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ADENOID CYSTIC CARCINOMA OF NASAL CAVITY: A CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT Adenoid cystic carcinoma (ACC) is relatively uncommon tumour, accounting for less than 1% of all head and neck malignancies. It is an asymptomatic malignant salivary gland tumour, characterized by wide local infiltration, local reoccurrence, perineural spread and rarely leads to distant metastasis. In this paper the authors present a case of adenoid cystic carcinoma affecting the nasal cavity with some intra-orbital and paranasal sinus extension in a 40-year-old female patient along with brief review of literature.

KEYWORDS : Adenoid cystic carcinoma, salivary gland tumour, perineural invasion

INTRODUCTION:

Adenoid cystic carcinoma can affect any salivary glands but more commonly it involves the minor salivary glands of the palate. The tumour is most often clinically deceptive due to its small size and slow growth which actually overlies its extensive subclinical invasion and marked ability for early metastasis making prognosis unfavorable. Clinical features of ACC are different from those of squamous cell carcinoma and thus we must be aware of its characteristics for diagnosis and treatment.

In the major salivary glands, ACC most commonly affect parotid and submandibular glands, while in the minor salivary glands, palate is the most common site involved; other sites are floor of mouth, tongue and lip.

Adenoid cystic carcinoma is the most common tumour of the sinonasal tract and accounts for 10-25% of all head and neck ACC.[1] 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. It was described as “one of the biologically destructive and unpredictable tumors of the head and neck” by Conley and dingman.

CASE PRESENTATION:

A 40 year old female patient presented with complaints of mass in left nasal cavity with difficulty in breathing from left side of nostril and swelling over left side of nose from last two years. She also complained of swelling over left side of cheek. The patient did not have any significant medical history and there was no history of fever and unexplained weight loss since the observation of the swelling by the patient.

There were no associated co-morbidities. On local examination, a mass over left side of dorsum of nose was palpated which was hard and non-tender measuring about 3x3cms with superior extension up to infraorbital margin (Figure 1a and 1b). 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 investigations complete haemogram and biochemical profile of the patient was found to be within normal limits.

CT Paranasal sinus revealed a expansile mass involving the anterior nasal cavity on left side causing its expansion along with expansion of left nasal bone and thinning of bony trabeculae with remodeling and pressure erosion with extension into prenasal soft tissue with erosion of left lamina papyracea with minimal intraorbital and maxillary sinus extension on left side (Figure 2a and 2b).

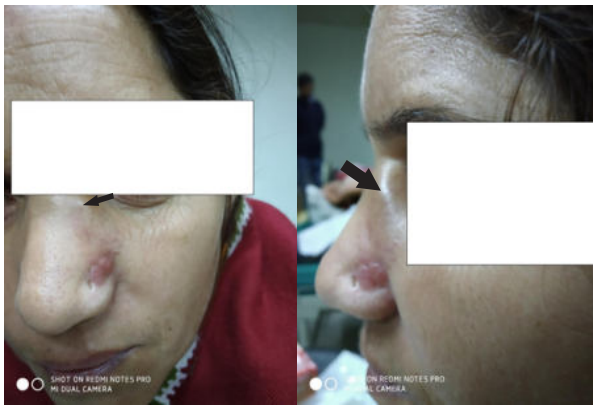
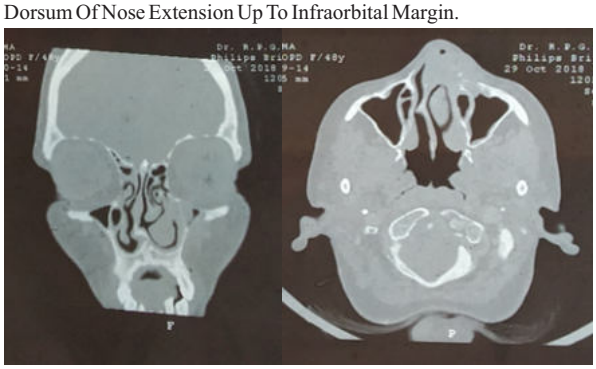


Figure 1a and 1b Showing Swelling (arrow) Over Left Side Of Dorsum Of Nose Extension Up To Infraorbital Margin.



(Figure 2a and 2b)
CT scan showing mass in left side anterior nasal cavity with some left side intraorbital and maxillary sinus extension.

Patient was taken up for elective surgery under general anesthesia. Tumour was approached by left Lateral Rhinotomy. The mass was delineated completely. The whole mass was removed in-toto and sent for histopathological examination (HPE). After removal of the mass, the bony walls were drilled thoroughly.

Histologically, excision biopsy showed the features of adenoid cystic carcinoma, with focal perineural invasion. There was no lymphovascular invasion.

The patient was sent for Radiation therapy following surgery after 3 weeks. She was recalled at every 6-month interval in view of the sinus involvement and propensity for distant metastasis.

DISCUSSION:

ACC is a relatively rare malignant salivary gland tumor comprising less than 1% of all malignancies of head and neck and was first described by Billroth in 1856.[1] It is most common 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] ACC may occur at any age although in most cases the patients age ranged from 24 to 78 years and have female predilection (F:M − 1.2:1).[3] Tumors involving the nose, paranasal sinuses and maxillary sinus have the worst prognosis as they are usually detected with higher stages at the time of diagnosis.[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.[7]

The cause of pain is due to neoplastic cell neurotropism and this was a significant complaint in our patient also.[8] Recurrence after surgery and decrease in survival rate is due to intraneural, perineural infiltration and positive margins.[9]

Three recognized histological patterns of ACC are cribriform, tubular and solid although, cribriform is the most commonest and solid is the least common. 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. Solid variant shows greater aggressiveness.[10]

The presenting case did not show any metastasis although literature reveals 25%-50% incidence of distant metastasis and lung is the most common involved site.[11]

CONCLUSION:

ACC is rather an uncommon salivary gland malignancy. It is unique for its peculiar histopathological features and tendency for perineural invasion. Poor prognostic indicators are solid histological subtype, high grading, skull base, and neural invasion. Lesions involving the sinus tend to have a poor prognosis due to its infiltrative growth and distant metastasis. High index of suspicion is required for early diagnosis and optimal management.

Declaration of Patient Consent:

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/ their consent for his/ her/ their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflict of interest: Nil

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