VOLUME-6 | ISSUE-4 | APRIL - 2017 • ISSN No 2277 - 8179 | IF : 4.176 | IC Value : 78.46

Adenoid cystic carcinoma of palate with simultaneous metastasis to lung: An unusual presentation

Pathology

KEYWORDS: Adenoid cystic carcinoma, metastasis, lung

HariRam	Professor, Department of Oral and Maxillofacial Surgery, Faculty of Dental Sciences, King George's Medical University, Lucknow. U.P.
ShilpiGangwar	Senior Resident, Department of Oral and Maxillofacial Surgery, Faculty of Dental Sciences, King George's Medical University, Lucknow. U.P.
SanjayKumar	Senior Resident, Department of Oral and Maxillofacial Surgery, Faculty of Dental Sciences, King George's Medical University, Lucknow. U.P.
Shadab Mohammad	Professor, Department of Oral and Maxillofacial Surgery, Faculty of Dental Sciences, King George's Medical University, Lucknow. U.P.
Neha Singh	Junior Resident , Department of Oral and Maxillofacial Surgery, Faculty of Dental Sciences, King George's Medical University, Lucknow. UP..

ABSTRACT

Adenoid cystic carcinoma (ACC) is a common malignant neoplasm of salivary glands. Slow growth, perineural invasion, potential local recurrence and late metastasis to lungs and bone are common characteristic of this tumor. Three histopathological patterns, cribriform, tubular and solid have been described in the literature. Solid type is related to a poor prognostic contrary to the cribriform type, which has a better prognosis. ACC patients have a poor prognosis, even after radical surgery and radiotherapy.

INTRODUCTION

The Adenoid cystic carcinoma (ACC) is a relatively rare epithelial tumor of the salivary glands. 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.[1] The minor salivary glands are affected most frequently in about 40% of cases, parotid gland in 30%, followed by the submandibular gland and in 1% cases sublingual glands are involved.[2] Most of ACC occur in fifth and sixth decades of life and females are slightly more affected than males.[3,4,5] Although the tumor grows slowly, but neural invasion, distance metastases and recurrences are common. Because of these features patients of ACC have a poor prognosis, even after radical surgery and radiotherapy.[6]

CASE REPORT

A 31 year old male reported to department of Oral and Maxillofacial Surgery with chief complaints of swelling and pain on right side of palate, which was gradually increasing in size since last 1 year. Patient had no history of any deleterious oral habits. On extra-oral inspection no obvious facial asymmetry was seen. The patient had adequate mouth opening and no restriction was felt on lateral movement. On palpation regional lymph nodes were not palpable.

On intra-oral inspection a well defined swelling measuring 3.5 x 2.5cm in size on right side of palate which was covering hard and soft palate both and was extending from maxillary right second premolar, upper right third molar and up to anterior tonsillar pillars(Fig.1). Swelling was ulcerated at top of posterior region and un-ulcerated mucosa was normal in colour and texture. On palpation, swelling was non tender and firm in consistency. There was no mobility and displacement of teeth.

Fig.1 Intraoral view.

Incisional biopsy was done under local anaesthesia. Sections stained with Hematoxylin and Eosin, revealed the presence of stratified squamous orthokeratinized epithelium overlying connective tissue stroma having the tumor islands. The tumor islands were comprised of proliferating neoplastic salivary epithelial cells arranged in cribriform, tubular and solid sheet configuration. Few areas showed hyalinization within the stroma. Eosinophillic coagulum within the tubules were also observed (Fig.2 &3). Based on these features a histological diagnosis of ACC was made.

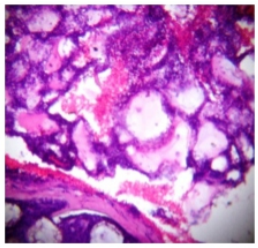

Fig.2 Histological slide

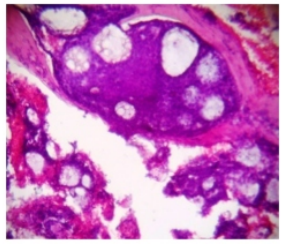

Fig.3 Histological slide

CT scan of maxilla revealed an ill defined heterogeneously enhancing soft tissue attenuation lesion in retromolar oropharyngeal soft tissue region at the level of floor of maxillary sinuses area on right side involving right lateral pharyngeal wall abbuting the right tonsillar pillar and adjacent part of base of tongue. The lesion was measuring approximately 3.2 X 2.5 X 1.2 Cm in size. Few small non enhancing hypodense areas were also noted within the lesion suggestive of necrotic changes. The lesion was also seen involving right medial pterygoid muscle. Interface of the lesion with adjacent part of soft palat was ill defined. Mild cortical irregularity was seen in maxilla adjacent to the lesion (Fig.4 & 5).

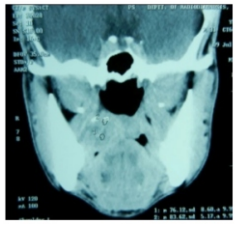

Fig. 4 CT Scan – Coronal view

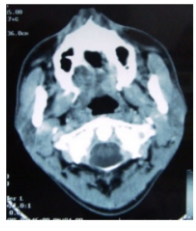

Fig.5 CT Scan – Axial view

X-ray of chest revealed multiple well defined round to oval radiodense opacities over bilateral upper and middle zone

suggestive of metastasis. Contrast enhanced CT scan of thorax revealed multiple hypodense lesions of subcentimeter sizes in both lung fields except one lesion of 1.5 cm in right middle lobe. Patchy areas of increased lung attenuation, suggestive of fibrosis were seen in apico- Posterior segment of left upper lobe.

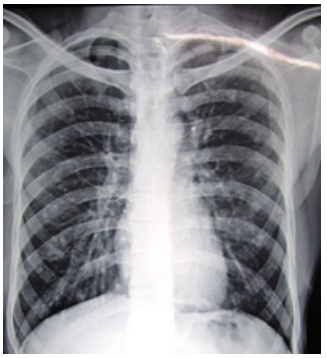

Fig.6 Chest x ray

On the basis of above findings diagnosis of adenoid cystic carcinoma of palate showing pulmonary metastasis to lung was made.

As per the diagnosis and stage of the disease patient was planned for chemotherapy. Combination chemotherapy comprising of carboplatin 600 mg iv and adriamycin 80 mg iv on day -1 were given for 3 cycles at 3 weekly interval after cardiac evaluation and fitness. After chemo therapy, slight reduction in size of tumour was seen. Three more cycles of the same chemotherapy were further given to patient. CT scan and X-ray of chest were done to evaluate status of disease and no increase in size was noted in a follow up period of two years.

Though metastasis is a common phenomenon in ACC, the mentioned case is presented here because of simultaneous distant metastasis to lung at the time of presentation and its aggressive behaviour.

DISCUSSION:

ACC is a malignant epithelial tumor characterized by slow growth, late onset of metastasis and poor prognosis[7]. In the reported case metastasis to lung was seen at the time of detection of ACC on the palate. We understand that the palate was the primary tumor from which subsequent haematogenous metastasis to the lungs took place. Theodor Billroth used the term "cylindroma" in 1859 to describe the histological standard of salivary gland tumors[1]. In 1954 Foote and Frazell renamed the lesion as adenoid cystic carcinoma[8].

ACC occurs most often in minor salivary glands and the submandibular gland, and less frequently in the major salivary (sublingual and parotid) glands. Other rare locations are aerodigestive tract, lacrimal glands and adenexal skin glands[4,5]. The reported case was present on the palate involving both soft and hard palate extending up to anterior tonsillar pillars. ACC has a wide spread age distribution and peak incidence occurs between the 5th and 6th decades of life [9]. Females are slightly more affected than males [5]. Evesson and Cawson[10] also observed predominance of ACC in females (F:M 1.2:1). However reported tumor was present in a 31 years old male.

Clinically, ACC is characterized as a painless slow growing mass with a propensity to invade peripheral nerves and having high recurrence rate with metastasis to other organs[11]. Perineural invasion is characteristic of these tumors [3,5] and occurs in up to 60% of cases[6]. Cervical metastasis is rare and occurs in only 8-13% of patients[5,12]. Distant metastasis may occur in up to 50% of ACC patients during the course of the disease, with the lungs and bones as the most common sites [12]. Pain was observed in reported case due to perineural invasion. Cervical lymphadeopthy was not seen. Lung metastasis was seen at the time of prentation of primary tumour. Metastasis to other organs was not seen.

There are three histological subtypes of ACC: cribriform (glandular), tubular and solid. They may occur either separately or together in the same tumor. The solid subtype is the most aggressive and has worst prognosis[3,4] Huang et al[13] observed a survival rate of 16.7 % after a 10 year treatment for cases where solid pattern was observed and 47.4% for lesions where cribriform and tubular pattern were found.

Prognostic factors are primary tumor (T), anatomical location, status of metastasis (M), neural invasion and histological grade of tumour[14]. Batsakis proposed that cribriform pattern has better prognosis. Without solid pattern it has 39% survival rate of 15 years; if solid pattern is present in 30% or less, has 26%survival of 15 years and tumours those are composed of mainly solid pattern have 5% of survival rate of 15 years [14]. All the three histological subtypes i.e. cribriform, tubular and solid were present in reported case.

ACC may be divided into two immunochemical groups. A group of neoplasm cells with prevalence of ductal formation positive to CEA, EMA, cytokeratin of low and high molecular weight and S-100 [11]. Other group of neoplasm express positively for actin of specified smooth muscle and cytokeratin of low molecular weight[15].

Metastasis in the lung with a primary tumor in the oral cavity is presented as multiple radiopaque shadows on the X-ray chest and CT [11]. In the present case also we observed multiple radiopaque shadows in the whole lung presuming that simultaneous presentation of metastasis along with primary which is rare and unique.

Treatments of ACC include four different modalities: surgical therapy, radiotherapy, chemotherapy and combined therapy (surgery and radiotherapy or radiotherapy and chemotherapy)[11,16]. ACC is locally aggressive, with high recurrence rate and late metastasis to lung, bone and brain, commonly leading to death between 10 and 20 years after initial treatment[17]. Compared to other malignant neoplasms, ACC is more difficult to fully remove, with frequently identified positive surgical margins[9]. Neck dissection is performed if cervical nodes are involved. Since metastasis was reported at the time of presentation, so chemotherapy was given to the patient under reporting.

CONCLUSION:

ACC is locally aggressive tumour, having high recurrence rate and late metastasis to lung, bone and brain. Local control, normal functionality and prevention of distant metastasis should be the goal of clinicians. In the present case, no regional lymph nodes were involved but metastasis to lung was seen simultaneously with detection of primary, showing a aggressive course of disease. Patient was given combination chemotherapy and no further increase in size of lesion (primary and lung) was seen during a period of 2 years after chemotherapy.

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VOLUME-6 | ISSUE-4 | APRIL - 2017 • ISSN No 2277 - 8179 | IF : 4.176 | IC Value : 78.46

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