Nasopharyngeal adenoid cystic carcinoma, suggestion of therapeutic innovations: A case report and review of literature

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ABSTRACT

Introduction: Nasopharyngeal adenoid cystic carcinoma is a rare tumor. Compared with others nasopharyngeal tumors, it is characterised by slow evolution but it is locally aggressive and has a high tendency to recurrences. Due to the rarity of cases, no consensus exists about treatment approaches.

Presentation of the case: We report the case of a 55-year-old-woman, with a locally advanced adenoid cystic carcinoma. The patient was operated by endoscopic surgery, received radiation and had a good objective response. The follow-up showed no local recurrence after one year.

Conclusion: The aim of this work is to review the literature concerning this rare malignancy, and discusses treatment approaches in initial situations and during recurrences. We supported the interest of the intraoperative neuronavigation system for surgical safety.

1. Introduction

Adenoid cyst carcinoma (ACC) is a rare tumor entity and represents approximately 1% of all carcinomas of the head and neck [1]. It is a malignancy of the nasopharynx, characterised by local infiltration and neural invasion. ACC tends to be locally aggressive and to demonstrate perineural invasion. This tumor is classified as a pathological form of adenocarcinoma.

ACC of the nasopharynx has particular characteristics on MRI which aids diagnosis.

Reports on large number of patients with ACC of the nasopharynx are lacking, which means that knowledge about its extension patterns is poor. We describe a case report in our ENT department, 20 August 1953 Hospital, Casablanca, Morocco.

This work has been written in accordance with the SCARE criteria [2].

2. Case report

It is about a case of 55 years old woman, living in Casablanca-Morocco, who presented in our ENT department for bilateral nasal obstruction, associated to hearing loss and otalgia, predominant on the left ear, with severe headache, continuous fatigue and deterioration of the general state. No cervical lymph nodes were found at the physical examination. No past medical or surgical history was notified.

Endoscopic examination was requested to objectify a brownish irregular mass measuring about 2 cm, depending on the nasopharynx, protruding at the left choanae (Fig. 1).

The biopsy and immunohistochemical study showed an adenoid cystic carcinoma of the nasopharynx (Fig. 2).

Facial MRI showed lesional process at the posterior wall of the nasopharynx in hyposignal T1, heterogeneous hypersignal T2, enhancing heterogeneously after injection of gadolinium.

On the other hand, this process moves toward the choanae and buds at the level of the left nasal cavity, in the inferior nasal cone. Laterally, it fills the fossa of Rosenmüller, close to the torus tubarius, filling them up.

At the top and back, this process extends to the clivus and the prevertebral muscles without an anomaly of detectable signal within them. Below, it buds in the oropharynx. Bilateral cervical adenopathies, the most voluminous of them is 11 mm long (Fig. 3).

The patient was scheduled at the operating room 2 weeks later.

The surgery was performed by a Professor in our department and was very instructive for residents. A "U-shaped" incision was made in the palate, 5 mm away from the maxillary alveolus to spare the greater palatine artery (Fig. 4). It was extended laterally and posteriorly to the anterior tonsillar pillar to give the flap adequate length. The flap was raised in the subperiosteal plane and reflected forward, with the soft

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palate centrally detached, exposing the edge of the hard palate and giving access to the nasopharynx. The posterior edge of the bony palate was “back bitten” and shortened with a drill and burr, providing circumferential control of the nasopharyngeal mass (Fig. 5).

A 3 cm mass was removed. The definitive biopsy confirmed the adenoid cystic carcinoma type. The histological sections on the choanae and the posterior wall of the nasopharynx are reached. (Fig. 6).

In view of the proximity of skull base, Neuronavigation system has been use for the safety of the surgical procedure. No anesthetic and surgical incidence occurred. She went out after 2 days with antibiotic treatment and local antiseptic with a nasogastric tube for 10 days.

She was sent to the radiotherapy department for supplemental care. She received 70 Gy of radiation, divided into 35 sessions. She had a good objective response and she was very satisfied of the result.

The follow-up showed no local recurrence after 1 year clinically, and after radiological check up.
Due to earlier diagnosis and masses in the neck, that appear much earlier in patients with NPC, ACC which develop primarily in the parotid gland or the palate, have site are thought to be the prognostic factors. It has been said that treatment.

Also, it is low sensitivity to radiotherapy and chemotherapy hinders the tumor resection without compromising patient safety. It well be useful, it is allows surgeons to optimise the resection to achieve better control of bleeding. In case of severe haemorrhage, muscle patch is the best way to control this complication. Padhve and al. studied 8 cases of internal carotid injury in endoscopic surgery, muscle patch was used to control haemorrhage, no deaths and neurological sequelae were noti ed [17]. Other advantages include less surgical time, less hospitalization time, no visible scars, avoiding complications such as epiphora, dysesthesia, trismus, and craniofacial deformities [13,14].

Therapeutic strategy depends on the pathophysiological characters of ACC, which tends to show local infiltration and neural invasion. A retrospective study suggested that surgery combined with radiotherapy should be performed on these patients [3]. Liu and Wen suggest that adjuvant radiotherapy improve the rate of local control [3,7]. Cao and al. propose that, in unresectable forms, the radiotherapy allow to decrease the tumoral volume and reduce the symptoms [15].

No evidence exists that chemotherapy would improve the prognosis of ACC patients after radiotherapy.

Concomitant chemoradiotherapy based on platinium alone, in combination with a taxane, has shown some benefit in terms of local control and appears to be an attractive therapeutic option in locally inoperable, locally challenged forms [16]. The addition of cetuximab to chemoradiotherapy has lead, in a phase II study, to two partial responses and to two complete responses [10].

Our patient didn't receive chemotherapy.

The prognosis of adenoid cystic carcinoma of the nasopharynx remains favorable. Survival rates at 5 years are 54.8%–100% compared to 50.6%–71% for other common forms of nasopharyngeal carcinomas [3].

Regarding to the rarity of this histological type, the number of cases included in the studies remains low for a multifactorial analysis of the prognostic factors. The solid component of the tumor, the advanced tumor stage and the perineural invasion appear to be associated with an unfavorable prognosis [3].

4. Conclusion

Adenoid cyst carcinoma of the nasopharynx is very rare. No consensus exists about treatment approaches. The diagnostic is endoscopic, histologic and molecular. MRI is the most efficient exam to study local invasion of the mass. Currently, the main treatment is total resection by

3. Discussion

Nasopharyngeal adenoid cystic carcinoma is a rare tumor and represents approximately 1% of all carcinomas of the head and neck [1]. It is classified as a pathological form of adenocarcinoma. Compared with others nasopharyngeal tumors, it is characterised by slow evolution but it is locally aggressive and has a high tendency to recurrences. Due to the rarity of cases, no consensus exists about treatment approaches. The tumor usually progresses slowly which results in delayed diagnosis. Epstein-Barr virus (EBV) doesn't seem to be involved in the pathogenesis of this histological type [3]. It is characterised by local infiltration and neural invasion. Adenoid cystic carcinoma (ACC) tends to involve into aggressive local infiltration and to extend along the cranial nerve canal, toward the orbital cavity, skull base, making all surgical approach hard and delicate [3,4]. Magnetic resonance imaging (MRI) is a particular method of interest because it detects perineural infiltration and bone marrow changes by providing satisfactory morphological information of the structures. It has been widely used for diagnosing and monitoring the response of treatment for carcinomas of the head and neck.

Extensions to the skull base and the deep facial space can be well illustrated using MR images. Navigation system during surgery may allow to decrease the tumoral volume and reduce the symptoms [15].

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Fig. 6. The mass of the nasopharynx after excision.
surgery, using endoscopic approach combined with radiotherapy. The endoscopic navigation system allows surgeons to optimise the resection to achieve tumor resection without compromising patient safety. Suggestions of therapeutic innovations were reported.

Reports on large number of patients with ACC of the nasopharynx are lacking, which means that knowledge about its extension patterns is poor.

Ethical approval

Written informed consent for publication of their clinical details and/or clinical images was obtained from the patient.

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Author contribution

Mohamed Moutaa Tatari: Corresponding author writing the paper.
Said Anajar: writing the paper.
Jawad Hasnaoui: writing the paper.
Khadija Salama: writing the paper.
Reda Abada: Correction of the paper.
Sami Rouadi: correction of the paper.
Mohammed Mahtar: correction of the paper.

Conflicts of interest

The authors declare having no conflicts of interest for this article.

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Consent

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