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Case Report

Nasopharyngeal adenoid cystic carcinoma presenting with exophthalmos: A case report ☆

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ABSTRACT

Adenoid cystic carcinoma is a malignant tumor involving the salivary glands, rarely developing in the nasopharynx. It is a slowly evolving entity with strong local aggressiveness and a high tendency to recurrence. We report the case of 23-year-old patient with adenoid cystic carcinoma of the nasopharynx presenting with exophthalmos in which radiation therapy is the sole therapeutic option.

Introduction

Formerly called “cylindrome” by Bilroth in 1856, adenoid cystic carcinoma is a malignant tumor involving the salivary glands, nasopharyngeal localization is uncommon with a slow-growing but locally aggressive, and therefore, subject to recurrence. Another important feature is its tendency to infiltrate neural structures and spread perineurally [1]. We report the case about this rare localization with intracranial and orbital extension.

Case report

A 23-year-old patient, with no particular history, who gradually complained a year ago of headaches and right earaches, resistant to symptomatic treatment, aggravated a month ago by the appearance of a right exophthalmos. Orbitocerebral magnetic resonance imaging (MRI) was performed.

Axial MRI T2-weighted (Fig, 1a) T1-weighted contrast-enhanced FAT suppressed sequences (Fig. 1b) demonstrates an irregular thickening of the right posterolateral wall of the nasopharyngeal in homogeneous isosignal T2, heterogeneous enhancement which demonstrates internal cystic components associated with infiltration of the right parapharyngeal space, ptéroigoid muscle pterygopalatine fossa and choana.

☆ Competing interest: No conflict of interest.

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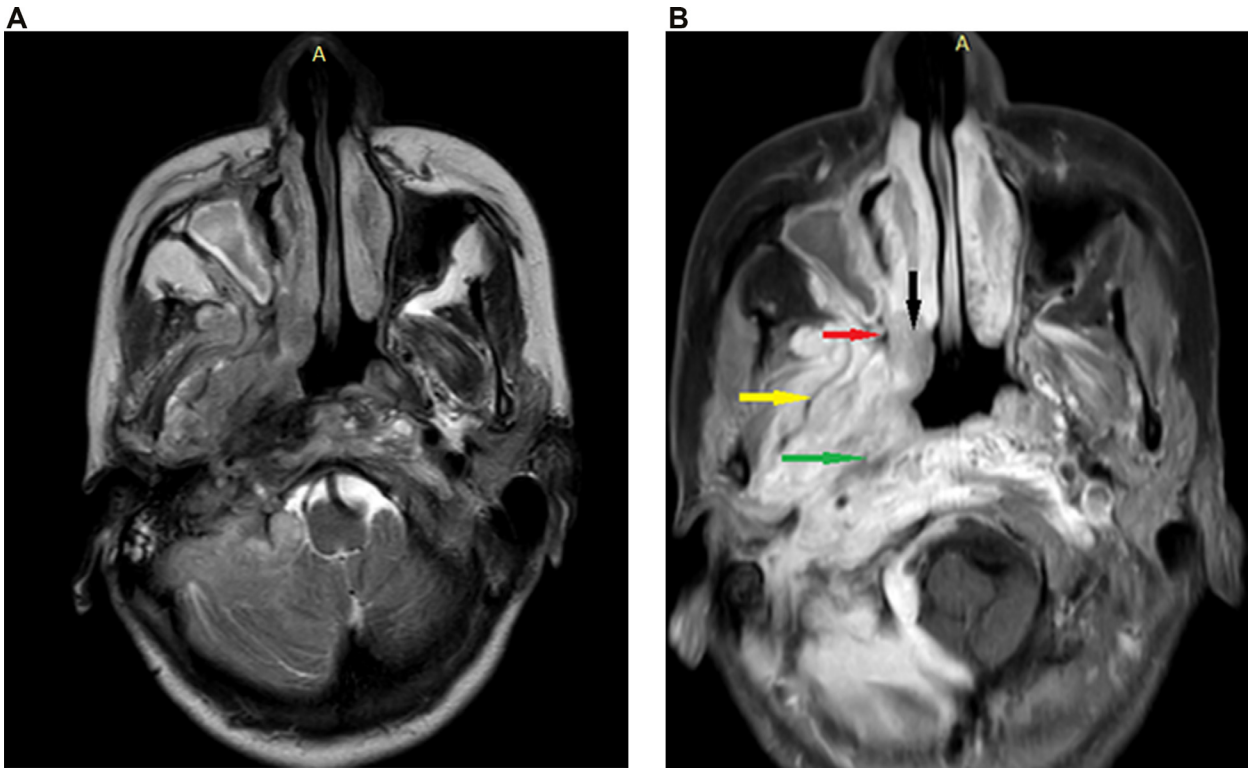

Fig. 1 – Orbitocerebral MRI, Axial T2 weighted (a) T1-weighted contrast-enhanced FAT suppressed sequences (b) demonstrates an irregular thickening of the right posterolateral wall of the nasopharyngeal in homogenous isosignal T2, heterogeneous enhancement with demonstrates internal cystic components associated with infiltration of the right parapharyngeal space, green arrow) ptéroigoid muscle (yellow arrow), pterygopalatine fossa (red arrow) Choana (black arrow). (Online version of figure is available online.)

MRI axial (Fig, 2a) and coronal (Fig. 2b) of T1-weighted contrast-enhanced FAT suppressed sequences showing the extend of the tumor through the inferior orbital fissure into the orbit associated with the invasion of the right internal carotid artery, cavernous sinus, bilateral optic nerve marked in the right with homolateral grade 3 exophthalmos. The tumor demonstrated an intracranial extension, in the right temporal lobe responsible of a temporal herniation.

The nasopharyngeal biopsy performed twice the first came back negative. The second biopsy with an immunohistochemical study concluded to an adenoid cystic carcinoma with cribriform and tubular patterns of the nasopharynx (Fig. 3).

Locally advanced irregular thickening of the right posterolateral wall of the nasopharyngeal with orbital extension responsible for grade 3 right exophthalmos and cerebral extension with temporal herniation in whom radiation therapy is the sole therapeutic option

Discussion

Adenoid cystic carcinoma of the nasopharynx is a rare entity accounting for 0.13%-0.48% of nasopharyngeal tumors [2]. it is characterized by slow evolution, but it is locally aggressive and has a high tendency to recurrences. Epstein-Barr virus (EBV) does not appear to be involved in the pathogenesis in this histological type [3]. It causes local infiltration and neural invasion tends to involve into aggressive local infiltration and to extend along the cranial nerve canal, toward the orbital cavity, skull base, making all surgical approaches very difficult and delicate [3]. Adenoid cystic carcinoma occurs primarily in adults, with a peak incidence in the fourth to sixth decade of life [4]. Also, it was more common in women in the literature [5]. The most common symptoms are epistaxis, progressive nasal stenosis, dysfunction of the Eustachian and, in relation to of the base of the skull, ocular motility disorders, diplopia, facial pain, dysfunction of the IX, X, XI, and XII cranial nerve pairs and, more rarely, Horner syndrome [6].

Histologically, these tumors are classified into 3 types with different prognosis: tubular (50% of cases), cribriform (30%), and solid (less than 10%). The presence of areas of necrosis, a solid component, or perivascular and perinervous invasion is histological factors predictive of a poor prognosis [7]. In our clinical case, it was a cribriform and tubulotrabecular form that seemed to have a more favorable prognosis.

MRI is fundamental in the initial extension assessment, making it possible to precisely characterize the local, perineural, and lymph node tumor extension as well as for post-treatment bone control. As for computed tomography scan, it mainly allows the study of bone destruction.

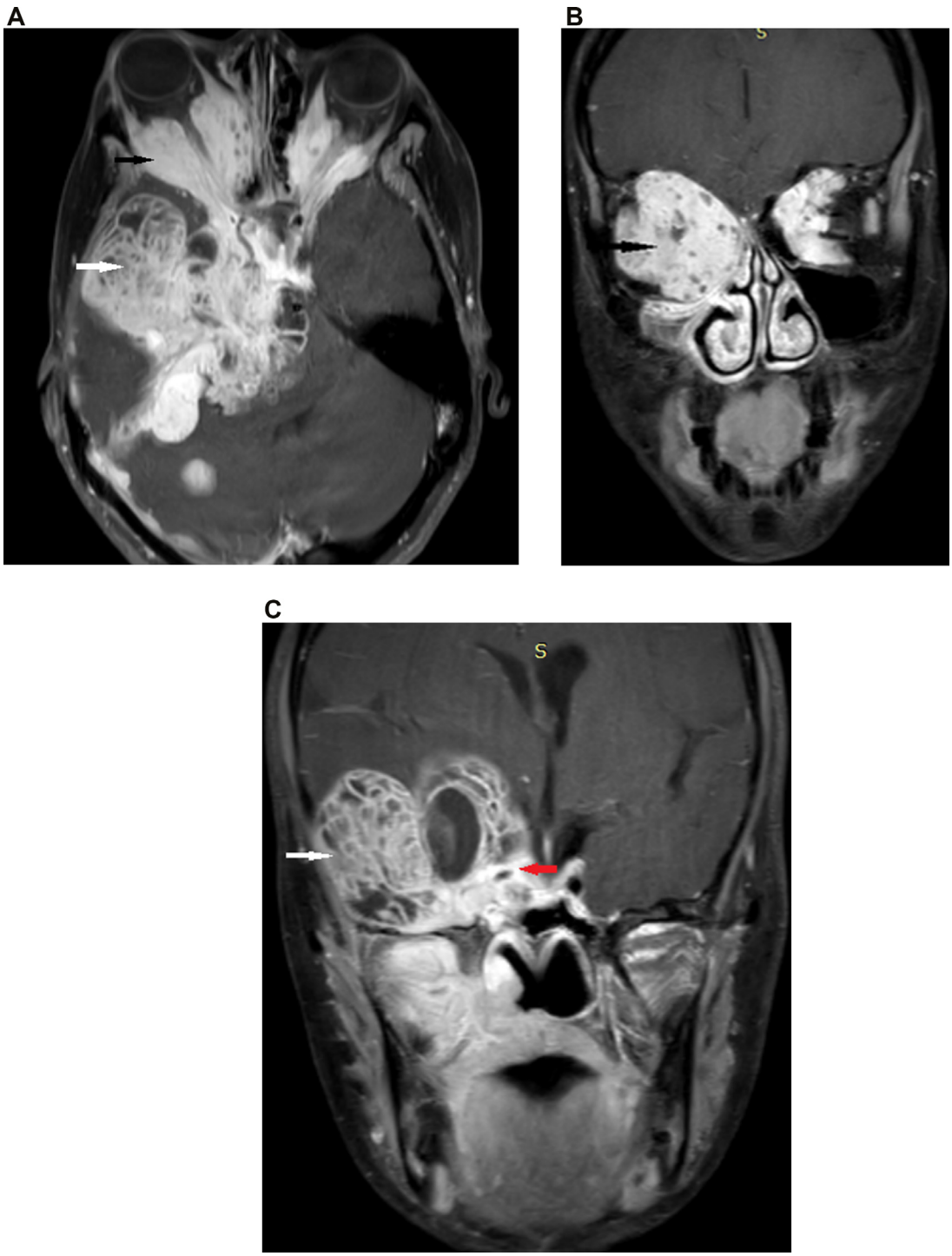

Fig. 2 – Orbitocerebral MRI, Axial (a) and coronal (b) of T1-weighted contrast-enhanced FAT suppressed sequences showing the extend of the tumor through the inferior orbital fissure into the orbit associated to the invasion of the right internal carotid artery, cavernous sinus, (red arrow) bilateral optic nerve marked in the right with homolateral grade 3 exophthalmos (black arrow). The tumor demonstrated an intracranial extension, in the right temporal responsible of a temporal herniation (white arrow). (Online version of figure is available online.)

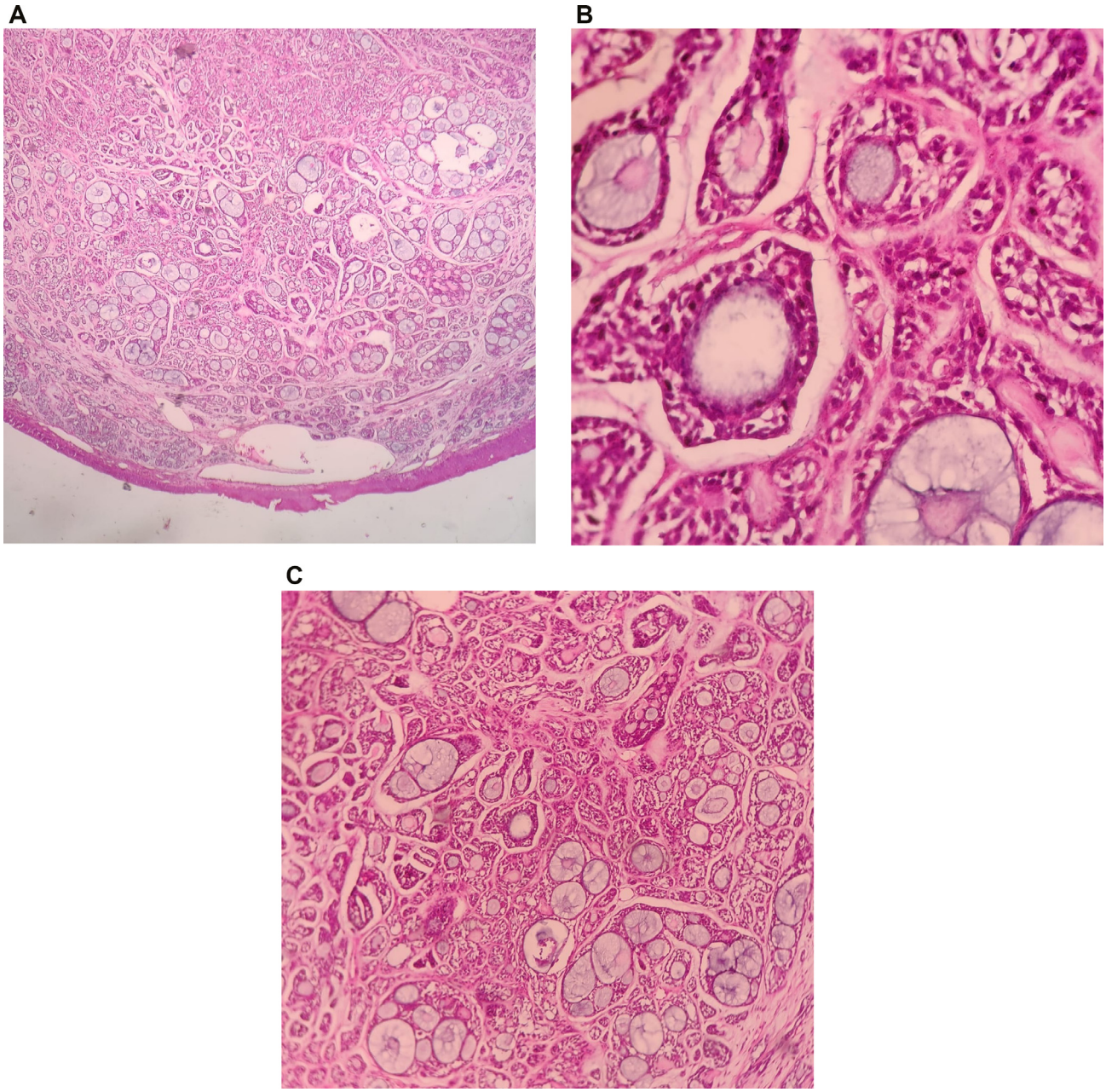

Fig. 3 – (a) Carcinomatous tumor proliferation made up of cribriform, tubular and pseudo cystic structures (HE ×100). (b) Structures bordered by cells with essentially myoepithelial differentiation (HE ×400). (c) Cribriform and tubular structures viewed at medium magnification. (HE × 200).

Dissemination by the lymphatic route is fairly low with a rate of distant metastases comparable to that of other nasopharyngeal tumors. When these occur, they mainly affect the lungs (70%), the liver, and the bone [8].

Regarding management, surgical treatment is indicated for tumors classified as stage I, II, and III. However, the particular anatomical site of the nasopharynx as well as the infiltrating and extensive nature of the tumor makes complete resection in healthy margins is often illusory [9]. Adjuvant radiotherapy is indicated to improve the rate of local control in unresectable forms, radiotherapy makes it possible to decrease tumor volume and reduce symptoms [10].

Conclusion

Adenoid cystic carcinoma of the nasopharynx with intracranial and orbit extension remains a rare pathology, revealed in our patient by exophthalmos in whom radiation therapy is the sole therapeutic option

Patient consent

The patient declares his consent for the production of his case

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