Solid Adenoid Cystic Carcinoma of the Maxilla: A Case Report

Solid Adenoid Kistik Karsinoma: Olgu Raporu

*Kaan ORHAN DDS, PhD, **Yavuz YÜKSEL DDS, PhD, **Durmuş İker GÖRÜR DDS, PhD

*Ankara University, Faculty of Dentistry, Department of Oral Diagnosis and Radiology
** Ankara University Faculty of Dentistry, Department of Oral and Maxillofacial Surgery

ABSTRACT

Adenoid cystic carcinoma is a rare malignant tumor that affects the major and minor salivary glands, the lacrimal glands, the ceruminous glands and occasionally the excretory glands of the female genital tract. %30-40 of these carcinomas occurred as head and neck tumors. Approximately 50 to 70% of reported cases occurred in the minor salivary glands. The most common location is the palate, generally in the area of the greater foramen. Adenoid cystic carcinoma is generally characterized by a slow growth rate, and it is often present for several years before patient seeks treatment. The solid central salivary gland tumour should be considered in the differential diagnosis of the aggressive lesions in the maxilla. In this paper solid adenoid cystic carcinoma which originated from right maxilla premolar-molar region in a 53-year-old woman is presented, the treatment modalities and the differential diagnosis are discussed.

ÖZET


KEYWORDS
Adenoid cystic, Carcinoma, Maxilla, Salivary gland neoplasms

ANAHTAR KELİMELER
Adenoid kistik, Karsinoma, Maksilla, Tükürük bezi neoplasmları
INTRODUCTION

Adenoid cystic carcinoma (ACC) is a rare malignant tumor that affects major and minor salivary glands, lacrimal glands, ceruminous glands and occasionally excretory glands of the female genital tract\(^1\)\(^-\)\(^15\). ACC are uncommon tumors, representing about 10% to 15% of the histology of head and neck tumors. They are 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\(^2\)\(^-\)\(^6\). They can also arise in other sites of the head and neck except those accounted above, including the tracheobronchial tree, and the esophagus\(^2\). ACC is well known for its prolonged clinical course and the tendency for delayed onset of the distant metastases. The distant metastases and regional lymphatic spread may even occur as late sequela, with the patient often dying of disease, sometimes many years after the original diagnosis\(^2\)\(^-\)\(^3\). The lungs are the sites of predilection for distant metastases. Metastases to bone, liver and brain have been also report in previous articles\(^6\). Regional lymphatic spread is rather uncommon. The clinical course is often relentless and fatal if this occurs. Long term survival can be achieved particularly with combined surgery and radiotherapy\(^4\)\(^-\)\(^6\). The tumor displays a distinctive cribriform histomorphology often described previously as “Swiss Cheese” or “Sievelike”; but this terminology has proved inadequate in expressing the wide spectrum of histologic diversity that may be seen. In addition to the more characteristic cribriform architecture, areas of tubular differentiation or of solid cellular growth may be observed\(^3\).

The purpose of this paper is to report a case of adenoid cystic carcinoma along with an analysis of the literature in order to make a contribution to the pathogenesis, treatment and differential diagnosis of the adenoid cystic carcinoma.

CASE REPORT

A 53-year-old woman presented in August 2004 having been referred by a general dental practitioner who had noticed a hard bony swelling in the right quadrant of the maxilla (Figure 1). The patient was complaining of a slowly growing, painless mass. She stated that this swelling had been present for several years but over the preceding 4 months had increased in size. She also stated that she had never been a smoker or drank alcohol and living as a housewife in a village for 42 years. The head and neck examination revealed no evidence of paresthesia or motor nerve deficiency, but submandibular and cervical lymphadenopathy was detected. Intraoral examination also showed a firm mass in the right maxilla starting from central incisor to first molar area. There was no overlying mucosal ulceration (Figure 2). Firstly, a panoramic radiograph was taken. This radiograph showed a unilocular radiolucency in the right maxilla, the cortical plate above the lesion appeared as involved (Figure 3). It was decided to perform a computed tomography (CT) scan for obtaining a more precise location and definition of the pathologic features. CT imaging demonstrated an enhanced 4.5x3.5x3.5 cm mass in the right maxilla, the cortical plate above the lesion appeared as involved (Figure 3). It was decided to perform a computed tomography (CT) scan for obtaining a more precise location and definition of the pathologic features. CT imaging demonstrated an enhanced 4.5x3.5x3.5 cm mass in the right maxilla with destruction of maxillary sinus. CT scans also showed, the infiltration of the mass through the adjacent areas especially to the muscles, and cervical lymphadenopathy approximately 10mm in size (Figure 4,5). To examine the lesion in detail, Magnetic Resonance Imaging (MRI) was performed in addition to CT. A coronal fat-suppressed T\(_1\) weighted image with gadolinium-DTPA enhancement (Gd-T1WI) showed a high intensity tumor in the right palatal region continuing to buccal region of the alveolar bone. The high intensity area continued to the major zygomatic and orbicularis oris muscle. The mass also infiltrated through the right maxillary sinus (Figure 6). An axial Gd-T1WI showed the high intensity area on the right hard palate long through the major palatine nerve (Figure 7).

With the patient under local anesthesia, an incisional biopsy was performed. The tissue was submitted for microscopic examination, and an adenoid cystic carcinoma was diagnosed. The
FIGURE 1

Photograph showing the extra-oral view of the patient

FIGURE 2

Intra-oral photograph showing a firm mass with no overlying mucosal ulceration

FIGURE 3

Panoramic radiograph demonstrates the unilocular radiolucency in the right maxillary quadrant. The cortical plate above the lesion appears involved

FIGURE 4

An axial CT scan showing enhanced, soft tissue mass filling the hard plate and buccal region of the maxilla and the maxillary sinus (arrows)

FIGURE 5

A coronal CT image also demonstrating the mass infiltrating through the orbicularis oris and major zygomatic muscles (arrows)

FIGURE 6

A coronal fat-suppressed T1 weighted image with gadolinium-DTPA enhancement (Gd-T1WI) showing the high intensity tumor (arrows)
tumor was resected surgically. It filled the maxillary antrum, but had not eroded the midline of the hard palate, and extended to the orbicularis oris muscle and major zygomatic muscle. A right maxillectomy with neck dissection was performed. Cervical lymph nodes also resected because of infiltration. No extensive perineural invasion was present with tumor at the resection margins. The total resected specimen was sent again for the histopathological examination and the final diagnosis was solid adenoid cyst carcinoma (Figure 8). A dental obturator was fitted subsequently in order to promote the patient’s life quality. The maxillary cavity received post-operative radiotherapy of 60 Gy in 30 fractions over 30 days. The follow-up after 1 year since the initial diagnosis showed no sign of recurrence.

**DISCUSSION:**

ACC was first described by Billroth in 1856, and called ‘cylindroma’ due to its characteristic histological appearance. ACC are uncommon tumors, representing about 10% to 15% of the histology of head and neck tumors. They are 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. Characteristic features include aggressive, slow growth, with insidious destruction of surrounding tissues and perineural invasion. The palate is the most commonly affected site as in our case and these tumors have the best prognosis. Three histological patterns of growth have been described: solid, cribriform, and tubuloductal. The World Health Organization (WHO) histological definition is ‘an infiltrative malignant tumor having a very characteristic cribriform appearance. The tumor cells are arranged as small duct-like structures or larger masses of myoepithelial cells disposed around cystic spaces to give a cribriform or lace-like pattern.” The solid growth architecture portends an extremely poor prognosis in adenoid cystic carcinoma. It is the exception to the indolent growth pattern and usually demonstrates a highly aggressive clinical course, with early and more frequent recurrence (nearly 100%). However, in contrast to this statement, our case did not show any recurrence still after one year. On the other end of the spectrum is the tubuloductal architecture, which is associated with the more-typical growth rate and the lowest rate of recurrence at 59%. Between these extremes lies the most common pattern, the cribriform architecture, which is associated with a recurrence rate of 89%.

There are no universally accepted agreed-upon predisposing factors for the development of ACC, and there seems to be no family tenden-
There is a very slight female preponderance, but in a recent study it was found a 1:1 ratio for gender distribution\(^4\). Patients are typically present with a slowly enlarging mass that, because of its indolent growth pattern, can be quite large. The mass is usually asymptomatic, painless, although bone invasion or perineural spread can cause pain or hypoesthesia. Most ACC are submucosal and appear as smooth, domed swellings without overlying ulceration. They are encapsulated but frequently well circumscribed; this appearance is misleading because these lesions have an insidiously infiltrative growth pattern. The incidence of cervical lymphatic metastasis is low; about 8% at presentation and 7% later\(^9\). Although spread to regional lymph nodes is rare, distant metastases, particularly to lungs and bone, are more common and often unpredictable. The distant metastases and regional lymphatic spread may even occur as late sequela, with the patient often dying of disease, sometimes many years after the original diagnosis\(^2,3\). CT and MRI are useful in surgical planning, especially with regard to the submucosal extend and perineural spread, which can be difficult to assess clinically. These lesions are generally poorly defined and have infiltrative margins. The center of the lesion, is usually low density on CT, a characteristic that can help differentiate it from a primary squamous cell carcinoma. ACC is suggested to occasionally occupy almost the whole area of a sparse component without influence on surrounding structures. The changes on MR images are characterized solely by an increase of intensity with preservation of the shape\(^{16-17}\). The differential diagnosis for ACC should include mucoepidermoid carcinomas, acinic cell carcinoma and malignant mixed tumors, adenocarcinoma and squamous cell carcinoma\(^{18}\).

The treatment of ACC involves surgical resection with wide margins. The surgeon must pay particular attention to obtaining clear margins around regional nerves. The surgical ablation must be individualized because radically mutilating surgery does not appear to improve the prognosis in highly aggressive tumors. Neck dissection like in our case should be performed who have clinical and radiological evidence of cervical lymphatic metastasis. Postoperative radiation therapy enhances local and regional control in ACC. Radical surgery followed by postoperative therapy results in 5- and 10-year survival rates 77 and 57% respectively\(^3,4,12\).

In our patient, we performed a neck dissection and maxillectomy with using a dental obturator right after the surgery which provided functional result. Postoperative radiation therapy was administered to the primary site and to ipsilateral neck. The histological findings in this case were of ACC of the maxilla with areas of cystic and solid pattern. No extensive perineural invasion was present with tumor at the resection margins. The patient is free of disease at 1-year follow-up.

In conclusion, this case demonstrates the value of diagnosis and treatment of cystic carcinoma as these tumors should be considered in the differential diagnosis of aggressive lesions in the maxilla. Although in our case there is no recurrence after one year, we are calling patient for follow-up at every 6 months because our case was a solid form of ACC which can show recurrence frequently (nearly 100%). Hence, long-term follow-up is essential for ACC in order to avoid recurrence of this lesion.

**REFERENCES**


CORRESPONDING ADDRESS

Kaan ORHAN DDS, PhD
Ankara University, Faculty of Dentistry, Department of Oral Diagnosis and Radiology 06500 Beşevler/Ankara
Tel: 0 312 212 62 50/331, Faks: 0 312 212 39 54, E-mail: call52@yahoo.com