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ORIGINAL ARTICLE

Adenoid cystic carcinoma of the paranasal sinuses: Retrospective series and review of the literature

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KEYWORDS
Adenoid cystic carcinoma;
Paranasal sinuses;
Nasal cavities;
Facial pain;
Epistaxis

Summary
Objectives: Adenoid cystic carcinomas (ACC) are rare malignant tumours arising in the major and minor salivary glands. Involvement of the nasal cavity and paranasal sinuses is rare and poorly described. The purpose of this study was to define the clinical and prognostic criteria of ACC of the paranasal sinuses based on the review of a series of 25 cases.
Material and methods: Single-centre retrospective study of 25 cases of ACC of the paranasal sinuses managed between 1998 and 2011, evaluating epidemiological, clinical, diagnostic and prognostic criteria. Factors influencing survival (Kaplan—Meier/Log Rank test) and the patient's quality of life (EORTC QLQ-C30 questionnaire) were also analysed.
Results: Most patients (72%) had a locally-advanced tumour (stage T3 or T4) at diagnosis. Tumour sites, in decreasing order of frequency, were the maxillary sinus, nasal cavities and ethmoid sinus. The most common presenting complaints were maxillary pain or heaviness, unilateral blocked nose, and repeated epistaxis. When the tumour was resectable, treatment comprised a combination of surgery and adjuvant radiotherapy. The 5-year overall survival rate was 63% and the 5-year disease-free survival rate was 43%. The TNM stage at diagnosis ($P = 0.03$), the histological subtype ($P = 0.023$), the possibility of combined surgery and radiotherapy ($P = 0.03$), and local control ($P = 0.05$) were significant factors of improved 5-year overall survival. Positive surgical margins were associated with a trend towards poorer 5-year disease-free survival (ns).
Conclusions: ACC are rare malignant tumours associated with a poor prognosis, characterized by a high recurrence rate. Recommended treatment is a combination of surgery and adjuvant radiotherapy whenever possible. Five-year survival varies as a function of TNM stage, histological subtype, treatment options and local control.

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Introduction

Adenoid cystic carcinomas (ACC) are rare malignant tumours, corresponding to 5% of all paranasal sinus malignant tumours [1], which represents less than 0.15% of all malignant head and neck tumours, regardless of site and histology [2]. ACCs usually arise in the major and minor salivary glands, but can occur in all sites comprising secretory glands (breast, cervix, colon, prostate).

ACCs are tumours with slow and insidious growth, often discovered at a late and locally advanced stage. Paranasal sinus tumours are associated with a poorer prognosis and surgery of these sites is often complicated by the extent of local progression. Recurrences are frequent and occur late, sometimes many years after initial management [3].

The purpose of this study was to analyse the clinical and prognostic criteria of this rare and poorly known entity, based on the study of 25 patients with ACC of the paranasal sinuses in the light of the most recent knowledge based on a review of the literature.

Material and methods

This single-centre retrospective study analysed all cases of ACC of the paranasal sinuses and nasal cavities observed between 1998 and 2011. Cases of ACC were identified by computerized coding (C30.0 and C.31.0, 1, 2, 3, 9) of the International Classification of Diseases (ICD-10, revised in 2011). Patients with an ACC involving a site other than the paranasal sinuses and nasal cavities or with a different histopathology were excluded.

The following parameters were evaluated for each patient:

- tumour stage according to the AJCC 2007 TNM staging system taking into account the classification indicated in the patient's medical charts based on physical examination, imaging and histological examination of the operative specimen;
- histological type and subtype (tubular, cribriform, solid or mixed) according to the initial histology report when it was indicated, or retrospectively by review of the slides;
- presenting symptoms and symptoms related to post-treatment sequelae;
- initial therapeutic management, comprising the type of surgery, examination of surgical margins, any adjuvant therapy.

Local recurrence was evaluated by calculating the time to onset following completion of first-line treatment. When this interval was less than 6 months, the patient was considered to present progressive disease. Overall survival and disease-free survival probabilities were calculated according to the Kaplan—Meier method; prognostic factors were analysed by Log Rank test with a limit of significance of $P < 0.05$.

The validated standardized EORTC QLQ-C30 quality of life questionnaire (qualitative self-assessment scale of severity) was prospectively sent to 13 patients after having obtained their agreement by telephone. Only one patient did not return the questionnaire. The questionnaire was not sent to 12 patients of the study (nine deceased patients, three patients lost to follow-up). The EORTC QLQ-C30 quality of life questionnaire was used to calculate a global quality of life score: a high score indicated a good perceived quality of life, a functional score (Functional Scale): a high score indicated limited functional impairment, a general symptom score (Symptomatic Scale): a low score indicated few symptoms.

Statistical analysis was performed with SPSS 20 software.

Results

Twenty-five patients with ACC of the paranasal sinuses or nasal cavities were diagnosed and managed between January 1998 and December 2011. This series comprised 12 women and 13 men with a mean age at diagnosis of 57 ± 14.4 years. The two most recent patients of this series had a follow-up of 9 months and 12 months, respectively. All patients underwent contrast-enhanced CT-scan of the facial bones, neck and chest. MRI of the facial bones, which is now systematically requested, was not performed in seven patients (due to changing practices during the course of this retrospective series). PET-scan has become systematic in recent years as part of the initial work-up and for follow-up in the presence of signs of progression. Table 1 presents the distribution of the patients of the series according to age, tumour site, histological subtype and TNM stage.

The majority of patients (72%) had a locally advanced, TNM stage T3 or T4 tumour at the time of diagnosis. Five of the eight patients with a T4 tumour were classified as stage T4a and three were classified as stage T4b. Only five patients had a stage T2 tumour, and no T1 tumours were observed in this series. The most common tumour site was the maxillary sinus (48%), followed by the nasal cavity (24%), ethmoid sinus (16%) and sphenoid sinus (12%). Only one patient presented lymph node invasion at diagnosis, classified as N1.

The most common presenting symptoms were feelings of maxillary heaviness, unilateral blocked nose and recurrent epistaxis. Horner's syndrome and unilateral seromucous otitis were observed in two of the three patients with a tumour of the sphenoidal sinus. Pain was the symptom most commonly reported by patients (64%) regardless of tumour site.

The histological type and subtype of ACC were reported on the initial histology report for nine patients. Review of the histology slides defined the precise histological subtype for another 13 patients. The slides were not available for review for three patients (patients referred by other centres). These 22 cases of ACC comprised seven cribriform (32%), eight mixed (36%), two solid (9%) and five tubular (23%) subtypes. Cribriform and tubular subtypes were often associated and were then classified as mixed subtype.

All patients with tumours considered to be resectable after discussion in multidisciplinary meetings were treated by a combination of surgery and adjuvant radiotherapy. This initial surgical management concerned 17 patients (68%); an 81-year-old patient with poor performance status did not receive adjuvant radiotherapy. The surgical procedures most commonly performed were maxillectomy ($n = 8$) with varying degrees of resection of the ethmoid sinus or floor of the orbit, followed by paranasal maxillo-ethmoidal resection ($n = 5$), extended to the skull base when necessary ($n = 2$).