■ CLINICAL INVESTIGATION

Primary Tumours of the External Auditory Canal. Our Experience in 34 Patients

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Objectives: Malignant neoplasms of the external auditory canal (EAC) and middle ear are rare but have a poor prognosis. The aim of this study is to identify the variables associated with worse prognosis.
Patients and method: Thirty-four patients were treated in our department between 1990 and 2006 for EAC and middle ear tumours. The patients were staged according to the 1990 Pittsburgh classification. In most cases, surgery was followed by post-operative radiotherapy.
Results: The overall disease-free survival was 49 % after 5 years. It reached 87 % in stages I and II, whereas the survival for stages III and IV was 21 % (*P* = .001). Pre-operative facial nerve paralysis (*P* = .03), lymph node metastasis (*P* = .01), and dural extension (*P* = .02) were associated with decreased survival rates.
Conclusions: In carcinomas of the EAC and middle ear, lymph node involvement, facial nerve palsy, and dural extension were associated with a poorer outcome. For tumours in advanced stages, new therapeutic protocols should be evaluated.

Key words: External auditory canal. Basal cell carcinoma. Epidermoid carcinoma. Temporal bone surgery.

Tumores primarios del conducto auditivo externo. Nuestra experiencia en 34 pacientes
Objetivo: Los tumores malignos del conducto auditivo externo (CAE) y el oído medio son poco frecuentes, pero se asocian a una alta morbimortalidad. El objetivo de este estudio es identificar los factores que se asocian con un peor pronóstico.
Pacientes y método: Presentamos un estudio retrospectivo de 34 pacientes con tumores del CAE y el oído medio tratados en nuestro servicio entre los años 1990 y 2005. Los pacientes fueron clasificados siguiendo el sistema de estadificación de Pittsburgh de 1990. El tratamiento en la mayoría los casos fue quirúrgico con radioterapia postoperatoria.
Resultados: La supervivencia libre de enfermedad a los 5 años para la serie total fue del 49 %. Agrupados por estadios, en estadios I y II fue del 87 % y en estadios III y IV del 21 % (p = 0,001). Se asociaron a un peor pronóstico la parálisis facial preoperatoria (p = 0,03), la metástasis ganglionar (p = 0,01) y la invasión de la duramadre (p = 0,02).
Conclusiones: En los carcinomas de CAE y oído medio, la enfermedad ganglionar, la parálisis facial y la extensión intracraneal se asocian a un peor pronóstico. Dado que la supervivencia a estos tumores en estadios avanzados es escasa, sería necesario considerar nuevos protocolos terapéuticos.

Palabras clave: Conducto auditivo externo. Tumores del temporal. Carcinoma epidermoide. Cirugía del hueso temporal.

INTRODUCTION

From an epidemiological standpoint, malignant tumours of the temporal bone represent 1 in 5,000 to 20,000 visits to the otological clinic[1,2]. The incidence is from 1 to 6 patients per million inhabitants and year[3-5]. They mostly affect patients in the fifth or sixth decade of life, with a similar distribution by gender or a slight predominance among men[6]. Otorrhea is the first symptom in most of the series published. Diagnosis is often delayed because the symptoms are very simiular to those of other benign otological processes (chronic otitis, cholesteatoma, etc.). Its pathogeny has been blamed on exposure to ultraviolet rays (pabellón and exposed EAC), long-standing chronic otorrhea, chemical substances such as chlorinated disinfectants, genetic predisposition or prior radiotherapy[1]. In middle-ear tumours, infection by the human papiloma virus has been identified as a predisposing factor[7]. From the point of view of anatomical pathology, the most frequent tumours are epidermoid, followed by adenocarcinoma, cystic adenoid, basocellular and melanomas of different types according to the various studies. Basocellular carcinoma is more frequent in the concha than in the EAC[8,9].

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Tumours of the temporal bone are locally aggresive, erode the bone and follow the route of nerves and blood vessels, invading neighbouring anatomical structures. The most widespread treatment is surgery, on its own for very early stages (T1), and accompanied by adjuvant radiotherapy in more advanced stages (T2, T3 and T4)[10].

The factors that have been associated with a worse prognosis of the disease are extension of the tumour, the presence of positive ganglia, positive margins[11], facial paralysis[2] and involvement of the dura mater[12], as well as other cranial pairs or moderate-severe pain on diagnosis[13-15]. Mortality has declined since the series reported by Lewis et al[16] to the most recent ones from Moffat et al[17] or Yin et al[6].

The general survival rates range between 50 and 70% after 5 years and is around 20% in the case of advanced stages. Mortality generally occurs due to local recurrence rather than regional or remote metastasis[17-20]. The initial surgical treatment proposed should ensure free margins, given the tumours' tendency to recur locally after more conservative surgery.

PATIENTS AND METHOD

We present a retrospective study of 34 patients with tumours in the EAC an the middle ear treated at our hospital between 1990 and 2005. A total of 20 males and 14 women between 38 and 84 years of age (mean 65 years) were treated. No patient had bilateral involvment.

Patients were excluded from this study if their tumours were not primary but invaded the temporal bone through contiguity or were metastatic tumours of the temporal bone, auricular concha, glomus or mesenchyme.

The particulars analyzed for each patient were age, gender, clinical presentation, stage in connection with treatment and survival.

The patients were classified using the Pittsburgh staging system described by Arriaga et al[21] in 1990: T1, tumour limited to the EAC, without bone erosion or extension to soft tissues; T2, bone erosion not affecting the entire thickness of the wall or < 0.5 cm extension into soft tissues; T3, complete erosion of the bone wall and < 0.5 cm extension into soft tissues or extension of the tumour to the middle ear, mastoid or facial paralysis; and T4, tumour eroding the cochlea, petrous apex, medial wall of the middle ear, carotid duct, jugular orifice or dura mater, or > 0.5 cm extension into soft tissues; N0, absence of ganglia, and N+, presence of ganglia. The stage is shown in Table 1.

Among their personal history, 8 patients (24%) had received radiotherapy in the area due of the other cutaneous and non-cutaneous tumours of the neck and head (between 2 and 24 years prior to the presentation of the tumour; mean, 10 years); 6 patients (18%) had a prior history of chronic suppurative otitis media; 5 patients (15%) had presented other cutaneous epithelial epidermoid or basocellular tumours and 9 (26%) were smokers. Some patients presented several of these combined factors.

Table 1. Distribution of Patients by Stages According to the Classification of Arriaga et al[21]

	N0	*N+*
T1	I (6)	III (1)
T2	II (7)	IV
T3	III (2)	IV
T4	IV (12)	IV (5)

The duration of the symptoms varied between a few weeks and more than a year, although most of them presented symptoms from 3 to 6 months earlier. The most frequent symptom was otorrhea, which appeared in 19 patients (56%). Other frequent symptoms were otalgia, hypoacusia and otorrhagia. Less frequent were the presence of a mass in the EAC, a pre-auricular mass or facial paralysis.

Radiological studies were made using computerized tomography (CT) and/or magnetic resonance (MR) pre- and post-operatively in all patients.

The stage of the tumour at the moment of its presentation predetermined the surgical approach adopted as reflected in a previous article[22]. We have adopted the classification of Saunders and Medina[23]. T1N0 tumours were handled using a type I lateral temporal resection; T2N0 tumours, by means of a type II or III temporal resection; the tumours in stages III or IV were approached using an extended temporal resection (sub-total or total) as a *of novo* treatment or as salvage surgery, when they had previously been treated previously with surgery and/or radiotherapy; in stages III and IV parotidectomy was performed preserving the facial nerve when it was not affected. The Eustachian tube was obliterated with bone, muscle and/or fascia. The defect was closed with a flap of temporal muscle or with a free skin graft.

RESULTS

From the anatomical pathology viewpoint, the most frequent tumour was epidermoid carcinoma (19 patients), followed by basocellular carcinoma (7 patients). Others included cystic adenoid tumours (4 cases), adenocarcinoma (2 cases), melanoma or warty carcinoma (1 case each).

Six patients (18%) presented at stage I; 7 (20%), at stage II; 4 (12%), at stage III, of whom 3 were T3N0 and 1 was T1N1. Finally, stage IV was represented by 17 patients (50%), 12 T4N0, 4 T4N1 and 1 T4N2 (Table 1).

With respect to the surgery performed (23 cases), 11 patients were given type I lateral resection, 6 received a type II lateral resection, 7 a type III lateral resection; in 2 cases resection was sub-total and in 6 it was total. Superficial parotidectomy was performed in 4 patients (1 at stage I, 1 at stage III, and 2 at stage IV, who also had their facial nerves removed), and total parotidectomy in 8 patients (1 at stage III and 7 at stage IV; in 2 patients it was possible to preserve the facial nerve).

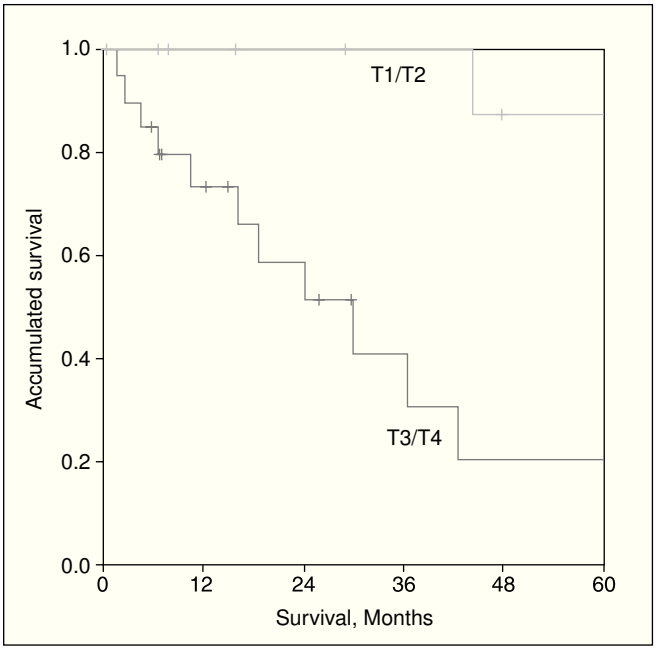

Figure 1. Disease-free survival after 5 years, grouped for stages I and II and for stages III and IV. Statistically significant difference (p = 0.001).

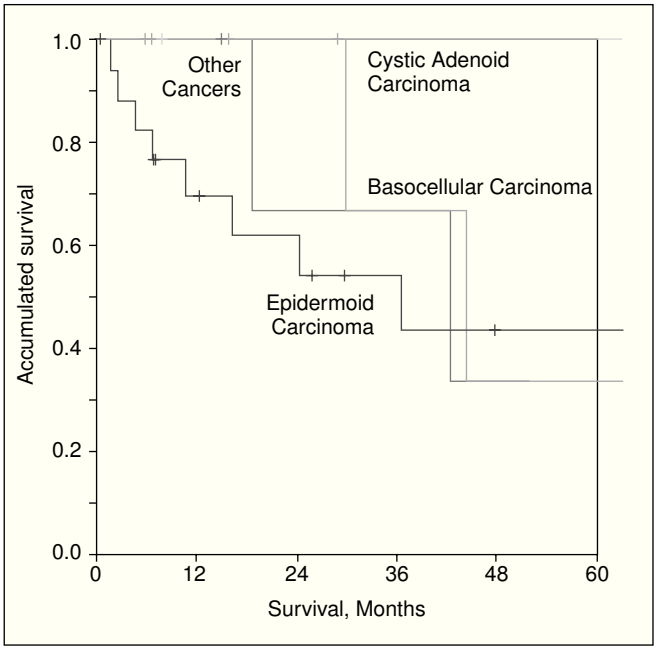

Figure 2. Survival after 5 years grouped by histological sub-type.

The defect was rebuilt using a free skin graft in 8 cases, temporal muscle in 18 cases, with abdominal fat in 3 cases and with a parascapular flap in 2 cases; 16 patients received post-operative radiotherapy. One patient received radiotherapy plus chemotherapy following surgery (total temporal salvage resection following radical mastoidectomy and prior radiotherapy). Two patients were treated palliatively in view of the advanced state of the illness, their age and baseline status.

With respect to complications, 5 patients presented a CSF fistula, 2 of which were resolved with postural measures, 2 by means of a lumbar drain and in one case it was necessary to close the fistula with abdominal fat one month after the initial surgery. Other complications were dehiscence and infection of the suture, pseudo-membranous enterocolitis due to metronidazole, seroma of the temple, acute epididymitis in connection with the vesical catheter and acute myocardial infarction. One patient died in the post-operative period after suffering a tronco-encephalic cerebrovascular accident with acute pulmonary oedema.

Disease-free survival after 5 years was 49% for the whole series. Grouped by stages, it was 87% for stages I and II, and 21% for stages III-IV, statistically significant differences (p = 0.001) (Figure 1).

With regard to the histology of the tumours, survival after 5 years was 43% for epidermoid carcinomas, 33% for basocellular carcinomas and 100% for cystic adenoid tumours, although one patient died because of the tumour after 68 months (Figure 2).

Twelve patients died in the course of the study, 2 due to inoperable tumours, 7 from cervical or local relapse and 3 due to remote or locoregional metastases.

Of these 12 cases, 50% died during the first year after surgery and another 25% during the second year, all of them from epidermoid tumours except for one melanoma. The patients with basocellular carcinoma who died from this disease did so after 30 and 44 meses and the patients with adenocarcinoma after 6 and 42 months. Of the 6 patients with positive ganglia, 4 have since died. The survival of N0 patients was 61% after 3 years and 54% after 5 years, whereas in stage N+ it was 20% after 3 years and 0% after 5 years, a statistically significant difference (p = 0.013) (Figure 3).

Five patients presented paresia or facial paralysis on diagnosis and the survival of this group of patients (20 % after 5 years) was significantly lower than in the rest of the patients in the series (41%) (p = 0.028).

Invasion of the dura mater was also associated with a worse prognosis, with survival of 0% after 48 months when it was affected and 60% after 5 years when it was not (p = 0.02).

In 8 patients it was necessary to resect the facial nerve due to infiltration of the extrapetrous portion of the nerve, which was rebuilt in 5 cases (3 with the great auricular nerve, 1 with rami of the cervical plexus and 1 with spinofacial anastomosis). In 2 cases the intrapetrous portion of the facial nerve was infiltrated and in both it was rebuilt using hypoglossofacial anastomosis.

DISCUSSION

If we consider the demographic variables, we find a slight male dominance coinciding with other series published, although this trend has not been seen in larger-scale revisions[1,5,6]. Other data such as age, initial symptoms or the

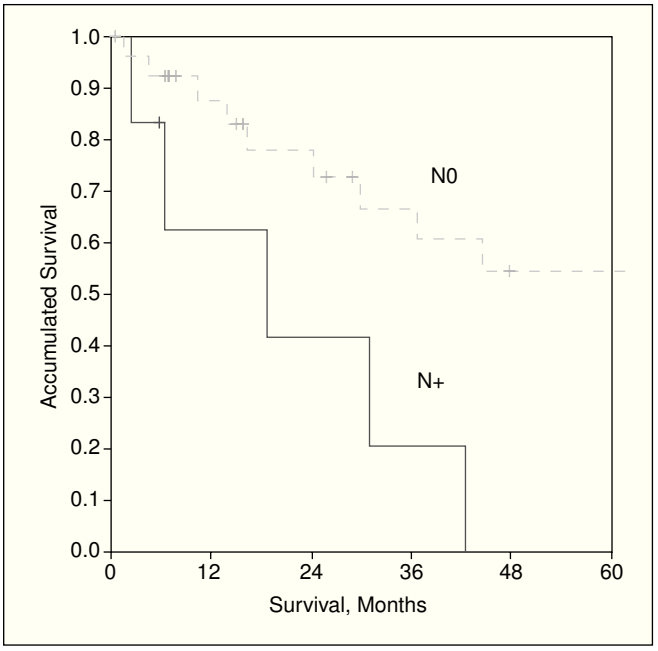

Figure 3. Disease-free survival in N0 and N+ patients. Statistically significant difference (p = 0.013).

histological strain of the tumour were not significantly different. The existence of a history of prior radiotherapy in 8 cases is noteworthy; the tumours developing in the ears of these patients were epidermoid carcinomas. Overall survival in our series was 49% after 5 years. The unfavourable factors (all statistically significant) in connection with survival include the presence of ganglia (18% presented positive ganglia, and all except one patient lost in the follow-up are known to have died from this disease), the involvement of dura mater (only 1 of 4 patients with intracranial extension is still alive and disease-free after 7 months) and the presence of facial paralysis at the time of the diagnosis. In addition, grouping the stages shows that stages I-II present a better prognosis than stages III-IV. For this reason it is very important to obtain an early diagnosis and an aggressive initial treatment for these tumours in order to offer the greatest possibilities of a cure. When the initial removal is not complete or is inadequate, survival rates fall dramatically. Moffat et al[17] found a higher survival among patients treated *of novo* than those salvaged, which may be due to the fact that the latter present more aggressive tumours or that they were not radically treated at the outset. This same trend was seen in our series: of the patients in stage IV, the two with the longest survival received a total resection of the temporal bone. Those same authors recommend carrying out superficial parotidectomy, systematic extirpation of the head of the mandible and functional voiding of the neck ganglia, as the extensions into the infratemporal fossa are radiologically under-diagnosed[17]. Without performing this procedure on all patients, we have obtained similar survival rates in our series, so we believe that it is not necessary to adopt such a radical attitude (e.g. extirpation of the mandibular condyle) untill there is sufficient scientific evidence to back it up.

The most recent papers recommend the use of complementary radiotherapy in all cases, as this provides better results when the resection margins have been negative[24]. It does not seem prudent to entrust proximal or affected margins to radiotherapy. We have not encountered significant differences in our series, but only a tendency towards a higher survival rate in the group treated with surgery and radiotherapy than in the group treated only with surgery in stages III and IV. Some authors propose chemotherapy in stages III and IV[6].

Epidermoid tumours generally behave more aggressively and have a worse pronosis. In our series, the survival was 43% after 5 years: 50% of the patients who died did so in the first year, with locoregional relapse, and 15% also presented remote metastasis.

The biological behaviour of basocellular tumours makes their prognosis more positive. However, it should be pointed out that 2 patients in our series died because of the tumour, although progression was slow. Other studies also point to the possibility that the survival of patients with basocellular tumours in advanved stages is poor[25]. Basocellular tumours in the auricular concha have a very good prognosis; when they take root in the EAC, however, the prognosis may be less positive, as they are disseminated widely through the neighbouring structures and resection may be incomplete if an adequate surgical technique is not applied.

Patients with cystic adenoide tumour progressed much more favourably, with longer survival. The follow-up of these patients must however continue for life, as relapse or metastases are commonplace many years later. If we consider patients with other histological strains of tumour in our series, as in most of the series reported, it is not possible to draw reliable conclusions in view of the small number of patients. It would be of interest to conduct prospectiv and multi-centric studies into these tumour types.

Malignant tumours in the EAC and the middle ear are infrequent, but they are associated with a high degree of morbi-mortality. The most frequent is epidermoid carcinoma, which almost always starts in the EAC. These tumours have a poor prognosis, particularly in advanced stages, which is when most of them are diagnosed because their clinical symptoms overlap with other benign processes and due to the absence of clear anatomical barriers preventing progression[8,18,26]. The treatment of these tumours is based on the combination of surgery plus radiotherapy. Prognosis depends on the tumour staging, the presence of positive ganglia, facial paralysis or intracranial extension with involvement of the dura mater. An early diagnosis and aggressive initial treatment are necessary with these tumours to offer the greatest possibilities of a cure.

These tumours can also be studied at the molecular scale to identify patients with more aggressive tumours who would benefit from new treatments[27].

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