Malignant Tumors of the Ear and Temporal Bone: A Study of 27 Patients and Review of Their Management

Pablo Martinez-Devesa, F.R.C.S.Ed. (ORL-HNS),¹ Martyn L. Barnes, M.R.C.S.Ed.,² and Chris A. Milford, F.R.C.S.¹

ABSTRACT

Objective: To evaluate the management of patients with malignant tumors of the ear and temporal bone. **Design:** Retrospective analysis of data. **Setting:** Radcliffe Infirmary, Oxford, United Kingdom. **Participants:** Twenty-seven patients were classified into two groups according to the site of origin of the tumor: (1) superficial (17 tumors): tumors arising from the skin of the pinna, parotid, and temporomandibular joint area; (2) deep (10 tumors): tumors arising in the ear canal and temporal bone. **Main Outcome Measures:** Treatment modality, complications, recurrence rate, disease-free interval, and survival. **Results:** The mean follow-up period was 25 months (0 to 60), and the median overall survival 46 months (0 to 102). Complications occurred in 6 patients (22%). The 3-year survival was 38% (95% confidence interval [CI], 19 to 58%), and the 5-year survival 19% (95% CI, 3 to 35%). **Conclusions:** There were insufficient data to demonstrate any difference in survival or disease-free interval related to the site of tumor origin (superficial versus deep tumors). There were independent differences in survival in favor of both performing parotidectomy and using postoperative radiotherapy, but neither reached significance at the 0.05 level.

KEYWORDS: Malignant tumor, ear, temporal bone, treatment

Malignancies of the ear and temporal bone are rare and aggressive types of tumors. The incidence has been reported to be between

one and six cases per million population years, which is less than 0.2% of all tumors of the head and neck.¹⁻³

¹Department of Otolaryngology, John Radcliffe Hospital, Oxford, United Kingdom; ²Department of Otolaryngology, Raigmore Hospital, Inverness, United Kingdom.

Address for correspondence and reprint requests: Pablo Martinez-Devesa, F.R.C.S.Ed. (ORL-HNS), Specialist Registrar, Department of Otolaryngology, West Wing, John Radcliffe Hospital, Headley Way, Headington, Oxford OX7 9DU, United Kingdom

(e-mail: devesa@doctors.org.uk).

Skull Base 2008;18:1-8. Copyright © 2008 by Thieme Medical Publishers, Inc., 333 Seventh Avenue, New York, NY 10001, USA. Tel: +1(212) 584-4662.

Received: June 24, 2007. Accepted: August 2, 2007. Published online: November 6, 2007.

DOI 10.1055/s-2007-992766. ISSN 1531-5010.

It is estimated that 1 case of middle ear carcinoma occurs for every 5000 to 20,000 cases of ear disease.^{4,5}

These tumors spread mainly by direct invasion into the temporal bone and neighboring structures⁶ (parotid, infratemporal fossa, dura, brain). Lymphatic metastases are uncommon (10%) and distant spread extremely rare.⁷

Computed tomography (CT) and magnetic resonance imaging (MRI) techniques⁸⁻¹⁰ are complementary and play a vital role in staging and prognosis.¹⁰⁻¹²

Currently neither the International Union Against Cancer (UICC) nor the American Joint Committee on Cancer (AJCC) has produced a staging system for ear and temporal bone tumors. The most common staging system being used in publications is from the University of Pittsburgh¹¹⁻¹⁴ (Table 1).

Table 1 University of Pittsburgh TNM Staging System Proposed for the External Auditory Canal

T Status

T1: tumor limited to the external auditory canal without bony erosion or evidence of soft-tissue involvement

T2: tumor with limited external auditory canal bone erosion (not full thickness) or limited (< 0.5 cm) soft-tissue involvement

T3: tumor eroding the osseous external auditory canal (full thickness) with limited (< 0.5 cm) soft-tissue involvement or tumor involving the middle ear and/or mastoid

T4: tumor eroding the cochlea, petrous apex, medial wall of the middle ear, carotid canal, jugular foramen, or dura, or with extensive soft-tissue involvement of the temporomandibular joint or styloid process, or evidence of facial paresis

N Status

Involvement of lymph node metastases is a poor prognosis finding; any node involvement should automatically be considered as advanced disease (i.e., T1N1 = stage III and T2, T3, T4, N1 = stage IV).

M Status

Distant metastases indicate a very poor prognosis and should be considered as stage IV disease.

In the absence of metastatic lymph nodes or distant metastases, T status of the tumor defines the clinical stage.

Surgical treatment of malignant tumors of the ear and temporal bone is not universally agreed upon. In general lesions localized to the external auditory canal (EAC) (T1 lesions) are treated with a *limited resection* (i.e., local resection, radical mastoidectomy), whereas more advanced lesions (T2 to T4) are treated by *en bloc resection* (partial temporal bone resection, subtotal petrosectomy, total petrosectomy^{10,12,13,15}).

Radiotherapy is advocated as an adjunct to surgery or for palliation rather than as a curative treatment in itself.^{16,17}

Overall survival rates are good for T1 and T2 lesions, less good for T3 lesions, and poor in T4 tumors.^{2-5,10,12,13,15}

MATERIALS AND METHODS

Between 1997 and 2005, 27 patients undergoing treatment for malignant tumors of the ear and temporal bone were identified from the Otolaryngology and Skull Base Department database at the Radcliffe Infirmary, Oxford, UK. The medical records of these patients were reviewed for age, sex, diagnosis, treatment (surgery, radiotherapy, and/or chemotherapy), complications, follow-up, recurrence, disease-free interval, and survival rate.

Patients were classified into two groups:

1. Superficial: those arising from the skin of the pinna, parotid, and temporomandibular joint (TMJ) area
2. Deep: those arising in the ear canal and temporal bone

They were staged according to the University of Pittsburgh TNM system (Table 1) for the deep tumor group ($n = 9$) and according to the UICC system for the superficial group ($n = 17$). One patient (with a deep angiosarcoma) was excluded from classification by either system.

Five types of surgical procedures were performed, in two broad categories:

1. Limited resection:
 - Local canal resection (LCR) and
 - Radical mastoidectomy (RM)
2. En bloc resection:
 - Lateral temporal bone resection (LTBR, consisting of a formal removal of the bony and cartilaginous external auditory canal, tympanic membrane, malleus, and incus with identification and preservation of the facial nerve along its vertical segment)
 - Subtotal petrosectomy (SP, involving the removal of the lateral temporal bone with preservation of the petrous apex and carotid artery), and
 - Total petrosectomy (TP, involving the removal of the entire temporal bone with or without carotid artery sacrifice)

Kaplan-Meier survival analyses were used, and tests were conducted on the survival effects of tumor origin, the inclusion of superficial parotidectomy in the operative procedure, and the use of postoperative radiotherapy.

RESULTS

Data were obtained for 27 patients, 18 male and 9 female, a ratio of 2 to 1 (sex distribution did not differ by group; chi square $p=0.696$). The median age was 66.5 years (range, 34 to 81). The deep group median age was 62.6 (34 to 77 yrs) and the superficial group 74.5 (64 to 81 yrs). This age difference was statistically significant (independent samples t -test and Mann-Whitney U tests, both $p < 0.005$). Diagnosis, staging, and management data are provided in Table 2.

DIAGNOSTIC DATA

Histological diagnoses were squamous cell carcinoma (SCC) (20 cases; 74%), basal cell carcinoma

(5 cases; 18%), angiosarcoma (1 case; 4%), and anaplastic carcinoma (1 case; 4%).

Seventeen tumors were classified as superficial and 10 deep, although the angiosarcoma case (deep group) was excluded from the statistical analysis as this tumor cannot be staged either with the Pittsburgh (solely for SCC) or the UICC (for superficial skin tumors) staging systems.

The distribution of the tumors ($n=26$) was T1, 3 cases; T2, 4 cases; T3, 4 cases; and T4, 15 cases. Clinically evident node-positive disease was present in 3 of these cases, and distant metastases in 1. Classification in stages was Stage I, 3 cases; Stage II, 4 cases; Stage III, 10 cases; and Stage IV, 9 cases.

MANAGEMENT DATA

Twenty-five patients underwent surgical treatment, with 17 receiving adjuvant radiotherapy postoperatively. One patient received radiotherapy (45 Gy in 10 fractions) as the only form of treatment and another patient had radiotherapy and chemotherapy (cisplatin and 5-fluorouracil) for palliation.

The surgical procedures performed were local canal resection (2 cases), radical mastoidectomy (1 case), lateral temporal bone resection (16 cases), subtotal petrosectomy (3 cases), and total petrosectomy (3 cases).

Also, 15 patients underwent superficial parotidectomy, 3 patients had a neck dissection performed (2 modified radical and 1 selective neck dissection), and 9 patients had tissue transfer reconstruction (2 pedicle flaps, 5 free flaps, 1 local flap, and 1 facial nerve graft). One patient had resection of the TMJ and another had resection of intracranial disease.

Six patients suffered complications: one patient died postoperatively due to respiratory distress syndrome. One patient developed osteoradionecrosis of the external ear canal, which

Table 2 Diagnosis, Staging, and Management Data

Sex/ Age	Diagnosis	Pittsburgh Stage	UICC Staging	Surgery	Radiotherapy	Chemo	Complications	Follow Up (mo)	Disease Free (mo)	Recurrence	Survival (mo)	Dead (cause) / Alive
1 F/34	SCC	T2 NO M0/II		LTBR	Yes	No	Osteoradionecrosis EAC	60	60	No	60	Alive
2 M/59	SCC	T1 NO M0/I		LTBR	No	No	No	60	60	No	60	Alive
3 M/65	BCC	T2 NO M0/II		LTBR + parot + free flap	Yes	No	Revision of flap	60	60	No	60	Alive
4 F/72	SCC	T1 NO M0/I		No	Yes	No	No	48	48	No	48	Alive
5 M/75	Skin SCC		T4 NO M0 / III	LCR + parot	Yes	No	No	24	24	No	24	Alive
6 F/72	SCC	T3 NO M0 / III		LTBR + parot	Yes	No	No	16	16	No	16	Dead / arteriopathy
7 M/59	SCC	T4 NO M0 / IV		LTBR + parot	Yes	No	No	60	60	No	60	Alive
8 M/63	SCC	T3 N1 M0 / IV		LTBR + parot + SND	Yes	No	No	45	45	No	45	Alive
9 M/81	Skin SCC		T4 NO M0 / III	LTBR + parot	Yes	No	No	14	9	Yes/9 months	14	Dead
10 F/72	Skin BCC		T4 NO M0 / III	LTBR + parot	Yes	No	Facial palsy	40	40	No	40	Alive
11 M/74	Skin SCC		T4 NO M0 / III	LTBR + parot + free flap	Yes	No	No	37	37	No	37	Alive
12 M/62	SCC	T4 NO M0 / IV		SP + free flap	Yes	No	Revision of flap anastomosis	4	0	Never clear	4	Dead of disease
13 M/69	SCC	T4 NO M0 / IV		No	Yes	Yes	No	9	0	Never clear	9	Dead of disease
14 M/76	Skin BCC		T4 NO M0 / III	LTBR + parot + pedicle flap	No	No	No	5	0	Never clear	5	Dead of disease
15 M/81	Angiosarcoma	*	*	LTBR	Yes	No	Temporary facial palsy	3	0	Never clear	3	Dead of disease
16 M/79	Skin SCC		T4 N1 M0 / IV	SP + MRND	No	No	No	3	0	No	3	Dead of disease
17 F/77	SCC	T2 NO M0 / II		LTBR	Yes	No	No	8	0	Never clear	8	Dead of disease
18 M/64	Skin SCC		T4 N1 M1 / IV	TP + parot + MRND + pedicle flap	Yes	No	No	46	5	Yes/5 months (Neck and lung metastases)	46	Dead of metastatic disease
19 M/72	SCC	T4 NO M0 / IV		SP	Yes	No	No	8	0	Never clear	8	Dead of disease
20 F/60	SCC	T3 NO M0 / III		TP + parot	No	No	ARDS/Death	0	0	Never clear	0	Dead postoperatively / septicaemia and ARDS
21 F/64	Skin SCC		T4 NO M0 / III	Mastoidectomy	No	No	No	18	6	Yes/6 months	18	Dead of disease
22 M/75	Skin BCC	T1 NO M0 / I		LCR + local flap	No	No	No	60	60	No	60	Alive
23 F/68	Skin BCC		T4 NO M0 / III	LTBR + parot + free flap	No	No	No	12	0	Never clear	12	Dead of disease
24 M/58	SCC	T2 NO M0 / II		LTBR + parot	Yes	No	No	7	7	No	7	Alive
25 M/65	SCC	T4 NO M0 / IV		LTBR + parot + Excision TMJ + VII graft	Yes	No	No	7	0	Never clear	7	Alive with disease
26 F/52	SCC	T3 NO M0 / III		LTBR + parot	Yes	No	No	3	3	No	3	Alive
27 M/40	Anaplastic SCC	T4 NO M0 / IV		TP + intracranial resection + free flap	No	No	No	2	2	No	2	Alive

*Inappropriate to stage by either system.

SCC, squamous cell carcinoma; LTBR, lateral temporal bone resection; EAC, external auditory canal; BCC, basal cell carcinoma; parot, superficial parotidectomy; LCR, local canal resection; SND, selective neck dissection; SP, subtotal petrosectomy; MRND, modified radical neck dissection; TP, total petrosectomy; ARDS, adult respiratory distress syndrome; TMJ, temporomandibular joint; VII, facial nerve VII.

was managed conservatively. Two patients suffered facial nerve palsies (one temporary which recovered fully and one permanent, Grade IV House-Brackmann classification¹⁸). Two flaps required revision of their vascular anastomoses.

SURVIVAL DATA

The overall mean follow-up period was 25 months (0 to 60 mos), and for surviving cases was 37 months (2 to 60 mos).

Of the 26 staged patients (excluding the angiosarcoma), 9 were never disease free and 3 had recurrence. The proportion uncured by stage at presentation was Stage I, 0%; Stage II, 29%; Stage III, 50%, and Stage IV, 60%.

The estimated median overall survival was 46 months (0 to 102 mos).

Of 27 patients, 13 remained alive without disease, 1 alive with disease, and 13 died (1 in the postoperative period, 1 of an unrelated cause, 10 from effects of local disease, and 1 from metastatic disease).

The overall 3-year survival was 38% (95% CI, 19 to 58%), and the 5-year survival 19% (95% CI, 3 to 35%).

Estimated median (95% CI) survivals by stage from the Kaplan-Meier analyses were Stage I, no deaths; Stage II, >24 months; Stage III, 16 months (10 to 22 mos), and Stage IV, 9 months (7 to 11 mos).

There were insufficient data to demonstrate any difference in survival or disease-free interval related to the site of tumor origin (superficial versus deep groups). A Kaplan-Meier survival plot by site of origin is given in Fig. 1.

Similarly, although there was a trend toward improved survival when a parotidectomy was performed, the study was underpowered to demonstrate statistical significance. Estimated median overall survival (95% CI) for both groups with parotidectomy was 46 months (0 to 102 mos), and without parotidectomy 9 months (0 to 24 mos).

Figure 1 Kaplan-Meier analysis. Effect of site of origin of the tumor on survival.

A Kaplan-Meier survival plot comparing inclusion or omission of parotidectomy in the operative procedure is given in Fig. 2. The plot is reproduced and stratified by tumor origin (superficial or deep) in Fig. 3.

Also, estimated median overall survival for the superficial group with the administration of radiotherapy was 46 months versus 12 months when radiotherapy was not administered

Figure 2 Kaplan-Meier analysis. Effect of parotidectomy on survival (independent of site of origin of the tumor).

Figure 3 Kaplan-Meier analysis. Effect of parotidectomy on survival (stratified by site of origin of the tumor). (A) Deep tumors. (B) Superficial tumors.

(Fig. 4). The data regarding radiotherapy for the deep group were insufficient for analysis.

To exclude the possibility of a confounding relationship between treatment with radiotherapy or stage of disease and the effect of parotidectomy, a further survival analysis (Cox regression) was conducted using survival models for all factors. Consistent with the findings in general, disease stage was the only factor to affect survival ($p < 0.05$), but both radiotherapy ($p = 0.29$) and parotidectomy ($p = 0.12$) led to improvements that did not reach statistical significance.

DISCUSSION

Malignant tumors of the ear and temporal bone are very rare, and the lack of prospective trials (randomized or not) poses a problem in establishing the correct management of this type of tumor. The management of these tumors relies only on the available evidence of clinical experience with small isolated or grouped series. SCC has been the most studied and reported of malignant tumors of the ear and temporal bone, but most of the other tumors behave in a similar manner

Figure 4 Kaplan-Meier analysis. Effect of radiotherapy on survival (stratified by site of origin of the tumor). (A) Deep tumors (radiotherapy effect $p = 0.63$). (B) Superficial tumors (radiotherapy effect $p = 0.18$).

and share the generally poor outcome of this disease.¹⁰

One of the main problems in the management of these tumors has been the lack of consensus in staging the disease, a process made difficult due to the close anatomical relationship to structures in the infratemporal fossa and skull base.

The University of Pittsburgh TNM staging system¹¹ has been widely accepted within the published literature in the field. The importance of this classification is that a T2 or T3 N1 tumor is considered to be stage IV, in contrast with the AJCC staging system (sixth edition) in which a T2 or T3 N1 "skin" tumor would be a stage III.

We think that part of the staging problem is that some tumors originate in the skin and then invade deeper structures, whereas others originate in the ear canal and temporal bone and spread laterally to the overlying appendage and adjacent skin. However, we have been unable to demonstrate a statistically significant difference in the survival and recurrence patterns of the two groups, so this hypothesis remains the authors' experience only.

In terms of treatment options, the available evidence supports surgery for primary treatment with curative intent and radiotherapy for adjuvant or palliative treatment.^{16,17} A literature search found no studies on the effect of chemotherapy in these tumors.

A systematic review performed by Prasad and Janecka² in 1994, reviewing 26 series with 144 patients treated for temporal bone SCC, suggested that for T3 and T4 tumors (those involving the middle ear and beyond), subtotal petrosectomy (preserving the petrous apex and carotid artery) seems to improve survival.

Also in this review, resection of involved dura did not appear to improve survival and there were insufficient data to comment on survival after treatment in patients with involvement of the petrous apex, internal carotid artery, or brain.²

With regard to the treatment of stage I and II (T1 and T2, N0) tumors, there are three main management options available in the published literature:

1. A more "aggressive" surgical treatment in the form of en bloc resection^{3,11,13,15}
2. A more "conservative" local resection^{2,10} and limiting radiotherapy to those cases of incomplete resection (except in adenoid cystic carcinoma)¹⁰
3. And finally, the "intermediate" position of selective petrosectomy with preservation of neural and vascular structures, as well as the otic capsule, because this treatment in some surgeons' experience does not alter the long-term outcome, in particular of aggressive basal cell carcinomas without deep invasion.¹⁹

In our series, one stage I tumor, and most of the stages II, III, and IV tumors, were treated by en bloc resection. Parotidectomy was included as part of the surgical procedure (as reported in other series^{3,11,13,15}) in cases of confirmed or suspected involvement of the parotid gland.

Although the disease is rare, and numbers have been inadequate to demonstrate statistically significant differences, we did observe improved survival for both superficial and deep tumors when parotidectomy was performed. We therefore recommend a low threshold to include this as part of the surgical treatment of all temporal bone tumors.

Guidelines from the American Society for Head and Neck Surgery and the Society of Head and Neck Surgeons²⁰ recommend postoperative radiotherapy when: (A) resection margins are close (less than 5 mm) or when proximity of tumor to important structures such as internal carotid artery or facial nerve precludes wide margins; (B) tumor is demonstrated at resection margins; (C) postoperative histology reveals perineural invasion. These conditions apply to the majority of resections for temporal bone carcinoma, thus indicating postoperative radiotherapy in most cases,²¹ although the above-mentioned systematic review (Prasad and Janecka²) showed no statistical significant difference in survival in patients who underwent lateral temporal bone resection and radiotherapy (48% overall 5-year survival) compared with patients who underwent lateral temporal bone resection alone (44% 5-year survival).

Our study shows that there is a trend toward increased overall survival in the superficial group of

tumors if radiotherapy is used postoperatively, although this result was not statistically significant ($p = 0.18$). There were insufficient data to analyze the effect of radiotherapy in the deep group.

Finally, the survival of these tumors has been variously reported in 2-, 3-, or 5-yearly intervals. Stage I and II tumors have been reported, depending on the series, to have a 50%² to 100%^{10,11} survival. Stage III and IV survival varies from 0%¹⁰ to 50%.^{13,15} Our overall 3-year survival was 38%, and the 5-year survival 19%, perhaps due to the greater number of large (T3 to T4) tumors in this study.

Recurrence has been reported to be anywhere from 25% for stages I and II to 100% for stages III and IV.¹⁰ Our study achieved lower recurrence rates for stages III and IV (50 to 64%).

CONCLUSIONS

Malignant tumors of the ear and temporal bone are rare and there is no consensus in their staging and management. We were unable to demonstrate any difference in survival or recurrence for superficial or deep tumors of the temporal bone. Parotidectomy seemed to improve survival for both superficial and deep tumor groups, but statistical significance was not reached. Radiotherapy also seemed to improve survival for the superficial group, but again, statistical significance was not reached.

REFERENCES

1. Kuhel WI, Hume CR, Selesnick SH. Cancer of the external auditory canal and temporal bone. *Otolaryngol Clin North Am* 1996;29:827–852
2. Prasad S, Janecka IP. Efficacy of surgical treatment for squamous cell carcinoma of the temporal bone: a literature review. *Otolaryngol Head Neck Surg* 1994;110:270–280
3. Moody SA, Hirsch BE, Myers EN. Squamous cell carcinoma of the external auditory canal: an evaluation of a staging system. *Am J Otol* 2000;21:582–588
4. Lewis JS. Squamous carcinoma of the ear. *Arch Otolaryngol* 1973;97:41–42
5. Arena S, Keen M. Carcinoma of the middle ear and temporal bone. *Am J Otol* 1988;9:351–356
6. Michaels L, Wells M. Squamous carcinoma of the middle ear. *Clin Otolaryngol* 1980;5:235–248
7. Sasaki CT. Distant metastases from ear and temporal bone cancer. *ORL J Otorhinolaryngol Relat Spec* 2001;63:250–251
8. Phelps PD, Lloyd GAS. The radiology of carcinoma of the ear. *Br J Radiol* 1981;54:103–109
9. Friedman DP, Rao VM. MR and CT of squamous cell carcinoma of the middle ear and mastoid complex. *AJNR Am J Neuroradiol* 1991;12:872–874
10. Nyrop M, Grontved A. Cancer of the external auditory canal. *Arch Otolaryngol Head Neck Surg* 2002;128:834–837
11. Arriaga M, Curtin H, Takahashi H, Hirsch BE, Kameron DB. Staging proposal for external auditory meatus carcinoma based on preoperative clinical examination and computed tomography findings. *Ann Otol Rhinol Laryngol* 1990;99:714–721
12. Gillespie MB, Francis HW, Chee N, Eisele DW. Squamous cell carcinoma of the temporal bone: a radiographic-pathologic correlation. *Arch Otolaryngol Head Neck Surg* 2001;127:803–807
13. Austin JR, Stewart KL, Fawzi N. Squamous cell carcinoma of the external auditory canal: therapeutic prognosis based on a proposed staging system. *Arch Otolaryngol Head Neck Surg* 1994;120:1228–1232
14. Zhang B, Tu G, Tang P, Hu Y. Squamous cell carcinoma of the temporal bone: reported on 33 patients. *Head Neck* 1999;21:461–466
15. Moffat DA, Grey P, Ballagh RH, Hardy DG. Extended temporal bone resection for squamous cell carcinoma. *Otolaryngol Head Neck Surg* 1997;116:617–623
16. Sinha PP, Aziz HI. Treatment of carcinoma of the middle ear. *Radiology* 1978;126:485–487
17. Birzgalis AR, Keith AO, Farrington WT. Radiotherapy in the treatment of middle ear and mastoid carcinoma. *Clin Otolaryngol* 1992;17:113–116
18. House JW, Brackmann DE. Facial nerve grading system. *Otolaryngol Head Neck Surg* 1985;93:146–147
19. Pensak ML, Gleich LL, Gluckman JL, Shumrick KA. Temporal bone carcinoma: contemporary perspectives in the skull base surgical era. *Laryngoscope* 1996;106:1234–1237
20. The American Society for Head and Neck Surgery and The Society of Head and Neck Surgeons. Clinical practice guidelines for the diagnosis and management of cancer of the head and neck. The American Society for Head and Neck Surgery and the Society of Head and Neck Surgeons; 1996
21. British Association of Otolaryngologists Head and Neck Surgeons. Effective Head and Neck Management. Third Consensus Document. British Association of Otolaryngologists Head and Neck Surgeons; 2002