Bilateral Squamous Cell Carcinoma of the Temporal Bones

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ABSTRACT

We report a case of bilateral squamous cell carcinoma of the temporal bones in a 66-year-old woman who underwent a left subtotal petrosectomy and a right lateral temporal bone resection. Hearing rehabilitation was successfully achieved by fitting a bone-anchored hearing aid (BAHA) on the right side. We feel that in cases where the extent of the tumor permits a lateral temporal bone resection in at least one side, fitting of a BAHA can achieve a good outcome without resorting to tympanoplasty techniques that may compromise the medial end of the surgical resection.

KEYWORDS: Squamous cell carcinoma, bilateral, temporal bone, external auditory canal, bone-anchored hearing aid

Squamous cell carcinoma of the temporal bone is a rare and invariably aggressive neoplasm, the surgical treatment of which remains a challenge in the hands of skull base surgeons. The prognosis for patients with this disease has remained poor despite advances in surgical and radiotherapeutic techniques.^{1,2} While survival is related to stage at presentation, some patients with seemingly incurable disease survive and others with less advanced disease succumb.

We report a patient who had bilateral temporal bone carcinomas and discuss their management and the associated dilemmas posed by this type of tumor. Such cases are extremely uncommon and bring with them additional management problems. This article focuses on options of hearing rehabilitation. To our knowledge, this is only the ninth case of bilateral squamous cell carcinoma of the external auditory meatus that has been recorded in the literature

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and the majority of these were reported more than 30 years ago (Table 1).³⁻¹⁰

CASE REPORT

A 66-year-old woman was referred to our department with a long history of bilateral otorrhea. Over the previous 6 weeks she had experienced left-sided otalgia associated with a blood-stained discharge. On examination the right ear canal was slightly infected. In the left ear there was a friable mass arising from the deep part of the bony external auditory canal. No cranial nerve deficits were evident and no nodes were palpable in the neck. There was no trismus. Her pure tone audiogram showed an age-related, mild, sensorineural hearing loss in the right ear of 20 dB and a 55- to 120-dB mixed hearing loss in the left ear with a 40-dB air-bone gap. A biopsy confirmed the diagnosis of squamous cell carcinoma and both CT and MR scans of the temporal bones showed the tumor filling the ear canal, eroding the anterior wall and infiltrating the temporomandibular joint. There was no obvious abnormality in the right ear (Fig. 1).

The patient was advised to undergo a subtotal temporal bone resection. At surgery the tumor was found to have eroded the tympanic

plate anteriorly, infiltrated the parotid gland, and transgressed into the middle ear cavity. A subtotal petrosectomy was performed that included the cochlea, the labyrinth, and the tympanic and mastoid segments of the facial nerve. The majority of the tumor was removed en bloc with the remnants cleared in a piecemeal fashion. The glenoid fossa and disk of the temporomandibular joint were also resected. On completion, all that remained was the apical bone and the skeletonized internal carotid artery. Fat from the anterior abdominal wall was harvested and used to fill the resection cavity. Temporalis muscle was reflected inferiorly to act as a bed for a full-thickness skin graft that effectively closed the former external auditory meatus. A hypoglossal-facial nerve anastomosis to the lower division of the facial nerve was undertaken and a cable graft placed between the geniculate ganglion and upper division of the facial nerve in the parotid. In this way we hoped to restore volitional movement to the upper face. A lateral tarsorrhaphy was subsequently performed.

Histological examination of the operative specimen confirmed the diagnosis of a moderately differentiated squamous cell carcinoma and the final pathological stage was T₃N₀M₀ (Stage III) according to the revised Pittsburgh classification.^{11,12} Although postoperative radiotherapy was advised, the patient found the procedure too claustrophobic and refused further treatment.

A few months later the patient developed severe pain in her right ear, which appeared to be caused by otitis externa but failed to settle with antibiotics. Granulation tissue was present in the external auditory canal that extended to the annulus. Biopsy of this tissue revealed squamous cell carcinoma. A CT scan showed soft tissue thickening within the external auditory meatus together with focal bone erosion. There was no evidence of recurrence on the left side (Fig. 2).

The patient underwent a lateral temporal bone resection and superficial parotidectomy with preservation of all branches of the facial nerve. A bone-anchored hearing aid (BAHA) was fitted over the right mastoid as a single-stage procedure.

Figure 1 T1-weighted magnetic resonance image of the left temporal bone. Tumor fills the middle ear cavity and extends beneath the skull base.

Figure 2 Bone-windowed computed tomographic scan of the right temporal bone showing bone erosion of the lateral part of the bony external auditory meatus.

The temporalis muscle was made to act as a bed for a full-thickness skin graft to close the external auditory canal. Pathological examination of the operative specimen confirmed a moderately differentiated squamous cell carcinoma and determined the stage: T₂N₀M₀ (Stage II) according to the revised Pittsburgh classification. The report also showed a small focus of tumor at the excision margin. However, it is our practice to remove additional tissue following the resection of the main block as an additional safety margin. The patient's postoperative pure tone audiogram showed that she had a bone conduction threshold of 10 dB. The BAHAs were fitted 3 months following surgery.

Unfortunately, neither nerve graft on the left side worked. Consequently, the patient later had a fascia lata static sling inserted through a facelift incision. At the same time a gold weight was placed in her left eyelid and a dermodesis browlift was performed.

Four years later she presented with a level III right neck node. A fine-needle aspiration biopsy (FNAB) produced a mixed lymphoid smear and the lymphadenopathy subsequently resolved completely over a period of 3 months without any treatment at all. Six months later she developed two more nodes at level III on the right side. This time the FNAB was positive for metastatic squamous cell carcinoma. A type II right modified radical neck dissection was performed and histo-

pathological examination confirmed squamous cell carcinoma at levels II and III. She subsequently had a course of radiotherapy to the neck (50 Gy in 20 fractions). Her radiotherapy was complicated by the development of xerostomia, depression, and later still, the onset of polymyalgia rheumatica. Despite all this she remains free of disease 7 years after the development of her first tumor.

DISCUSSION

Squamous carcinoma of the temporal bone is a rare but aggressive tumor which usually has a poor prognosis. Survival is related to the stage of the tumor at presentation but is independent of histological grade.¹³ Patients who have positive nodal disease or recur inevitably succumb to their disease, often quickly. Outcome data are difficult to interpret because until recently there was no universally accepted staging system. The two main staging systems that are quoted in the literature are Clarke's modification of Stell's proposal^{14,15} and the revised Pittsburgh classification system.^{11,16} Furthermore, the majority of series are flawed or difficult to interpret because of their small size. Another problem is the fact that the majority of clinical studies regarding temporal bone carcinoma consist of mixed case loads of histologically different tumors and not solely squamous cell carcinomas.

Most studies would advocate some form of surgical resection of the tumor, with adjuvant radiotherapy for advanced stage disease. The extent and type of surgery has been debated in the literature and ranges from mastoidectomy to total petrosectomy.^{1,17,18} The only meta-analysis on the treatment of temporal bone carcinomas, Prasad and Janecka,¹⁹ showed that survival for advanced stage disease is poor, despite the extent of primary surgery. A recent analysis of the patients treated in our department determined that the overall 5-year survival was 45.3% (mean survival, 20.5 months) in our hands (Bibas and associates, unpublished data).

Table 1 Cases of Bilateral Temporal Bone Carcinomas Identified in the Literature (Staging of Tumors Based on Information Given in Case Reports and According to the Revised Pittsburgh Classification)

Study	Age at Presentation	Interval between Presentation of Tumors (months)	Right Temporal Bone	Left Temporal Bone	Follow-Up (months) since Diagnosis of Second Tumor (overall since first tumor, in brackets)	Status at Last Follow-Up
Juby ³	41	54	Chronic suppurative otitis media Stage: T ₃ N ₀ M ₀ Treatment: Radical mastoidectomy + radiotherapy (no information on dose) Complications: Total hearing loss	Chronic suppurative otitis media Stage: T ₄ N ₀ M ₀ Treatment: Radical mastoidectomy + radiotherapy (no information on dose)	No information	No information
Lederman ⁴ Hakata et al ⁵	No information 32	96	Stage: Not enough information Treatment: Radical mastoidectomy + brachytherapy (10 mCu) + radiotherapy (5400 rads with ⁶⁰ Co) Complications: Facial nerve palsy, total hearing loss	Previous mastoidectomy Stage: Not enough information Treatment: Preoperative radiotherapy (3200 rads with ⁶⁰ Co) + local excision + brachytherapy (10 mCu radium) + postoperative radiotherapy Follow-up: Left facial nerve palsy, Residual tumor on biopsy, radon brachytherapy.	18 (114)	Died from disease
Brookes ⁶	62	228	Chronic suppurative otitis media Stage: T ₃ N ₀ M ₀ Treatment: Extended radical mastoidectomy + postop DXT (telecobalt 5500 rads) Follow-up: Free of disease 16 years post-treatment	Neck mets, pulmonary mets Chronic suppurative otitis media, Waldenstrom's macroglobulinemia Stage: T ₃ N ₀ M ₀ Treatment: Extended radical mastoidectomy + postop DXT (telecobalt, no information on dose) Follow-up: Transient facial nerve palsy 10 days postop	6 (235)	Alive; no disease

Milford and Violaris ⁷	55	276	Previous subtotal perforation Stage: T ₃ N ₀ M ₀ Treatment: Radical mastoidectomy + postop DXT (no information on dose) Chronic suppurative otitis media Stage: T ₄ N ₀ M ₀ Treatment: Mastoidectomy + postop DXT (no information on dose)	Stage: T ₄ N ₀ M ₀ Treatment: Modified radical mastoidectomy + postop DXT (no information on dose) Chronic suppurative otitis media Stage: Not enough information Treatment: Radiotherapy (no information on dose) Complications: Total hearing loss Stage: T ₁ N ₀ M ₀ Treatment: Radiotherapy (66 Gray for both ears) Follow-up: Progression of tumor Stage: T ₃ N ₀ M ₀ Treatment: Subtotal petrosectomy + parotidectomy + reconstruction with abdominal fat graft and temporalis musculofascial flap + postop DXT (6300 cGy) Stage: T ₃ N ₀ M ₀ Treatment: Subtotal petrosectomy	0 (276)	No information
Snyman and Claassen ⁸	66	No information	No information	No information (24)	No information	Alive; no disease
Munk-Nielsen and Hansen ⁹	65	Synchronous	Stage: T ₂ N ₀ M ₀ Treatment: Radiotherapy (66 Gray for both ears) Follow-up: Regression of tumor Stage: T ₁ N ₀ M ₀ Treatment: Lateral temporal bone resection + tympanoplasty	No information	No information	Died with disease present in left ear
Wolfe et al ¹⁰	69	1.5		15.5 (17)		Alive; no disease
Bibas and Gleeson (current study)	66	6	Stage: T ₂ N ₀ M ₀ Treatment: Lateral temporal bone resection Recurrence: At 3 years in neck; treated with modified radical neck dissection and postoperative DXT	59 (65)		Alive; no disease

DXT, radiotherapy.

Hearing rehabilitation for these patients is difficult, as surgery inevitably results in considerable conductive hearing loss, at a minimum. After all, the primary intention of treatment is cure and any resection has to be radical. In one previous case, reported by Wolfe and colleagues,¹⁰ a lateral bone resection was undertaken on one side with preservation of the ossicular chain and tympanoplasty. The patient was left with a mastoid cavity. The mastoid and the area over the parotid and temporomandibular joint were skin grafted. This resulted in an air-bone gap of 0 to 25 dB 4 months after surgery. In our case, we performed a lateral temporal bone resection, removing both malleus and incus, and a full-thickness skin graft was used to cover the temporalis muscle that had been reflected inferiorly to reconstruct the defect. The pinna was retained and the patient was fitted with a BAHHA with very good results as the unmasked bone conduction threshold postoperatively at the time of fitting was 10 dB. Hearing rehabilitation can thus be optimized without potentially compromising the medial end of the surgical resection margin. It is our opinion that this should be the treatment of choice. For those patients whose pinna must be removed as part of the resection, a combined osseo-integrated pinna prosthesis and hearing aid should be provided.

CONCLUSIONS

Bilateral temporal bone carcinoma is a rare but aggressive tumor with poor prognosis despite aggressive treatment. Hearing rehabilitation is a secondary consideration, although it is an extremely important issue, particularly for those with impaired hearing on the contralateral side. We feel that in patients in whom it is possible to preserve cochlear function, BAHHA can give excellent results without compromising the extent of surgical excision, and this should be the preferred method of rehabilitation.

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