# Adenoid Cystic Carcinoma Arising From Minor Salivary Glands Of Palate Involving Maxillary Sinus

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**Abstracts**: Adenoid cystic carcinoma (ACC) was first described by Billroth in 1856 and was called 'cylindroma' due to its characteristic histological appearance. ACC is the most common malignant neoplasm of the lacrimal gland, and the second most common type of carcinoma arising in the salivary glands, following mucoepidermoid carcinoma. Palate is the most commonly affected site followed by parotid gland, submandibular gland, antrum & tongue. Characteristic features include aggressive, slow growth, with insidious destruction of surrounding tissues, perineural invasion, prolonged clinical course and the tendency for delayed onset of the distant metastases which worsens the prognosis. Long term survival can be achieved particularly with combined surgery and radiotherapy. The most common pattern is the cribriform architecture. Histopathology is the gold standard for the diagnosis of ACC. CT & MRI are considered to be of almost similar significance in detection of perineural spread with preference to MRI because of its high soft tissue contrast. Here we have mentioned a case of Adenoid Cystic carcinoma arising from minor salivary glands of palate. [Shah J et al NJIRM 2014; 5(2) :115-118]

**Key Words**: Adenoid cystic carcinoma, cylindroma, minor salivary gland tumor, Asymptomatic palatal growth, perineural spread.

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**Introduction** ACC (Adenoid cystic carcinoma) is an unusual malignant epithelial tumor which was first described by Billroth in 1856 and was called 'cylindroma' due to its characteristic histological appearance. This neoplasm shows an adenoid cystic histological pattern and may arise in a variety of anatomic sites like major and minor salivary glands, lacrimal glands, ceruminous glands, mucous glands of upper respiratory and digestive tract, breast and occasionally excretory glands of the female genital tract[1,2]. More than 70% of the tumors arise in minor salivary (mucous) glands. The palate is most commonly affected which could also be a site of presentation for other minor salivary gland malignancies like squamous cell carcinoma (SCC) and mucoepidermoid carcinoma[1,3,4]. Parotid gland, submandibular gland, antrum & tongue are the other sites affected[1,3].

An asymptomatic lump or swelling is the major presenting complaint. Occasionally it may present as a painful swelling. Sometimes patient may complain of pain in the form of either throat discomfort or referred facial pain. The unusual variety of other presenting complaints are nasal obstruction, headache, facial pain, hoarseness of voice, bleeding, oroantral fistula, depending on the site involved [1,2,3,5].

Characteristic features include aggressive, slow growth, with insidious destruction of surrounding tissues, perineural invasion, prolonged clinical course and tendency for delayed onset of the distant metastases. The distant metastases and regional lymphatic spread may even occur as uncommon late sequel, lungs being the common site of predilection. Three histological subtypes are described based on growth patterns: tubular, cribriform and solid. These subtype distinctions are important because the solid form shows a much worse prognosis than either cribriform or tubular[1,6].

Histopathology is the gold standard for the diagnosis of ACC. CT & MRI are considered to be of almost similar significance in detection of perineural spread with preference to MRI because of its high soft tissue contrast[2].

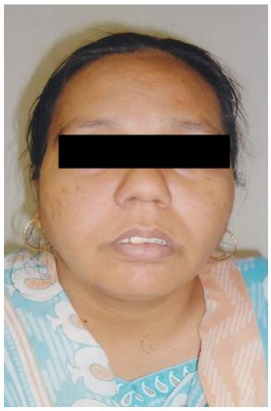
Prognosis is influenced by numerous factors i.e. complicated anatomy of the nose and paranasal sinuses prevents complete exenteration of the disease resulting in poor outcome and high morbidity. Hence early diagnosis and treatment is of immense value in decreasing the morbidity and mortality. Surgery is the main stay of treatment in early stages and surgery followed by chemoradiotherapy is the treatment of choice for

advanced malignancies. Radiotherapy can be given followed by salvage surgery for recurrence. Patients should be on regular follow up with nasal endoscopy and radiological imaging[5].

**Case report** : A 42 year old female reported at Oral Medicine And Radiology department with complain of pain and swelling of right side of upper face region since 6 months associated with nasal stuffiness and pain. For the same patient consulted an ENT doctor at that time, who advised CT scan and biopsy but patient did not undergo any investigation. No history of trauma, discharge from eyes or nose / paresthesia or difficulty in vision reported. Before 10 years patient had undergone extraction of upper left posterior teeth because of caries and pain. Upper right posterior tooth was missing since beginning. Medical or family history was not significant.

Extraorally, a single diffuse swelling of size approximately 3×4 cm was present at right side of maxilla extending anteroposteriorly from ala of nose upto lateral canthus of the eye and from superioinferiorly from infraorbital region upto smile line (Fig.1). Overlying skin appears normal in colour, no ulceration or bleeding or discharge present. Obliteration of right nasal cavity, deviated nasal septum towards left side, elevated level of right infra orbital margin seen. No discharge from eyes or nose was noted.

**Fig.1. Shows diffuse extraoral swelling over right middle third of face.**



Intraorally, a single ill defined tender, firm and fluctuant swelling of size approximately 1×4 cm was present obliterating right upper buccal vestibule extending from right central incisor region upto 2nd molar with tensed, normal coloured overlying mucosa without ulceration, bleeding or discharge present. Single well defined nodular swelling of size approximately 1.5×1.5 cm was found at the midline of palate extending 1 cm on both sides and anteroposteriorly from premolar region upto 2nd molar region with normal overlying mucosa and no ulceration or bleeding or discharge present (Fig. 2). Missing 14 and 26, carious 17, with no mobility of any tooth or discoloration seen.

**Fig.2. Shows intraoral swelling over buccal vestibule and palate with tensed overlying mucosa in buccal vestibular swelling.**

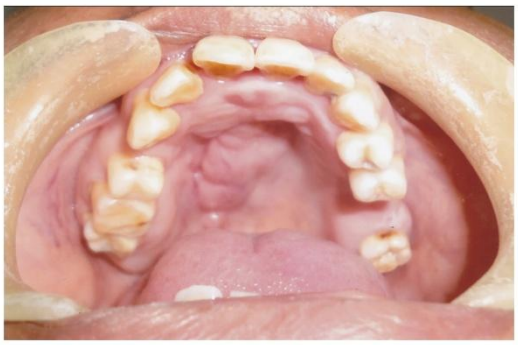
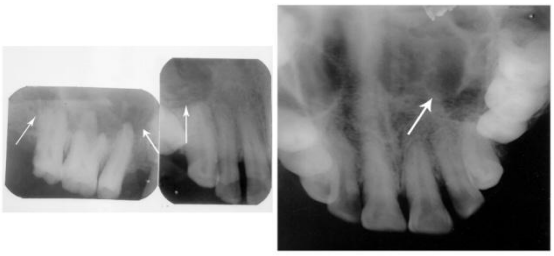


**Fig.3.Intraoral periapical views and occlusal radiograph showing presence of well defined radiolucency over right maxilla with non corticated, scalloped border, loss of lamina dura of the involved part of teeth with no root resorption. (white arrow).**



Clinical features were in favor of any growth arising from maxilla or maxillary sinus. Various investigations were done to reach a proper diagnosis. Intraoral radiographs of upper right teeth region showed a single well defined radiolucency with non corticated and scalloped border with loss of lamina dura in the apical 1/3rd of the roots of involved teeth seen with no evident root resorption (Fig. 3). Orthopantomograph & PNS view showed cloudy appearance of right maxillary sinus with destruction of roof and its lateral

border. Also involves right nasal cavity with elevated floor and displacement of nasal septum towards left side (Fig.4 & 5).

**Fig.4.Paranasal Sinus view showing cloudy appearance of right nasal cavity, maxillary sinus with destruction of all walls, nasal septum deviation towards left side.(white arrow).**

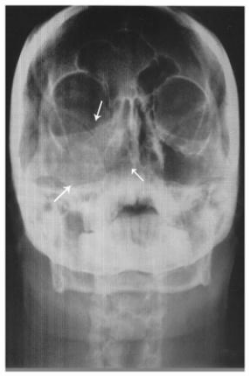
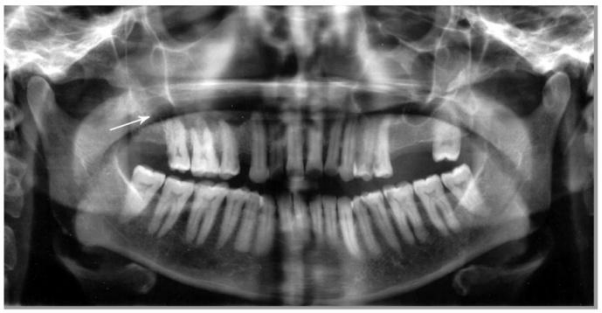


**Fig.5.Orthopantomograph shows features like, large radiolucency in periapical region of molars, cloudy appearance of right maxillary sinus with destruction of roof and its lateral border, with elevated right nasal floor and septum deviation towards left side.(white arrow).**



On CT evaluation evidence of 50×48×45mm sized polypoidal soft tissue density lesion noted arising from right maxillary sinus, showing heterogenous post contrast enhancement with non enhancing necrotic areas within it causing erosion of anterior medial and lateral wall of maxillary sinus. Mucosal thickening involving right frontal, left maxillary, left ethmoidal and bilateral sphenoid sinuses ( Fig.6 ). These findings were suggestive of malignancy of either maxillary sinus or maxilla. Thereby squamous cell carcinoma, adenoid cystic carcinoma, mucoepidermoid carcinoma were considered as differential diagnoses. Biopsy was done from right maxillary sinus through right nasal cavity which consisted of multiple irregular, flat, grey white to brown soft to firm tissue. H&E staining of which showed basaloid cells arranged in cribriform pattern and adenoid cystic pattern with punched out spaces filled with secretion. The cells show large nuclei & high N/C ratio with scant cytoplasm suggestive of adenoid cystic carcinoma of minor salivary glands (Fig.7).

**Fig.6. CT Showed heterogenous post contrast enhancement with non enhancing necrotic areas within it causing erosion of anterior medial and lateral wall of maxillary sinus. (white arrow).**

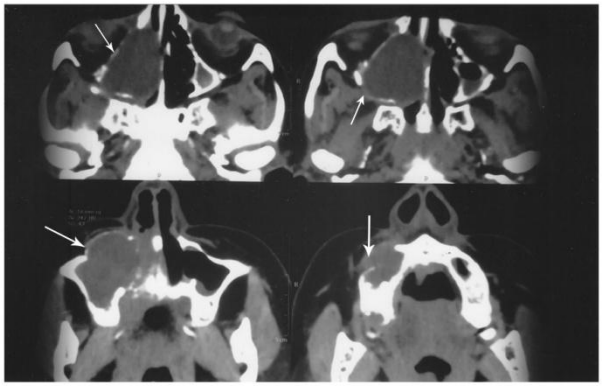
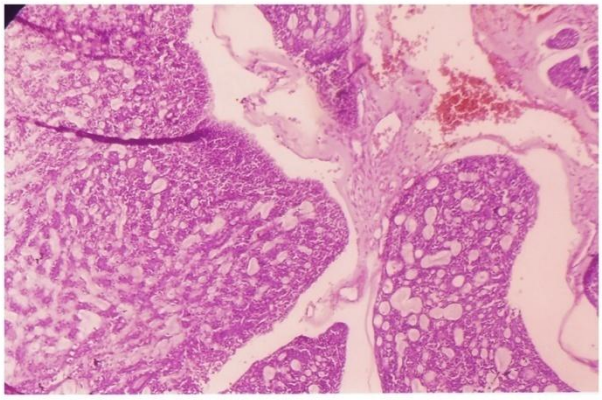


**Fig.7. Showing basaloid cells arranged in cribriform pattern and adenoid cystic pattern with punched out spaces filled with secretion.**



**Discussion:** The above mentioned case is of a 42 yr old female having a firm, tender, diffuse swelling of right maxilla obliterating right nasal cavity and deviation of nasal septum towards left side with elevated right infraorbital margin. Swelling was present in the buccal vestibule and the palate. Radiographic features revealed destruction of all the walls of maxillary sinus with hazy appearance and nasal septum deviation.

Clinical and radiographic features were suggestive of any malignant growth of maxilla/ maxillary sinus. Hence squamous cell carcinoma, adenoid cystic carcinoma and mucoepidermoid carcinoma were considered in differential diagnosis.
Mucoepidermoid carcinoma arising within the jaws as primary central bony lesion is extremely rare with its usual radiographic presentation being multilocular cyst like radiolucency[4].

SCC constitutes about 80% of all malignancies that arise in the nasal cavity and paranasal sinuses, whereas ACC is of salivary origin and is the 2nd most common sinonasal malignancy accounting for 10% of cases. SCC may present as a nasal mass or obstruction, rhinorrhoea, epistaxis, cranial neuropathies or pain. Long standing lesions may alter patient's facial features causing asymmetry/ proptosis, nasal obstruction, hyposmia, visual disturbances and paraesthesia. Sometimes the mass may be so large that it may affect the floor of the maxilla as well as the hard palate [1,2,3,5].

ACC usually presents with almost similar features. Mostly it is a slowly growing mass that is asymptomatic and painless though bone invasion and perineural spread can cause pain/ hypoesthesia[1]. Clinically and radiographically, this case presented with the same features. Histologically the diagnosis was confirmed as cribriform variety of ACC arising from minor salivary glands of palate. Sinonasal malignancies are mostly associated with exposure to smoking, formaldehyde, asbestos, wood or leather dust, patient's history did not reveal any of the etiological factors[4].

Perineural spread (PNS) occurred more often in palatal malignant tumors, as upper invasion of palatal malignant tumor is easily demonstrated because of contiguity of palatal bone and its thinness[2].Our case did not present with PNS even though the tumor was involving the palate. CT and MR images are very useful for detection of PNS in ACC. CT findings showed absence of PNS with low density area in the centre of the lesion suggestive of ACC[1].

Considering the CT and histopathological findings, treatment of ACC was planned out which included surgical resection of the lesion with wide margins obtaining clear margins around regional nerves followed by postoperative radiation therapy that enhances local and regional control in Adenoid cystic carcinoma[1, 5].

**Conclusion:** Though not as common as Squamous cell carcinoma, Adenoid cystic carcinoma should also be considered in differential diagnosis when there is presence of an asymptomatic growth in buccal as well as in palate region. CT scan can help in differentiating SCC from ACC but histopathological examination is mandatory in differentiating between the two, as well as in determination of type of ACC as its three different histological variants affect the treatment plan and moreover the prognosis.

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