ADENOID CYSTIC CARCINOMA OF THE NASOPHARYNX: A CASE REPORT WITH REVIEW OF LITERATURE

CARCINOME ADÉNOÏDE KYSTIQUE DU NASOPHARYNX: A PROPOS D'UN CAS AVEC REVUE DE LA LITTÉRATURE

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ABSTRACT

Introduction: Adenoid cystic carcinoma (ACC) rarely develops in the nasopharynx. Compared with other nasopharyngeal tumors, it is characterized by a slow progression, a local aggressivity and a high tendency to recurrence.

Case presentation: We report the case of a 71-year-old patient diagnosed with locally advanced ACC of the nasopharynx treated by concomitant radio-chemotherapy.

Conclusion: Clear consensus is not available for the most appropriate management strategy for nasopharyngeal ACC.

Key words: Adenoid cystic carcinoma; Nasopharynx; Radiotherapy; Chemotherapy

RÉSUMÉ

Introduction: Le carcinome adénoïde kystique (CAK) se développe rarement dans le nasopharynx. Comparativement aux autres tumeurs du nasopharynx, il se caractérise par une progression lente, une agressivité locale et un fort potentiel de récidive.

Observation: Nous rapportons le cas d'un patient âgé de 71 ans diagnostiqué avec un CAK localement avancé du nasopharynx traité par radio-chimiothérapie concomitante.

Conclusion: Un consensus clair n'est pas disponible concernant la stratégie de gestion la plus appropriée pour le CAK nasopharyngé.

Mots clés: Carcinome adénoïde kystique; Nasopharynx; Radiothérapie; Chimiothérapie,

INTRODUCTION

Adenoid cystic carcinoma (ACC) of the nasopharynx is a very rare histological type accounting for less than 1% of all nasopharyngeal tumors [1]. This tumor is characterized by local aggressiveness, slow progression, and distant metastasis [2]. Due to the rarity of this tumor and the lack of clinical studies, there is no consensus regarding therapeutic strategies [1].

The objective of our work is to discuss, through this case with a review of the literature, clinical particularities, therapeutic and evolutionary features of this tumor type.

CASE REPORT:

We report the case of a 71-year-old hypertensive male patient, previously operated for a frontal meningioma. He presented with a 6-month history of headaches without epistaxis or other associated rhinological signs. The patient also reported a recent right-sided hearing loss. Nasal endoscopy showed a suspicious tumor process in the posterior wall of the nasopharynx lateralized on the right, bleeding on contact with filling of the right Rosen Müller fossa (Fig.1 A). Otoscopy showed a right seromucous otitis (Fig.1B). On cervical examination, there was no palpable cervical lymphadenopathy. Neurological examination did not reveal any involvement of the cranial pairs. An endoscopic-directed biopsy was performed. The final pathological examination substantiated the presence of the cribriform pattern of nasopharyngeal ACC.

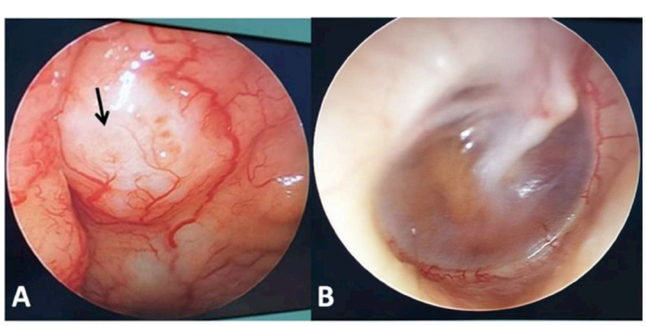

Figure 1: A-Nasal endoscopy showing a bulging of the posterior wall of the cavum lateralized to the right in relation to a CAK of the cavum B-Aspect of right otitis media

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CT scan and Magnetic resonance imaging (MRI) including unenhanced axial, sagittal, and coronal T1-weighted sequences, axial T2-weighted sequences, and contrast-enhanced axial, sagittal, and coronal T1-weighted sequences, showed a tumor process in the upper and lateral right wall of the nasopharynx filling the Rosen Müller fossa and obstructing the right eustachian tube with lateral extension to the right infra temporal fossa and invasion of the medial and lateral pterygoid muscle; an invasion of the right foramen lacerum and ovale and of the cavernous sinus with osteolysis of the carotid canal and envelopment of the carotid artery in its intra petrous and intra cavernous portions (Fig.2).

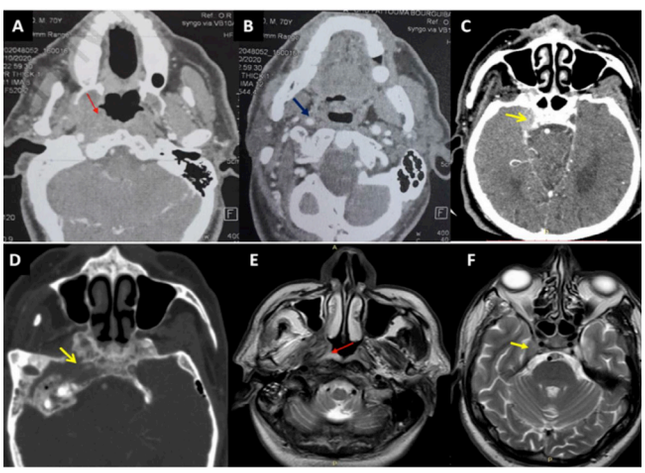

Figure 2: CT (A, B, C, D) of the facial mass in the parenchymal window after injection of contrast product (A, B, C) and bone window (D) and MRI of the facial mass in T2 sequence fat sat (F, E) passing through the cavum showing on axial sections a tumor process of the upper and right lateral wall of the nasopharynx filling the fossa of Rosen Muller and obstructing the eustachian tube on the right (red arrow) with extension in lateral to the FIT medial and lateral pterygoid muscle invasion (red star). We noted an invasion of the lacerum and ovale foramen and of the right cavernous sinus (arrow yellow) and a sheath of the carotid in its intra petrous and intracavernous portions (blue arrow).

A thoracic-abdominal-pelvic CT scan and a bone scintigraphy were performed and did not reveal any secondary lesions. The tumor was classified as T4N0M0. The patient underwent concomitant radio-chemotherapy. Our patient received external beam radiotherapy (conventional 3D radiotherapy) with a total dose of 70 Gy. Radiation was delivered in 2-Gy daily fractions. Regarding chemotherapy drug used, it was a combination of cisplatin and 5-fluorouracil. After 2 years of follow-up, nasal endoscopy and MRI (Fig.3) suspected tumor recurrence, which was confirmed by biopsy. The patient is currently presented to our multidisciplinary consultation meeting.

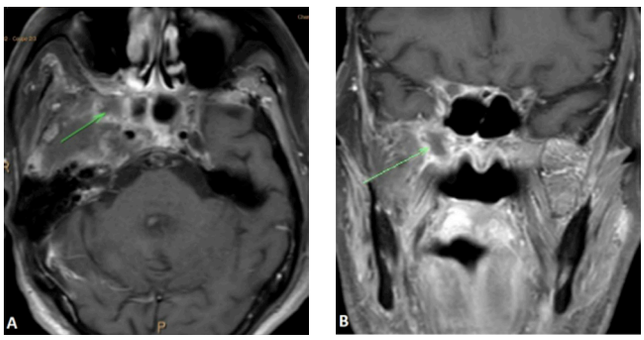

Figure 3 (A,B): Follow up MRI of the neck passing through the cavum showing a tumor process of the right lateral wall of the nasopharynx (green arrow).

DISCUSSION

Nasopharyngeal ACC is a rare entity, accounting for 1% of nasopharyngeal tumors [1]. It has a very slow clinical course, often leading to a late diagnosis [1,3]. A female predominance has been noted [4]. In our case, it was a male patient. In fact, Dong et al reported 18 cases with ACC of The nasopharynx including 8 male and 10 female patients [5]. Onset of the disease usually occurs between the third and sixth decades [5]. Epstein-Barr virus (EBV) does not seem to play a role in its pathogenesis [1,2]. This histological type is characterized for its high local aggressiveness wi th frequent peri-neural invasion and involvement of the cranial nerves that may extend to the orbital cavity and skull base [4,6]. The peri-neural extension is along the V2, V3 respectively via the foramen round and ovale, cavernous sinus or trigeminal ganglion via the foramen lacerum [7].

There is a low rate of lymphatic dissemination, not exceeding 15% [4, 8]. Our patient had no cervical nodes despite an advanced tumor stage. The incidence of distant metastases is comparable to that of other nasopharyngeal tumors [8,9].

The revealing symptoms are not very specific. It may include epistaxis, unilateral nasal obstruction, signs of tubal dysfunction, diplopia, or headache [5]. Nevertheless, the painful character and the presence of neurological symptomatology should draw attention. Nasal endoscopy with guided biopsies plays a vital role in the establishment of an early diagnosis, and, therefore, an accurate and timely treatment planning. There are three histological subtypes of ACC: tubular, cribriform and solid [8]. Our patient had a cribriform subtype.

Differentiating ACC of the nasopharynx from nasopharyngeal carcinoma is difficult. The only way to identify the disease is histological study. MRI is the optimal imaging technique for the study of cavum tumor pathology. Special characters on MRI images of ACC of the nasopharynx,are a valuable aid to diagnosis.

In T2-weighted sequences, the signal is variable according to the histological type [10]: the cribriform form is reflected by a hyper signal whereas the solid form is reflected by a hyposignal [10]. In addition, as seen on MRI images, ACC of the nasopharynx is characterized

by a high incidence of perineural invasion, frequent and aggressive local infiltration, and infrequently, lymph node metastasis. Therefore, the main interest of MRI lies in the detection of this peri-neural extension thanks to its high spatial resolution. MRI is superior to CT in the determination of peri-neural extension especially in asymptomatic patients [11].

The complex anatomical location of the nasopharynx and the infiltrative and extensive nature of this tumor make a complete resection with healthy margins often impossible.

Primary carcinomas of the nasopharynx are usually treated with concomitant radiotherapy or radiochemotherapy [12]. Surgical treatment such as endoscopic or external nasopharyngectomy has a place in localized carcinoma, which is considered less sensitive to radiotherapy, but is associated with several difficulties and high morbidity [12]. Adjuvant radiotherapy is indicated to improve the local control rate [4]. In unresectable forms, radiation therapy can decrease tumor volume and reduce symptoms [2]. Concomitant chemoradiation combining platinum with a taxane has shown some benefit in terms of local control and seems to be an interesting therapeutic option in locally advanced inoperable forms [11], as in our patient's case. Recent data indicate that a high proportion of ACCs stain positively for epidermal growth factor receptors (EGFR) and may respond to agents that act as EGFR inhibitors [12].

The overall response rate with monotherapy varies between 6% and 70% in favor of cisplatin. Multidrug therapy combining platinum derivative, doxorubicin and 5-fluorouracil achieves better response rates but at the cost of higher toxicity and without benefit in terms of overall survival [13].

The evolutionary potential of ACC requires long-term monitoring. The prognosis of nasopharyngeal ACC remains favorable. Five-year survival rates are 54.8% to 100% versus 50.6% to 71% for other common forms of nasopharyngeal carcinoma [4]. However, patients with advanced tumors, including intracranial involvement or skull base invasion had bad prognosis [13]. Solid component of the tumor, advanced tumor stage, and peri-neural invasion [3,4].

This observation emphasizes the rarity of adenoid cystic carcinoma in this location, and underlines the difficulties of management of locally advanced cases as well as the management of recurrences.

CONCLUSION

ACC of the nasopharynx remains a rare entity. The lack of randomized clinical trials explains the lack of consensus and the complexity of the management of these tumors. A better molecular understanding could lead to the emergence of new targeted therapies.

Compliance with ethical standards

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