State of the art in temporal bone malignancies

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Purpose of review
To discuss the histological variants, tumor staging, work up and the latest trends in the treatment of malignancies of the temporal bone.

Recent findings
Because of the rarity of this subset of tumors, there has been no serious attempt to study tumor histologies of the temporal bone other than the squamous cell carcinomas (SCCs). The modified Pittsburgh tumor staging, though popularly used, was primarily developed only for SCC of the external auditory canal. Recent studies have shown that this staging is not without faults. There is also divergence of opinions regarding the surgical procedures to be adopted in treating temporal bone carcinomas. Moreover, the role of radiotherapy and chemotherapy has not been clearly defined.

Summary
In this review, we analyzed all the histological varieties of tumors that can arise from the temporal bone and classified them. The merits and demerits of the modified Pittsburgh tumor staging has been discussed outlining the need for further refining this system. The surgical approaches and their applications with respect to the extent of the tumor have been defined. The role of parotidectomy, neck dissection and adjuvant radiotherapy has been discussed.

Keywords
external auditory canal, facial nerve, radiotherapy, squamous cell carcinoma, temporal bone, temporal bone malignancies

INTRODUCTION

Temporal bone malignancies (TBMs) refer to a rare but distinct set of neoplasms originating within the confines of the temporal bone that account for approximately 0.2% of all head and neck malignancies. The annual incidence of such tumors is estimated to be between 1 and 6 per million population [1▪▪]. The incidence of TBMs at the Gruppo Otologico (Piacenza-Rome), which is a quaternary referral center for diseases of the skull base, is 130 (0.5%) cases for about 25 000 otological inpatient admissions over three decades. The rarity of these tumors can be adjudged by the fact that, in the last 15 years, there have been just about 20 series published dealing with squamous cell carcinomas (SCCs) of the temporal bone with a study population of over 20 patients, the largest series being that of Yin *et al.* [2] with 95 individuals. Nonsquamous TBMs are even more rarely reported, most of them being limited to case reports. Because of the rarity of these tumors, it has been difficult for a single institution to analyze the data and formulate an optimal evaluation and treatment strategy [3]. Another impediment to the study of TBMs is the lack of a universally accepted staging system. There is no recognized American Joint Committee on Cancer or International Union against Cancer staging system [4]. The T staging that is currently available and still in the process of evolution, was adopted from a series of articles that were published by the Pittsburgh University [5–9] for SCC of the external auditory canal (EAC). A literature review indicates that unlike SCC of the EAC, there has been no serious attempt to classify and stage TBMs of other histologies and sites of origin other than the EAC.

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Curr Opin Otolaryngol Head Neck Surg 2014, 22:000–000
DOI:10.1097/MOO.0000000000000035

KEY POINTS

- This article presents an overview of the present management approaches to TBMs.
- There is need to classify nonsquamous TBMs and malignancies arising from sites other than the EAC.
- The modified Pittsburgh classification needs to be analyzed in greater detail and possibly revised.
- TTBR may provide the same benefit as STBR followed by radiotherapy but involves higher risk to the patient.
- Surgery followed by postoperative radiotherapy is the mainstay of treatment for advanced lesions.

However, over the years, morbidity and mortality in temporal bone resections have reduced significantly because of advances in neuroimaging, skull base surgery and neuroanesthesia. Moreover, the otologist/skull base surgeon of today is more compliant with the general oncological principles and practices than a few decades earlier. From very poor prognosis a few decades ago, using a combination of surgery and adjuvant radiotherapy, up to 100% 5-year survival rates have been achieved by many authors for T1 and T2 tumors and up to 86 and 48% respectively in T3 and T4 tumors, which indicates the progress made globally in the control of this disease.

The temporal bone is host to both primitive and metastatic neoplasms. Most of the 'primitive' neoplasms affecting the temporal bone are epithelial neoplasms arising from the middle and inner ear including SCCs (Fig. 1), endolymphatic sac tumor (Fig. 2) and adenoid cystic carcinomas (Fig. 3) [10–12]. Secondary tumors may take origin as a direct extension from auricle, central nervous system, pharynx or salivary glands or even by blood-borne distant metastases. A classification of malignancies that can arise within the temporal bone based on cellular histology is presented in Table 1.

Origin and spread of tumors

TBMs have been reported to arise from practically all parts of the temporal bone (Table 2) including the EAC, middle ear, mastoid, endolymphatic sac, petrous apex and the internal auditory canal (IAC) [13–49,50▪,51–71].

TBMs can be locally aggressive due to the presence of numerous pathways in the temporal bone along which the tumor can spread from the site of origin. Instead of creating a barrier to the tumor's diffusion, the temporal bone in fact acts as a medium for microscopic diffusion through bony canals and intraosseous vessels [72▪] and nerves. The petrosquamous suture line, the fissures of Santorini, the foramen of Huschke, the stylomastoid foramen, the round and oval windows, the mastoid air cells system and the tympanic membrane all provide pathways for tumor spread. The bone plates over many structures like the jugular bulb, carotid, tegmen, fallopian canal and the labyrinth are thin and are hence vulnerable to tumor erosion.

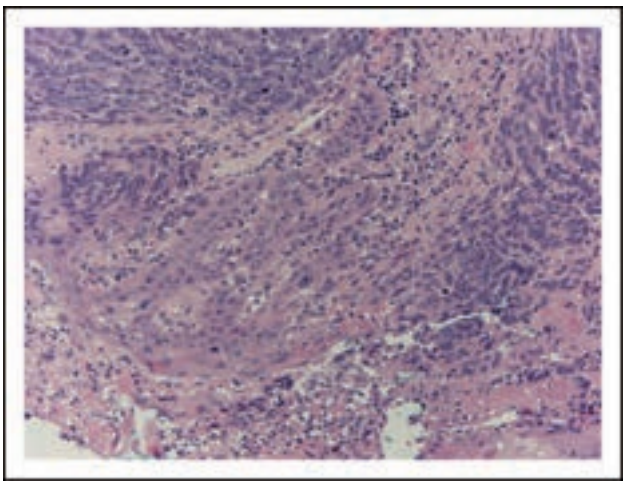

FIGURE 1. Squamous cell carcinoma of the external auditory canal showing moderately differentiated cells with keratinization in the center and areas of basaloid appearance (★) (200×).

Staging of temporal bone malignancies

Staging of TBMs is important for determining prognosis and outcome after treatment. However, a standardized staging system has not yet been

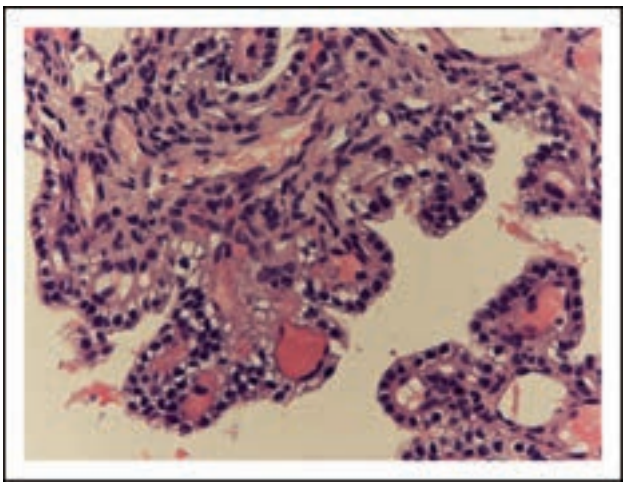

FIGURE 2. Endolymphatic sac tumor showing papillary and cystic pattern with bland clear cells (400×).

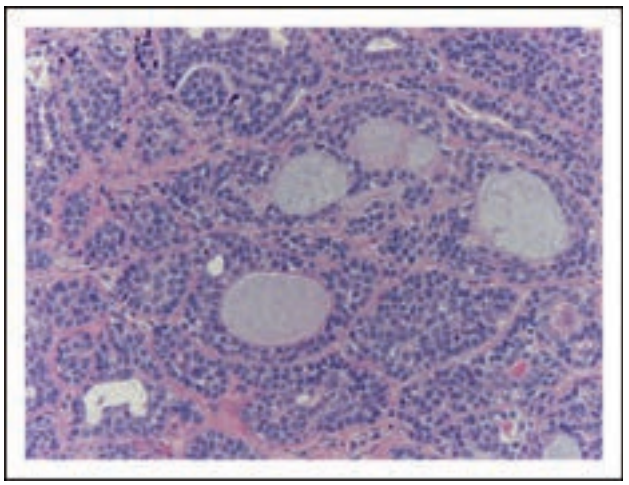

FIGURE 3. Adenoid cystic carcinoma showing prominent solid and cribriform pattern with a few pseudocystic structures (200×).

accepted internationally [73▪]. In 1980, Goodwin and Jesse [74] divided SCC of the temporal bone into three sites based on their origin: concha and cartilaginous meatus, osseous meatus, and middle ear. This is valid as malignancies arising from each of these subsites have different tumor behavior and it would be unwise to compare them using the same parameters. For instance, tumors arising from the concha and the cartilaginous meatus tend to invade the soft tissues earlier than bone and behave more like cutaneous malignancies in other parts of the body. The tumors involving the osseous meatus

Table 1. Classification of malignancies that arise within the temporal bone

Tissues of origin	Histologic type
Epithelial neoplasms	Squamous cell carcinoma
	typical
	verrucous carcinoma
	basaloid
	spindle cell (sarcomatoid)
	Basal cell carcinoma
	Ceruminal gland adenocarcinoma
	Adenoid cystic carcinoma
	Schneiderian carcinoma
	Endolymphatic sac tumor
Neuroendocrine neoplasms	Neuroendocrine carcinoma
	Carcinoid
Mesenchymal neoplasms	Rhabdomyosarcoma
	Fibrosarcoma
	Osteosarcoma
	Chondrosarcoma
	Malignant fibrohistiocytic neoplasm
	Malignant schwannoma (MPNST)
	Sarcoma (not otherwise specified)
Hematological neoplasms	Non-Hodgkin's lymphomas
	Plasmacytoma
Others	Langerhans cell histiocytosis
	Melanoma
	Metastases

MPNST, malignant peripheral nerve sheath tumor.

Table 2. Reported tumors in various subsites of the temporal bone

Subsite of TBMs	Tumors
EAC	Squamous cell carcinoma
	Verrucous carcinoma [13]
	Adenoid cystic carcinoma [14–16]
	Acinic cell carcinoma [17]
	Mucoepidermoid carcinoma [18,19]
	Merkel cell carcinoma [20]
	Malignant cylindroma [21]
	Ceruminous adenocarcinoma [22,23]
	Ductal carcinoma [24]
	Metastasis [25,26]
Tympanic membrane	Squamous cell carcinoma [27–30]
	Lymphoma [31]
	Metastasis [32]
Middle ear	Squamous cell carcinoma
	Adenocarcinoma [33–35]
	Amelanotic melanoma [36]
	Lymphoepithelial carcinoma [37,38]
	Hemangiopericytoma [39,40]
	Carcinoid [41–44]
	Malignant inverting papilloma [45,46]
	Langerhans cell histiocytosis [47,48]
	Metastasis [49]
Mastoid	Endolymphatic sac tumors [50▪,51,52]
	Langerhans cell histiocytosis [47,48]
	Plasmacytoma [53]
	Lymphoma [54]
	Sarcoma [55,56]
	Metastasis [57,58]
Facial nerve	Malignant schwannoma [59]
	Metastasis [60,61]
Petrous apex	Chondrosarcoma [62,63]
	Langerhans cell histiocytosis [64]
	Metastasis [57,65]
IAC	Squamous cell carcinoma [66]
	Epidermoid carcinoma [67,68]
	Metastasis [69–71]
Labyrinth	Metastasis [57]

EAC, external auditory canal; IAC, internal auditory canal; TBM, temporal bone malignancy.

tend to involve the bone anteriorly and posteriorly because of the thin skin. Similarly, tumors arising from the middle ear may involve the deeper parts of the temporal bone sparing the EAC. The prognosis and management of tumors of each of these areas are different. Over the years, many classifications have been proposed [5–9,75–77], but only the modified Pittsburgh classification for T staging for SCCs of the EAC has gained acceptance. There is still no consensus among authors for tumor staging in other parts of the temporal bone.

Many studies in the past have validated the usefulness of the modified Pittsburgh T staging and reported a good correlation between disease stage and prognosis [1▪▪,72▪,78–80]. However, recent data suggest that such staging is not perfect. According to the classification, facial nerve paralysis as a single factor could upgrade tumors to T4, but Moffat and Wagstaff [81] in their series did not find facial nerve involvement to be a significant prognostic factor (as also supported by other studies) [78,82,83]. The same authors also emphasize that no meaningful comparison can be drawn between different series unless the degree of histopathological differentiation is taken into account [84]. Similarly, Dean *et al.* [85] reported that perineural invasion of the facial nerve did not significantly influence disease-free survival following facial nerve resection. Ito *et al.* [80] in their study found that although extensive ($\geq$0.5 cm) soft tissue involvement was considered as T4 disease in the Pittsburgh staging system, this was not a prognostically significant factor. Zanoletti *et al.* [72▪] reported a better prognosis for T4 tumors extending into the anterior soft tissues (parotid and condyle), as opposed to T4 tumors spreading medially, posteriorly and inferiorly. In view of all these reports, it is clear that the modified Pittsburgh classification needs re-evaluation.

Another limitation of the Pittsburgh T-staging system is that it is not possible to apply it for tumors of other subsites of the temporal bone including concha, auricle and periauricular area as tumors in such subsites vary in presentation, extensions, histology and management. It is also uncertain whether the Pittsburgh classification, which was initially proposed for SCC of the EAC, is applicable to other histological types of malignancies arising in the EAC. In our series of 130 TBMs, malignant tumors other than SCC of the EAC accounted for 47% of all lesions (Table 3). Taking into consideration the vast number of tumors that go unstaged, there is presently also a need for a separate staging system for squamous and nonsquamous malignant tumors of other subsites of the temporal bone.

Preoperative management

In the following sections, the discussion is primarily directed toward SCC as this is the most common tumor of the temporal bone and it would be impossible to discuss management of all the histological variants of TBMs in this context.

Imaging is important in TBMs for accurate tumor and node staging as many patients have

Table 3. Histological varieties of temporal bone malignancies (n = 130) in our series

Type	Site of origin	No (%)
Squamous cell carcinoma and its variants	EAC, middle ear, auricle	69 (53.1%)
Chordoma	Clivus, petrous apex, cerebellopontine angle, middle ear, mastoid, tympanic bone, jugular bulb, occipital bone	18 (13.8%)
Chondrosarcoma	Petrous bone, clivus, jugular foramen	13 (10%)
Adenocarcinoma	Middle ear/mastoid, endolymphatic sac	11 (8.4%)
Adenocystic carcinoma	EAC, IAC, middle ear	7 (5.4%)
Sarcoma	Middle ear/mastoid, petrous bone	3 (2.3%)
Metastasis	IAC, auricle, geniculate ganglion	3 (2.3%)
Hemangiopericytoma	Middle ear/mastoid	1 (0.8%)
Ceruminous gland carcinoma	Mastoid	1 (0.8%)
Melanoma	Middle ear/mastoid	1 (0.8%)
Lymphoma	Middle ear/mastoid	1 (0.8%)
Localization of acute promyelocytic leukemia	EAC	1 (0.8%)
Aggressive papillary tumor	Petrous bone	1 (0.8%)

EAC, external auditory canal; IAC, internal auditory canal.

limited findings on physical examination. High-resolution computed tomography of the temporal bone offers the most accurate method for the evaluation of bone erosion due to malignancies. However, a reported limitation of CT is its inability to distinguish between tumor and fluid in the middle ear, soft tissue or mucosal thickening in the absence of bone erosion. Also, spread along fascial planes and neurovascular structures can be difficult to detect. MRI can provide excellent differentiation between soft-tissue tumor margin, muscle and soft-tissue infiltration, and can help in distinguishing tumor from obstructive inflammatory changes. In addition, obstruction of the sigmoid sinus and encasement of the petrous internal carotid artery are better detected on MRI than CT, because of the vascular signal void seen on precontrast MRI and the flow enhancement of the sigmoid sinus seen on postcontrast MRI [86▪]. Tumor extension, specially cranial spread into the middle and posterior cranial fossa and caudal spread into the infratemporal fossa, is also better detected on MRI. Enhanced T1-weighted spin-echo images with fat-signal suppression are most suitable for this purpose. MRI has also made it possible to confidently diagnose perineural spread of malignancies [87,88]. Fat saturation gadolinium-enhanced magnetic resonance (MR) scans are often capable of detecting subtle tumor tracking along the fifth and seventh cranial nerves, as well as other nerves that travel through the many foramina of the skull base, before the lesions have grown sufficiently large to affect the surrounding bone. Dental implants and densely calcified or ossified cartilages usually do not significantly degrade MR images as in CT [89]. Despite the advances in imaging, it is still a challenge to exactly determine the size and extensions of TBMs. In their comparison of preoperative radiographic with intraoperative findings in 26 patients, Leonetti *et al.* [75] reported that anterior and inferior growth pattern were accurately assessed radiologically, while underestimations of tumor extent were seen with posterior, superior and medial tumor extensions. Another study showed statistical evidence that erosions of less than 2 mm on the anterior wall of the EAC are not recognized preoperatively by CT [90]. In our experience, we have also often observed that peritumoral inflammatory changes show a much larger lesion on MRI than it actually is.

Surgical techniques for temporal bone malignancies

Surgery is the primary treatment of choice in TBMs. Radiotherapy is used as an adjuvant treatment to surgery except in advanced tumors requiring palliation. The role of chemotherapy is not established in TBMs. The surgical procedures employed for the resection of TBMs are briefly described below [91].

Lateral temporal bone resection

This is the primary surgery of choice in T1 and T2 tumors. The approach entails a complete canal wall up mastoidectomy with an extended facial recess opening. The EAC is resected en bloc along with the tympanic membrane, the malleus after disarticulation and removal of the incus, with the medial limit defined at the level of the incudostapedial joint [1▪▪] (Figs. 4 and 5). Some authors advocate that routine superficial parotidectomy be done with lateral temporal bone resection (LTBR), specially in T2 tumors [84,92,93].

Subtemporal bone resection

This is used in T3 and T4 tumors and is an extension of the LTBR. After the steps of LTBR are performed, this procedure extends medially in a piecemeal fashion and includes IAC identification, facial nerve exposure and removal of the otic capsule with preservation of the petrous apex. Care must be taken to remove adequate bone around the tumor. The capsule of the temporomandibular joint and, if necessary, the condyle of the mandible is resected when found involved. If the tumor extends into the mastoid and dural involvement is suspected, middle and posterior fossa craniotomies might be necessary to achieve adequate exposure. Generally, the dura is a good barrier for tumor spread and it must be removed only when necessary. If the dura is found infiltrated, its incision is undertaken and it is excised until free margins are reached. If the facial nerve is invaded by the tumor, it should be included in the specimen. The facial nerve may otherwise be

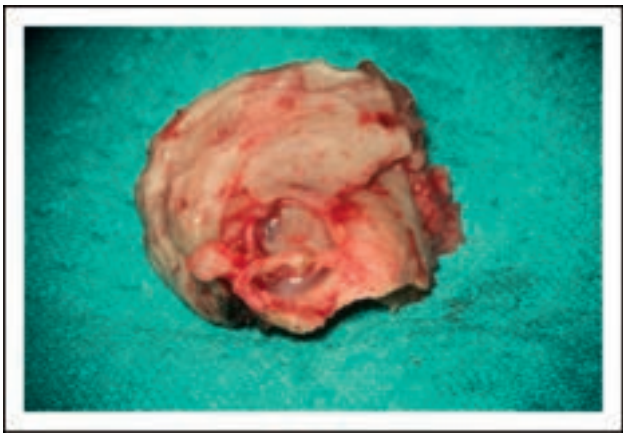

FIGURE 4. The specimen resected en bloc includes the external auditory canal, tympanic membrane and the malleus.

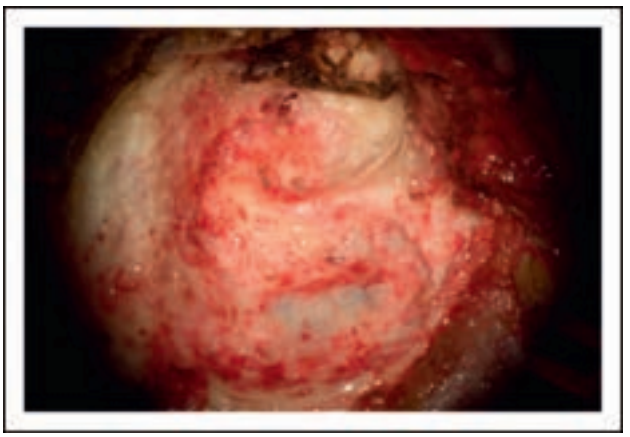

FIGURE 5. The temporal bone cavity after lateral temporal bone resection.

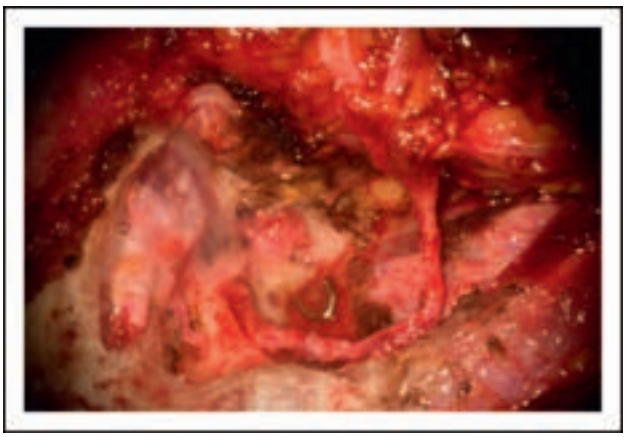

FIGURE 6. Subtemporal bone resection with exposure of the middle fossa dura, mobilization of the facial nerve, superficial parotidectomy. The jugular bulb was not resected because of its dominance on the affected side as seen in Fig. 7. A vascular clip was placed at the area of the jugular bulb where there was an element of suspicion of residual tumor to target the area during postoperative radiotherapy.

re-routed to give access to the tumor. The sigmoid sinus and jugular bulb are preserved unless infiltrated. If there is any area of uncertain tumor clearance around the jugular bulb or the lower cranial nerves it is advisable to leave a vascular clip *in situ* for the site to be identified and targeted during postoperative radiotherapy (Figs. 6–8).

Total temporal bone resection

This procedure is used in advanced T4 tumors. It may be performed with or without resection of the pinna. Bone is resected superiorly for 3 cm above the temporal line to expose the middle fossa dura and behind the sigmoid sinus by a similar amount to leave a residual margin of healthy bone. Medial dissection extends through the labyrinth and exposes the intrapetrous carotid artery. Inferiorly, the sigmoid sinus and jugular bulb are mobilized from surrounding bone. The sternocleidomastoid and digastric muscles are freed from the mastoid tip. At this stage in the procedure, the ascending ramus of the mandible is transected with a Gigli saw or a drill, and this and the head and coronoid process are dissected free and removed. A total parotidectomy is completed, and the specimen is removed en bloc. The residual tip of the petrous bone is then removed with a high-speed drill [84]. Resection of the carotid artery can also be accomplished if the contralateral cerebral blood supply has been proven to be adequate by angiography and preoperative balloon occlusion [94,95▪▪].

Selection of surgical procedure

The optimal surgical strategy for SCC of the temporal bone remains controversial. A review of the literature (including articles with a study population of 15 and over) shows that many authors often adopt an individualistic approach to primary tumor and neck nodes management (Table 4) [1▪▪,3,78,80,90,92,93,96–99].

Sleeve resection with retention of the pinna is still supported as a treatment of T1 tumors of the

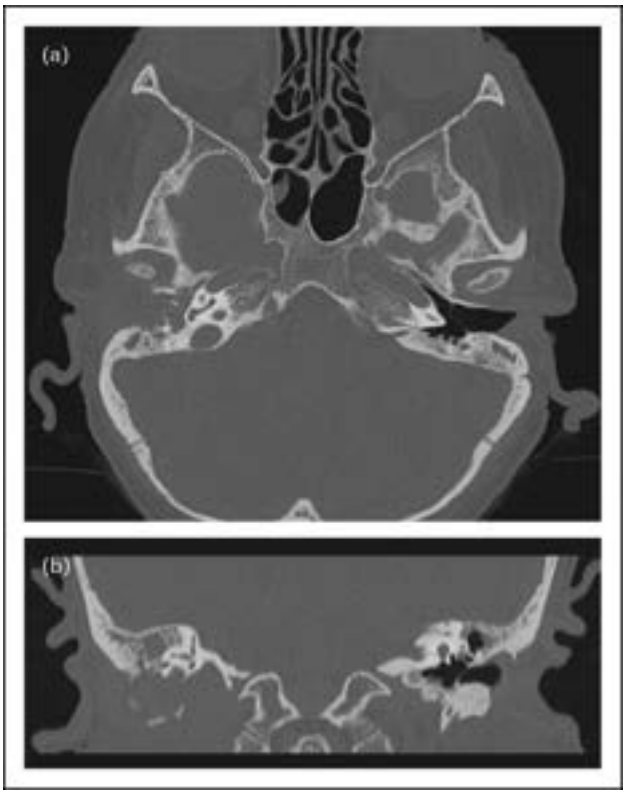

FIGURE 7. Preoperative axial (a) and coronal (b) views of the same patient as in Fig. 6 with T3 tumor of the external auditory canal.

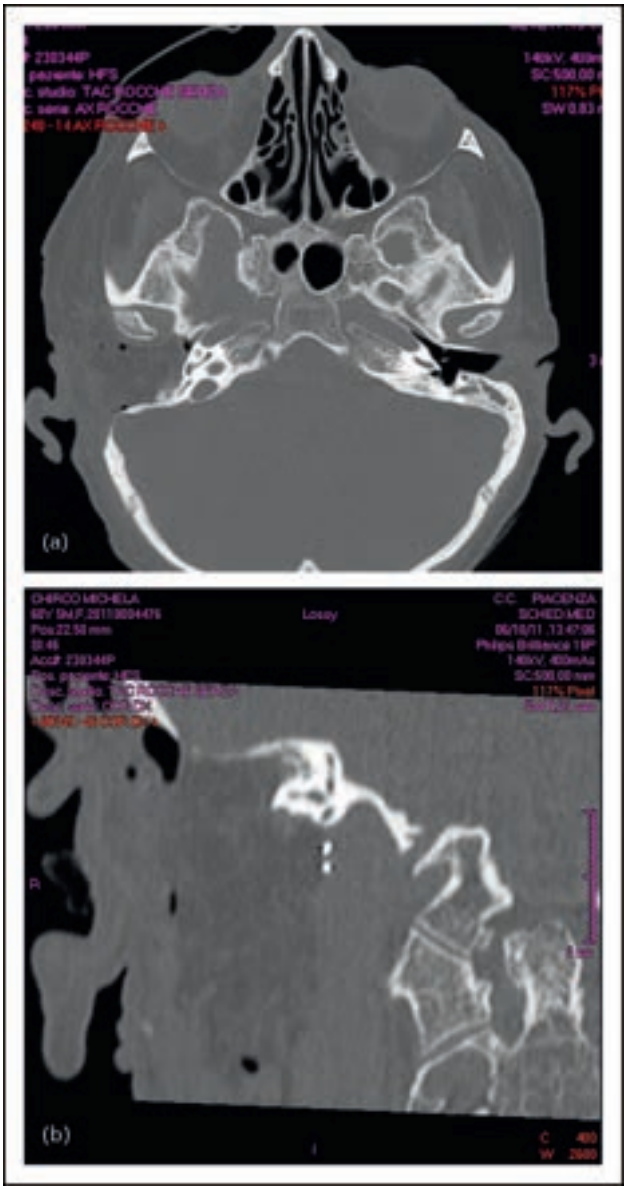

FIGURE 8. Postoperative axial (a) and coronal (b) views of the same patient as in Figs. 6 and 7 after subtemporal bone resection. The vascular clip can be identified near the jugular bulb in the coronal plane (b).

EAC [73▪,98], but it is difficult to believe that excision of the cancer field is not compromised [84]. Zhang *et al.* [73▪] have reported recurrence rates of 46.2% with sleeve resection for T1 and T2 lesions. We do not perform SRs at our center where LTBR is the accepted mainstay for T1 and T2 tumors. Although most authors [1▪▪,3,92] agree that LTBR alone or with superficial parotidectomy is adequate for T1 and T2 tumors, Moffat *et al.* [84] believe that LTBR with excision of the entire pinna, head, and ascending ramus of the mandible and with superficial parotidectomy is mandatory for these tumors, but their assumption is based on a limited number of cases. Both LTBRs and subtemporal bone resections (STBRs) are accepted techniques in T3 tumors [1▪▪,84,96]. In T4 tumors, although STBR is the surgery of choice [1▪▪,90,92,99], the application of total temporal bone resection (TTBR) in advanced T4 tumors is a matter of debate.

The management of more advanced tumors (T4) is particularly challenging as a result of the complex anatomy and the proximity of intracranial structures. In early studies, the surgical attempt to remove these lesions was associated with high rates of morbidity and mortality [100,101], but this is no longer true in the context of modern skull base surgery. The palliative benefits of TTBR like decreased pain and improved hygiene in comparison with radiotherapy have also been emphasized by Moffat *et al.* [102]. Although recent studies reported an improvement in terms of surgical morbidity and survival in patients who have undergone TTBR [94,102,103], this procedure is still associated with significant postoperative deficits. As previously reported by several authors, the surgical morbidity of STBR is usually limited to facial nerve palsy, and loss of hearing and balance, whereas additional morbidities of TTBR include potential damage to the cavernous sinus and internal carotid artery and postoperative deficits involving the third, fourth, fifth and sixth cranial nerves [96,104,105]. In agreement with other authors [8,82,106–109], we believe TTBR is unjustified because of the increased risk of morbidity and no proven survival benefit. STBR performed by a combination of en bloc and piecemeal resection techniques followed by postoperative radiotherapy could be a reasonable choice in patients with T4 tumors [82,96,105]. Prasad and Janecka [110], in their review of the English literature, reported that patients with carcinomatous invasion of the petrous apex, IAC, dura and brain had a poor estimated survival rate, although TTBR or dural excision was used.

En bloc versus piecemeal resection

Performing an en bloc resection according to fundamental oncological principles to obtain clear negative tumor margins would involve more danger to the surrounding intracranial structures and the cranial nerves while the facial nerve would have to be sacrificed. The alternative is to do a piecemeal resection wherein drilling is continued around the tumor till healthy bone appears after gross tumor removal. This is our preferred treatment policy in T3 and T4 tumors. The advantage of a piecemeal resection is that while the same amount of bone could be removed as in en bloc resection, exposure is better

Table 4. Surgical management of SCC of the external auditory canal by various authors

Author	No (n) and staging[a]	Surgical technique for tumor				Routine neck dissection				Routine SP/TP				Postoperative RT/CRT				Overall survival			
		T1	T2	T3	T4	T1	T2	T3	T4	T1	T2	T3	T4	T1	T2	T3	T4	T1	T2	T3	T4
Bacciu *et al.* [1▪▪]	45 (5 T1, 6 T2, 15 T3, 19 T4)	LTBR	LTBR	LTBR/ STBR	STBR ± MCR	–	–	–	+	–	±	±	+	–	–	+	+	100%	100%	86.2%	48.7%
Leong *et al.* [92]	35 (4 T1, 1 T2, 2 T3, 27 T4)	LTBR	LTBR	ETBR	ETBR	–	+	+	+	+	+	+	+	+	+	+	+	100%	100%	100%	41.4%
Lassig *et al.* [93]	17 (4 T1, 3 T2, 3 T3, 7 T4)	LTBR	LTBR	LTBR	LTBR	+	+	+	+	+	+	+	+	–	+	+	+	100%	100%	63%	54%
Hosokawa *et al.* [90]	15 (4 T1, 4 T2, 4 T3, 3 T4)	TCR	TCR	LTBR+ Cy	STBR+ Cy	–	–	–	–	–	–	–	–	–	+	+	+	NA			
Chi *et al.* [96]	25 (15 T1, 3 T2, 19 T3, 35 T4)	LTBR/LR	LTBR/ LR	STBR/ LTBR	STBR/ LTBR	–	–	–	–	–	–	–	–	–	+	+	+	100%	66.7%	21.1%	14.3%
Gidley *et al.* [78]	124 (24 T1, 14 T2, 2 T3, 31 T4)	LTBR	LTBR	LTBR	STBR after ICT	NA	NA	NA	NA	+	+	+	+	+	+	+	+	48%	28%		
Ito *et al.* [80]	16 (3 T1, 2 T2, 3 T3, 8 T4)	LCR/LTBR	LTBR	LTBR	LTBR/ STBR	–	–	–	–	–	–	–	–	+	+	+	+	NA			
Bibas *et al.* [97]	18 (3 T2, 3 T3, 12 T4)	–	LTBR	LTBR/ STP	STP	NA	NA	NA	NA	–	–	–	–	+	+	+	+	–	100%	59%	
Kunst *et al.* [98]	28 (15 T1, 2 T2, 2 T3, 8 T4)	LR	LTBR	LTBR	LTBR/ STBR	–	–	–	–	–	–	–	–	+/–	+	+	+	85%	46%		
Nakagawa *et al.* [3]	25 (1 T1, 3 T2, 5 T3, 16 T4)	LTBR	LTBR	LTBR	LTBR/ STBR	+	+	+	+	–	–	–	+	–	Preop CRT	100%	100%	80%	35%		
Lavieille *et al.* [99]	30 (12 T1+T2, 6 T3, 12 T4)	LTB±MCR	LTBR	STBR+MCR	STBR+ MCR	+	+	+	+	+	+	+	+	+	+	+	+	82%	67%	17%	
Moffat *et al.* [84]	39 (0 T1, 2 T2, 6 T3, 31 T4)	–	LTBR	LTBR/ STBR	TTBR	+	+	+	+	+	+	+	+	+	+	+	+	100%	100%	50%	34%

CRT, chemoradiotherapy; Cy, condylectomy; ETBR, extended temporal bone resection; ICT, induction chemotherapy; LCR, local canal resection; LTBR, lateral temporal bone resection; MCR, mandibular condyle resection; NA, not available; RT, radiotherapy; SP, superficial parotidectomy; TCR, total canal resection; TP, total parotidectomy.

under a microscope with less hazards to the cranial nerves. It is not always possible to obtain clear margins with piecemeal resection but this does not compromise tumor clearance if adequate bone is removed around the lesion. Though many reports [1▪▪,8,84,96,111] indicate poorer survival rates in resections with positive margins, this must not be viewed as a factor against piecemeal resection but more as a result of advanced nature of the disease. Further, Zanoletti *et al.* [72▪] raise the questions of how 'free' negative margins must be in order to be considered oncologically well tolerated in the case of SCC of the temporal bone, to what extent the surgery must be enlarged beyond the presumably free margins to ensure oncological radicality and whether 'free' margins can really be considered negative in bone. These are pertinent issues which future studies must focus upon.

Role of parotidectomy

Tumors of the EAC can involve the parotid gland either by direct extension or through nodal dissemination of the disease because one of the first echelon [84] lymph nodes involved in SCC of the temporal bone is the intraparotid or periparotid node. Preformed pathways around the EAC like the cartilaginous fissures of Santorini, the petrosquamous suture line and the bony foramen of Huschke are suspected to facilitate easy spread of tumor anteriorly. This is the main reason why some surgeons [83,99,112] prefer to associate superficial parotidectomy with LTBRs, specially if tumor is found to be eroding the anterior wall of the EAC. The incidence of parotid involvement in SCC of the EAC has been reported to be between 10 and 62% of patients [73▪,75,113–115]. But there is no recorded benefit of performing a routine superficial parotidectomy in terms of better survival rates. It is well known that nodal dissemination in TBMs is uncommon and hence many authors avoid performing a routine superficial parotidectomy for the sake of nodal clearance [79]. We do not include superficial parotidectomy as a routine procedure in all T2 cases and prefer to perform it in T2 tumors only if there is evidence of involvement of the anterior wall of the EAC. However, in T3 tumors, we routinely perform a superficial parotidectomy. In T4 tumors and when there is evidence of involvement of the parotid gland per se, we perform a total parotidectomy.

Role of neck dissection

Lymph nodes draining the EAC and the middle ear are the parotid and periparotid, pre and postauricular, submandibular, upper deep cervical and the retropharyngeal lymph nodes. Though lymph nodes involvement is relatively rare (10–36%) [8,84,85,92,116] in TBMs, the presence of nodal metastasis, seen in advanced stages, has an adverse outcome on survival. Although some studies have reported a poor survival rate in the presence of nodal metastasis [72▪,79,83,85], others have not found this to be statistically significant [1▪▪,4,92,117]. With this background it is difficult to justify a routine neck dissection in a N0 neck, specially in T1 and T2 tumors. A positive neck at presentation is a sign of aggressiveness of the primary tumor. Therefore, when there is clinical or radiological evidence of neck nodes involvement one must pursue an aggressive neck dissection. Zanoletti and Danesi [118] concluded that the prognosis of the clinically positive neck is bad even though failures never occurred in the neck but at the site of the primary. Hence, they recommend a more extended and adequate approach to the primary tumor. In N0 neck, we perform a frozen section of the level II lymph nodes and proceed with neck dissection only when this is positive for metastasis. In N+ necks, we perform routine neck dissection to involve the parotid and the level II lymph nodes.

Role of adjuvant therapy

Although radiotherapy has proven to be an effective adjuvant therapy, its role as a primary treatment modality for TBMs is not established. Postoperative radiotherapy is used to improve local and regional control of the disease. It is indicated in T3 and T4 tumors and also in T1 and T2 tumors where there is evidence of bone invasion, a positive margin, perineural invasion or nodal metastasis. In T3 and T4 tumors, many studies have reported better outcomes with adjuvant radiotherapy when compared with surgery alone [8,73▪,84,85,97,119,120]. Moreover, patients whose advanced tumors could not be fully resected showed little benefit from adjuvant radiotherapy in terms of survival [111]. Cristalli *et al.* [119] applied intraoperative radiotherapy and reported a high global local control rate of 73.3%. In important studies by Shiga *et al.* [121] and Sugimoto *et al.* [122] where concomitant chemoradiotherapy was used to treat advanced tumors, 5-year survival of 78% and a 2-year survival of 80% were reported, respectively. Despite the disadvantage of toxicity associated with this protocol, the high survival rates in advanced cancers indicate that this could be an effective treatment modality and further studies could be pursued in this direction.

Our therapeutic guidelines according to T staging is compared with those proposed by the Belgium Consensus Conference of March 2002 and by Kunst *et al.* [98] in Table 5.

Table 5. Treatment strategies for squamous cell carcinomas of the external auditory canal

Tumor staging	Belgium Consensus Conference	Kunst *et al.* [98]	Our guidelines
T1	LTBR and ND	LTBR and RT if margins are positive	LTBR and RT if margins are positive
T2	LTBR and ND followed by RT	LTBR and RT	LTBR and SP if tumor breaches periosteum of AW-EAC, followed by RT
T3	STBR and ND followed by RT	LTBR/STBR and RT. If N+ then ND; if parotid+ then TP	STBR+SP and RT. If N+ then ND; if parotid+ then TP
T4	Palliative treatment	Palliative treatment (T4b and c) or in T4a and some b; tumors: STBR/TTBR and SP, followed by RT. If N+ then ND; if parotid+ then TP	T4a and b–STBR with TP and ND, if necessary MCR or skin excision & reconstruction, followed by RT; T4c-Palliative treatment. *No TTBR for petroclival extension*.

AW-EAC, anterior wall of external auditory canal; LTBR, lateral temporal bone resection; MCR, mandibular condylar resection; ND, neck dissection; RT, radiotherapy; SP, superficial parotidectomy; STBR, subtemporal bone resection; TP, total parotidectomy; TTBR, total temporal bone resection. T4a indicates involvement of extracranial infratemporal fossa, skin, parotid; T4b, involvement of intrapetrous bone and extradural extension; T4c, meningeal or intradural involvement.

CONCLUSION

Despite the fact that significant attention has being focused on TBMs in the last few decades, there is still need to comprehensively study the entire gamut of histological varieties, refine the existing tumor staging system or develop novel ones and standardize treatment protocols. Radical surgery is the mainstay of treatment of TBMs in all tumor stages. Advances in neuroradiology, skull base surgical techniques and neuroanesthesia have made surgical resection of even advanced tumors in this area feasible with minimal morbidity. Adjuvant radiotherapy is indicated in T2, T3 and T4 tumors and is considered in T1 only in case of incomplete resection.

Acknowledgements

None.

Conflicts of interest

There are no conflicts of interest.

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