Case Report

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Nasopharynaeal Adenoid Cystic Carcinoma, A Rare Disease: A Case Report and Review of Literature

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ABSTRACT

Nasopharyngeal adenoid cystic carcinoma is a rare tumor. Compared with other nasopharyngeal tumors, it is characterized by slow evolution but is locally aggressive and has a high tendency to recurrence. Due to the rarity of cases, no consensus exists about treatment approaches. We report the case of 42-year-old man with a locally advanced adenoid cystic carcinoma. The patient received concurrent chemoradiation and had a good objective response. After one year, he developed lung metastasis and received platin-based chemotherapy. The aim of this work is to review the literature concerning this rare malignancy and discusses treatment approaches, both for initial management and locoregional/metastatic relapses.

Keywords : Adenoid Cystic Carcinoma ; Nasopharynx; Chemoradiation; Hadron Therapy; Prognostic

Introduction

Adenoid cystic carcinomas of the head and neck are rare. These tumors are characterized by their slow but aggressive evolution. Nasopharyngeal localization of adenoid cystic carcinoma accounts for less than 1% of all nasopharyngeal tumors. Given the rarity of this tumor group and the absence of clinical studies, there is no consensus on the therapeutic strategy. We report an observation about this localization.

Observation

A 42-year-old man, with no particular pathological history, a former smoker, presented for consultation with the persistence of right otalgia and the appearance of nasal obstruction. A cervicofacial MRI showed an expansive process of the right nasopharynx, involving the left peristaphilin muscles of the petrous apex, the right sphenoid sinus. The tumor was classified as T4N0M0. A nasofibroscopy and biopsy with histological and immunohistochemical study confirmed the diagnosis of solid adenoid cystic carcinoma. Within two months of diagnosis, the patient received concurrent chemoradiotherapy, using an intensity-modulated conformal radiation therapy (IMRT) technique. The total dose delivered in the macroscopic tumor volume was 70 GY in 34 fractions and 50 days. The anatomoclinical target volume of the nodal areas received on both sides 65 GY in zones 2 and 50 GY in zones 3, 4, and 5. Chemotherapy consisted of two cycles of 100mg/m2 of cisplatin at three-week intervals. At the end of this treatment, the disease was considered in complete clinical and radiological remission. After one year of follow-up, slowly evolving micronodules appeared. Two and a half years after the initial diagnosis, faced with symptomatic progression with the appearance of other bilateral lung lesions, chemotherapy based on cisplatin 75mg/m2 and doxorubicin 50mg/m2 every three weeks was initiated. After three courses, the clinical and radiological evaluation of the lesions showed progression. Due to the deterioration of the patient's general condition, supportive care was offered. The evolution is marked by the death of the patient 6 months later.

Discussion

Adenoid cystic carcinoma of the nasopharynx is a rare entity representing 0.13 to 0.48% of nasopharyngeal tumors, histologically classified into 3 types (tubular (50%), cribriform (30%) and solid (10%) [1]. It presents a very slow clinical evolution, often leading to a late diagnosis. This entity is characterized by great local aggressiveness with the predominance of perineural invasion and damage to the cranial nerves which can extend to the orbital cavity and the base of the skull, making any surgical approach very delicate [2,3]. There is a low rate of lymphatic dissemination, not exceeding 15% in the various series reported [4]. The incidence of distant metastases is comparable to that of other nasopharyngeal tumors. The therapeutic management of locoregional disease (stages 1, 2 and 3) is based on surgery. However, the complex anatomical location of the nasopharynx and the infiltrating and extensive nature of this tumor makes a complete resection at clear margins often illusory. Adjuvant radiotherapy is indicated to improve the rate of local control. For unresectable forms, radiotherapy reduces tumor volume and reduces symptoms [5]. Concomitant platinum-based chemoradiotherapy alone or in combination 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. Our patient also responded very well to concomitant platinum-based chemoradiotherapy with complete remission [6]. A new therapeutic approach for the locally advanced stages and in the case of local recurrence is hadron therapy.

The rationale for hadron therapy with protons and carbon ions stems from ballistic properties that allow precise targeting of tumor volume while preserving healthy tissue [7]. With regard to metastatic disease, the efficacy data with regard to chemotherapy, mainly come from small series of phase 2 studies that included adenoid cystic carcinoma of the head and neck, in the global sense. The overall response rate with monotherapy varies between 6 and 70% in favor of cisplatin. Polychemotherapy combining platinum salts, doxorubicin, and 5FU achieves better response rates but at the cost of higher toxicity [8]. As for the use of anti-EGFR targeted therapy in this particular histological group, cetuximab tested in a phase 2 study in 14 metastatic patients in combination with 5FU and cisplatin showed 5 partial responses. The prognosis depends on the histological type, thus the solid type has an unfavorable prognosis. In case of tumor without a solid component, the 15-year survival rate is around 39% and if the solid component is predominant, the 15-year survival is 5%.

Conclusion

Adenoid cystic carcinoma of the nasopharynx remains a rare entity and a difficult tumor to treat given its invasive character and its high tendency for local and metastatic recurrences. The lack of randomized clinical studies explains the lack of consensus and the complexity of the management of these tumors. Considering the radiosensitive nature, new irradiation technique have appeared with encouraging results. Ongoing studies for a better molecular understanding could lead to the emergence of new targeted therapies for the management of this orphan neoplasia.

Conflicts of Interest

The authors declare no conflicts of interest in relation to this article.

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