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OTOLOGY

Clinical outcome and prognostic factors in adenoid cystic carcinoma of the external auditory canal: proposal for a refined T-stage classification system

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Abstract

Purpose Although adenoid cystic carcinoma (ACC) of the external auditory canal (EAC) has a different pathophysiology from squamous cell carcinoma, the same staging system is used. The aim of this study was to propose a refined staging system, which is more suitable for ACC of the EAC.

Methods A total of 25 patients who were diagnosed with ACC of the EAC were reviewed. The modified Pittsburgh staging system (mPSS) that is universally used for temporal bone malignancy was refined for ACC (rPSS). The limited (< 0.5 cm) lateral soft tissue involvement was classified as T1 and extensive (≥ 0.5 cm) lateral soft tissue involvement as T2. The disease-free survival rate (DFSR) was assessed in the patients who underwent surgical treatment according to two staging systems; mPSS and rPSS.

Results When staging using mPSS, most patients (96.0%, $n = 24$) were classified as T4. However, when rPSS was used, T1, T2, T3, and T4 stage occupied 36.0% ($n = 9$), 40.0% ($n = 10$), 12.0% ($n = 3$), 12.0% ($n = 3$), respectively. There was no difference in DFSR according to the T stage using mPSS ($p = 0.466$). However, when rPSS was used, the DFSR showed significant correlation with the T stage ($p = 0.032$).

Conclusions Clinical T stage of mPSS was not sufficient to predict survival rate in ACC of the EAC, and we propose that the information on the lateral soft tissue involvement needs to be added to the existing staging system.

Keywords Adenoid cystic carcinoma · External auditory canal · Modified Pittsburgh staging system · Disease-free survival rate

Introduction

Carcinoma of the external auditory canal (EAC) is extremely rare, only occurring in approximately 1% of the head and neck cancers [1]. Most common histologic type of EAC malignancy is squamous cell carcinoma (SCC), and adenoid cystic carcinoma (ACC) accounts for 5% of EAC malignancy [2]. In usual, ACC shows a relatively slow clinical course, but long-term prognosis is relatively poor due to its high frequency of loco-regional recurrence and distant metastasis [3].

To the best of our knowledge, there has been few studies of ACC from the EAC (ACC-E). Among various treatment modality, complete resection of tumor followed by radiotherapy (RT) is a common treatment for ACC-E. Nevertheless, the recurrence occurs several years after surgery, and survival has been mainly dependent on significant distant metastasis, especially to the lung. Prognostic factors known for the recurrence of ACC-E include advanced T stage, duration of symptoms, pathologic findings including perineural or bony invasion, resection margin, and extent of surgical resection [4].

There are two main staging systems for SCC of the EAC, the modified Pittsburgh staging system (mPSS) and the American Joint Committee on Cancer (AJCC) 8th edition. In the AJCC 8th edition, cutaneous SCC of the head and neck was updated to include SCC of the EAC. The mPSS have been more preferred because it takes into account the unique anatomical constraints of the ear and temporal bone,

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allowing a better understanding of the patterns of spread and resectability [5]. Although the mPSS was developed and validated mainly for SCC of the EAC [6] and there is a significant difference between SCC and ACC in pathophysiology and prognosis, it was often used for ACC mainly because there has not been a validated staging system for ACC. Furthermore, T4 stage in the mPSS covers a wide range from extensive soft tissue extension (≥ 0.5 cm) and extension to medial wall of middle ear or skull base. Because the ACC arises from ceruminous glands that are not present in the bony EAC but in the cartilaginous EAC [7], the ACC has a greater tendency to show lateral soft tissue extension than the SCC. The direction of soft tissue extension (anterior, posterior, medial, or lateral) has a critical impact on prognosis, but mPSS does not differentiates the direction of extension in detail [8]. Therefore, it is required to make a refinement of this staging system that can effectively represent the clinical severity and prognosis of ACC-E.

We aimed to analyze the clinical characteristics of 25 patients with ACC-E who were managed with various treatment modality, and to propose a refined staging system which is more suitable for ACC-E.

Materials and methods

Study design and patients

Between October 1, 1997, and June 30, 2021, a total of 25 patients who were diagnosed with ACC-E in a tertiary referral center were included in this study. All patients underwent enhanced temporal bone computed tomography (CT), magnetic resonance imaging (MRI) and positron emission tomography to assess the extent of the lesions.

All tumors were staged according to the mPSS [6] and refined Pittsburgh staging system (rPSS). The mPSS classified the limited (< 0.5 cm) soft tissue involvement as T2 and the extensive (≥ 0.5 cm) soft tissue involvement as T4 without any directional information. The rPSS added an information on the lateral soft tissue involvement including periauricular region. We classified the limited (< 0.5 cm) lateral soft tissue involvement from the bony EAC as T1 and extensive (≥ 0.5 cm) lateral soft tissue involvement as T2 (Table 1). Except for the lateral soft tissue involvement, the mPSS was applied as is. For instance, the presence of anterior extensive soft tissue involvement such as temporomandibular joint (TMJ) was classified as T4 stage.

The histologic types of ACC, evidence of perineural or lymphovascular invasion, parotid invasion, and resection margin were determined by an expertise pathologist. The grading system of histopathologic type by Szanto [9] and Spiro [10], which represents the amount of the solid type, was also determined. Institutional review board was approved (IRB No. 2021-11-081) and the requirement to obtain informed consent was waived.

Treatment modality

Our basic policy for treatment was to perform surgery for all resectable cases that consented to treatment. Lateral temporal bone resection (LTBR) and superficial parotidectomy was performed in all cases with or without evidence of parotid gland invasion. In addition, selective neck dissection of level II (SND II) was conducted in most cases. Post-operative adjuvant RT was also performed in most cases using a dose of 50–60 Gy for the tumor target area. Otherwise, in cases with distant metastasis, chemoradiation was first administered for the primary and metastatic lesions.

Survival rate

We evaluated overall survival rate (OSR) and disease-free survival rate (DFSR) in all patients. We also evaluated which clinical factors were associated with the survival rate.

Table 1 Refinement of mPSS for ACC of the EAC

Stage	CT or MRI findings	Refined criteria
T1	Tumor limited to the EAC without bony erosion or evidence of limited (< 0.5 cm) lateral soft tissue involvement	Limited (< 0.5 cm) lateral soft tissue involvement
T2	Tumor with limited EAC bone erosion (not full thickness) or extensive (≥ 0.5 cm) lateral soft tissue involvement	Extensive (≥ 0.5 cm) lateral soft tissue involvement
T3	Tumor eroding the osseous EAC (full thickness) with limited (< 0.5 cm) soft tissue involvement, or tumor involving the middle ear and/or mastoid	None
T4	Tumor eroding the cochlea, petrous apex, medial wall of the middle ear, carotid canal, jugular foramen, or dura, or with extensive (≥ 0.5 cm) soft tissue involvement not laterally, such as involvement of TMJ or styloid process, or evidence of facial paresis	Extensive (≥ 0.5 cm) soft tissue involvement not laterally

mPSS Modified Pittsburg staging system, *ACC* adenoid cystic carcinoma, *EAC* external auditory canal, *CT* computed tomography, *MRI* magnetic resonance imaging, *TMJ* temporomandibular joint

Finally, the DFSR was assessed in the patients who underwent surgical treatment according to two staging systems: mPSS and rPSS.

Statistical analysis

All statistical analyses were conducted with SPSS Statics ver. 21 (SPSS, Chicago, IL, USA). The Kaplan–Meier method was applied to evaluate survival rates, and the log-rank test was used to investigate significant factors for survival rates. The cutoff for statistical significance was set at $p < 0.05$.

Results

Characteristics of patients

A total of 25 patients (13 male, 12 female) were included in the study. The mean age of patients at the time of diagnosis was 51.55 years (range 33–74 years). The most common clinical presentation was otalgia ($n = 12$, 48.0%), followed by mass in the EAC ($n = 10$, 40.0%), otorrhea ($n = 2$, 8.0%), and facial palsy ($n = 1$, 4.0%). The mean symptom duration was 40.60 months (range 1–240 months). The mean follow-up period from diagnosis until last visit was 69.5 months (range 2–246 months).

Among 25 patients, 21 (84.0%) underwent primary surgery. Other two patients (patient-10 and -11) could not undergo operation because of the advanced stage with distant metastasis. Patient-2 preferred RT for primary treatment due to poor general condition and patient-14 refused all treatments.

There were other loco-regional invasion including parotid gland ($n = 3$, 14.3%), middle ear ($n = 2$, 9.5%), mastoid ($n = 7$, 33.3%), dura ($n = 1$, 4.8%), and auricle ($n = 12$, 57.1%) in the patients who underwent surgical treatment. We defined auricular involvement when tumor shows a positive margin laterally more than 0.5 cm from the bony EAC, that is T2 stage in rPSS, and it comprised 57.1%.

Treatment modality

Table 2 presents the detailed information of patients. LTBR was performed in all of the 21 patients who underwent surgery. Further resection was required in 12 patients (57.1%) because of the positive resection margin in intra-operative frozen section biopsy. Superficial parotidectomy was conducted in all patients, and SND II was performed in 18 (85.7%) patients.

Among 21 patients who underwent surgery, 17 (80.9%) received adjuvant RT after surgery. The mean duration between surgery and adjuvant RT was 29.5 days (range 21–39 days). Of the remaining four patients who did not receive adjuvant RT after surgery (patient-4, -12, -13, -21), patient-4 showed distant lung metastasis and patient-13 showed loco-regional recurrence (post auricular mass) in follow up period. After recurrences, patient 4 received palliative chemotherapy for lung metastasis and patient-13 received adjuvant RT after complete excision of recurrence site. Patient-12 refused adjuvant RT due to poor general conditions and patient-21 did not require adjuvant RT as the small primary lesion was completely removed by LTBR with negative resection margin.

Among patients who could not undergo surgery, patient-2 received primary RT. The patient showed distant lung metastasis after 14 months and received palliative chemotherapy. Other two patient (patient-10 and -11) could not undergo surgery due to distant metastasis (patient-10, lung metastasis; patient-11, multiple bone metastasis). They were recommended for palliative chemoradiotherapy but refused it due to poor general condition.

Histopathology

Among three histopathologic patterns of ACC, the most common histological subtype was cribriform pattern, which was found in 20 (95.2%) patients. The solid pattern was present in 7 (33.3%) and the tubular pattern in 11 (52.3%) patients. When the Szanto grade was applied, there were 14 (66.7%), 4 (19.1%), and 3 (14.2%) patients corresponding to grade I, II, and III, respectively, and for Spiro grade, there were 14 (66.7%), 7 (33.3%), and no patients corresponding to grade I, II, and III, respectively. Perineural and lymphovascular invasion were found in 11 (52.4%) and 2 (9.5%) patients, respectively.

Recurrence and survival rates in all patients

Among 25 patients, one patient (patient-13) had loco-regional recurrence and underwent further resection of primary site with post-operative RT. Two patients (patient-10 and -11) already had distant lung metastasis at diagnosis, and five patients (patient-1, -2, -4, -5, and -6) developed distant lung metastasis during the follow-up period. All of them received subsequent chemotherapy, but all died with disease.

The OSR and DFSR of all patients was 48.2% and 47.5%, respectively. The patients who received primary surgical treatment showed significantly higher OSR and DFSR than those who did not. The OSR was 60.0% for patients who received primary surgical treatment and 0.0% for those who did not ($p < 0.001$). Likewise, the DFSR was 52.3% for patients who received primary surgical treatment and 25.0% for those who did not ($p = 0.016$).

Table 2 Detailed demographics in 25 patients of the study

Patients number	Sex/age (years)	Symptom	TNM stage (rPSS)	Symptom duration (months)	Initial treatment	Adjuvant RT	Recur duration	Distant metastasis	F/U (months)	Status
1	F/33	Otalgia	T1N0M0	48	LTBR + SP	+	57	Lung	246	DWD/DM
2	M/74	EAC mass	T3N0M0	12	RT		14	Lung	59	DWD/DM
3	M/46	Otorrhea	T1N1M0	36	LTBR + SP + SND	+	–	–	190	A
4	M/57	EAC mass	T2N0M0	120	LTBR + SP	–	42	Lung	131	DWD/DM
5	M/65	Otalgia	T2N0M0	24	LTBR + SP + SND	+	24	Lung	88	DWD/DM
6	F/42	EAC mass	T3N0M0	60	LTBR + SP + SND	+	3	Lung	72	DWD/DM
7	M/65	Otalgia	T1N0M0	48	LTBR + SP + SND	+	–	–	138	A
8	F/40	Otalgia	T3N0M0	24	LTBR + SP + SND	+	–	–	139	A
9	M/65	Otalgia	T2N0M0	12	LTBR + SP + SND	+	–	–	126	A
10	M/59	FN palsy	T4N0M1	1	None	–	–	Lung (at diagnosis)	27	DWD
11	M/67	Otalgia	T4N1M1	48	None	–	–	Lung, bone (at diagnosis)	2	DWD
12	F/73	Otalgia	T2N0M0	24	LTBR + SP + SND	–	–	–	54	A
13	M/51	EAC mass	T2N0M0	240	LTBR + SP	–	23	–	46	A/LR
14	F/59	Otalgia	T2N0M0	24	None	–	–	–	47	AWD
15	M/38	Otorrhea	T4N0M0	48	LTBR + SP + SND	+	–	–	42	A
16	M/40	Otalgia	T1N0M0	48	LTBR + SP + SND	+	–	–	34	A
17	F/61	Otalgia	T2N0M0	48	LTBR + SP + SND	+	–	–	32	A
18	F/48	EAC mass	T1N0M0	1	LTBR + SP + SND	+	–	–	28	A
19	M/42	EAC mass	T1N0M0	84	LTBR + SP + SND	+	–	–	26	A
20	F/55	EAC mass	T2N0M0	24	LTBR + SP + SND	+	–	–	23	A
21	M/36	EAC mass	T1N0M0	12	LTBR + SP + SND	–	–	–	22	A
22	F/77	Otalgia	T2N0M0	12	LTBR + SP + SND	+	–	–	16	A
23	F/57	EAC mass	T2N0M0	2	LTBR + SP + SND	+	–	–	4	A
24	F/69	Otalgia	T1N0M0	12	LTBR + SP + SND	+	–	–	7	A
25	F/62	EAC mass	T1N0M0	3	LTBR + SP + SND	+	–	–	7	A

EAC external auditory canal, *LTBR* lateral temporal bone resection, *STBR* subtotal temporal bone resection, *SP* superficial parotidectomy, *SND* selective neck dissection of level II, *DWD* dead with disease, *DM* distant metastasis, *A* alive without disease, *LR* loco-regional recurrence, *AWD* alive with disease

Factors related to survival rates in the patients who underwent surgical treatment

Regarding histopathological patterns, presence of solid subtype showed lower DFSR (16.7%) than absence of solid component (100%) ($p = 0.001$). The presence of cribriform ($p = 0.565$) or tubular ($p = 0.302$) subtype did not show significant association with DFSR. Likewise, high-grade histopathological type of Szanto grade III and Spiro grade II showed poor DFSR ($p < 0.001$, and $p = 0.001$). In addition, perineural invasion and lymphovascular invasion were not correlated with DFSR. Adjuvant RT after surgery showed higher DFSR (72.4%) than surgery only (33.3%, $p = 0.181$), and the patients who underwent SND II showed higher DFSR (87.7%) than those who did not (0%, $p = 0.019$). Finally, the patients who had symptoms less than the median duration (40.60 months) showed significantly higher DFSR (100.0%) than those who had symptoms longer than median duration (22.2%) ($p = 0.008$) (Fig. 1).

Modified staging system and the survival rates

We analyzed clinical T stage according to both mPSS and rPSS (Table 3). Then, clinical T4 stage occupied 96.0% ($n = 24$), and there was only one patient with T1 stage according to the mPSS. In the rPSS, T1, T2, T3, and T4 stage comprised 36.0% ($n = 9$), 40.0% ($n = 10$), 12.0% ($n = 3$), 12.0% ($n = 3$), respectively. Among T4 stage ($n = 24$) in mPSS, 18 cases (75.0%) were down-staged to T1 or T2 stage in the rPSS. Also, three patients with mastoid involvement and extensive lateral extension were down-staged to T3 stage. There were three patients with T4 stage in new staging system: one showed soft tissue involvement to TMJ, the other showed dural involvement, and the third had ipsilateral facial paresis.

Among 21 patients who underwent surgical treatment, DFSR was 100.0% in T1 ($n = 1$), 56.4% in T4 ($n = 20$)

Table 3 Clinical T stage according to the mPSS and rPSS

TNM staging	mPSS Cases (percentage)	rPSS Cases (percentage)
T1	1 (4.0%)	9 (36.0%)
T2	0 (0%)	10 (40.0%)
T3	0 (0.0%)	3 (12.0%)
T4	24 (96.0%)	3 (12.0%)
Total	25 (100.0%)	25 (100.0%)

mPSS modified Pittsburgh staging system, *rPSS* refined Pittsburgh staging system for adenoid cystic carcinoma

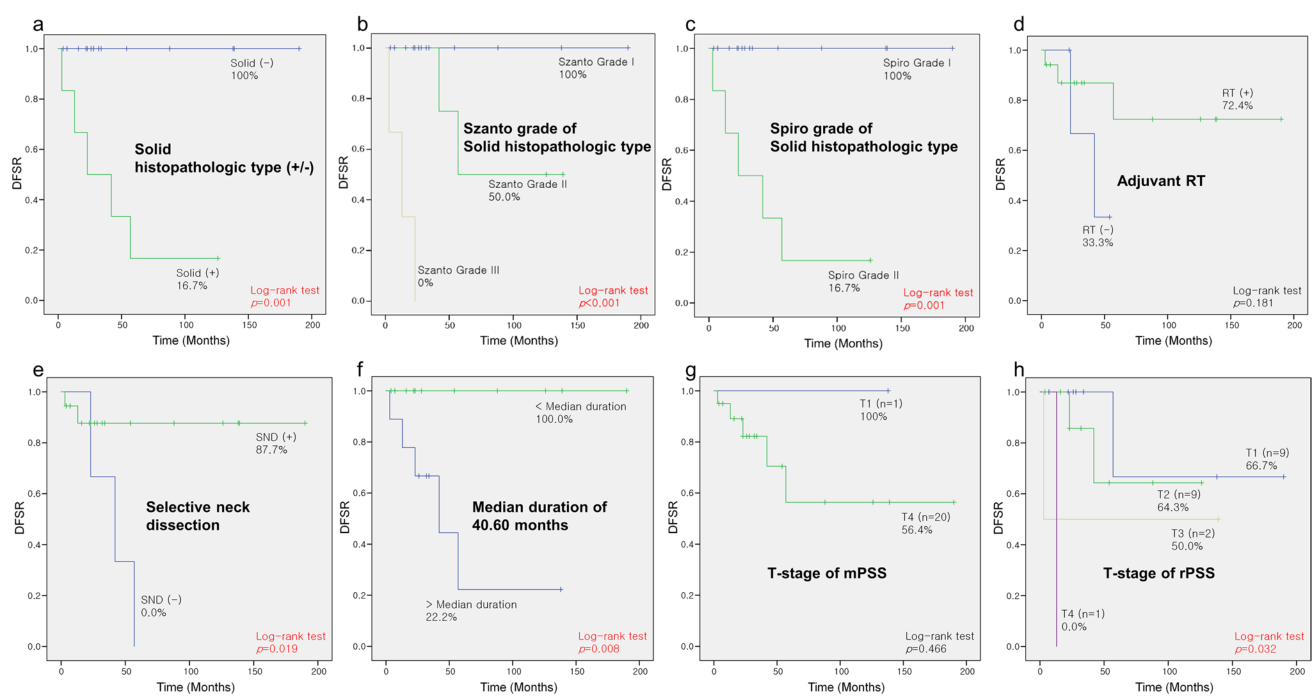

Fig. 1 DFSR according to various clinical factors. Solid histopathologic type, SND II, and prolonged symptom duration were significantly associated with DFSR. The DFSR was also significantly different according to T-stage of rPSS. **a** Solid histopathologic type. **b** Szanto grade. **c** Spiro grade. **d** RT. **e** SND II. **f** Symptom duration. **g** T-stage of mPSS. **h** T-stage of rPSS. *DFSR* disease free survival rate, *SND II* selective neck dissection of level II, *RT* radiotherapy, *mPSS* modified Pittsburgh staging system, *rPSS* refined Pittsburgh staging system

($p = 0.466$) when mPSS was used. However, DFSR was significantly different according to the clinical T stage of rPSS ($p = 0.032$) (Fig. 1). There was also significant difference of DFSR between early stage (T1 and T2, 63.3%) and advanced stage (T3 and T4, 33.3%) in the rPSS ($p = 0.041$).

Discussion

The staging system for SCC of the EAC has been established and further modified by Pittsburgh group [6]. The mPSS was utilized mainly for SCC of the EAC in most previous studies. Although the mPSS has also been applied to ACC-E, there are distinct differences in pathophysiology and disease progression between SCC and ACC. Thus, we propose a refined staging system for the ACC, which was partially modified from the mPSS.

The most common clinical symptom in our series was otalgia, accounting for 50% in accordance with other studies [4, 11], and the mean duration of otalgia was 31.0 months (range 12–48 months). Among them, some patients ($n = 6$, 54.5%) visited primary physicians for otalgia but only painkillers were prescribed. Ting et al. reported that 40.9% of patients with EAC carcinoma were misdiagnosed at initial [12]. In addition, ACC-E is hard to diagnose in an early stage until they become large enough to be easily seen because it is often subcutaneously grown. Since the tumors also have a typical indolent course, diagnosis is often delayed without a meticulous examination of the EAC [13]. Because our study shows that DFSR significantly worsened with longer symptom duration ($p = 0.008$), active identification of the cause through biopsy or imaging is required in patients with long-term otalgia that does not improve with conventional management.

We conducted superficial parotidectomy for all patients. In previous studies, Gou et al. suggested that superficial parotidectomy is necessary for all patients with ACC of the EAC, even it is early stage or there is no parotid involvement [14]. Jiang et al. also recommended superficial parotidectomy for ACC of the EAC due to close histological association between the EAC and the parotid gland [4]. The ACC-E tends to invade the parotid gland through the Santorini fissure even without bony invasion, not through metastatic spreading into the parotid lymph node. As preoperative radiographic imaging underestimates the extension of the tumors [15], superficial parotidectomy would increase the chance of a cure for ACC-E.

The node metastasis is rare in ACC of the head and neck area [16]. There were only 2 cases (8.0%) with clinical node metastasis among 25 patients in this study. Other studies showed similar rate (2.3–4.3%) of node metastasis in ACC-E [4, 14]. Min et al. reported that the node metastasis decreased survival rate in ACC of the salivary gland [17]. Conversely, Green et al. reported that the node metastasis in ACC-E did not have a negative impact on survival rate in multivariate analysis [18]. The prophylactic SND II for N0 neck was performed in 18 patients (85.7%), and the patients who underwent SND II showed significantly higher DFSR (87.7%) than those who did not (0%) in our study. Three patients with early-stage lesion (patient-1, -4, -13) did not undergo SND II, and all developed loco-regional recurrence or lung metastasis in follow up period, and our study revealed that the prophylactic SND II could increase DFSR without additional morbidity.

The evidence of adjuvant RT after surgical treatment in ACC of the EAC is not established yet. Generally, adjuvant RT in temporal bone carcinoma is indicated for nodal disease, extracapsular extension, margin positivity, and advanced disease of stage III/IV [19, 20]. However, Dong et al. suggested that adjuvant RT is necessary for all patients with ACC of EAC to reduce recurrence, given the high prevalence of perineural invasion [2]. Four patients (patient-4, -12, -13, and -21) did not undergo adjuvant RT after surgical treatment in our series. Among them, patient-4 and -13 developed recurrences in follow up period. Adjuvant RT after surgery showed higher DFSR (72.4%) than only surgery (33.3%) even though it did not show statistical significance.

Regarding histopathologic patterns, the presence of solid subtype showed lower DFSR (16.7%) than absence of solid component (100%) ($p = 0.001$). This result is consistent with previous studies on the prognosis of histopathologic patterns in ACC. Perzin et al. reported the solid subtype was associated with the poorest prognosis, followed by the cribriform and tubular patterns in ACC of the salivary gland [21]. Ishida et al. also reported that the solid subtype was observed in only advanced staged cases indicating poor outcomes in ACC of the head and neck area [16]. Therefore, when the histopathological type includes solid subtype, more active adjuvant treatment and close follow-up should be implemented compared to other histologic types.

Distant metastasis is relatively common in ACC-E, and it has a significant effect on survival. There were two patients with distant metastasis (8.0%) when the patients first presented, and distant metastasis developed after treatment in 5 out of 21 patients (19.0%). However, these patients all died with disease, in contrast to 100% OSR for patients without distant metastasis. Yibe et al. also reported that distant metastasis was the main cause of death in patients with ACC-E, because all patients without distant metastasis showed 100% survival rate in their study [22].

There have been several studies suggesting modification of the mPSS for SCC of EAC. Zanoletti et al. suggested subclassifying the T4 stage according to the direction of tumor invasion. Their results showed that tumors spreading anteriorly (parotid space and preauricular region) had a significant higher survival outcome compared to the tumors spreading to other directions (posterior, superior, inferior, or medial) in SCC of the EAC [23]. Otherwise, Breau et al. reported anterior canal wall tumors had a higher rate of local recurrence and decreased survival rate because the tumors on the anterior wall of the EAC have less impediment to spread beyond the temporal bone [24]. Shinomiya et al. proposed that T4 stage need to be subclassified into T4a and T4b (tumor extension to brain, internal carotid artery, or jugular vein) according to the prognosis [25]. Mazzoni et al. also suggested to divide T3 into T3a and T3b according to tumor extension [26]. All above literatures have focused on subdivision of T3 and T4 in the mPSS according to the prognosis because T3 and T4 includes a wide range of tumor extension.

In patients with ACC, since the tumor arises from ceruminous glands that are present in the cartilaginous EAC, lateral soft tissue expansion of 0.5 cm or more often occur to the auricle and peri-auricular region. In our study, the lateral extension showed a better prognosis than extension into other directions. For example, the patient-20 showed extensive lateral soft tissue involvement of tumor without medial or superior invasion. She underwent total auricular resection, followed by reconstruction with anterolateral thigh free flap (Fig. 2). When we added the directional information of soft tissue extension in the existing staging system, DFSR showed a significant difference according to the T stage. Thus, we propose a refined staging system (rPSS) for ACC-E, which adds a directional information of soft tissue extension to the mPSS.

Statistical analysis in this study is limited due to the small case series, but the case number is similar to other studies on ACC-E, which also have a low frequency [2, 4]. Our study is also limited by a relatively short mean follow-up period of 69.5 months. Ishida et al. reported both loco-regional and lung metastases may occur even 25 years after surgical treatment in ACC of head and neck area [16]. Jones et al. also reported that the local recurrence rate at 30 years was 100% in the retrospective study including the longest follow-up of 39.2 years in ACC of head and neck area [27]. Considering these characteristics, it is hard to cure, or even impossible to judge the cure from ACC. Therefore, a life-long imaging follow-up with gradual increase of interval may be recommended for ACC-E, because it take an extraordinarily long-term course.

Conclusion

ACC-E is a rare condition that can be misdiagnosed at initial. In our experience, complete resection accompanied with superficial parotidectomy, SND II, and adjuvant RT could improve the survival rate. Factors predictive of DFSR included solid histopathologic type, SND II and prolonged symptom duration.

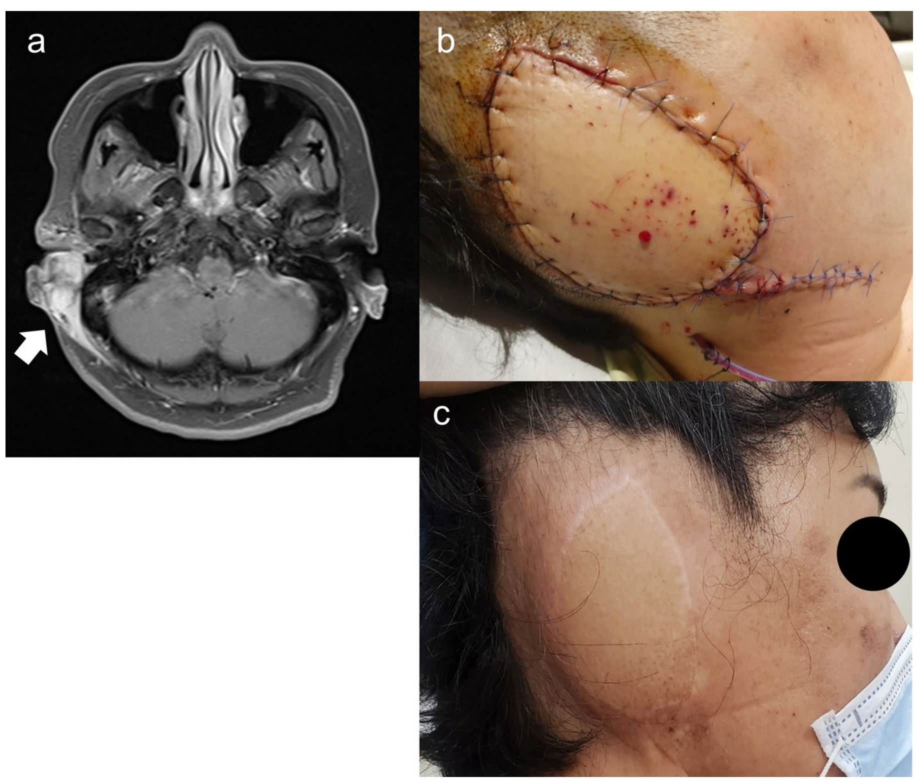

Fig. 2 Patient 20 who underwent total auricular resection and reconstruction with anterolateral thigh free flap. **a** T2-weighted MRI imaging showed extensive lateral soft tissue involvement of right side. White arrow, lateral auricular involvement of tumor. **b** Immediate post-operative wound. **c** Post-operative wound 2 years after surgery

In addition, clinical T stage of mPSS was not sufficient to predict survival rate in ACC-E. The information on the lateral soft tissue involvement needs to be added to previous staging system. Therefore, we propose a refined staging system that is more suitable for ACC-E.

Author contributions J-YL and Y-SC had full access to all the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis. Study concept and design: Y-SC; acquisition, analysis, or interpretation of data: all authors; drafting of the manuscript: J-YL; critical revision of the manuscript for important intellectual content: all authors; statistical analysis: J-YL; administrative, technical, or material support: Y-SC; study supervision: Y-SC.

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Data availability The data that support the findings of this study are available from the corresponding author upon reasonable request.

Declarations

Conflict of interest All authors have completed and submitted the ICMJE Form for Disclosure of Potential Conflicts of Interest and none were reported.

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