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Case Report

Naso-Sinus Extension of a Large Adenoid Cystic Carcinoma

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Abstract

Adenoid cystic carcinoma (ACC) of salivary glands is a slow-growing malignant tumor, characterized by wide local infiltration, perineural spread, a propensity to local recurrence and late distant metastasis [2].

The optimal treatment is generally considered to be surgery with postoperative radiotherapy to optimize local disease control.

We report the case of a 50-year-old patient who has a right jugular swelling with a bombardment of the palate for the right.

The biopsy a diagnosed adenoid cystic carcinoma

Keywords: *Adenoid Cystic Carcinoma; Hard Palate; Swellings*

Introduction

ACC is a rare malignant tumor that affects the major and minor salivary glands the lacrimal glands, ceruminous glands and occasionally the excretory glands of the female genital tract [1].

With a reported yearly incidence of 3 - 4.5 cases per million, ACC is an uncommon tumor, accounting for about 1% of all head and neck malignancies and about 10% of all tumors of the salivary glands [2].

ACC is a relentlessly growing tumor characterized by perineural invasion and multiple local recurrences. Regional lymph node metastases are conventionally regarded as rare, but these may be under-recognized due to potentially occult, clinically undetectable cervical metastases [2].

Current treatment recommendations for ACC include complete surgical resection and postoperative radiation therapy.

The recurrence rate of ACC is variable in the literature, with an estimated rate of 42% [1].

ACC is characterized by indolent evolution, and patients can survive for many years [3].

We report the case of a 58-year-old patient with adenoid cystic carcinoma of the right palate.

Case Report

This is a 58-year-old patient with no particular pathological history. The history of the disease dates back to 7 months by the appearance of a progressive right nasal obstruction which became bilateral with episodes of epistaxis and anosmia.

The clinical examination finds a patient in good general condition. Exo-oral examination revealed facial asymmetry with right jugular swelling (Figure 1).

The swelling was of firm consistency; there was no palpable lymphadenopathy. The endobuccal examination showed a tumor in the hard palate, elongated from front to back. The tumor was indurated, firm, macroscopically well delineated, and did not bleed on contact (Figure 1). Otherwise the patient presents a poor dental condition. Endoscopy highlighted a tissue process of the right nasal fossa.

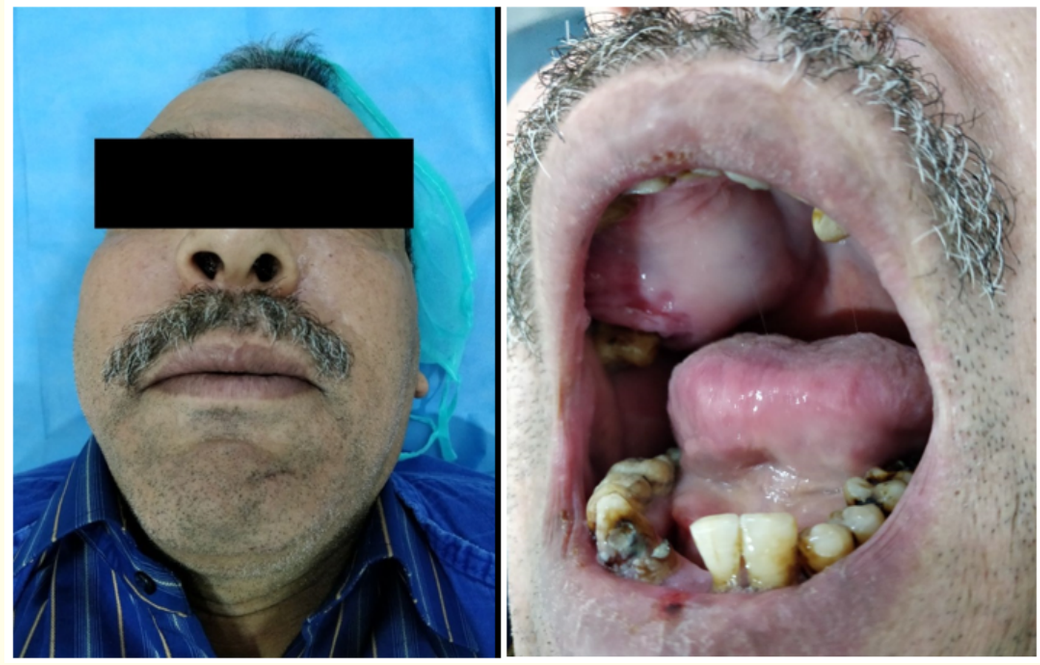

Figure 1: *Right jugular swelling on exo-buccal examination, hard palate tumefaction on endo-buccal examination.*

A computed tomography scan was requested, showing a hetero dense tissue process of the right maxillary sinus blowing the bone structures into contacts. It is heard in the left nasal fossa, by deviating the nasal septum, spreading to the ethmoid cells and coming into contact with the globe (Figure 2).

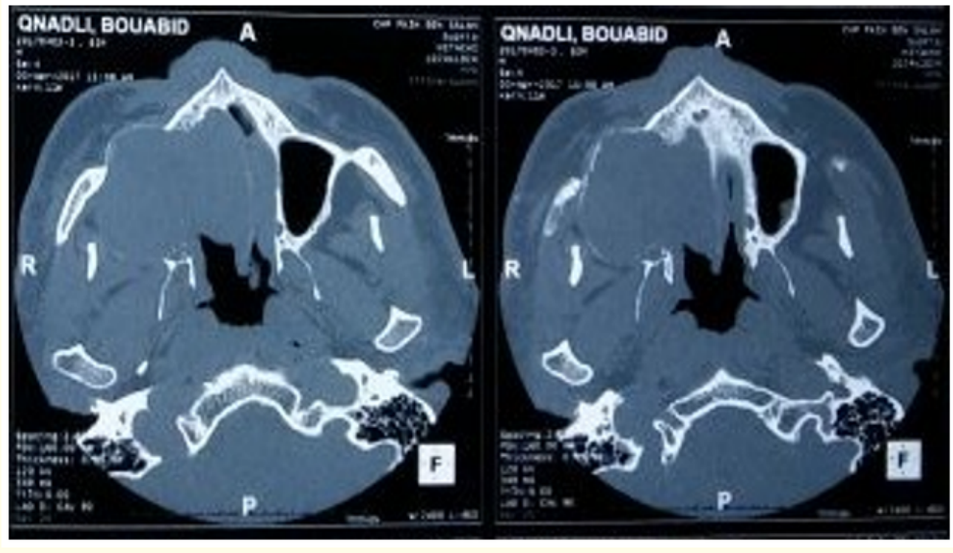

Figure 2: *CT scan shows a tissue process invading the right maxillary sinus.*

Magnetic resonance imaging showed a right maxillary sinus filling by tissue possess in iso signal T1 and hyper signal T2 with intense enhancement after injection of gadolinium product (Figure 3). A mass biopsy was done that diagnosed an infiltrant cystic adenoid carcinoma

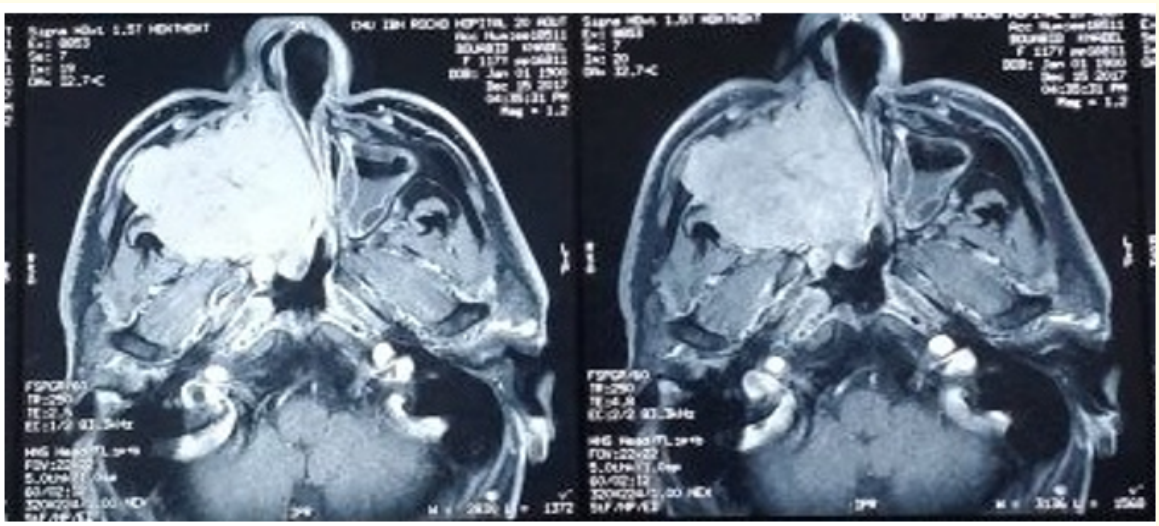

Figure 3: *MRI, T1 sequence shows intense enhancement after injection of gadolinium product of a process invading the right maxillary sinus.*

After the patient's consent, the treatment proposed consisted of a large surgical excision of the tumor under general anesthesia, followed by radiotherapy and a dentomaxillary prosthesis (Figure 4). At 3 months of supervision the patient is well.

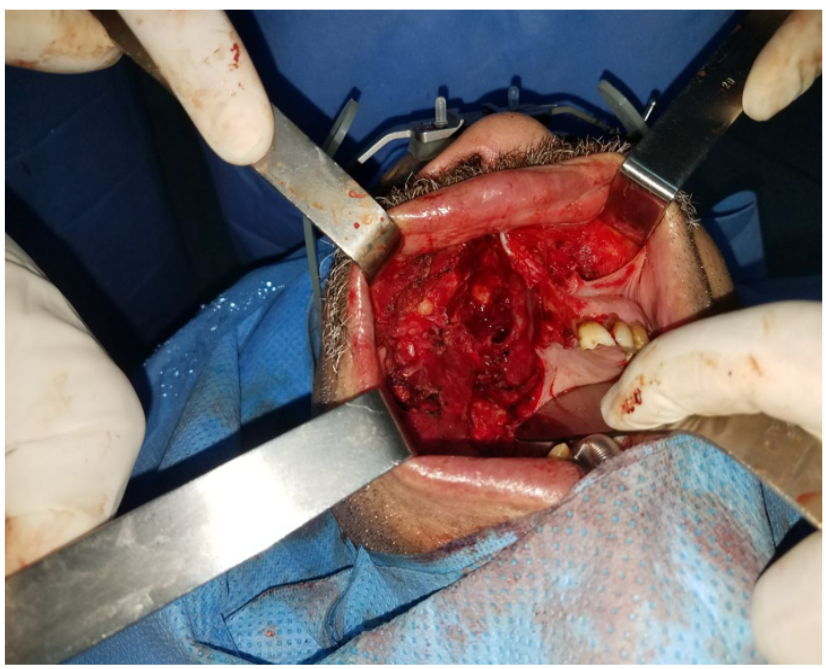

Figure 4: *Resection of the tumor with a right hemimandibulectomy.*

Discussion

ACC is one of the common and best recognized salivary gland malignancies [2]. The term "adenoid cystic carcinoma" was introduced by Ewing in 1954,this tumor was named as "cylindroma" earlier by Billroth in 1859 because the epithelial and connective tissue elements formed a system of intertwining cylinders, the term "basalioma" was coined by Krompecher in 1908, who considered this type of tumor to be of analogous nature to the basal cell growths of the skin, it is a relentless tumor that is prone to local recurrence and eventual distant metastases [2].

ACC is a rare tumor of the head and neck region; however, it is the most common malignant tumor of minor salivary glands of the palate, as in our case [1].

The tumor affects men and a woman equally, usually occurs in the fifth decade of life [1], the age of our patient joins the literature.

Clinical presentation is often as an asymptomatic mass, however, this tumor is more likely than others to present with pain or paresthesia and numbness, this tumor has a marked tendency to invade nerves and is seen in about 80% of all cases [1]. Nerve infiltration was not objectified in our case.

ACC has an aggressive long-term behavior, with persistent and recurrent growth pattern and late onset of metastases resulting in frequent eventual death It has been described as one of the most biologically destructive and unpredictable tumors of the head and neck [3].

In the study of Nascimento., *et al.* 98% of patients reported a mass, 48% had pain, 30% had ulceration, and one patient had facial nerve paralysis; these symptoms had been present from 1 month to 4 years. The presenting symptoms of ACC vary according to the site of disease. The location of the palate is frequent in the form of a mass [3].

Confirmatory diagnosis of ACC is primarily based on the characteristic histological features which play a significant role not only in diagnosing the tumor but also helps to determine treatment and prognosis [1].

Histological, ACC is composed of a mixture of epithelial and ductal cells which are arranged in three characteristic patterns - cribriform, tubular and solid - with most tumors being composed of a mix of these [2].

Radiological investigations, especially CT scans are very important to study the tumor and to make an assessment of extension, and to look for eventual recurrences after the surgery.

Treatment of ACC is influenced by location of the tumor, stage at diagnosis and biologic behavior as reflected in histologic grade, The gold-standard treatment for ACC, that is deemed as potentially resectable after extensive workup is radical surgical resection, ensuring free margins, and postoperative radiotherapy [3].

Distant metastasis is the most common presentation of treatment failure, the lung is by far the most common site of metastasis, with the liver being the second most common site, bone metastases usually indicate a fulminant clinical course, another unusual feature of ACC is that unlike most carcinomas, it seldom metastasizes to regional lymph nodes [2].

ACC has well-known prognosis profile, the 5-year survival rate after effective treatment is 75%, but long-term survival rates are low (10 years - 20% and 15 years - 10%), postoperative radiotherapy combined with more aggressive surgeries increases the long-term survivals to 30 - 40% range [2-7].

Conclusion

The early diagnosis of adenoid cystic carcinoma remains the key to ensuring correct management with prevention of distant metastases. The therapy involving combination of surgery and radiotherapy remains the modality of choice in most cases.

Informed Consent

The patient gave us informed consent for publication.

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