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ORIGINAL ARTICLE

Cancer of the external auditory canal

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KEYWORDS
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Summary
Introduction: Cancer of the external auditory canal is a rare tumour with an annual incidence of one per one million inhabitants. The objective of this study was to evaluate the 5-year overall survival and disease-free survival rates in a series of patients with carcinoma of the external auditory canal and to compare our results concerning the clinical presentation, management and survival with those of the literature.
Patients and method: Ten patients were included in this retrospective, single-centre study over a 20-year period. Data concerning age, symptoms, imaging, TNM stage according to the Pittsburgh classification, histology, management, sequelae, recurrences and survival were recorded.
Results: The mean age of the patients of this series was 60.7 years. Seven patients had a squamous cell carcinoma. The other histological types were undifferentiated carcinoma, adenoid cystic carcinoma and neuroendocrine carcinoma. Staging was based on the Pittsburgh classification with one stage I, one stage III and eight stage IV tumours. Five-year overall survival rates were 100%, 50% and 0%, respectively. The mean 5-year overall survival rate was 35% and the mean 5-year disease-free survival rate was 24%.
Conclusion: Carcinoma of the external auditory canal is a difficult diagnosis when the tumour does not present as a fungating mass protruding from the external auditory canal. The Pittsburgh classification was used for TNM staging of these tumours, allowing comparison of our results with those of the literature. The clinical findings and survival rates observed in this study are comparable to those reported in the literature. These tumours are associated with a poor prognosis on the basis of our results and published data.

Introduction

Cancer of the external auditory canal (EAC) is a rare tumour, representing less than 0.2% of all head and neck cancers [1]. Only 4% of cancers of the outer ear are confined to the EAC. The annual incidence of carcinoma of the EAC is estimated to be between one to six per million inhabitants [2] and the prevalence is one per million inhabitants [3,4], which explains the small sample sizes of the series published in the literature.

These tumours must be diagnosed at an early stage to avoid the mutilating surgery required at advanced stages. Otoscopy usually allows easy diagnosis of these tumours by providing direct vision of the lesion. In clinical practice, the

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lesion is usually superinfected, invasive and can be confused with chronic otitis externa [5]. When the lesion is not correctly diagnosed, it can spread locally to the middle ear and invade vital structures present in the temporal bone (internal carotid artery, facial canal, cochlea, vestibule).

There is no consensus at the present time concerning TNM staging of these tumours by the Union Internationale Contre Le Cancer (UICC) or the American Joint Committee on Cancer (AJCC). However, since the beginning of the 1990s, the Pittsburgh radioclinical classification of carcinoma of the EAC [6] has become widely used in the international literature and has been demonstrated to be reliable and reproducible [1,7].

The reference treatment for cancer of the EAC is surgery [8], which is often mutilating due to the anatomical site of these tumours. These operations were reputed to be associated with severe postoperative morbidity. However, progress in general anaesthesia, aseptic techniques and intensive care has allowed an improvement of postoperative survival following radical surgical procedures. The progress provided by new technologies, such as surgical navigation and intraoperative facial nerve monitoring, have allowed an improvement of the quality of otological tumour resection and improvement of the patient's quality of life by reducing the risks of sequelae. Despite these improvements, advanced tumours (stages III—IV) are associated with a poor prognosis with low disease-free survival and overall survival rates.

The most common histological type of carcinoma of the EAC is squamous cell carcinoma. The other histological types are basal cell carcinoma, malignant melanoma, Merkel cell carcinoma, angiosarcoma, adnexal carcinoma, including ceruminous adenocarcinoma and adenoid cystic carcinoma, and lymphoma [9]. The prognosis depends on the histological type [9].

The primary objective of this study was to evaluate the overall survival and disease-free survival rates of patients with a malignant tumour of the external auditory canal with metastatic potential. The results of this study are discussed in the light of the literature.

Patients and methods

This retrospective, single-centre study was based on review of the medical charts of a consecutive series of patients with a malignant tumour of the EAC managed between 1991 and 2011 in our otorhinolaryngology department.

Clinical data concerning age, gender, otological history, date and mode of diagnosis, site, histological type, management, and survival were recorded for each patient included. The date of diagnosis corresponded to the date of the first biopsy that confirmed the malignant nature of the lesion. Tumour site, lymph node status (cervical and parotid) and the presence of metastases were determined from clinical findings, computed tomography (CT) and/or magnetic resonance imaging (MRI) imaging and the operation report. Tumour stage was established according to the university of Pittsburgh Tumor, Node and Metastasis (TNM) classification for squamous cell carcinomas of the EAC [1,8] (Table 1).

The type of therapeutic management and the length of postoperative hospital stay in surgically treated patients were recorded. The histology reports for the operative specimens indicated the surgical margins, revised surgical margins, bone and cartilage invasion and the presence of lymph node invasion on lymph node dissection specimens.

Table 1 University of Pittsburgh TNM staging system for carcinomas of the external auditory canal (EAC) [1,6].

T
T1: tumour limited to the EAC without bony erosion or evidence of soft tissue involvement
T2: tumour with limited external auditory canal bone erosion (not full thickness) or limited (< 0.5 cm) soft tissue involvement
T3: tumour eroding the osseous bone EAC (full thickness) with limited (< 0.5 cm) soft tissue involvement or tumour involving the middle ear and/or mastoid
T4: tumour eroding the cochlea, petrous apex, medial wall of the middle ear, carotid canal, jugular foramen, or dura, or with extensive soft tissue involvement (> 0.5 cm), or evidence of facial paresis
N
N0: no regional lymph node metastasis
N1: a single regional metastatic lymph node < 3 cm
N2a: a single ipsilateral metastatic lymph node measuring 3—6 cm
N2b: several ipsilateral metastatic lymph nodes < 6 cm
N2c: contralateral metastatic lymph node
N3: metastatic lymph node > 6 cm
M: distant metastasis
Stages
Stage I
Stage II
Stage III
Stage IV: T4N0 or T × N+

Each case was discussed at a multidisciplinary consultation meeting. Surgical resection was systematically proposed except for locally advanced tumours with extension to the posterior cranial fossa, internal carotid artery or middle cranial fossa, in the presence of contraindications to general anaesthesia related to comorbidities or in the case of metastatic disease. Adjuvant radiotherapy was systematically proposed after surgical resection. Chemotherapy was considered in cases with positive surgical margins, local recurrence or distant metastases.

Post-treatment sequelae (peripheral facial nerve palsy, sensorineural hearing loss) and cosmetic sequelae (poor healing, amputation of the outer ear) were also recorded. The 5-year overall survival and disease-free survival rates were evaluated according to the Kaplan—Meier method.

Results

Ten patients with a mean age of 60.7 years (±7.2) and a sex ratio of 1 were included. Two patients had been followed for cholesteatoma. The presenting symptoms and signs were non-specific. Six (60%) patients reported otorrhoea and three of them (30%) had been followed for persistent