ORIGINAL ARTICLE

Evaluation and Management of Carcinoma in External Auditory Canal

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Abstract:

A retrospectively reviews of 11 patients treated for carcinoma of EAC at our department between January 2000 and December 2018 was conducted to evaluate outcomes in treating carcinoma of external auditory canal (EAC) and to analysis factors which effect the prognosis of this disease. All patients underwent surgical treatment and the diagnosis confirmed by pathological examination. Results: There were adenoid cystic carcinoma (ACC) in 6 patients, squamous cell carcinoma (SCC) in 3 patients, adenocarcinoma (AC) in one patient, and verrucous carcinoma (VC) in 1 patient. The tumors were classified as Stage I in 5 cases, Stage II in 2 cases, Stage III in 3 cases, and Stage IV in 1 cases. Four patients underwent extensive tumor resection (ETR), 2 patients underwent lateral temporal bone resection (LTBR), 2 patients underwent modified LTBR, one patient underwent subtotal temporal bone resection (STBR), and 2 patients underwent only open biopsy. Besides, adjunctive procedures, including neck dissection, parotidectomy and pinna resection were performed when indicated. Ten patients received postoperative radiotherapy. By the end of follow up, two patients had died of their disease, 2 lost to follow up, 2 survived with the disease, and the rest survived disease-free. The median follow-up period was 24 months. Complete tumor resection appears to be an effective treatment for carcinoma of the EAC. Patients with SCC seem to have worse prognosis than those with ACC. Radiation therapy seems less effective for the disease than surgical treatment.

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Introduction:

Carcinoma of the external auditory canal (EAC) is a rare disease, with an annual incidence of approximately one to six cases per million people which accounts for less than 0.2% of all cancers in the head and neck area.[1] These carcinomas can originate from EAC or be an extension of tumors from the auricle. Owing to the rarity of this disease, experiences and reports in diagnosing and managing this disease are scarce. Although cases of cancer of the EAC or temporal bone have been reported previously[2,3]. Most of the reports are limited to squamous cell carcinoma (SCC). Comprehensive analyses of this disease have been limited.

Moreover, there is not a standardized staging system for EAC cancer so far. Yet an clearly defined staging system is important for investigation of this disease. To date, the most widely used system is the University of Pittsburgh staging system[4,5] which was first described by Arriaga et al. (1990)[6] and

modified by Moody[7].

The system is valid with respect to preoperative assessment of tumor extent and survival prognosis. The optimal management of patients with tumor of EAC also remains a topic of debate and controversy. Resection surgery with or without adjuvant radiotherapy is advocated by most otorhino-laryngologist as the standard treatment for this is ease[8].

Methodology:

Sixteen patients were seen and treated for carcinoma of the EAC at our institution between January 2000 and December 2018. All medical records were retrospectively reviewed for patient age,sex, presenting symptoms, CT findings, disease stage, surgical approach, histologic diagnosis, radiotherapy and follow up findings. The tumors in the 11 patients were classified ac-cording to the modified Pittsburgh staging system (Table 1) (Morris et al., 2012; Moody et al., 2000).

In this study, the types of surgery performed on the 11 patients for the disease included open biopsy, extensive EAC tumor resection, lateral temporal bone resection (LTBR),modified LTBR and subtotal temporal bone resection (STBR).Besides, adjunctive procedures, including neck dissection, parotidectomy and pinna resection were performed when indicated. Extensive resection of EAC tumors demands that EAC skin, cartilages and a portion of bones which have the possibility of being involved by the tumor be excised and that the negative margins confirmed by intra operative frozen bi-opsy. The procedure of LTBR removes the bony canal en bloclateral to the facial nerve. The extent of this resection includes the tympanic membrane, malleus, and incus.

The stapes, facial nerve, and inner ear structures are preserved. A modified TBR removes the EAC, leaving the uninvolved tympanic membrane intact.

Table –I: Modified Pittsburg staging system (Morris et al., 2012; Moody et al., 2000;Moffat and Wag staff, 2003).

Tumor	Description
T1	Tumor limited to the EAC without bony erosion or evidence of soft tissue involvement.
T2	Tumor limited to the EAC with bone erosion (not full thickness) or limited soft tissue involvement (G0.5 mm).
T3	Tumor eroding the osseous EAC (full thickness)with limited soft tissue involvement (G5 mm) 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 tumor with extensive soft tissue involvement (95 mm), such as involvement of temporomandibular joint or styloid process; or with evidence of facial paresis.
Lymph nodes :	
N0	No regional nodes identified
N1	Single ipsilateral regional node < 3 cm in size
N2a	Single ipsilateral regional node 3 -6 cm in size.
N2b	Multiple ipsilateral nodes
N2c	Bilateral or contralateral nodes
N3	Node>6cm
Tumor stage	
Stage I	T1 N0
Stage II	T2 N0
Stage III	T3 N0
Stage IV	T4 N0 and T1-T4 N+

Subtotal temporal bone resection (STBR) extends the dissection into the labyrinth, the cochlea, or both. The frozen-section pathology was used to estimate the safe margin in all types of surgery.

The goal of the surgery was to extirpate disease, achieve a negative margin and minimize morbidity and mortality. The choice of surgical approach was based on the extent of tumor involvement determined via physical examination, imaging studies and pathology tests. Extensive tumor resection was chosen for patients without bone erosion. LTBR or modified LTBR was chosen for patients with limited EAC bone erosion(not full thickness) and no involvement of tympanic cavity.

When the medial border of tumor was not closely adjacent to the tympanic membrane, the modified LTBR was our first choice. STBR was performed when tumor extended into the tympanic cavity. Neck dissection, parotidectomy and pinna resection were selected if involvement of cervical lymph node, parotid gland or pinna was suspected. Beyond that, radio-therapy was chosen as an adjuvant therapy for patients with stage III/IV tumors. Furthermore, radiotherapy was also cho-sen as the main treatment for patients who refused or were unsuited for a resection surgery.

Results:

The characteristics of the 11 patients including age, sex, pathological diagnosis, presenting symptoms, CT findings, tumor stage, surgery approach and adjunctive therapy, as well as treatment outcomes, are described below.

There were 4 males and 7 females who were enrolled in this study. All of the tumors originated from the EAC, except 1which extended from the pinna. The age of these patients ranged from 28 to 72 years at the time of surgery, and the median age was 46 years. Most of them were older than 40 years, and only 2 patients were 40 years old or younger.

Table –II: Distribution of structures involved

Sl. No	**Structure**	**Number of cases**
1.	EAC only	2
2.	Bone erosion	1
3.	Soft tissue involvement (> 5mm)	2
4.	Soft tissue involvement (< .5mm)	3
5.	Proauricular lyneph mode involvement	3
6.	Osseous EAC	1
7.	Middle ear	1
8.	TMJ	1
9.	Mastoid bone	1
10.	PJarotid gladn	1

All patients received pathological examination. Tumor types included adenoid cystic carcinoma (ACC, n=6), (SCC n=3), adenocarcinoma (AC, n=1) and verrucous carci-noma (VC, n=1). Among the 6 ACCs, 2 were well-differentiated and 4 were moderately differentiated. Within the subset of 4 cases that received parotidectomy and/or neck dissection, 2 had cervical lymph node metastases, and none had pathologic evidence of parotid invasion.

Table-III: Distribution of histopathological diagnosis

Sl.No	Histologic findings	Number of cases (n=11)
1	ACC	6
2	SCC	3
3	AC	1
4	VC	1

ACC = Adenoid cystic carcinoma, SCC = Squamous cell carcinoma,
AC= Adenocarcinoma, VC = Verrucos carcinoma

Otorrhea was the most common presenting symptom, occurring in 9 patients, followed by otalgia (n=7), hearing loss (n=6), ear stuffiness (n=5), tinnitus (n=6), and headache (n=1). None had facial paralysis at the time of presentation. External auditory canal mass was discovered in all patients at physical examination. In the 6 patients with ACC, all had otalgia and 3 had otorrhea. In the 3 patients with SCC, all presented with otorrhea, but only one complained of otalgia.

Table-IV: Distribution of Clinical Presentation

S.l No	**Symptoms**	**Frequency**
1.	Otalgia	7
2.	Otorrhoea	9
3.	Ear stuffy	5
4.	Tinnitus	6
5.	Hearing loss	6
6	Headache	1

This series of 11 cases, 8 had been misdiagnosed in the past. two were diagnosed as otitis externa, 2 as chronic sup-purative otitis media and 1 as papilloma in spite of the attempted biopsy. Other misdiagnoses included cholesteatoma, EAC granulation and neuralgia. Six patients had been previ-ously surgically treated, of them 1 presented with recurrentcancer despite the "complete" resection, and 4 with persistent cancer following "incomplete" resection.

Tumor staging was in accordance to the modified Pitts-burgh staging system which determine tumor stage post-operatively using imaging studies, intraoperative findings, and pathology report. Tumor in our cases were Stage I in 5 cases, Stage II in 2 cases, Stage III in 3 cases, and Stage IV in 1 cases, with 2 also showing N1 nodes involvement. No patients had distant metastasis.

Management All but 2 patients underwent surgical tumor excision. Two patients refused resection surgery, and received an open biopsy and radiotherapy. Modified LTBR was performed in 2 cases Four cases of ACC required only extensive tumor resection. LTBR was performed in 1 SCC and 1 ACC case and Only 1 patients underwent STBR. Two patients (1 AC and 1 SCC)underwent neck dissection. Once patient (with AC) underwent superficial parotidectomy and 3 patients (1 with stage IV SCC,1 with stage III AC and 1 with stage IV ACC) underwent total parotidectomy. Radiotherapy was performed in 9 patients.

There were no perioperative deaths in this series. Upon their last follow-up, 2 patients with stage IV SCC relapsed and subsequently died of the disease at 9 and 25 months after the surgery, respectively. 7 patients were lost to follow-up. The 2 patients who had only open biopsy survived with disease by their last follow up to 24 months. Facial paralysis was seen in one of these two patients.

Discussion:

Malignancy of EAC is a rare disease. The otolaryngologist may encounter few or even none EAC tumors in his career. Patients of such disease often initially present with non specific symptoms, such as ear discharge and otalgia. Because of the lack of specific clinical manifestations and minimal experience in diagnosing and managing this disease among many ENT doctors, misdiagnosis of EAC carcinoma occurs frequently, especially during its early stages[10.]

Otorrhea and otalgia are the most common symptoms of temporal bone tumors[11]. Because these symp-toms are similar to those of otitis externa and chronic suppurative otitis media, EAC cancer is easily misdiagnosed as those common otologic diseases. In this group of patients, the rate of initial misdiagnosis was 55%, with the most common being otitis externa. Other misdiagnoses included chronic suppurative otitis media, cholesteatoma, papilloma, granulation and neuralgia.

Some articles suggest (Ting and Chun-fu, 2013; Gidley,2013) that biopsy should be chosen for a pathological diagnosis if EAC carcinoma is suspected, especially if a case of otitis media or otitis externa does not respond to standard therapy.

However, the credibility of biopsy is doubtful. In this study, 1 patients were initially misdiagnosed as papilloma by biopsy, but were ultimately admitted for cancer of EAC based on the surgical pathological findings. The reliability of biopsy is closely related to the location and depth of specimen collection. Another important element in the accurate biopsy identification of EAC carcinoma is specimen size that should be sufficient for accurate his to pathological examination. So we suggest that multiple biopsies or an open biopsy be conducted, if EAC carcinoma is highly suspected. Additionally, CT and MRI should be considered, which not only help to provide information that can lead to an early diagnosis but are beneficial to estimating the extend of disease. CT is good at discovering bone erosion and MRI is excellent at showing the soft tissue lesion. The two imaging modalities are complementary[12]. If temporal bone erosion is seen on a CT scan, the patient is highly likely to have a malignant carcinoma of the temporal bone. In this group of patients, bone erosion was seen in 1 patients

by CT. For patients without temporal bone erosion on CT scan, a temporal bone MRI on may still be necessary and can reveal potential soft tissue lesions. In this study, high-resolution temporal bone CT-scan in one patient showed a left external auditory canal filled with soft tissue with no obvious bone destruction, but pathology showed stage IV ACC because of the extensive soft tissue invasion (>5 mm).According to the literature Moffat and,[13,14] SCCs are the most commonly observed and account for 80% of the temporal bone tumors, followed by ACC. Interestingly the result of our study show that ACC was the most common (50%), with SCC being the second most common (31%). The cause of our different findings is unknown and may be related to the small study sample size or racial differences. Although recent studies reported that surgical resection was the main treatment for EAC carcinoma, what types of surgery should be used is still in debate. To ensure a negative margin, Moffat et al.[15 - 18] suggested that an aggressive primary surgical treatment should be used even in an early stage tumor. Although there is an improvement of prognosis in patients who have undergone TTBR, this procedure is associated with significant morbidity which can severely impair patient's quality of life. So our team does not use this technique.

References:

1. Ting, Z.H.A.N.G., Chun-fu, D.A.I., 2013. WANG Zheng-ming.The misdiag-nosis of external auditory canal carcinoma. Eur. Arch. Otorhinolaryngol.270 (5), 1607-1613.

2. Gidley, P.W., 2013. DeMonte F.Temporal bone malignancies. Neurosurg. Clin.N. Am. 24 (1), 97e110.

3. McRackan, T.R., FANG Te-yung, Pelosi, S., et al., 2014. Factors associatedwith recurrence of squamous cell carcinoma involving the temporal bone.Ann. Otol. Rhinol. Laryngol. 123 (4), 235-239

4. Morris, L.G., Mehra, S., Shah, J.P., et al., 2012. Predictors of survival andrecurrence after temporal bone resection for cancer. Head. Neck 34 (9),1231-1239.

5. Gidley, P.W., Roberts, D.B., Sturgis, E.M., 2010. Squamous cell carcinoma ofthe temporal bone. Laryngoscope 120, 1144-1151.

6. Arriaga, M., Curtin, H., Takahashi, H., Hirsch, B.E., Kamerer, D.B., 1990.Staging proposal for external auditory meat us carcinoma based on pre-operative clinical examination and computed tomography findings. Ann. Otol. Rhinol. Laryngol. 99, 714-721.

7. Moody, S.A., Hirsch, B.E., Myers, E.N., 2000. Squamous cell carcinoma ofthe external auditory canal: an evaluation of a staging system. Am. J. Otol.21, 582-588.
8. Bacciu, A., Clemente, I.A., Piccirillo, E., et al., 2013. Guidelines for treating temporal bone carcinoma based on long-term outcomes. Otol. Neurotol. 34(5), 898-907.
9. Moffat, D.A., Wagstaff, S.A., 2003. Squamous cell carcinoma of the temporalbone. Curr. Opin. Otolaryngol. Head. Neck Surg. 11 (2), 107-111.
10. Ting, Z.H.A.N.G., Chun-fu, D.A.I., 2013. WANG Zheng-ming.The misdiag-nosis of external auditory canal carcinoma. Eur. Arch. Otorhinolaryngol.270 (5), 1607-1613.
11. Gidley, P.W., Roberts, D.B., Sturgis, E.M., 2010. Squamous cell carcinoma ofthe temporal bone. Laryngoscope 120, 1144-1151.
12. Horowitz, S.W., Leonetti, J.P., Azar-Kia, B., et al., 1994. CT and MR oftemporal bone malignancies primary and secondary to parotid carcinoma.AJNR Am. J. Neuroradiol. 15 (4), 755-762.
13. Moffat, D.A., Wagstaff, S.A., 2005. The outcome of radical surgery andpostoperative radiotherapy for squamous carcinoma of the temporal bone.Laryngoscope 115 (2), 341-347.
14. Fang-lu, C.H.I., Feng-ming, G.U., Chun-feng, D.A.I., et al., 2011. Survivaloutcomes in surgical treatment of 72 cases of squamous cell carcinoma ofthe temporal bone. Otol. Neurotol. 32 (4), 665-669.
15. Moffat, D.A., Wagstaff, S.A., 2003. Squamous cell carcinoma of the temporalbone. Curr. Opin. Otolaryngol. Head. Neck Surg. 11 (2), 107-111.
16. Masterson, L., Rouhani, M., Donnelly, N.P., et al., 2014. Squamous cell car-cinoma of the temporal bone: clinical outcomes from radical surgery andpostoperative radiotherapy. Otol. Neurotol. 35 (3), 501-508.
17. Graham, M.D., Sataloff, R.T., Kemink, J.L., et al., 1984. Total en blocresection of the temporal bone and carotidartery for malignant tumors ofthe ear and temporal bone. Laryngoscope 94, 528-533.
18. Jimbo, H., Kamata, S., Miura, K., et al., 2011. En bloc temporal bone resectionusing a diamond threadwire saw for malignant tumors. J. Neurosurg. 114(5), 1386-1389.
Liu, S.C., Kang, B.H., Nieh, S., et al., 2012. Adenoid cystic carcinoma of theexternal auditory canal. J. Chin. Med. Assoc. 75 (6), 296-300.