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ADENOID CYSTIC CARCINOMA: CASE REPORT

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Abstract
Of all the tissues in the human body, perhaps the salivary glands have the most histologically heterogeneous group of tumours and the greatest diversity of morphologic features among their cells and tissue**.** Adenoid cystic carcinoma is a clinically and pathologically well-defined entity that has been described extensively in literature so far. It occurs primarily in minor salivary glands and relatively frequently in oral major salivary glands .The report of such a case in oral cavity is discussed here.

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Keywords: Adenoid cystic Carcinoma, Swiss- Cheese, minor salivary gland tumour, perineural invasion, hard palate

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INTRODUCTION

Adenoid cystic carcinoma is reported to be the common and best recognized epithelial salivary gland neoplasms. Historically, in 1859 Billroth was first to report this neoplasm and coined it 'cylindroma'.[1] Later in 1954, Ewing labeled it with the term 'adenoid cystic carcinoma 'on the basis of its histological appearance. It is a relentless tumor that is prone to local recurrence and eventual distant metastases [2] It is clinically innocuous by virtue of its small size and slow growth potential which depicts its extensive subclinical invasion and marked ability for early metastasis, the factors which make the prognosis of the neoplasm questionable. [3].Among all adenoid cystic carcinomas, 50% occur in intraoral sites, with definite predilection for the posterolateral portion of the hard palate[2]Approximately 31% of lesions affect minor salivary glands, particularly the palate, though they can also be observed in the submandibular and parotid glands[4]. 19.8% tumours occur in tongue as well of which about 85% of cases showed specificity to base of the tongue.[17] It is usually non-capsulated, slow growing lesion which metastasizes late and has a liking to invade the surrounding perineural spaces.[5]It accounts for about 5% to 10% of all salivary gland neoplasms, representing 2% to 4% of malignant occurrences of the head and neck area. The local recurrence rates vary widely from 16 to 67% and the incidence of distant metastases increases with time.Here, we report a case of Adenoid cystic carcinoma in the right posterolateral portion of the hard palate, in a 30 year old woman.

Case Report: A 30 yearold female patient visited the Department of Oral Medicine and Radiology, Sharad Pawar Dental College and Hospital, Sawangi, Wardha, with the chief complaint of swelling on the left posterolateral portion of hard palate since 2 months. The history revealed that swelling had started insidiously and has gradually increased to its present size since its onset. The swelling was associated with dull and continuous pain one month ago and no associated signs of pus discharge and bleeding were visible. Medical, dental, family and personal histories were not contributory. Patient was conscious, co-operative and well oriented to time, place and person. All vital signs were found to be within normal limits whereas extraoral examination revealed no abnormal findings.

Intraoral examination revealed a solitary, well defined swelling extending anteroposteriorly from 26 to 28 and mediolaterally from marginal gingiva of 26

to midpalatine raphe. The swelling was of size 3x5cm approximately, surface of which was smooth and colour was same as that of adjacent mucosa. On palpation all the inspectory findings were confirmed. The swelling was tender and firm in consistency. No regional lymphadenopathy was found. (FIG-1)

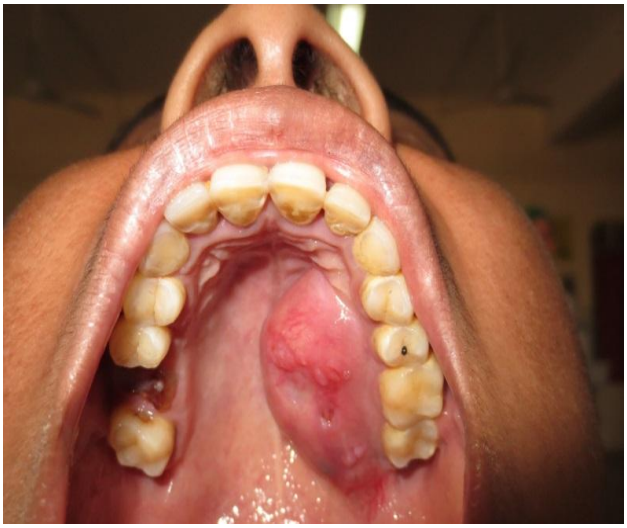

FIG-1: intraoral clinical photograph showing lesion on the hard palate

Clinical differential diagnosis included a benign or a low grade malignant neoplasm of minor salivary glands, reactive/inflammatory condition of minor salivary glands, a malignant growth of the maxillary sinus, benign mesenchymal neoplasm and much less likely a slow-growing malignant mesenchymal neoplasm.

Intraoral periapical and Orthopantomograph revealed a single well defined radiolucency with corticated border extending from mesial of 25 to mesial of 27 involving the floor of maxillary sinus of the same side whereas contralateral sides when compared appeared to be normal. (FIG-2)

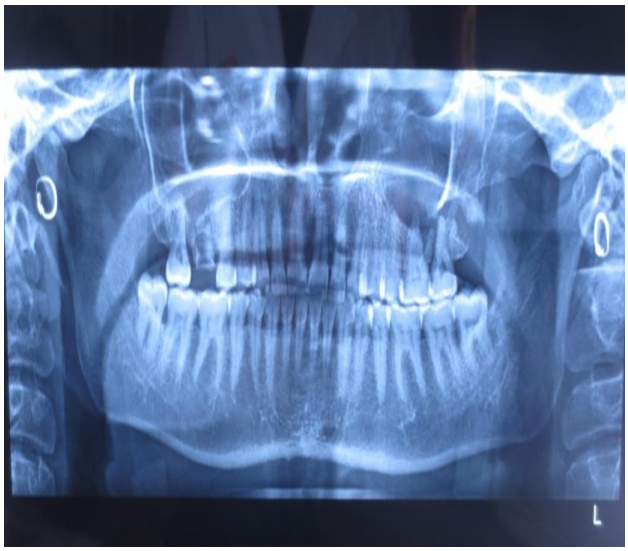

FIG- 2: Orthopantomograph of the case

PNS skull shows haziness in the left border of maxillary sinus. (FIG-3)

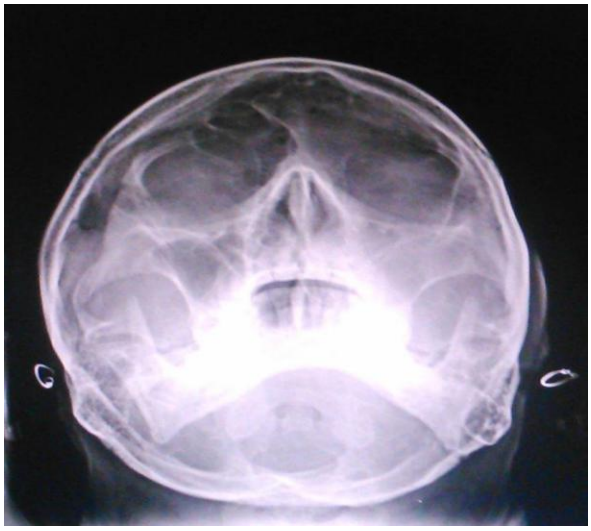

FIG-3: Posteroanterior view of skull

Axial CT scan revealed that lesion involved the left maxillary sinus.

Radiographic findings were suggestive of malignancy in maxillary sinus .(FIG-4)

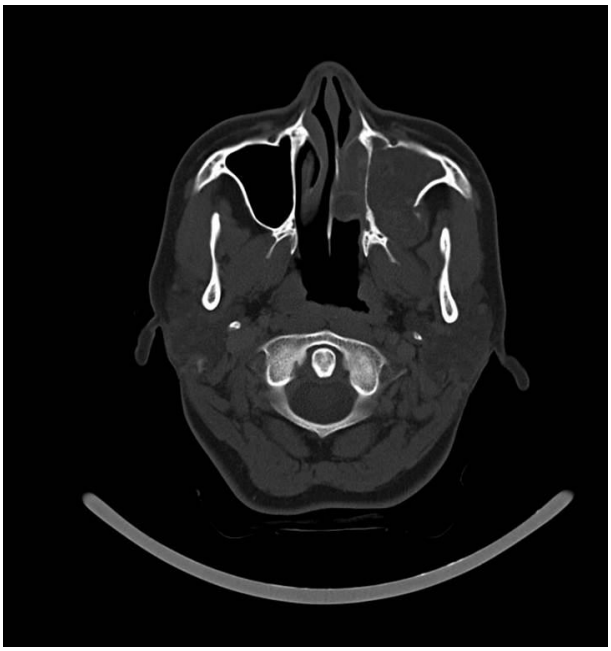

FIG-4: CT scan view of the lesion

The incisional biopsy was performed under local anaesthesia for histopathological diagnosis. The hematoxylin and eosin stained sections showed uniform cells arranged in cord like pattern, with deeply stained nuclei and with round to oval pseudocysts, containing pale pink granulofibrillar material, giving the entire structure a typical "swiss-cheese" appearance. Perineural invasion was also seen at some places. The histopathological impression was that of an adenoid cystic carcinoma of cribriform pattern.(FIG-6,7,8)

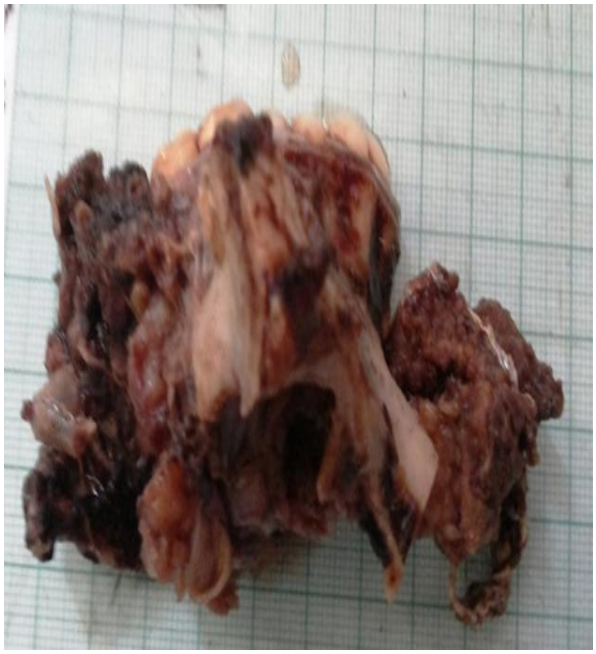

FIG -5: Gross Specimen of the lesion

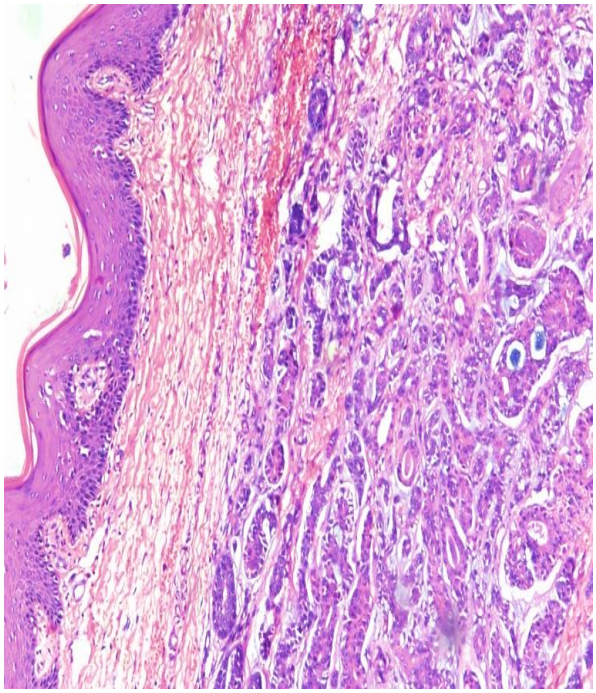

FIG-6: H& E stained section under low power view showing epithelium and glandular connective tissue

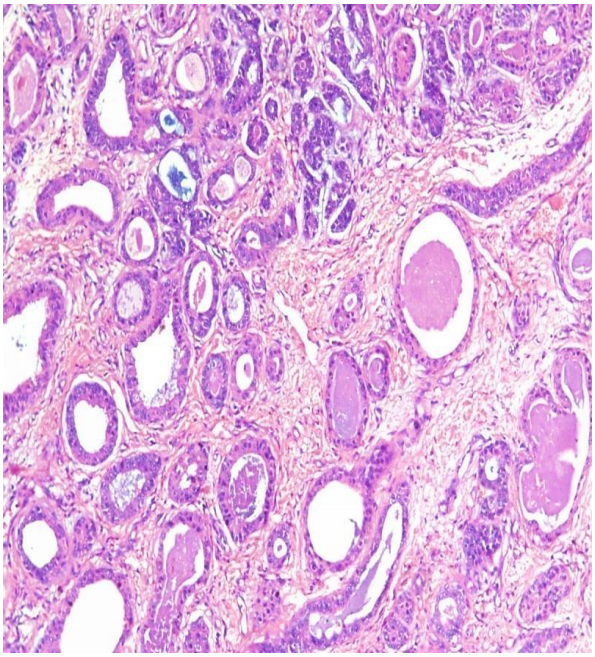

FIG-7:H&E stained section under low power view showing cribriform and swiss cheese pattern of the salivary gland acini

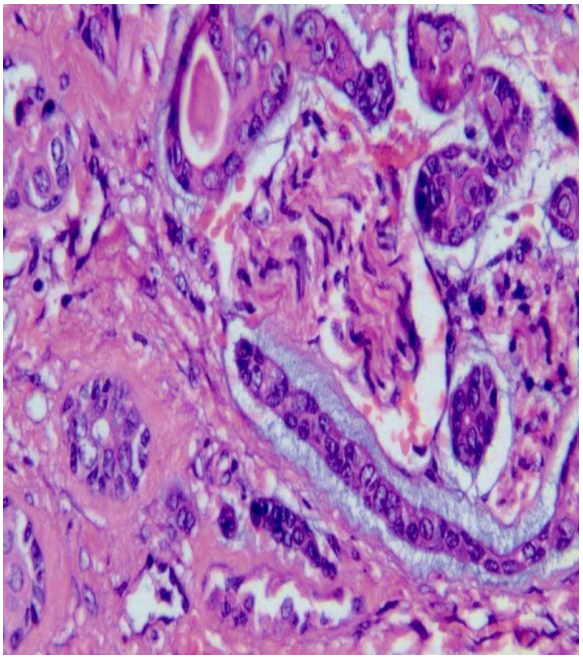

FIG-8: H&E stained section showing perineural invasion.

The patient then underwent wide local Selective neck dissection upto level III of left side along with hemi- maxillectomy of the same side followed by reconstruction with temporalis flap. The entire specimen was sent to department of Oral Pathology and Microbiology for histopathological diagnosis. The gross specimen was of size 7x8 cm. The part of hard palate and zygoma was also received. Histopathological findings were same as that of incisonal biopsy.

Adjuvant radiotherapy was planned for the patient after the diagnosis.(FIG-5) No local recurrence has been observed in 6 months and the patient is on regular follow up .

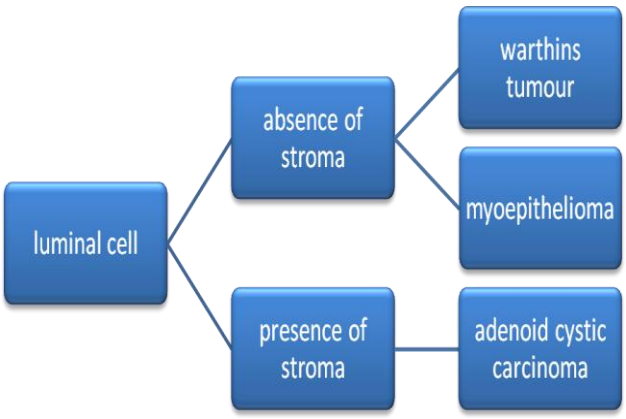

FIG-9: Histogenetic concept of adenoid cystic carcinoma in particular

Discussion:

The reported incidence is approximately 3% of all head and neck neoplasms [6].Tumours of minor salivary gland origin account for only 10–15% of all salivary gland neoplasms[7].Adenoid cystic carcinoma

occurs on the palate with the frequency of 8-15% of all the palatal salivary neoplasm.

[1]The term 'adenoid cystic carcinoma' was introduced by Ewing (Foote and Frazell) in 1941. This tumor was named as 'cylindroma' earlier by Billroth in 1859[8] because the epithelial and connective tissue elements formed a system of intertwining cylinders..The term 'basalioma' was coined by Krompecher in 1908[8]Who considered this type of tumor to be of analogous nature to the basal cell growths of the skin. Females are more commonly affected than males .It can occur at any site but in oral cavity minor salivary glands are more commonly affected than major salivary glands . It can also occur in parotid and submandibular gland as well .Perineural spread of ACC has long been recognized however it is not pathognomonic of the lesion.[9] Pain is usually a constant finding in the course of the disease and some of the cases affecting parotid gland may show signs of facial paralysis. In a study conducted by Buchner A et al, the relative frequency of intraoral minor salivary gland tumors was 0.4%, among these 41% were malignant. Among these malignant neoplasms, the most common was mucoepidermoid carcinoma (21.8%) followed by polymorphous low grade adenocarcinoma (7.1%), Adenoid cystic carcinoma was found to be third most common (6.3%)[10]

HISTOMORPHOLOGY AND GRADING OF TUMOUR –[11]Various growth patterns of adenoid cystic carcinoma have been recognised –

CRIBRIFORM VARIANT – Extensive sheets, uniform bands, or cribriform nests usually composed of relatively small, darkly stained, slightly separated basal/myoepithelial cells and small, at times inconspicuous ductlike structures, which may contain secretory products. Round to oval, often fairly uniformly sized intercellular spaces, termed

pseudocysts, containing pale grayish blue to pinkish granulofibrillar material at times with a reticular pattern, which develop in relation to the basal/myoepithelial cells.

TUBULAR VARIANT –Tubular pattern with bilayered ducts generally composed of tubular epithelial cells with moderate amount of eosinophilic cytoplasm .

Tubules are fairly placed with collagenous stroma producing architecture that can be characteristic .

SOLID VARIANT: Arranged as variable, at times fairly uniformly sized groups or as sheets of small, darkly stained tumor cells, those are excess proliferations of the basal/myoepithelial cell component. Small

duct like structures must be identifiable among the basaloid cells. Nests or sheets of basaloid cells with the above features from 30% or more of the neoplasm.

GRADING OF THE TUMOUR –[11]

a. GRADE I: The tumor consisting only of cribriform and tubular histomorphology.

b. GRADE II: A mixture of cribriform, tubular and solid growth patterns, with solid growth pattern less than 30% of the tumor.

c. GRADE III: Tumors with predominantly solid features (>30% or more of the tumor)

HISTOGENESIS OF SALIVARY GLAND NEOPLASMS- The concept of histogenesis lies in its pattern of differentiation based upon –[12]

- The organization of the tumour cells , their differentiation types,materials synthesized by cells and their proper placement within the tumour.

These factors are independent but highly intergrated and operate between various types of salivary gland tumours or within one type. Using the normal salivary gland ducto-acinar unit with its combination of duct luminal or acinar cells bordered externally by a row of myoepithelial and/or basal cells as a model, salivary gland tumors can be divided effectively into two broad categories: one a caricature of the normal gland with two basic cell types, that is, neoplastic luminal and myoepithelial or basal cells, and the other differentiating either neoplastic luminal cells or myoepithelial and/or basal cells. Primarily monocellular salivary gland tumors are composed either of luminal- type cells, with or withoutacinar differentiation, or of the neoplastic counterpart ofmyoepithelial and/or basal cells.In a nutshell potential interrelationship of salivary gland neoplasms and various taxonomic categories is described (FIG -9)

TREATMENT, PROGNOSIS AND SURVIVAL – Salivary gland ACC has indolent behaviour and is prone to have late recurrence. [13][14][15] The treatment requires the surgical excision of the lesion with wide margins. The tumour has propensity not only to invade perineurally but also has a tendency to spread [3]Grading of adenoid cystic carcinoma is suggested to have prognostic significance, but staging has been shown to be perhaps a more meaningful predictor of clinical outcome.

[16]Long term survival is particularly low in grade III tumors[11](Distant metastases occur in 25-50% of patients, even many years after the diagnosis, and lung is the most involved site.[16]

5 years survival rate after effective treatment is 75%, but long term survival rates are low (10 years – 20% and 15 years – 10%[3]

Postoperative radiotherapy, combined with more aggressive surgeries increases the long term survivals into the 30% to 40% range.Solid primary growth pattern is associated with worse prognosis. Clinical size greater than 4 cm indicates greater subclinical spread. Delayed diagnosis and delayed treatment also worsen the prognosis. Close or unclear surgical margins are indicators of the requirement of wider excision. Multiple local recurrences are also associated with grave prognosis.

CONCLUSION–Adenoid cystic carcinoma is an aggressive tumour occurring in salivary gland and quite innocuous in its presentation. Thus, meticulous observation and thorough examination helps us to reach the diagnosis of the disease entity.

CONFLICT OF INTEREST – None Declared

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