The tumor cells showed hyperchromatic ovoid nuclei and scanty cytoplasm with peripheral pallisading. Spaces in the tubules contained basophilic mucinous material. The tumor was infiltrating into the bone, surrounding adipose tissue, skeletal muscle and focally showed perineural invasion. Rare mitosis was seen. The tumor was staged as a Stage IV tumor (T4N0M0).

The patient was sent for Radiation therapy following surgery after 3 weeks. A total of 30 cycles (60Gy) was given over 6 weeks. After completion of radiotherapy he is on regular follow-up at monthly intervals. Patient is asymptomatic and post operative DNE shows nasal cavity free from tumor. There were no neck nodes palpable.

III. DISCUSSION

ACC is a minor salivary gland tumor that can arise in nasal cavity and paranasal sinuses and the second most common malignancy next to squamous cell carcinoma.[1, 2] Maxillary sinus is most common site for ACC followed by nasal cavity.[1, 3, 4] There are studies which indicate that surgery followed by radiotherapy is the most common treatment for patients with sinonasal ACC, which was the treatment given in our case. [5, 6] Though ACC is considered a radiosensitive tumor, it should be always combined with surgery because radiotherapy can give adequate clearance of positive margins left behind after surgery.[8] The recurrence rate after surgery is due to perineural invasion. Overall 5-year survival rates for patients with sinonasal ACC from 50% to 86% have been reported.[1, 4, 9]

Spiro et al observed patients with ACC of maxillary sinus had advanced disease due to tissue and bony invasion.[9] The cause of pain is due to neoplastic cell neurotropism and this was a significant complaint in our patient also.[10] Reccurence and decrease in survival rate is due to intraneural, perineural infiltration and positive margins.[11] Solid variant shows greater aggressiveness.[12] The Prognosis of ACC tumor is influenced by its location with tumors of maxillary region having the worst prognosis.[11]

Recently, numerous studies have shown benefit with locoregional control and long survival for ACC patients when surgery and radiotherapy are combined, especially for those with unfavorable prognosticators such as advanced lesion.[13, 14] Therefore, some authors postulate that postoperative radiation likely delays rather than prevents recurrence. [9, 15] The rate of recurrence for solid tumors is 100% and low grade variants are 59-89%. Irrespective of surgery and radiotherapy, the overall recurrence rate of sinonasal ACC is 65%.The distant metastasis is usually seen in lung and bone, which is around 35-50%. The 10 year survival rate is usually less than 20%.

CONCLUSION

Thus ACC is a rare tumor and most patients present in a late stage. The prognostic factors are based on site, stage, histopathological subtype of the tumor. Poor prognostic indicators are solid type, grading, skull base and neural invasion. High index of suspicion is required for early diagnosis and optimal management.

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