

Primary temporal bone cancer: A retrospective review

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INTRODUCTION

Primary temporal bone cancers (PTBCs) are rare and difficult to treat malignancies that remain poorly understood. They account for approximately 0.2% of all head and neck cancers, with an estimated incidence of 1 to 6 per 1,000,000 persons per year (1). Squamous cell carcinoma (SCC) is by far the most common type of malignant tumor of the temporal bone representing up to 80 percent of PTBCs (1-3).

Temporal bone squamous cell carcinoma (TBSCC) can present with common or non-specific otologic complaints such as otorrhea, otalgia or hearing loss. A more ominous presentation can involve facial nerve weakness/paralysis, parotid mass or neck mass. (3-6). While some of these symptoms are present in other conditions, it is important to have a high index of suspicion if they fail to respond to standard therapies. Facial paralysis and cervical lymphadenopathy have been found to be poor prognostic factors (2, 7). The initial step in the workup of TBSCC includes high-resolution imaging, such as CT and MRI, which aid in the three-dimensional characterization of the tumors as well. To date, there is no American Joint Committee on Cancer (AJCC)-approved staging system. However, the Modified Pittsburgh staging system is the most widely used system and it has been shown to predict survival (6, 8) (Table 1). It emphasizes the importance of nodal and facial nerve involvement as well as distant metastasis.

Management includes surgical resection, complex reconstruction, and adjuvant therapies.

To date, only a few centers have reported large experiences in the management of temporal bone cancer. The lack of large data sets makes it difficult to study and establish standardized management algorithms and guidelines. Combined efforts are desperately needed to better understand and treat this devastating disease.

The main goal of our study is to review our experience with the management of TBSCCs and to evaluate their outcomes in hopes to establish evidence-based institutional guidelines.

METHODOLOGY

Study Design: Retrospective case review

The study was conducted at a tertiary referral center. After IRB approval was obtained, adult patients with a diagnosis of primary temporal bone cancer from 2000 to 2015 were included in the study. The Modified University of Pittsburgh staging system (Table 1) was used to stratify patients by stage after final workup. Demographic, diagnostic, treatment data were reviewed and compared with existing literature.

A list of patients was generated using the Department of Pathology database by searching for keywords that included: ear, temporal, middle ear, ear canal. Next, the patients with electronic medical records were selected for the present study. The list was screened using the following inclusion and exclusion criteria.

Inclusion criteria

- Adult patients (age \geq 18 years old) diagnosed with pathology-proven primary temporal bone cancer
- Underwent treatment at Emory or affiliated institutions
- Had at least one follow-up visit after treatment

Exclusion criteria

- Cutaneous squamous cell carcinoma of the ipsilateral pre- or postauricular skin
- Cancer metastatic to temporal bone
- Patients with multiple primary cancers

RESULTS

TNM	Description
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 (<0.5 cm) soft tissue involvement
T3	Tumor eroding the osseous external auditory canal (full thickness) with limited (<0.5 cm) soft tissue involvement, or tumor involving middle ear and/or mastoid
T4	Tumor eroding the cochlea, petrous apex, medial wall of the middle ear, carotid canal, jugular foramen, or dura, or with extensive (>0.5 cm) soft tissue involvement; patients presenting with facial paralysis
N0	No regional nodes identified
N1	Single ipsilateral regional node <3 cm
N2a	Single ipsilateral regional node 3-6 cm
N2b	Multiple ipsilateral regional nodes \leq 6 cm
N2c	Bilateral or contralateral regional nodes \leq 6 cm
N3	Regional node >6 cm
M0	Absence of distant metastatic disease
M1	Presence of distant metastatic disease
Stage	
I	T1N0M0
II	T2N0M0
III	T3N0M0
IV	T4N0M0, T1-4N1M0, T1-4N2-3M1

Table 2 – Demographics and management

Total number of primary TBCs (N)	18
Age at presentation (years)	
Mean	70
Min	32
Max	79
Genre	% (n)
Males	66.7 (12)
Females	33.3 (6)
Risk factors	-
Tobacco	52.9 (9)
Alcohol	41.2 (7)
Preoperative imaging	-
CT	82.4 (14)
MRI	29.4 (5)
PET/CT	23.5 (4)
Treatment	-
Underwent treatment at our institution	94.4 (17)
Lost to follow up at our institution	5.5% (1)
Surgery	94.4 (16)
Chemotherapy	17.6 (3)
Radiation	52.9 (9)
Surgery alone	52.9 (9)
Chemotherapy alone	0
Radiation alone	5.9 (1)
Surgery + Chemotherapy	0
Surgery + Radiation	47.1 (8)
Surgery + Chemoradiation	17.6 (3)
Recurrence (1 year after treatment) (Stage III at presentation)	5.9 (1)

Table 3 – Symptoms at presentation

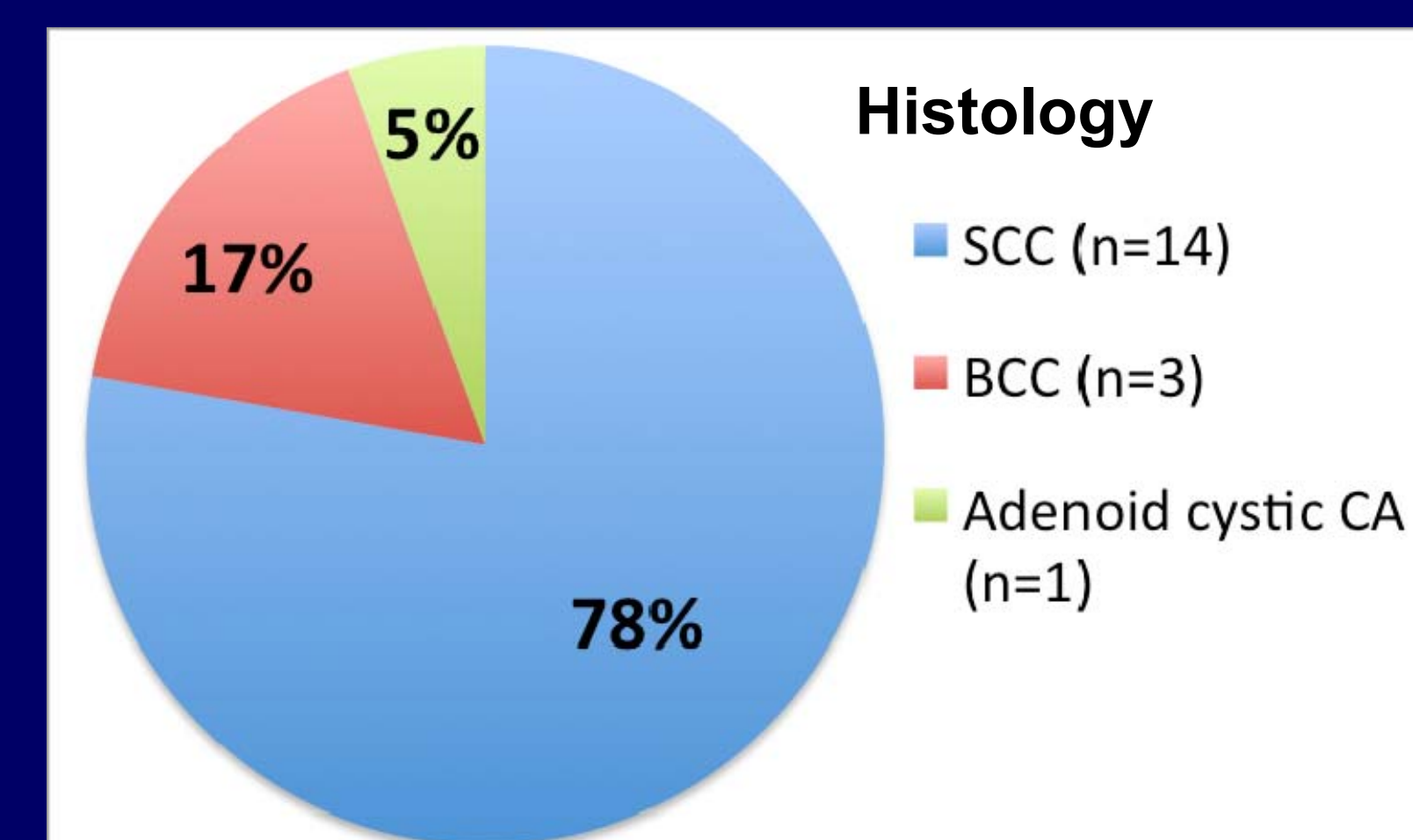
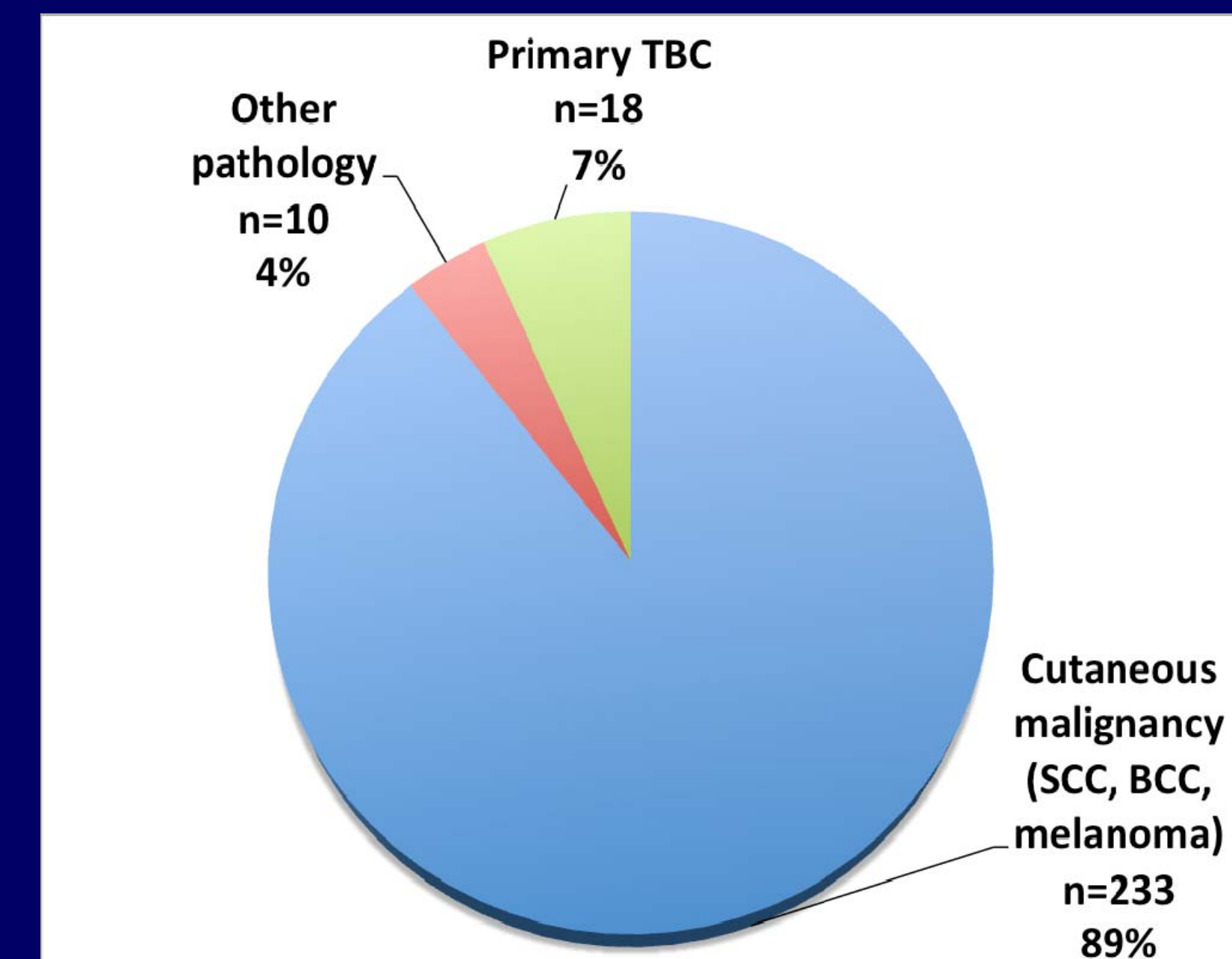
Symptom	% (n)
Hearing loss	58.8 (10)
Otalgia	35.2 (6)
Facial nerve paralysis	11.8 (2)
Neck lymphadenopathy	5.9 (1)
Parotid mass	5.9 (1)
Tinnitus	5.9 (1)
Vertigo	11.8 (2)
Otorrhea	64.7 (11)
Otorrhagia	11.8 (2)
Trismus	0
Dysphagia	0
Hoarseness	0
Tongue weakness	0
Odynophagia	0
Diplopia	0

Table 4 – Surgical treatment

Treatment	% (n)
Sleeve Resection	25 (4)
Lateral TB Resection	68.8 (11)
Total TB Resection	0
Petrosectomy	12.5 (2)
Neck dissection	18.8 (3)
Parotidectomy	43.8 (7)
Reconstruction	62.5 (10)
Free flap	6.3 (1)
Pedicled or rotational flap	56.3 (9)

Table 5 –Disease stage

Stage	% (n)
Stage I	22.2 (4)
Stage II	11.1 (2)
Stage III	22.2 (4)
Stage IV	38.9 (7)
Unable to assess	5.9 (1)



SUMMARY

- Our study confirms that primary TBC are rare tumors with only 18 patients found to have the disease over a 15 year period in a busy tertiary referral center.
- The majority of patients were of male gender (66.6%) with a mean age of 70 years of age.
- Approximately half of the patients had a history of either smoking cigarettes or drinking alcohol
- Otorrhea, hearing loss, and otalgia were the most prevalent presenting symptoms.
- The majority of the primary TBC found on this study were SCC (78%)
- Almost all patients underwent surgical intervention (94.4%)
- At least half of the patients underwent a combination of treatment modalities, with surgery followed by radiation being the most common combination (47.1%)
- The most common surgical management was LTBR followed by reconstruction with a pedicled/rotational flap.
- A large proportion of patients had late stage disease (Stage IV = 38.9%)
- Our data is consistent with existing literature

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