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Adenoid Cystic Carcinoma of Palate: A Case Report and Literature Review

Salma Adnane[1*], Mounia El Bouhairi[1], Kaoutar El Khalifa[1], Ihsane Ben Yahya[2]

[1]Department of Oral Surgery, Dental Consultation and Treatment Cente, Ibn Rochd University Hospital Center, Casablanca, Morocco
[2]Oral Surgery Unit, Dental Consultation and Treatment Center, Ibn Rochd University Hospital Center, Dental Faculty, The Mohammed VI University of Health Science, Casablanca, Morocco
Email: *sallyad993@gmail.com

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Abstract

Adenoid cystic carcinoma (ACC) represents 10% of all salivary gland neoplasms and is the most common malignancy of the minor salivary glands. 50% occur in intraoral sites, with a definite predilection for the hard palate. Other locations include the major salivary glands, particularly the parotid, aerodigestive tract, lacrimal glands, and adnexal skin glands. This tumor frequently occurs in the fifth decade of life with a slight female predominance. It's characterized by slow growth, a high risk of local recurrence, neural invasion and delayed distant metastasis which can lead to poor prognosis. Classical management involves surgery and subsequent radiotherapy. The histological stage, tumor location, size, and early diagnosis of the lesion are all important factors in successful treatment and patient survival. The objective of this article is to report the diagnostic and therapeutic attitude towards a palatal Adenoid Cystic Carcinoma diagnosed in a woman of 61 years old, treated in the oral surgery department of the dental consultation and treatment center in Casablanca and to make an update on the data of the literature concerning this pathology.

Subject Areas

Dentistry

Keywords

Adenoid Cystic Carcinoma, Hard Palate, Minor Salivary Gland, Oral Surgery

1. Introduction

Adenoid cystic carcinoma (ACC) is an uncommon tumor that may arise in a

wide variety of anatomical sites in the head and neck. It accounts for about 1% of all head and neck malignancies and about 10% of all tumors of the salivary glands.

Among the major glands, the parotid is the most common site of occurrence. Intraorally 50% of ACCs occur on the palate. The other less common sites are the lower lip, retromolar tonsillar pillar area, sublingual gland, buccal mucosa and floor of the mouth [1].

It is most commonly found in the 4th to 6th decade of life with a slight female predilection [2].

Clinically, the tumor is characterized by an indolent growth pattern but an aggressive progression, with local recurrence and distant metastasis. The most commonly reported symptom is a slowly growing mass, followed by pain attributed to its tendency for perineural invasion [3].

The slow growth and the absence or paucity of symptoms often result in delayed diagnosis and many patients present with advanced disease which can lead inevitably to a poor prognosis.

According to the National Comprehensive Cancer Network (NCCN) Guidelines of 2019, most patients will undergo extensive resection followed by postoperative radiation regardless of margin status. With this treatment modality, the rate of disease-free survival at 5 years is generally high [4].

The aim of this paper is to report the case of adenoid cystic carcinoma of palate diagnosed in a patient of 61 years old treated in the oral surgery department of the dental consultation and treatment center of Casablanca and to make an update on the current understanding of intraoral adenoid cystic carcinoma including a review of its epidemiology, clinical behavior, pathology, histopathology, diagnostic assessment, treatment and prognosis.

2. Case Report

We report the case of a 61-years-old woman, received in the consultation of the Oral Surgery Department of the Dental Consultation and Treatment Center of Casablanca complaining of palatal swelling that has been evolving for a few months.

The patient was a non-smoker, and her medical history indicated that she was taking medication for Arterial Hypertension.

The extra-oral examination was unremarkable. No regional lymphadenopathy was found.

The intra-oral examination revealed an edentulous posterior right maxilla and a well-limited swelling in the right posterolateral portion of the hard palate. It was non-tender, hard in consistency and immobile, painless on palpation, covered with a normal overlying mucosa which had been evolving for more than 10 months and which increased gradually in size.

The swelling was seen extending into the posterior right area of the palate but not crossing the midline (**Figure 1**).

The panoramic radiograph and the CBCT were unremarkable, no bony changes were observed and routine blood investigations were found to be normal (Figure 2).

On the basis of clinical and radiographic features, a diagnostic hypothesis of pleomorphic adenoma of minor salivary gland was established.

An incisional biopsy was performed under local anesthesia and was subjected to histopathological examination (Figure 3). This latter showed the strands of uniformly stained deeply basophilic cells along with pseudocysts containing eosinophilic granulofibrillar material (Figure 4).

A diagnosis of Adenoid Cystic Carcinoma was established. The patient was referred to the department of maxillofacial surgery, and an extension assessment was carried out. Fortunately, no evidence of distant metastasis was observed.

The patient was treated by wide surgical excision with clear margins. Adjuvant radiotherapy was also carried out.

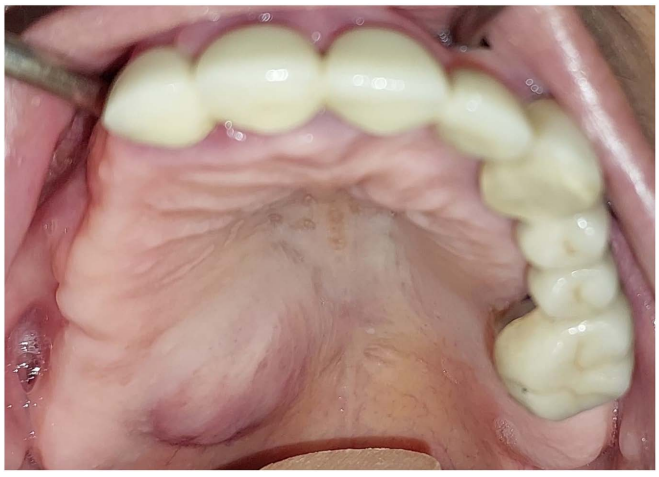

Figure 1. Preoperative Intraoral photograph showing a well-limited swelling in the right posterolateral portion of palate. Painless on palpation, covered with a normal-looking mucosa.

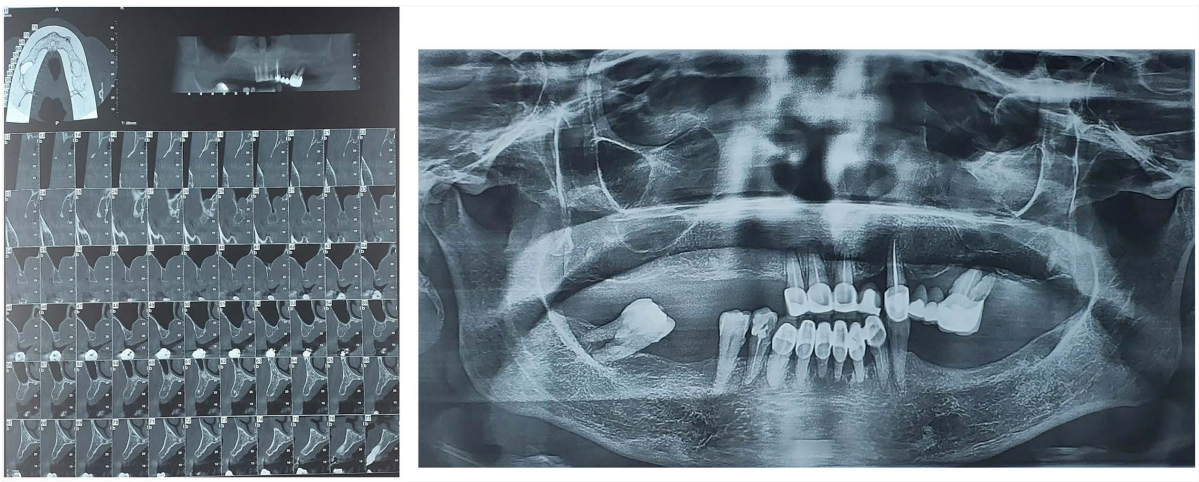

Figure 2. Pre-operative Radiographs showing a normal palatal bone.

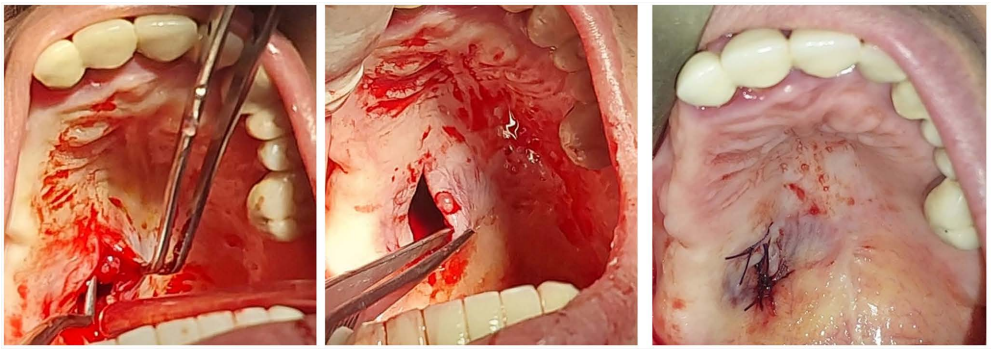

Figure 3. Intraoperative photographs: Incisional biopsy performed under local anesthesia.

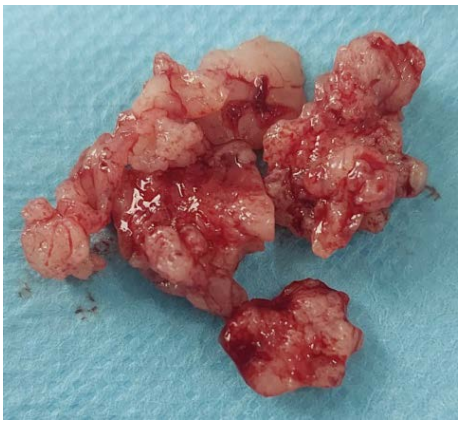

Figure 4. Surgical specimen.

3. Discussion

Adenoid cystic carcinoma (ACC) is a rare tumor, accounting for only 1% of all malignant tumors of the oral and maxillofacial region. It's one of the most common malignant tumors of the minor salivary glands. It has been described as one of the most biologically destructive and unpredictable tumors of the head and neck [5].

The palate is usually the most affected site, followed by the tongue. Other commonly affected sites include the floor of the mouth and the lip. ACC occurs most often in minor salivary glands and the submandibular gland, and less frequently in the sublingual and parotid glands [6].

It may occur at any age. In most cases, the patients are middle aged and are diagnosed with the disease in the fourth through sixth decades of life. The literature is inconsistent when it comes to gender predilection, with some authors reporting a male predominance and others finding a female or no gender predilection [7]. The exact etiology of ACC remains unknown. However, recent studies have revealed molecular abnormalities that are linked to ACC development and progression. Indeed, chromosome abnormalities and genetic deletions have been observed in samples of ACC through various studies. The p53 tumor suppressor gene seems to be inactivated in advanced and aggressive forms of this neoplasm, according to several authors. Otherwise, the specific molecular ab-

normalities that underlie this disease process are still unknown [8].

Clinically, It frequently presents with unspecific symptoms, small size initially, swelling, slow growth as seen in this case, perineural invasion as well. ACC is also characterized by local recurrence and distant metastasis. And it is often problematic to diagnose because of the difficulty in defining its clinical differentiation and distinguishing it from benign tumours making the prognosis questionable [1] [3].

Our patient initially presented a well-limited swelling in the right posterolateral portion of the hard palate. It was non-tender, hard in consistency and immobile, painless on palpation, covered with a normal overlying mucosa.

Histopathologically, 3 subtypes of ACC are known: cribriform, tubular and solid. They may occur either separately or together in the same tumor, and the solid subtype is the most aggressive and has the poorest prognosis [9].

The diagnosis is made by histopathological evaluation of a biopsy of a tumor mass. Besides the radiographic examination including panoramic radiograph, CT scans of head and neck region, the magnetic resonance imaging is an important diagnostic tool to assess the exact extent of the lesion. Perineural invasion is a common symptom of ACC. Hence, patients may present neuropathic symptoms. Examining the cranial nerves is essential when assessing these patients if recurrence is suspected.

The differential diagnosis of ACC is largely that of other benign and malignant neoplasms that arise in palate. These include pleomorphic adenoma (PA) and mucoepidermoid carcinoma (MEC). Patients with these types of palatal tumors present a mass with similar manifestations, such as swelling, a lack of tenderness and fluctuance, and occasional pain. When patients with palatal tumors have intact mucosa, making accurate differential diagnoses among these tumors on the basis of clinical examination is sometimes difficult [10].

Different treatment modalities have been established for ACC, including surgery, radiotherapy, chemotherapy, and combination therapy. Surgical resection with the widest margins possible is required because the tumor cells extend well beyond the clinical or radiographic margins. It is generally accepted as the primary treatment modality, followed by postoperative radiation regardless of margin status [11].

In the case reported in this article, the patient initially underwent diagnostic surgery, which consisted of an excisional biopsy. Histological examination confirmed the diagnosis of Adenoid Cystic Carcinoma. The patient was referred to the department of maxillofacial surgery. The extension assessment was unremarkable and confirmed the absence of distant metastasis. The patient was treated by wide surgical excision with clear margins. Adjuvant radiotherapy was also carried out.

The role of chemotherapy for adenoid cystic carcinoma is still controversial. The combination of surgery and radiotherapy is a common choice which has improved locoregional control of the disease. In fact, disease-free survival rates

at 5 years are generally high. In his study, Chang revealed that the 5-year survival rate of patients with ACC ranges from 67% to 73%. The 10-year and 20-year survival rates are lower because of delayed locoregional recurrence and distant metastasis [12]. The prognosis of ACC depends on several factors including the primary lesion size, the anatomical localization, the presence or absence of metastasis at diagnosis time, the presence or absence of perineural invasion and the histopathology grade [9].

Long-term follow-up is then necessary because of the high risk of locoregional recurrence and distant metastasis, which represent a surgical and systemic therapy challenge.

4. Conclusions

Adenoid cystic carcinoma is a very unpredictable tumor occurring in the 4th to 6th decade in the major and minor salivary gland tumors. These tumors are slow growing with a high incidence of local recurrence, regional and systemic metastasis. A delayed diagnosis can be associated with unfavorable outcomes.

For this reason, appropriate diagnostic measures, proper diagnosis, and prompt treatment are very important, in order to enable a more favorable prognosis and better quality of life.

The dentist has an important role to play in the early detection and diagnosis of malignant lesions and subsequent referral of patients to appropriate further therapy.

Conflicts of Interest

The authors declare that they have no conflict of interest.

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