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# Adenoid Cystic Carcinoma of the Nasal Fossa: A Case Report and Review of the Literature

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Adenoid cystic carcinoma; Nasal cavity; Treatment

#### 1. Abstract

Adenoid cystic carcinoma of the nasal cavity is a rare malignancy. It is a slowly progressive disease, characterized by local recurrences and distant metastases and requires early and adequate management. We report a case of Adenoid cystic carcinoma of the nasal fossa and will discuss the diagnostic and therapeutic characteristics in the highlight of a review of the literature.

## 2. Introduction

Adenoid cystic carcinoma (ACC) is a rare malignancy accounting for 1% of head and neck tumors and 10–15% of salivary tumors [1]. Localization in the nasal cavities remains rare. Its slow and painless growth often delays the diagnosis, which requires early, adequate and multidisciplinary management and oncological excision with wide and healthy excision margins, the only guarantee of optimal local control of the tumor [2].

## 3. Observation

This is a 52-year-old patient, with no notable pathological history and who has been presenting for more than a year since her first consultation, a feeling of nasal obstruction with clear left rhinorrhea, with some episode of scant epistaxis on the while evolving in a context of conservation of the general state. The naso-fibroscopy examination shows a tumoral process of the lateral wall of the left nasal fossa which completely fills it, hard on palpation, non-bleeding on contact. The patient underwent a facial MRI which revealed

a tumor occupying the entire left nasal cavity, invading the ipsilateral maxillary sinus with bone lysis without cervical adenopathy (Figure 1).

A biopsy of the tumor was made with anatomopathological study confirming adenoid cystic carcinoma. The patient was operated with excision of the tumor and left maxillectomy.

Anatomopathological examination of the surgical specimen confirms a cribriform CAK with the closest healthy limits at 5 mm. The patient received postoperative external radiotherapy on the tumor bed and the naso-sinus cavities at a dose of 66 Grays, at a rate of 2 Gy per session and 5 sessions per week (Figures 2 and 3). The patient is in good locoregional and remote control with a follow-up of 16 months.

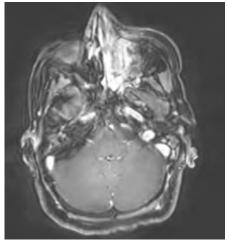


Figure 1: IRM faciale en coupe axiale T1 avec injection de gadolinium

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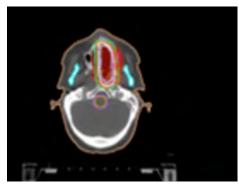


Figure 2: contourage des volume cibles et OAR

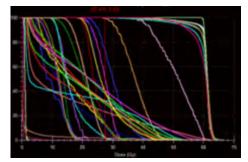


Figure 3: Histogramme dose volume DVH

## 4. Discussion

CAK is a rare malignant tumor, representing 1% of malignant head and neck tumors and 10% of salivary gland tumors. The localization at the level of the nasal cavities is exceptional. It usually occurs around the fifth decade [1] with a slight female predominance [2]. No risk factor for CAK is found in the literature and no association between the development of CAK and HPV human papillomavirus has been demonstrated [3].

Clinically, most of the initial clinical manifestations of CAK of the nasal cavities are nonspecific. It presents with a symptomatology common to any process filling the nasal cavity, such as nasal obstruction, rhinorrhea, epistaxis, anosmia. The diagnosis is often made at an advanced stage because it is a slowly growing, insidious and painless tumor [4].

On the radiological level, MRI is more sensitive than CT for assessing local extension and better detecting perineural extension. There is no specific tumor signal on CT or MRI. CAK results in an expansive tumor mass that is well or poorly limited, willingly heterogeneous. However, because of its nervous tropism, the perineural extension tracts must systematically be sought on MRI since they are present radiologically in 60% of patients with CAK [5].

Histologically, the CAK is a tumor made up of tubular cells and modified myoepithelial cells. Three morphological profiles exist which can be associated with each other, with a predominance of one of the three profiles: cribriform, tubular and solid [6].

Imaging is a valuable aid in the diagnostic process, the remote extension assessment must be exhaustive in search of cervical lymphadenopathy or distant metastases. It must include an abdominal cervico-thoracic CT scan. The PET-scanner alone is not the reference examination for the extension assessment and the follow-up of a CAK of the nasal cavities.

Therapeutically, tumor excision surgery associated with postoperative adjuvant radiotherapy is the gold standard in the treatment of CAK of the nasal cavity [7-9].

Although this approach remains the treatment of choice, exclusive radiotherapy, in inoperable cases, offers a valid therapeutic alternative to guarantee good control of the disease.

The surgery must be as wide as possible to obtain healthy resection margins even if there is no obvious consensus concerning the limits and depth of resection [10]. However, even incomplete excision in very locally advanced tumors improves overall survival compared to non-operated patients receiving non-surgical treatment. Cervical lymph node dissection is not systematic given the low lymphophilia of CAKs [11,12].

Postoperative radiotherapy is systematic for patients with risk factors for local recurrence, the presence of deep infiltration (bone, cartilage, muscle, etc.), the presence of clinical or radiological lymph node metastasis, a high-grade tumor, positive or insufficient resection margins (<5mm), or an advanced tumor stage [5,13].

It is recommended to perform adjuvant radiotherapy using an intensity modulation technique with an irradiation dose of between 60 and 70 Gy on the tumor bed, depending on the quality of excision and lymph node status, administered in fractions of 1 .8 to 2 Gy per day, 5 days per week [7-9,14]. The perineural invasion specific to CAK makes it necessary to include in the irradiated volumes the paths of the adjacent cranial nerves up to their emergence from the base of the skull. In the current state of knowledge, there is no argument to recommend systematic prophylactic treatment of cervical lymph node areas in patients without adenopathy.

Radiotherapy by proton therapy and carbon ions are therapeutic options in development for inoperable CAKs, resistant to conventional radiotherapy or with macroscopic remnants [15,16].

Due to the low rates of response to chemotherapy, it is not recommended to offer concomitant adjuvant radio-chemotherapy in the curative treatment of CAKs in the nasal cavity [4,14].

Brachytherapy appears to be a treatment option for recurrent or inoperable CAKs [17]. The implanted dose depends on the tumor volume, with a target margin of 0.5-1 cm, it varies between 100 and 160 Gy.

Systemic treatments have shown little efficacy in the treatment of CAKs. The choice of treatment should be guided by the patient's comorbidities, therapeutic history and the toxicities of the various chemotherapies. Based on the data available in the literature, no standard chemotherapy regimen could be recommended [18].

The advent of immunotherapy also makes it possible to broaden the therapeutic possibilities in the treatment of CAKs and studies Volume 10 Issue 9 -2022 Case Report

have shown the efficacy of Nivolumab and Pembrolizumab in locally evolved or recurrent CAKs alone or as adjuvants [1,19,20].

# 5. Prognosis

The 5-year prognosis of CAK of the nasal fossa varies according to the TNM stage, histological subtype, therapeutic possibilities and locoregional control [21,22].

Being a slowly progressive disease with frequent recurrences and sometimes late metastases, long-term follow-up of patients is necessary. Monitoring must be long, clinical and paraclinical: an ENT examination with cervical palpation and naso-fibroscopy, an MRI of the facial bones every 6 months then every year and a thoracic-abdominal CT scan every year [22].

### 6. Conclusion

CAK of the nasal cavity is a rare malignant tumor. The reference treatment remains wide excision surgery followed by postoperative adjuvant radiotherapy. Targeted therapies and immunotherapy remain a promising area of research. Long-term follow-up is recommended because of the late onset of local and distant recurrences.

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