ADENOID CYSTIC CARCINOMA OF PALATE IN YOUNG - A RARE SIGHT

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ABSTRACT

Adenoid cystic carcinoma is rare malignant neoplasm of salivary gland accounting for 1% of all malignant tumor of the oral and maxillofacial region and 6% of all salivary gland tumors. Typically it presents as slow growing painful mass affecting predominantly women in 5th to 6th decade of life. Known for prineural invasion, if not treated early it may prove fatal. Although much has been documented about the lesion since its discovery, Adenoid cystic carcinoma remains a topic of controversy regarding its origin, clinical course, treatment modality and recurrence. This case report documents yet another peculiar presentation of an aggressive Adenoid cystic carcinoma in a 30 year old woman. A detailed clinical differential diagnosis is also discussed.

KEY WORDS : Adenoid, Cystic, Carcinoma, Palate.

INTRODUCTION

Adenoid cystic carcinoma (Adenocystic carcinoma) are rare clinical entity marked by cylinders or bands of hyaline or mucinous stroma separated or surrounded by nests or cords of small epithelial cells, occurring in the mammary glands, salivary glands, and mucous glands of the respiratory tract. Because of its typical histopathological presentation, Billroth in 1859 coined the term cylindroma to name this clinical entity, however its preferred to avoid using this terminology as the same term is used for a skin adnexal tumor that has a markedly different clinical presentation and prognosis.[1] Foote and Frazell, in 1953, renamed the lesion adenoid cystic carcinoma ( ACC).[2] ACC is a rarity, comprising about 1% of all malignant tumor of the oral and maxillofacial Region.[3] ACC also accounts for about 6% of
all salivary gland tumors, 15–30% of submandibular gland tumors, 30% of minor salivary gland tumors, and 2–15% of parotid gland tumors. ACC typically are seen in middle aged individuals and exhibit slight female predilection. ACC is the second most common malignant neoplasm of salivary gland next only to mucoepidermoid carcinoma. It usually occurs in the 5th and 6th decade of life and is rare in people younger than age 20. Isacsson and Shear reported that palate is the most common site of occurrences of ACC followed by floor of the mouth, tongue and gingiva in decreasing order. Parotid tumors may cause facial nerve paralysis and palatal tumors may result in paresthesia due to palatine nerve invasion. As this tumor has a propensity for perineural invasion, the tumor tissue often can extend far beyond the obvious tumor margin. ACC usually presents as a slowly growing firm mass that may ulcerate in the oral cavity. Pain is a common and important finding, often occurring before a noticeable swelling or ulceration. Patients often complain of a constant, low-grade, dull ache, which gradually increases in intensity. Loco-regional lymphatic disease is uncommon; late distant metastases and local recurrences are relatively frequent. We report here a rare case of stage IVa adenoid cystic carcinoma in a 30 year old women which presented as an aggressively growing lesion of palate with a history of one month.

CASE REPORT
A 30 year old female patient reported to the department of oral medicine and radiology with complaint of swelling and pain in the palate on left side. She gave history of noticing a peanut sized swelling on the palate one month back which gradually increased to attain the present size. Initially asymptomatic, the swelling became tender as it increased in size. She also noticed a tender extraoral swelling on the left ala of the nose since the past 15 days. General physical examination revealed that she was moderately built and nourished, alert, responsive and cooperative. Vital signs were all within normal range. Left submandibular lymphnode was palpable, mobile and nontender. A solitary illdefined swelling was present in the region of left ala of the nose, measuring approximately 2.5 cm in diameter, causing mild asymmetry of face. The skin over the swelling appeared normal. Swelling had resulted in the mild uplifting of the left ala of nose. The nostril on the left side seems to be partially obliterated due to the swelling, although there was no history of paresthesia, epistaxis or nasal stuffiness. Intraorally a well defined swelling was present on the left side of palate, crossing the midline. Swelling was smooth anteriorly with bluish tinge and rough and erythematous posteriorly. The lesion was approximately 3cm x 2 cm, extending from rugae to the soft palate and midpalatine to alveolus. The swelling was soft to firm, tender, not
fluctuant, not mobile, not reducible, not compressible, with no paraesthesia. The associated teeth (24 and 25) were mobile but not tender. Also a second swelling was seen intraorally on the labial aspect of 21 and 22, which was about 1.5 cm, partially obliterating the labial sulcus. The mucosa over the swelling was normal. The swelling was non tender and bony hard. Considering the clinical features of the rapidly growing mass in palate which had perforated the cortices to involve the nasal cavity and resulting in mobility of involved teeth, a clinical diagnosis of malignant neoplasm minor salivary gland was given. The differential diagnosis considered were necrotizing sialometaplasia, pleomorphic adenoma, mucoepidermoid carcinoma of salivary gland, adenoid cystic carcinoma, Polymorphous low grade adenocarcinoma, lymphoma and carcinoma of maxillary sinus. Electric pulp vitality testing revealed that 23, 24 and 25 were non vital. Routiene blood investigations gave normal blood picture. A screening panoramic radiograph revealed an illdefined radiolucency in the right maxilla, measuring approx 4cm x 2.5cm, extending from distal root surface of 21 to 26 extending into the maxillary sinus resulting in haziness of maxillary antrum. Maxillary occlusal radiograph showed that the lesion was extending from the periapical region of anterior teeth till the mesial aspect of 28, sparing midpalatine suture. Intraoral periapical radiograph revealed an ildeffined, hazy radiolucency with irregular, infiltrating borders, associated with the periapical region of 23, 24, 25 and 26. Also evident was radiopaque specks suggestive of bony spicules present within the radiolucency. Root of 25 was resorption. Interdental bone between 24, 25, 26 was spared. The floor of the maxillary sinus was completely destroyed. Chest radiograph revealed no abnormality. CT Scan with contrast revealed a heterogeneously enhancing mass lesion with hypodense areas within it suggestive of necrosis in the region of left hard palate measuring 4cm x 3.5 cm in size and causing destruction of hard palate and erosion of medial wall of left maxillary sinus.(Fig. 3) Bony spicules were noted within the mass lesion. Also evident was the destruction of nasal septum and inferior turbinate. The mass lesion was seen to extend into the left maxillary sinus, left nasal cavity with minimal contralateral extension in right nasal cavity.(Fig.4) Pterygoid plates, pterygoid muscles, right maxillary sinus, walls of orbit, parapharyngeal space appears normal. The radiographic picture supported the presence of a malignant mass. Considering a massive invading lesion which has completely destroyed the palatal cortices and invaded the entire left maxillary sinus, nasal cavity and nasal septum, a Stage IVa (T4AN0M0) salivary gland neoplasm was hypothesized. Incisional biopsy was performed intraorally and soft tissue measuring about 1.0cm x 1cm x 0.5 cm, yellowish brown in colour, soft to firm in consistency was taken as specimen for histopathological examination. Subsequent
microscopical examination of specimen revealed tumor tissue arranged in varying pattern in the connective tissue stroma. Tumor tissue showed cells with hyperchromatic nucleus and little cytoplasm, surrounded by parakeratinized stratified squamous epithelium. (Fig. 5) The patterns seen were predominantly cribriform with area also showing ductal, solid and hylinised areas. Cystic and duct like space showed mucus like material within it. (Fig. 6) Tumor tissue invaded the bone and surrounding connective tissues. The histopathological features led us to give a final diagnosis of cribriform variety of adenoid cystic carcinoma of minor salivary gland of palate. Wide excision of the intra oral lesion with hemimaxillectomy of left side and extraction of all the maxillary teeth was performed under GA. (Fig. 7) Two weeks post operatively a temperory obturator was given. After 6 months of follow up when no complication was encountered a permanent obturator with complete denture was given. Patient is under regular follow up. There is no signs and symptoms of recurrence of neoplasm from the past one year since the surgery. (Fig. 8)

DISCUSSION
ACC is an uncommon form of malignant neoplasm that arises most commonly in the major and minor salivary glands of the head and neck. ACC is considered a slow growing aggressive tumor affecting individuals in fifth or sixth decade. [12] Our case was a unusual as it occurred in a 30 year old female and the time period between the first symptom and the final outcome was less than a month. Also though the tumor invaded into the maxillary sinus, destroying it completely, there were no signs clinical or histopathological) of perineural invasion (except the dull pain which can be multifactorial), making the case even more peculiar. In this case the presentation of the lesion was that of a aggressive mass causing bicortical expansion and was primarily seen as palatal swelling. The differential diagnosis considered were necrotizing sialometaplasia, pleomorphic adenoma, mucoepidermoid carcinoma of salivary gland, adenoid cystic carcinoma, Polymorphous low grade adenocarcinoma, lymphoma and carcinoma of maxillary sinus. Necrotizing sialometaplasia is a locally destructive inflammatory condition of salivary gland affecting palatal swelling. The differential diagnosis considered were necrotizing sialometaplasia, pleomorphic adenoma, mucoepidermoid carcinoma of salivary gland, adenoid cystic carcinoma, Polymorphous low grade adenocarcinoma, lymphoma and carcinoma of maxillary sinus. Necrotizing sialometaplasia is a locally destructive inflammatory condition of salivary gland affecting palatal salivary glands most frequently. Although it may occur at any age, it affects adult males more commonly. It affects hard palate more frequently then soft palate. It presents initially as a non ulcerated swelling which converts into craterlike proliferative ulcer mimicking a malignant lesion. Pleomorphic adenoma is the most common salivary gland tumor. Most Pleomorphic adenoma present as a smooth, dome shaped, firm submucosal mass or nodule with growth rate which is usually indolent. It can occur at any age but is seen in middle aged adults with
slight female predeliction. Carcinomas are known to occur in middle and elderly individual, causing rapid destruction of jaw bone and present as firm swelling associated with mobile teeth. Mucoepidermoid carcinoma is the most common malignant salivary gland neoplasm. Minor salivary glands of palate is the second most common site of occurrence after parotid gland. It usually presents as asymptomatic slow growing swelling which are sometime fluctuant to firm with a blue or red colour. Adenoid cystic carcinoma occurs most frequently in the palate. It presents as a painfull, slow growing mass affecting the surrounding structures in middle aged adult individuals. Polymorphous low grade adenocarcinoma occurs most commonly in palate and usually presents as asymptomatic slow growing swelling. It usually affects older adults and has definite female predeliction. Extra nodal lymphoma may present in the palate as nontender, diffuse swelling with boggy consistency although buccal vestibule is the most common site. Rarely carcinomas of maxillary sinus (mucoepidermoid carcinoma, adenoid cystic carcinoma, adenocarcinoma) may perforate the hard palate and present intraorally as diffuse swelling or as an ulcerated lesion. ACC is a slowly growing but highly invasive cancer with high recurrence rate. Lymphatic spread to local lymph nodes is rare. Hematogenous spread, however, occurs often in the course of the disease. Metastases into the lung are more common than regional lymph node metastasis. In our case clinical examination ruled out lymphnode involvement and metastasis to lungs were also ruled out by chest x-ray. Histopathologically, ACC presents three patterns, cribriform, tubular and solid. The cribriform (glandular) pattern is the most classic and best recognized appearance, characterized by islands of basaloide epithelial cells that contain multiple cylindric, cyst-like spaces resembling swiss cheese. Szanto et al. have defined a three tier grading system which takes into account the histomorphologic pattern and percentage of the solid component. This has been regarded as a useful prognostic indicator. Grade 1 ACCs are well differentiated and composed of tubular and cribriform patterns without solid components; grade 2 ACCs are characterized by a pure cribriform pattern or mixed with less than 30% of solid areas; and grade 3 ACCs are tumors with marked predominance of the solid pattern. The case reported here was classified as grade 1 ACC. Possible treatments of ACC include four different modalities; surgical therapy, radiotherapy, chemotherapy and combined therapy. Surgical excision with wide margins is the treatment of choice. Avery and Miglianico recommend postoperative radiation since radiation often produces tumor regression and relieve symptoms. Postoperative radiotherapy is also indicated for cases of tumor location close to the base of the cranium with the presence of neck lymph node metastasis and perineural invasion, the solid histological subtype, and in the case of recurrent tumors. Since our case
was a grade 1 ACC, an exclusive surgical approach was adapted which showed fairly good result. The presence of cervical lymphatic metastasis at the time of diagnosis, an advanced tumor stage, the presence of affected surgical margins, solid histological type or histopathology grade and macroscopic perineural invasion indicates poor prognosis of ACC. [21,22,23] The histological subtypes of low malignancy levels (tubular and cribriform) have a better prognosis than those of a high-degree of malignancy (trabecular and solid). Since our case was cribriform variety of ACC, a good prognosis was predicted.

CONCLUSION
ACC is a relatively rare neoplasm, the diagnosis of which if not made early can prove fatal. Although ACC is known to us from a long time yet it is surrounded by contravercies. Right from the evolution to the clinical behaviour to the most effective treatment, ACC has always remained the topic of debate. There is need for more clinical trials to investigate the various clinical course of this neoplasm and to evaluate the most effective treatment modalities via lifelong follow up of such patients. This case presents yet another unique presentation of adenoid cystic carcinoma and demands documentation. So also it suggests that ACC should be considered in the differential diagnosis of swellings associated with orofacial region especially the palate.

REFERENCES
