Nasopharyngeal adenoid Cystic carcinoma: About tow cases and review of the literature

O. Masbah, MD[1], D. Filali, MD[1], I. Bekkouch, MD[1], K. Rami, MD[1], N. Mellas, MD[1], N. Ismaili, MD[1], L. Kanouni, MD[1], K. Hassouni, MD[1], T. Kebdani, MD[1], N. Benjaafar, MD[1], Bk. Elgueddari, MD[1]

(1) Department of radiotherapy, Faculty of Medicine of Rabat, University Mohamed V, Morocco.

✉ *Corresponding Author: Dr Ouafe Masbah, MD*
Department of radiotherapy, Faculty of Medicine of Rabat, University Mohamed V, Morocco.
E-mail: masbahouafae1979@yahoo.fr

Key words: Adenoid Cystic Carcinoma, Nasopharynx, Surgery, Radiotherapy, Chemotherapy.

Submitted: March 2010 - Accepted: 30 May 2010

ISSN: 2070-254X

Abstract

Nasopharyngeal adenoid Cystic carcinoma is a rare tumour; few cases were reported in the literature. It is characterized by slow growth rate, high tendency to local recurrence and metastatic spread. The corner stone of its therapeutic management is surgery when is possible, unfortunately because of difficulty of access to the nasopharynx; surgery is more often limited; so radiotherapy seems to remain the ideal treatment. The chemotherapy is indicated in the metastasis situations.

We report two cases aged respectively: 50, and 46 years old. The first patient presented with diffuse pulmonary metastasis; the treatment combined chemotherapy, based on cisplatin and adriamycin drugs, and palliative radiation therapy at the dose of 30 Gy. The patient died 6 months after diagnosis. The second patient, presented with locally advanced disease, was treated by external beam radiotherapy at the dose of 70 Gy. 36 months after the end of the treatment, she presented a local recurrence treated by re-irradiation of the nasopharynx at the dose of 60 Gy. The patient is still alive without disease 12 months after the treatment completed.

Introduction

Previously termed "cylindroma" by Bilroth in 1856, the adenoid cystic carcinoma (ACC) of the nasopharynx is a rare disease with a slow growing, but locally aggressive and thus prone to recurrence. Another important characteristic is its tendency to infiltrate neural structures and to spread perineurally [10]. We report two cases about this rare localization.

Case 1: A 50 years old female admitted to our Hospital with headache and epistaxis for 5 months duration. Otorhinolaryngological examination showed a nasopharyngeal tumour extended to the oropharynx without palpable lymph nodes. Computed tomography scan of the head and neck showed a big tumor on the right side wall of the nsopharynx. This tumour was extended to the right pterygoïd fossa and towards the sphenoïdal sinus, and filling the ethmoïdal cells, without endocranial extension (fig1). The nasopharyngeal biopsy revealed invasive adenoid cystic carcinoma (solid type). The radiological assessment of the disease found diffuse pulmonary metastasis (fig 2). The patient was managed with combined chemotherapy based on cisplatin 100mg/m ² and doxorubicin 50mg/m ² repeated every 3 weeks. However, the assessment of the disease after 2 cycles of chemotherapy showed tumour progression in the nasophrynx and in the lung. Then, the treatment consisted of palliative nasopharyngeal radiotherapy (30Gy) in 10 fractions, and best supportive care. The patient died six months later with severe haemoptysis.

Case 2: A 46 years old female without any medical history, admitted to our institution with nasal obstruction, epistaxis and right hypoacousia during 8 months. Otorhinolaryngological examination showed tumour of the right superior-side wall of the nasopharynx without palpable adenopathy. The CT scan of the head and neck revealed a tumour at the nasopharyngeal superior and right walls (fig 3). Biopsy of the nasopharynx found adenoid cystic carcinoma infiltration. Radiological assessment of tumour with chest radiography and abdominal ultrasound imaging showed no abnormalities. The patient was treated successfully with radiotherapy (70 Gy) in 35 fractions. However, she presented a local recurrence 36 months later. Then, the management consisted of reirradiation at the dose of 60 Gy in 30 fractions. The patient is still alive without evident disease 12 months after the treatment completed.

Discussion

Adenoid cystic carcinoma (ACC) is a malignant tumour of the exocrine glands. It most commonly arises in the salivary glands, even if localizations have been described in prostate, lacrymal glands, uterine cervix, breast and bronchial mucosa [7]. Nasopharyngeal localization is uncommon, accounting for 0.5% to 4% of all nasopharyngeal carcinomas and for 2.4% to 3.7% of all head and neck ACC [1].

In the nasopharynx, ACC occurs most frequently in the 5th decade of life, without sex predilection [5]. The symptoms most commonly found are epistaxis, progressive nasal stenosis, dysfunction of the Eustachian tube and, in relation to the invasion of the skull base, disorders of ocular motility, diplopia, facial pain, dysfunction of IX, X, XI and XII pairs of cranial nerves and, more rarely, Horner's syndrome [8].

Imaging of ACC is based on computed tomography (CT) scan, particularly helpful in detecting bony erosions of the skull base, and on Magnetic Resonance Imaging (MRI) with gadolinium, effective in demonstrating possible involvement of infra-temporal fossa, cavernous sinus, and perineural or

perivascular infiltration [8].

Histoligically, three subtypes have been described by Perzin and al [5] in 1978, reflecting various degrees of progression of cellular differentiation, as well as aggressiveness of biologic behaviour. The tubular subtype, which is the most differentiated form and the 2nd most frequent (30%), presents a recurrence rate more than 50% and an overall 9-year survival. The cribriform subtype, which is less differentiated than the tubular and the most common (50%) for most authors, presents a 90% recurrence and an 8-year overall survival. The cribriform and tubular subtypes manifest a tendency for local infiltration. The less differentiated and thus the most malignant and aggressive is the solid subtype, accounting for 10% of the cases. It often gives distant metastases (70% overall) and mainly invades lung, brain and bones and has by far the poorest prognosis [5].

As Adenoid Cystic Carcinoma has low sensitivity to Radiotherapy [4,6], surgical treatment is the main treatment policy for patients with limited NACC mainly in stage I, II, and III [2,9]. Wen et al. [10] showed the 5-year OS of the surgical treatment group was 50.0%, which was significantly higher than that of the radiotherapy group (38.5%). Mendenhall et al.(16) studied the treatment results of 101 patients with head and neck adenoid cystic carcinoma, and he found the 10-year rates of local control of the radiotherapy group and the surgery plus radiotherapy group were 43% and 91%, respectively; moreover, the 10-year absolute survival rates were 42% and 55%, respectively. Therefore, the optimal treatment policy for patients with NACC may be surgery plus radiotherapy. Patients with incomplete resection or with advanced tumors had bad prognosis. However, recent study from Schramm and Imola [9] had demonstrated that combined extensive surgical resection and radiation therapy might achieve survival outcomes in nasopharyngeal ACC comparable with treatment results reported about Adenoid Cystic Carcinoma of other sites in head and neck region. Chemotherapy has a limited role in the ACC of the nasopharynx and its use is still a matter of discussion. Cisplatin, 5-Fluorouracil, Doxorubicin and others are used in combination with radiotherapy, with reports of some success and remissions [5].

Conclusion

Nasopharyngeal adenoid Cystic carcinoma is rare. Its biological behaviour is characterized by slow growth, high tendency to local recurrence and metastatic spread. Its histological features are particularly important for prognostic prediction: solid pattern has the worst outcome. The corner stone of its therapeutic management is surgery. Radiotherapy improves the local control of the disease and chemotherapy is helpful in treating the metastatic disease.

References

1. Aloulou S, Merad-Boudia Z. (2002) Adenoid cystic carcinoma (cylindroma) of the nasopharynx with extension into the cavernous sinus. Presse Med ; 31:1653-6.

2. Dai.T, Satoshi and all (2001): clinical study of adenoid cystic carcinoma of the head and neck. Auris Nasus Larynx 28: S99-S102

3. Mendenhall WM et al.(2004) Radiotherapy alone or combined with surgery for adenoid cystic carcinoma of the head and neck. Head Neck; 26:154–162

4. Mendenhall WM, Morris CG, Amdur RJ, Werning JW, Hinerman RW, Villaret DB. (2004) Radiotherapy alone or combined with surgery for adenoid cystic carcinoma of the head and neck. Head Neck; 26:154–162.

5. Papadas T, Chorianopoulos D, Mastronikolis N. (2007): Nasopharyngeal adenoid cystic carcinoma: a rare nasopharyngeal tumor. European Review for Medical and Pharmacological Sciences; 11: 55-57

6. Pfeffer MR, Talmi Y, Catane R, Symon Z, Yosepovitch A, Levitt M. (2007) A phase II study of Imatinib for advanced adenoid cystic carcinoma of head and neck salivary glands. Oral Oncol; 43:33–36.

7. Saadi I, El Marfany M, Hadadi K, Amaoui B, Kebdani T, Errihani H, et al. (2003) Adenoid cystic carcinoma of the nasopharynx: a case report. Cancer Radiother; 7: 190-4.

8. Soprani F, Armaroli V, Venturini A, Emiliani E, Casolino D (2007). A rare case of adenoid cystic carcinoma of the nasopharynx manifesting as Horner's syndrome: discussion and review of the literature. Acta Otorhinilaryngologica Italica; 27: 216-219

9. Tian-Run Liu et al. (2008) Adenoid Cystic Carcinoma of the nasopharynx: 27-Year Experience. The laryngoscope Nov; 118(11):1981-8

10/ Wen SX, Tang PZ, Xu ZG, Qi YF, Li ZJ, Liu WS.(2006) Therapeutic modalities of nasopharyngeal adenoid cystic carcinoma. Zhonghua Er Bi Yan Hou Tou Jing Wai Ke Za Zhi 2; 41:359–361.

Figures

Figure 1: CT scan showing a tumor at the right side wall of the nasopharynx

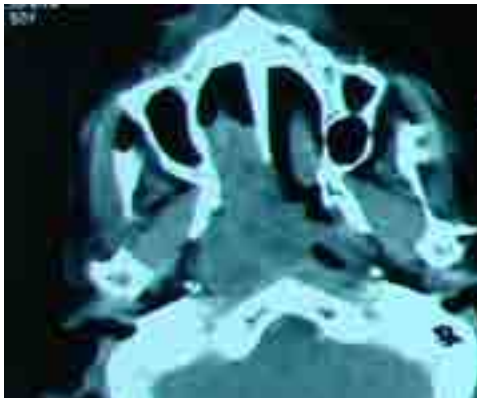

Figure 2: Chest radiography showing diffuse metastasis in both lungs

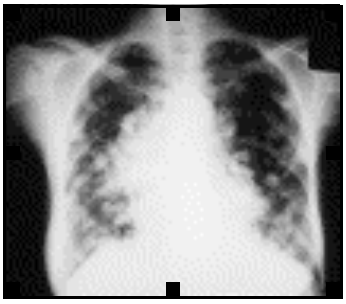

Figure 3: Axial CT showing a tumor of the right side wall of the nasopharynx