# Trigeminal neuralgia revealing adenoid cystic carcinoma of the infra-temporal fossa

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## ABSTRACT

**Objective**: Adenoid cystic carcinoma (ACC) is a rare malignant neurotropic tumor characterized by slow growth, frequent local recurrences, and high incidence of distant metastasis. We report a case of trigeminal perineural invasion revealing a rare location of ACC and we review through literature its clinical features and therapeutic modalities.
**Observation**: we report a case of a male presenting a right facial neuralgia. The anamnesis revealed recent right nasal obstruction with a reflex right ear pain. Clinical examination revealed altered sensation in the right V1 and V2 territories. A sub-mucosal tumoral process was discovered at the nasal endoscopy. CT scan and MRI showed an aggressive process of the infra-temporal fossa. The biopsy concluded to the diagnosis of adenoid cystic carcinoma.
**Conclusion:** Local invasion and perineural spread of head and neck ACC worsen prognosis and can alter the surgical options and therapeutic approach. Detection on imaging is vital to allow the patient to undergo appropriate treatment.
**Key words:** Trigeminal neuralgia, Adenoid cystic carcinoma, Infratemporal fossa

## INTRODUCTION

Adenoid cystic carcinoma (ACC) is a rare malignant tumor representing about 1% of all malignant tumor of the oral and maxillofacial region [1]. Perineural spread in the head and neck region is frequently associated with neurotropic tumors. ACC, followed by squamous cell carcinomas (SCCs) are the tumors with the highest prediction of nerve invasion [2].
We report a case of trigeminal perineural invasion revealing a rare location of ACC and we review through literature its clinical features and therapeutic modalities.

## OBSERVATION:

A 43-year-old North African male presented a 7-month history of right facial neuralgia, associated recently with right nasal obstruction, hyposmia, reflex right otalgia and intermittent headaches. His past medical history does not report recurrent naso sinusal infections or bleeding.
Clinical examination revealed altered sensation to light touch over the territory of the right V1 and V2, with preserved facial and ocular muscular functions. Nasal endoscopy showed a deviation of the nasal septum on the left with a submucosal tumor process opposite the sphenopalatin foramen. No palpable cervical lymph nodes were found.
CT-scan showed a tumoral process centered on the right pterygo-palatal fossa to the pterygomaxillary cleft and the pterygoid canal causing lysis of the postero-medial and postero-superior walls of the right maxillary sinus, as well as the base of the skull (Figure.1).

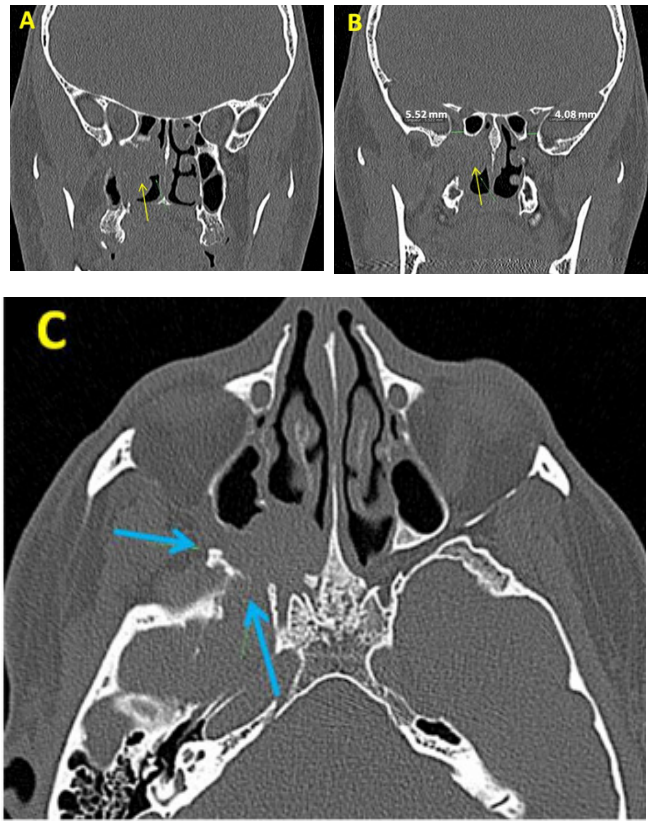


**Figure 1**: Coronal (A and B) and axial (C) CT scan showing Tumor process (A, B, thin arrow) centered on the right pterygo-palatal fossa with extension to the nasal cavity responsible for enlargement of the superior orbital fissura (B, fine line) measured at 5.52 mm in the right side Vs 4.08 mm in the left one) as well as the base of the skull (C).

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Biopsy concluded to adenoid cystic carcinoma. No distant metastasis was assessed on the pelvic thoraco-abdominal CT scan.
Exclusive radiotherapy was indicated but died 2 years after the end of treatment.

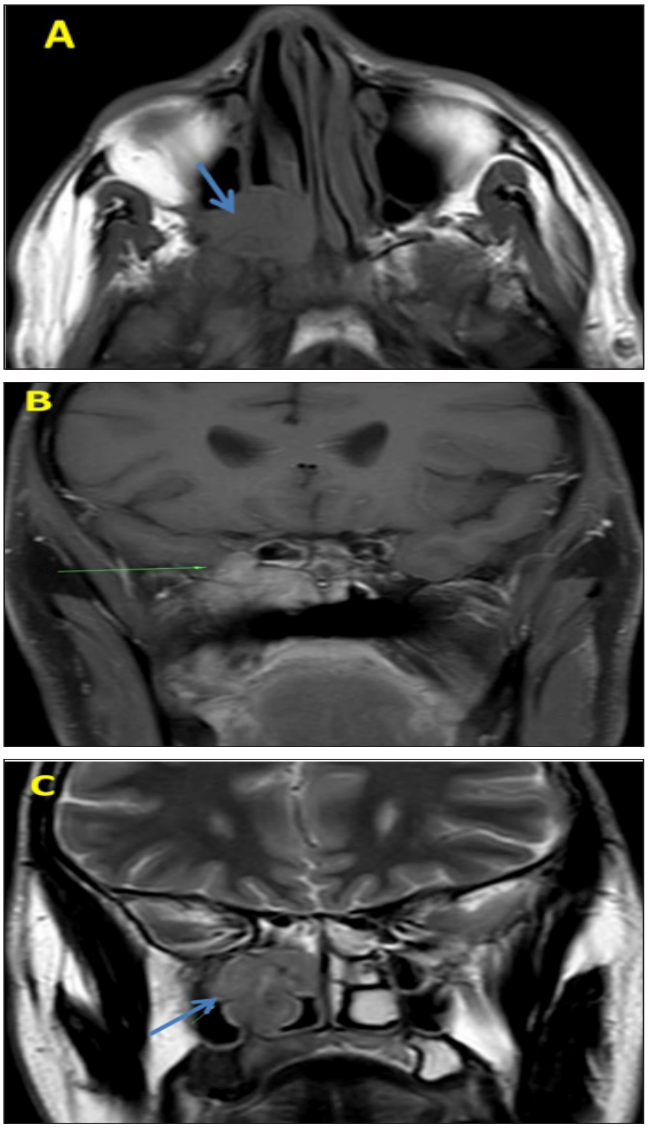


**Figure 2**: MRI in T1 axial section (A) and T1 Gado sequence in coronal section (B), T2 coronal section (C) showing an infiltrative tumor process (thick arrow)centered on the right pterygo-palatine fossa which is in discrete hyposignal T1, hypersignal T2 intensely enhanced after injection of gadolinium, with endocranial extension (Thin arrow)

MRI showed an infiltrative tumoral process centered on the right pterygo-palatine fossa appearing in low-T1 signal, high T2 signal, and intensely enhanced after injection of gadolinium, with endocranial extension evoking a malignant tumor (Figure 2).

## DISCUSSION:

ACC is a rare tumor representing fewer than 10 to 22% of all salivary gland neoplasms [3]. It mostly affects adults in the 4th–6th decades and occurs more frequently in females [4].

ACC is the most common cancer in minor salivary glands, where the proportion of ACC ranges from 32% to 71%. ACC is most commonly found in the palate, followed by the paranasal sinuses (14–17%) and other sites of the oral cavity. Localization at the infra-temporal fossa (FIT) is rare [4].
ACC is the most common primary tumor to spread via the perineural mechanism with an incidence of perineural spread widely variable and ranging from 20% to 80% [1, 5, 6]. In fact, Perineural invasion is defined as tumor cell invasion in, around, and through the nerves. Some authors use the terms perineural invasion and spread interchangeably, and others refer to invasion as the microscopic process and spread as the macroscopic result seen on imaging such as MRI [7]. Perineural spread (PNS) of head and neck tumors most commonly occurs along the branches of the trigeminal and facial nerves [4, 6].
Clinical signs and symptoms depend on the site of the lesion. In the nose and paranasal sinuses, nasal obstruction, deep facial pain, epistaxis and eye symptoms are frequent. In our case, the revealing symptom was trigeminal neuralgia. The maxillary (V2) and mandibular (V3) branches of the trigeminal nerve are most frequently involved in perineural spread and allow a route of tumor infiltration into the pterygopalatine fossa (PPF), Meckel's cave, and the cavernous sinus [7].
According to literature, the time between onset of neurological signs and symptoms, and the time of diagnosis, ranges between few months to three years [4]. Seven months was the time consultation for our patient.
In terms of imaging, literature revealed that MRI was the optimum imaging technique due to its high soft-tissue contrast; nevertheless it is not specific for distinguishing ACC from other head and neck primary tumors [7]. Routine use of intravenous gadolinium and fat-suppression sequences aids detection of locoregional spread. Axial and coronal planes are most useful for assessing skull base involvement. Unenhanced T1-weighted (T1W) images are important for assessing fat planes and fat-containing structures such as the pterygopalatine fossa and bone marrow for low signal tumor infiltration [8, 9].
Primary ACC tumor can have a variable appearance at MRI. It can present as a well-defined mass or an ill-defined mass with diffuse infiltration of surrounding structures with a tendency to invade fat and bone. ACC typically enhances homo-geneously, although heterogeneous enhancement and necrosis can also be a feature of this tumor. The signal on T2W is variable and lower signal is habitually described in the solid form and carries a worse prognosis [7].
Compared to CT, MRI has been proven to be more sensitive for detecting perineural spread. Sensitivity ranged from 95 to 100%. Although, some authors [6] showed that CT and MR images were found to be equally useful for the detection of PNS of ACC in the oral and maxillofacial regions.

CT is complementary to MRI for assessing local bone changes such as widening of the skull base foramen [7].
Treatment of ACC is influenced by location of the tumor, stage at diagnosis and biologic behavior as reflected in histologic grade [6]. Many authors confirmed that the gold standard treatment of head and neck ACC is based on complete surgical resection. Only surgical removal with clean surgical margins or radiotherapy treatment can reduce the possibility of recurrence and metastasis in cervical lymph nodes, lungs, bones and brain [10].
Postoperative radiotherapy is recommended for patients with perineural invasion and inclusion in the irradiated volumes of the paths of the adjacent cranial nerves until their emergence from skull base is required [4, 11].
In our case, the tumor was deemed unresectable and the patient had exclusive radiotherapy after a multidisciplinary committee meeting.
In patients with adverse prognostic factors, with poorly controlled disease or affected with symptomatic metastases, chemoradiotherapy using various agents may be considered. Targeted molecular therapies has been extensively studied in patients with advanced ACC, but the rather indolent course of the disease makes it difficult to observe clinical responses [4].
Prognosis of ACC of the head and neck is rather poor and many clinicians assume that cure is never achieved in ACC [4]. In fact, the incidence of PNS in patients with head and neck cancer dramatically alters the treatment and prognosis since it is associated with local recurrence and higher incidence of positive margins [5, 6].
The risk of distant metastases is high (48%), it can usually occur within 8 years of surgery.
Lymphatic spread is rare. Hematogenous spread appears often in the course of the disease. Metastases to the lung are more frequent than regional lymph node metastasis [1].
According to Ouyang et al [12], patients with local-regional recurrence show a 10-year worse survival rate comparing to those with no local-regional recurrence (32.5% vs 72.2%).
In case of recurrence, a chemotherapy treatment is debatable. Recent studies showed a low response rate.

## CONCLUSION:

We report a case of ACC arising from the infratemporal fossa invading the pterygoid canal and maxillary sinus as well as the base of the skull. Local invasion and perineural spread of head and neck ACC is frequent andworsen prognosis and can alter the surgical options and treatment approach. Early diagnosis of ACC results in better quality of life and higher survival rate.

### Compliance with ethical standards

**Conflict of interest:** The authors stated that there is no conflict of interest.
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