

Malignant Tumors of Temporal Bone in Songklanagarind Hospital: Management and Results

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Objective: To evaluate the clinical course and treatment in patients with carcinoma of temporal bone

Material and Method: The authors retrospectively reviewed the patients treated for primary carcinoma of temporal bone in Otolaryngology Head and Neck Surgery department, at Songklanagarind Hospital between 2002 and 2013. Thirty-two patients, including 10 cancer of pinna and 22 carcinoma of the External Auditory Canal (EAC), were included in this study. The treatment modality was lateral temporal bone resection and postoperative radiotherapy and palliative radiotherapy for advanced stage tumors (T4). Twelve patients were excluded due to difference in modality. Twenty patients remained in the study.

Results: Twenty patients had carcinoma of EAC, early stage (T1, T2) 36.4% and late stage (T3, T4) 63.6%. The overall survival of this group was 46.9%. The median survival period was 22 months (0.286-0.769, 95% CI).

Conclusion: The complete resection is the treatment of choice in primary cancer of temporal bone. Postoperative radiotherapy for a microscopic residual tumor is highly recommended. The most important thing is early diagnosis.

Keywords: Carcinoma of external auditory canal, Management, Results

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Primary cancer of the temporal bone (pinna, external auditory canal, middle ear, and temporal bone) are extremely rare, with the incidence estimated to be one to six per million per year in both United States and England⁽¹⁻⁹⁾. The most common type is squamous cell carcinoma⁽¹⁰⁾. The risk factors are infection, repeated trauma, and previous radiotherapy⁽³⁾. The diagnosis was usually delayed because of the clinical presentations were the same as an infection of the external ear and middle ear such as chronic suppurative ear disease and otitis externa. The types of surgery are controversial. They are local excision, sleeve resection, en-bloc resection, and aggressive surgery (such as subtotal or total temporal bone resection). Aggressive surgery has a significant morbidity and mortality rate because the temporal bones are close to vital organs such as great blood vessels or the cranial nerve. The best treatment is en-bloc resection with free surgical margin. Postoperative radiotherapy is controversial.

The purpose of the present article is to review and evaluate the clinical course, surgical treatment,

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and radiotherapy in patients with carcinoma of external auditory canal in our hospital during a 12 years period.

Material and Method

This was a retrospective study of patients treated for primary carcinoma of the temporal bone at the Otolaryngology Head and Neck Surgery Department, at Songklanakarind Hospital between 2002 and 2013. The study included 32, 10 cancer of pinna and 22 carcinoma of the external auditory canal (EAC).

Medical records were retrospectively reviewed. Data of age, sex, clinical presentation, computed tomographic of temporal bone, histopathological diagnosis, treatment modalities (type and extent of operation), and outcomes of treatment were collected and evaluated.

The tumor stage was determined preoperatively by imaging using computed tomography (CT). TNM staging was proposed by Arriaga et al and revised by the Pittsburgh group in 2000⁽³⁾ for external auditory canal and middle ear tumor.

Ten patients had carcinoma of the pinna, and two patients had temporal bone cancers associated with nasopharyngeal cancer. They were excluded because

of the difference in presentation, prognosis, and treatment outcomes. Thus, 20 patients were included in this study.

The treatment modality was mastoidectomy for T1 tumors at the posterior canal wall, and lateral temporal bone resection for T1, T2, and T3 tumors. All patients underwent postoperative radiotherapy, and palliative radiotherapy for advanced stage tumors (T4). Superficial parotidectomy was performed in some patients who had parotid node metastasis or tumor in the parotid gland. Neck dissections were performed when the patients had a palpable lymph nodes or positive imaging.

Overall survival was defined as the time from completed treatment up to the latest follow-up or death. Overall survival analysis was generated using the Kaplan-Meier method. The log-rank test was used to test the statistical significance of the differences between the curves. The analysis was performed with R Statistical Software (ver.2.15.1; Foundation for Statistical Computing, Vienna, Austria), *p*-value <0.05 were considered statistically significant.

The present study was approved by the Ethics Review Committee of the Institution.

Results

Twenty patients (10 women and 10 men) had carcinoma of the EAC. Histopathological examinations confirmed 17 (85%) squamous cell carcinomas (SCC), one (5%) adenoid cystic carcinoma (ACC), one (5%) sebaceous carcinoma, and one (5%) ceruminous adenocarcinoma. The mean age was 62.2 years (range 27-88 years, SD 15.46).

The median follow-up duration was 17 months, ranging from three to 84 months.

The presenting symptoms were otorrhea, otalgia, hearing loss, and facial palsy (Table 1).

Table 1. Clinical presentation of 20 patients with malignant tumors of temporal bone

Symptom	No. of patients (%)
Otorrhea	15 (75)
Otalgia	15 (75)
Hearing loss	17 (85)
Facial palsy	3 (15)
Trismus	1 (5)
Mass or ulcer or granulation tissue in external auditory canal	20 (100)

Physical finding included ulcers, mass, or granulation tissues in all patients. The authors found bilateral carcinoma in one patient with history of chronic otitis media in childhood. Most of the patients were initially diagnosed and treated as infection, such as otitis externa and chronic otitis media (longer than 3 months). Five patients had infection on the right side of their head, 14 on the left side, and one patient had bilateral ear carcinoma from young age and had history of chronic otitis media.

The authors staged our patients according to TNM staging system proposed by Arriaga et al and revised by the Pittsburgh group in 2000⁽¹¹⁾ for external auditory canal and middle ear tumors to make our results comparable to other groups, both squamous cell carcinoma as well as adenoid cystic carcinoma (Table 2). Eight patients were staged T2, four were T3 and 10 were T4. Early stage (T1, T2) were 36.4% and late stage (T3, T4) were at 63.6%.

Most of the patients were treated by surgery and postoperative radiotherapy. Three types of surgical resection according to the Kuhel classification⁽²⁾ were performed in our department. They included sleeve resection, local resection of EAC skin and cartilaginous part of EAC, modified or radical mastoidectomy, lateral temporal bone resection (LTBR), en-bloc resection of the entire EAC, the tympanic membrane, the malleus, and the incus through a mastoid approach. The LTBR may be combined with a parotidectomy or a neck dissection if the extension of disease to parotid or neck lymph node was suspected.

One patient underwent mastoidectomy, 12 patients underwent lateral temporal bone resections, and seven had palliative radiotherapy. A summary of patients was provided in Table 3.

For patients with carcinoma in the early stage of (T1, T2), confined to the EAC, the lateral temporal bone resection is the treatment of choice. If we can resect the entire tumor, the prognosis is very good, showing no evidence of residual and recurrence of disease. If the patients had a delayed diagnosis, carcinoma beyond middle ear in late stage (T3, T4), the operation would not be able to remove the entire tumor (residual tumor), the tumor could re-occur and showed poor prognosis, even though the patients were followed with postoperative radiotherapy.

The overall survival of the patients who had stage I to stage III (disease) was 100% (Fig. 1). The 13 patients having stage IV disease had an overall survival rate of 46.2%. The median survival period was 15 months (range 3-35).

Table 2. University of Pittsburgh TNM staging system proposed for external auditory canal carcinoma

T status	T1: tumor limited to the external auditory canal without bony erosion or evidence of soft tissue extension T2: tumor with limited external auditory canal bony erosion (not full thickness) or radiographic finding consistent with limited (<0.5cm) soft tissue involvement T3: tumor eroding the osseous external auditory canal (full thickness) with limited (<0.5cm) soft tissue involvement, or tumor involving middle ear or mastoid, or patients presenting with facial paralysis T4: tumor eroding the cochlear, petrous apex, medial of middle ear, carotid canal, jugular foramen or dura, or with extensive (>0.5cm) soft tissue involvement
N status	Involvement of lymph nodes is a poor prognostic finding and autonomically places the patient in an advanced stage (i.e. stage III [T1N1] or stage IV [T2, T3 and T4])
M status	Distant metastasis indicates a poor prognosis and immediately place patients in the stage IV category In the absence of metastatic lymph nodes or distant metastases, T status of the tumor defines the clinical stage

T = tumor; N = node; M = metastasis

Table 3. Summary of 20 patients with temporal bone carcinoma

No.	Age	Sex	Side	T status	Surgery	RT	Pathology	Outcome, n (%)	Outcome measurement
1	51	F	Lt	T2	LTBR	Yes	SCC	5 (100)	NED >10 years
2	33	M	Lt	T4	LTBR®	Yes	SCC	12 (0)	AD
3	61	M	Lt	T4	LTBR®	Yes	SCC	12 (0)	AD
4	27	M	Bilat	T4	M.®	Yes	SCC	12 (0)	AD
5	67	M	Lt	T4	LTBR®	Yes	SCC	12 (0)	AD
6	58	M	Rt	T3	-	Palliative	SCC	12 (0)	AD
7	88	F	Lt	T4	-	Palliative	SCC	12 (0)	AD
8	49	M	Lt	T4	-	Palliative	SCC	12 (0)	AD
9	82	F	Lt	T1	-	Yes	SCC	1 (100)	NED >6 years
10	52	M	Lt	T4	-	Palliative	SCC	12 (0)	AD
11	70	M	Lt	T4	-	Palliative	SCC	12 (0)	AD
12	56	F	Rt	T2	LTBR	Yes	Ceruminous	5 (100)	NED >3 years
13	62	F	Lt	T3	LTBR	Yes	Adenoca	2 (50)	NED >3 years
14	76	M	Lt	T4	-	Palliative	SCC	12 (0)	AD
15	59	M	Lt	T4	-	Palliative	SCC	12 (0)	AD
16	76	M	Lt	T4	-	Palliative	SCC	12 (0)	AD
17	58	M	Rt	T4	LTBR®	Yes	SCC	12 (0)	AD
18	79	F	Rt	T2	LTBR	Yes	SCC	5 (100)	NED >2 years
19	66	M	Rt	T2	LTBR	Yes	SCC	5 (100)	NED >1 year
20	74	M	Lt	T2	LTBR	Yes	SCC	5 (100)	NED >1 year

T = tumor; RT = radiotherapy; F = female; M = male; Lt = left; Rt = right; Bilat = bilateral; LTBR = lateral temporal bone resection; SCC = squamous cell carcinoma; NED = no evidence of disease; AD = active disease; M. = mastoidectomy; ® = residual tumor; Adenoca = adenocarcinoma

The overall survival of this group was 46.9%. The median survival period was 22 months (95% CI 0.286-0.769) (Fig. 2).

Discussion

Carcinoma of the external auditory canal and temporal canal is extremely rare. Of all malignant

tumors of the ear, 60% are auricular tumors⁽³⁾. The most common histologic type was squamous cell carcinoma at 83%, the rest was glandular tumors such as adenoid cystic carcinoma, adenocarcinoma, mucoepidermoid carcinoma, ceruminous adenocarcinoma, and sebaceous carcinoma^(2,3,12). No gender difference was found, except with adenoid cystic carcinoma, which

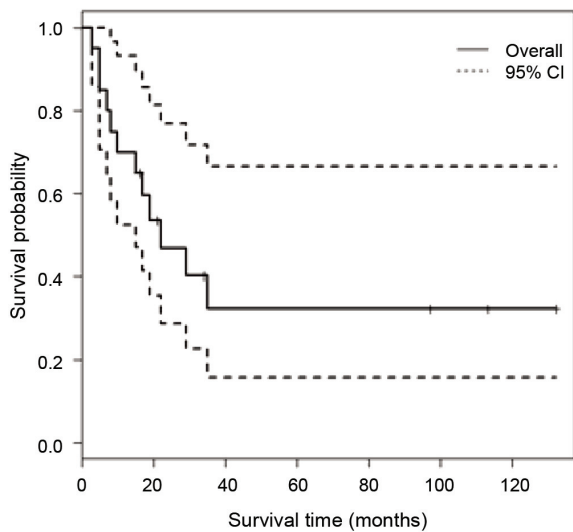


Fig. 1 The overall survival Kaplan-Meier curve. X-axis indicates overall survival. Y-axis indicates survival time in months.

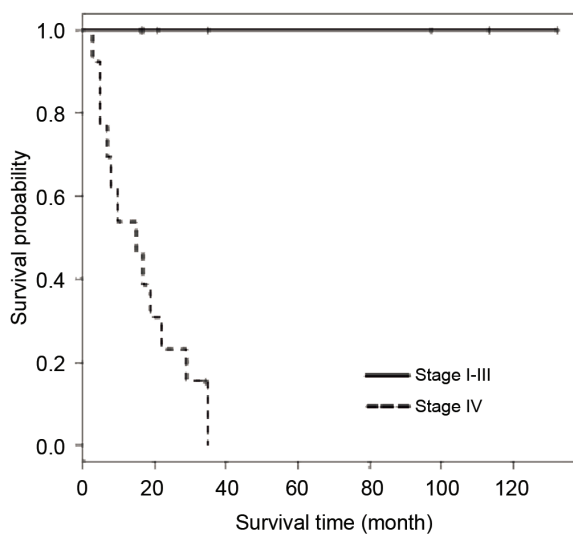


Fig. 2 The overall survival of the patients based on tumor staging. Stage I-III vs. stage IV, $p < 0.001$.

was higher in females than males, with a ratio of 1.58:1⁽¹⁾. The peak incidence was in the sixth decade of life (54.5%). In the present study, only 10 patients (29.4%) had carcinoma of pinna and 85% were of squamous cell carcinoma.

The diagnosis was often delayed because of the coexistence of infection of the external ear and middle ear, such as chronic suppurative ear disease and otitis externa. In other studies the patients with an early diagnosis (stage I and II) was 59% and late stage

disease (stage III and IV) was 41%⁽²⁾. In the past, most patients presented as stage T4, which mean that we could not remove the entire tumor. Now, while many patients are diagnosed early, as stage T2, it is still only 36.4% of the population. In this study, almost two thirds of patients were T3 or T4 stage at presentation (63.6%) and considered to have a poor prognosis. They could not being cured by surgical treatment. This suggests the importance of early diagnosis and treatment.

The most common presentations are hearing loss, otorrhea, and otalgia. The patients in the present series were treated for chronic ear infection by topical eardrops and antibiotics for more than six months. The awareness of carcinoma was very low due to very low incidence. Therefore, we should be suspicious for malignancy when chronic otorrhea do not respond to standard treatment, or the patients have other symptoms and signs such as severe pain at night, facial palsy, bloody discharge, and parotid swelling. This indicates that it is very late for surgical treatment.

Early diagnosis is the most important factor because all of the tumor can be removed by surgery. A worse prognosis happen when the entire tumor cannot be removed. Prasad and Janecka⁽¹³⁾ had reviewed a 5-year survival rate for patients with tumors confined to the EAC (T1, T2); it was at 50%. In the patients with carcinoma involving the middle ear (T3), the overall 5-year survival was 20 to 30%, averaging only five months^(2,3,13,14). The most adverse prognostic factors are invasion of the otic capsule, parapharyngeal space invasion, and intracranial involvement⁽⁶⁾.

If the patients had an early diagnosis as stage T1 and T2, the patients could be cured by surgery such as mastoidectomy, and lateral temporal bone resection. Postoperative radiotherapy in T1 disease is still controversial. It has been suggested in patients where the tumors could not be radically resected or positive margins. In our patients, we give postoperative radiation because it is very difficult to decide if we could remove the entire tumor from tympanic bone, or if residual microscopic tumor has been left by en-bloc resection. This is because positive margin or microscopic margin may not be corrected. In the present study, we did not find tumor recurrence after a combined treatment.

In T3 or T4 patients, if we could not remove the entire tumor at the time of the operation, then some residual tumor could recur and could not be controlled by radiotherapy. Therefore, in those cases, the prognosis was very poor. Poor outcomes can occur in tumors that involve the facial nerve, dura, cranial nerve,

positive margin, and residual tumor⁽⁹⁾. The prognosis was most likely related to the extension of the tumor.

The authors performed lateral temporal bone resections in late case that is beyond surgery. It had high morbidity and high mortality. This has been confirmed by Lassig et al⁽¹⁵⁾. In addition, the extended temporal bone resection (ETBR) was described by Leong et al⁽¹⁶⁾. ETBR included a LTBR with additional resection of the adjacent involved structure.

A superficial parotidectomy has been suggested in occult parotid node metastasis, or tumors that are close to the anterior wall of EAC for adequate safety margin, but if the tumor involved the parotid gland, a total parotidectomy may be done⁽¹¹⁾.

Neck metastases are rare in carcinoma of the temporal bone. The incidence of neck node metastasis is 10 to 23%⁽¹¹⁾. Therefore, neck dissection should be performed if the neck node was palpable or the imaging was positive.

Conclusion

Patients with early-stage cancer can benefit from less aggressive surgical interventions without significant morbidity or mortality. The complete resection is the best treatment of choice, and postoperative radiotherapy for a microscopic residual tumor is highly recommended. In the advanced stage, if the surgery could not remove the entire tumor, it may have recurrence and the prognosis may not be good. As a result, the overall survival rate was 46.9% and the survival of stage I to stage III was 100%. Therefore, when treating infections of the external auditory canal, if they do not cure in one or two months, the doctor should look for the carcinoma of the external auditory canal for early diagnosis.

What is already known on this topic?

Primary cancer of the temporal bone is very rare. Currently, there is no staging system for temporal bone carcinoma accepted by AJCC. Therefore, the management is in controversy. It varies from local excision, sleeve resection, en-bloc resection, to aggressive surgery (such as subtotal or total temporal bone resection).

The aggressive surgery has a significant morbidity and mortality rate because the temporal bones are close to vital organs such as great blood vessels or the cranial nerve.

The best treatment was en-bloc resection with free surgical margin. The postoperative radiotherapy is still in controversy.

What this study adds?

Patients with early-stage temporal cancer can benefit from less aggressive surgical interventions without significant morbidity or mortality. The complete resection is the treatment of choice, and postoperative radiotherapy for a microscopic residual tumor is highly recommended. In the advanced stage, if the surgery could not remove the entire tumor, it may have recurrence and the prognosis is not good.

The most important is the early diagnosis. Most of the patients were initially diagnosed and treated as infection, such as otitis externa and chronic otitis media (longer than 3 months). Therefore, if the infectious disease of the external auditory canal did not respond to treatment in one or two months, the doctor should look for carcinoma of the external auditory canal for early diagnosis.

Potential conflicts of interest

None.

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มะเร็งของกระดูกเทมพอร์ลในโรงพยาบาลสงขลานครินทร์: การรักษาและผลการรักษา

วันดี ไช่มุกด์, ยุวดียา ปลอดภัย

วัตถุประสงค์: เพื่อศึกษาการดำเนินโรคและการรักษาผู้ป่วยมะเร็งของกระดูกเทมพอร์ล

วัสดุและวิธีการ: เป็นการศึกษาย้อนหลังในผู้ป่วยมะเร็งของกระดูกเทมพอร์ลที่ได้รับการรักษาในโรงพยาบาลสงขลานครินทร์ พ.ศ. 2545 ถึง พ.ศ. 2556 ผู้ป่วยทั้งหมด 32 ราย เป็นมะเร็งที่ใบหู 10 ราย มะเร็งของช่องหูชั้นนอก 22 ราย รักษาด้วยการผ่าตัดกระดูกเทมพอร์ลออกบางส่วน (*lateral temporal bone resection*) และตามด้วยรังสีรักษา และให้รังสีรักษาเพื่อบรรเทาอาการเพียงอย่างเดียวในมะเร็งระยะสุดท้าย

ผลการรักษา: ผู้ป่วยมะเร็งช่องหูชั้นนอกทั้งหมด 20 ราย คิดเป็นมะเร็งระยะแรก (*early stage; T1, T2*) ร้อยละ 36.4 และเป็นมะเร็งระยะท้าย (*late stage; T3, T4*) ร้อยละ 63.6 อัตราการรอดชีวิตร้อยละ 46.9 โดยมีค่าเฉลี่ยการอยู่รอดเท่ากับ 22 เดือน

สรุป: การรักษาที่ดีที่สุดของมะเร็งคือการผ่าตัดก่อนมะเร็งออกให้หมด และการให้รังสีรักษาในกรณีที่เหลือเนื้อเยื่อมะเร็ง สิ่งที่สำคัญที่สุดคือการวินิจฉัยให้รวดเร็วที่สุด
