Squamous Cell Carcinoma and Basal Cell Carcinoma of the Ear Canal and Temporal Bone

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Introduction

The most common primary tumor of the ear canal and temporal bone is squamous cell carcinoma. It accounts for approximately 60–70% of all tumors affecting the temporal bone. Basal cell carcinoma of the ear canal is in a distant second place compared to squamous cell carcinoma.

Primary temporal bone carcinoma is very rare, and it accounts for only 0.2% of tumors in the head and neck [1, 2]. A myriad of tumor types affects the temporal bone [3]. The literature on temporal bone cancer is difficult to interpret since many papers combine multiple tumor types or multiple primary sites (ear canal, auricular, periauricular, or parotid) in order to demonstrate a substantial clinical volume [4–14]. Different tumor histologies have very different natural histories and survival outcomes. Squamous cell carcinoma (SCC), for example, has a much worse prognosis than either adenocarcinoma or basal cell carcinoma [8]. Additionally, most papers contain small numbers of patients, which prohibit drawing impactful conclusions, or survey long-time periods, over which diagnostic imaging and surgical techniques vary [15]. Some papers combine cases with different staging systems, e.g., the Pittsburgh staging system and the AJCC system for cutaneous or parotid malignancies [4, 16]. While these published reports do contain important observations, conclusions drawn must be tempered by remembering that these are tumors with widely varying natural histories, and these staging systems have widely varying criteria (e.g., a T2 parotid tumor is not equal to a T2 non-melanoma cutaneous malignancy nor is it equal to a T2 temporal bone tumor). As far as possible, this chapter will attempt to tease out the important clinical facts regarding squamous cell carcinoma and basal cell carcinoma in the ear canal and temporal bone.

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Incidence

In general, the cancer incidence rate is the number of new cancers at a specific site occurring in a specified population during a year. Exact numbers for the incidence of middle ear and temporal bone cancer have been elusive. From the University of Michigan at Ann Arbor, Furstenberg in 1924 made an extensive literature search and gathered 75 cases; he estimated the incidence of temporal bone cancer to be 1 case in 20,000 cases of aural conditions [17]. Tabb et al. reported 9 patients with cancer of the ear canal among 8500 consecutive otologic patients admitted to Charity Hospital in New Orleans over a 15-year period [18].

Lodge et al. estimated an incidence of 6/1,000,000 in 1955 [19]. This number was based on their experience with six cases over a 3-year period of time in Halifax, England, two men and four women, with ages ranging from 36 to 65 years. Despite these figures being over 60 years old, they are repeated frequently throughout the literature and often with incorrect citation.

In 1984, Morton et al. calculated the incidence of middle ear and mastoid cancers in the UK at around 1/1,000,000 persons per year [20]. The incidence of middle ear cancer is estimated at 1 case per 8,000–10,000 patients hospitalized with otologic disease [21, 22]. The incidence of aural cancer in otologic practice has been reported to be 1 in 20,000 patients [19]. These older series include many different tumor types, including paraganglioma. Although these papers are frequently cited, their results must be viewed with caution by contemporary readers.

Arena and Keen (1988) reported that the estimated incidence of temporal bone and middle ear cancer in the US is 0.08 cases/100,000 US standard population [23]. Their incidence is based on SEER data. Their paper makes note of a decline in the number of temporal bone resections performed: 3.8/year in the 1960s and 1970s and 1.2/year in the 1980s. They offered several possible explanations for the decline: decreased incidence of chronic suppurative otitis media, decreased use of head and neck radiation for benign

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disease, and an increase in fellowship-trained head and neck surgeons, leading to a dilution of cases.

In 1998, Mandolis et al. reported an updated report on the SEER database, revealing that there are roughly 200 new cases of temporal bone cancer per year in the US [14]. Manolidis et al. reported the incidence of temporal bone malignancy to be 1 in every 432 new patients seen by a mature skull base group and that tumors of epithelial origin were 1 in every 1167 new cases seen by the group [14]. Their series contained patients with many different tumor histologies and many different primary locations.

Contemporary twenty-first century data place the incidence of temporal bone cancer between 1 and 2 cases/1,000,000 persons/year. Chee et al. reported that the incidence is slightly higher at 2.1 cases/million population/year in Singapore [24]. A nationwide, retrospective study from Denmark of primary external ear canal and middle ear cancers published in 2008 revealed an incidence of 1.3 cases per million people per year [8]. While it is a small country (population 5.3 million at the time of publication), this represents the most accurate and up-to-date incidence in the literature.

The incidence of temporal bone cancer increases in patients who have received head and neck radiation. Lo et al. found an incidence of 0.15% (11 patients) of external auditory canal cancer in a cohort of nasopharyngeal cancer patients ($N = 7442$) who had received radiotherapy as part of treatment for nasopharyngeal tumor [25]. This rate is 1000 times higher than in the general population.

Squamous cell cancer of the middle ear is exceedingly rare. In a large, single institution review, Gidley et al. found only 3 cases of middle ear SCC out of 71 cases of SCC of the ear canal and temporal bone [5]. Using the Surveillance, Epidemiology, and End Results (SEER) database, Gurgel et al. reported on 135 patients with middle ear SCC over a 32-year span (or roughly 4 cases/year) [26].

Etiology

The exact etiology of cancers in the ear canal and temporal bone remains elusive. Unlike traditional head and neck cancer, smoking and alcohol use are not strong etiologic factors for development of ear canal and temporal bone cancer [27]. While both squamous cell and basal cell carcinomas are known to be caused by sun exposure, this cause surely could not account for cancers within the canal or middle ear.

Chronic otitis externa and malignant otitis externa have been linked to the development of SCC [28, 29]. Nyrop et al. reported that 30% of their patients had a prior history of chronic otitis externa [12]. Yin et al. reported that 12.6% of their patients had recurrent or long-term otitis externa and otitis media [30].

SCC developing in mastoid cavities has been documented in a few case series [31–33]. Monem et al. reported a case of SCC developing in a mastoid cavity of a person who used a chlorinated disinfectant (Eusol—Edinburgh Solution of Lime) to irrigate his mastoid cavity and theorized that this chemical compound might be a carcinogenic etiologic agent [34]. Meiteles and Conley described a case of SCC arising in a fenestration cavity [35].

Long-standing otorrhea (averaging 20 years or more in duration) is reported frequently in association with SCC and has been implicated as a potential cause of middle ear and temporal bone cancer [19, 21, 36–41]. Bradley and Maxwell (1954) found 35 cases of carcinoma in 7287 cases of chronic otitis media, i.e., 1 in 208 cases [42]. Chronic otitis media and externa have been reported in 7–61% of patients with SCC of the ear canal, middle ear or mastoid (Fig. 7.1) [2, 39, 43–48].

Cholesteatoma has been associated with squamous cell carcinoma of the middle ear and mastoid in a few case reports and small case series [31, 49, 50]. Intermittent chronic drainage of long-standing duration was common in these patients. Many patients did not seek additional treatment until facial paralysis prompted consultation with an otologist [31, 41]. Vikram et al. studied 3 cases of middle ear SCC out of 225 cases of chronic suppurative otitis media (CSOM) in India, and they found cholesteatoma associated with 2 of these cases and another presenting with aural polyp. Their report highlights the fact that cancer should be suspected when CSOM is suddenly associated with new symptoms such as severe earache, bleeding, or facial palsy [51]. A case of squamous cell carcinoma arising from external auditory canal cholesteatoma has been reported in the Japanese literature [52].

More recently, there has been research into the presence of human papillomavirus (HPV) in middle ear and temporal bone cancers [53, 54]. Jin et al. first reported the presence of HPV-16 in 11 of 14 patients with middle ear squamous cell carcinoma [55]. A history of long-term (more than 20 years) chronic otitis media with otorrhea was present in 13 of these patients. It should be noted that HPV DNA has been found in 36% of middle ear cholesteatomas [56]. Masterson et al. identified high-risk human papillomavirus (HPV16, 18, 31, and 45) in 3 of 14 patients with squamous cell carcinoma of the temporal bone [57]. The presence of HPV was not shown to produce a difference in disease-specific survival when compared to other temporal bone cancer patients without HPV.

Radiation is a known cause of malignancy. Exposure to radium in watch dial painters has been described in two case reports [58, 59]. Therapeutic radiation to the head and neck can produce secondary temporal bone malignancies [60]. Both squamous cell carcinoma and sarcomas are described as occurring in the temporal bone following radiation [25, 39, 60–64]. Goh et al. reported on seven patients who had nasopharyngeal cancers treated with radiotherapy who later developed a temporal bone cancer (five SCC, one osteogenic sarcoma, and one chondrosarcoma) [63]. Lo et al. reported that 0.19% of their nearly 8000 patients with nasopharyngeal cancer developed a temporal bone malignancy [25]. This indicates a relative incidence of 15 cases/10,000 patients,

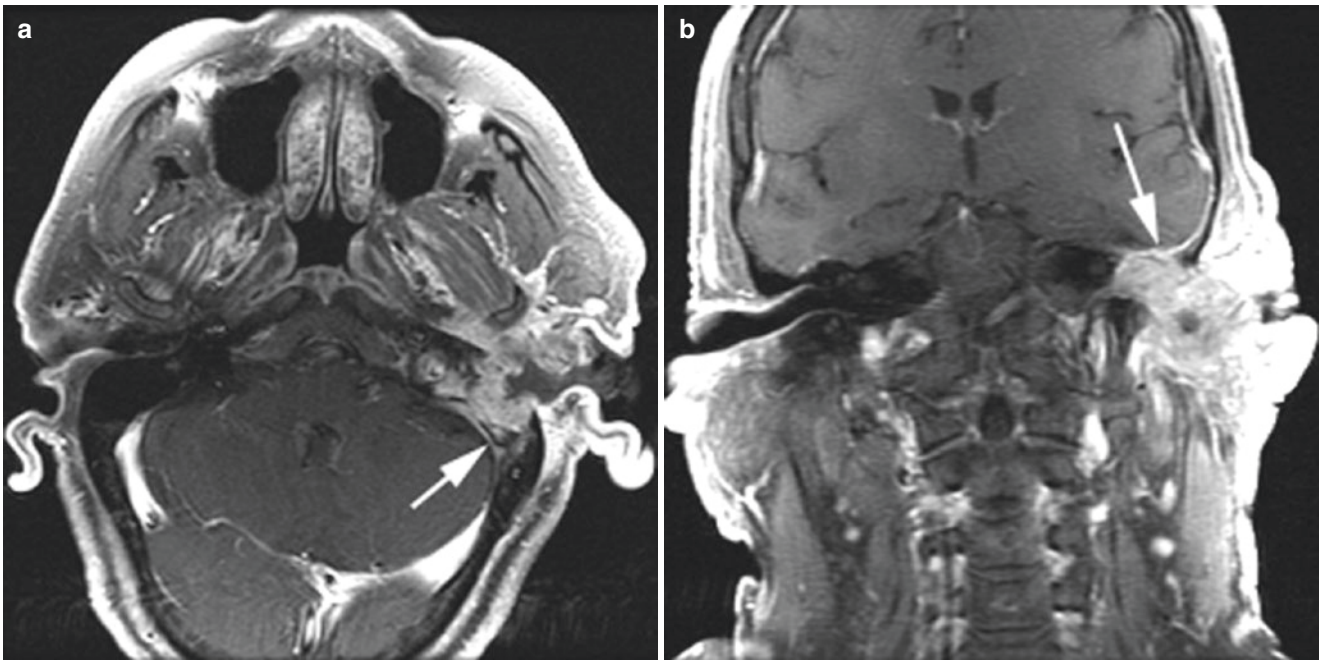

Fig. 7.1 Squamous cell carcinoma arising in a mastoid cavity. This 59-year-old man suffered from lifelong chronic otorrhea. Contrast-enhanced axial T1 MRI clearly showing dural enhancement (arrow) marking advanced-stage disease. (**a**) Axial. (**b**) Coronal

which is considerably higher (1500×) than the background incidence. Lambert et al. reported 13 cases of radiation-associated malignancy (RAM) involving the ear canal and temporal bone; 10 patients had SCC, while 3 had sarcoma [60]. Several reports include patients with a distant, prior history of radiotherapy [24, 47, 65].

Epidemiology

Squamous cell carcinoma of the ear canal, middle ear, and temporal bone typically occurs in older patients. Most large number studies have a mean age in the seventh decade of life. Depending on the study, the age can range from 21 to 92 years [5, 66]. Men are typically more commonly affected than women, but this finding does vary among recent studies. Higgins and Moody looked at 21 studies covering 348 patients, and they found an average age of 61.9 years [67]. In their study, the percentages of men to women were 60–40%, respectively [67].

Clinical Presentation

Cancers of the ear canal and temporal bone present with common symptoms such as otorrhea (24–100%), otalgia (19–81%), and hearing loss (2–75%) [5, 8, 15, 39, 46, 68–70]. The classic triad of otorrhea, otalgia, and hearing loss is found in only 10% of patients with cancer of the temporal bone. Other symptoms, such as trismus, facial weakness, dysphagia, and hoarseness, are seen much less commonly and are usually associated with advanced-stage disease.

Given that these symptoms are commonplace and usually caused by infection, many patients demonstrate symptoms for many months prior to diagnosis [5, 71]. Madsen et al. reported symptom duration was on average present for 13 months (range 1–74 months, median 6 months) [8]. Gidley et al. reported symptoms were present for 1–120 months prior to presentation, with ear pain having a shorter time to presentation (mean 5 months) than facial paralysis (mean 6 months), otorrhea (mean 9 months) or hearing loss (mean 37 months) [5]. Chang et al. reported symptom duration median of 12 months, with a range of 2–60 months [6]. Only four (out of 12) of their patients had a history of chronic otitis media [6].

Squamous cell carcinoma involving the ear canal will have an exophytic or ulcerated appearance (Fig. 7.2a) and can be mistaken for erythematous skin and granulation tissue. Basal cell carcinoma typically has an ulcerated appearance with rolled edges (Fig. 7.2b). Adenoid cystic carcinoma in its early stage is often subcutaneous and easily missed on a cursory ear examination (Fig. 7.2c). Melanoma in the ear canal is usually amelanotic (Fig. 7.2d).

These signs and symptoms can be confused with benign disease, such as otitis externa, otitis media, or cholesteatoma [72]. The differential diagnosis for disease in the ear canal should include osteomyelitis of the skull base (also called malignant otitis externa), pseudoepitheliomatous hyperplasia, and carcinoma [73, 74]. The differential diagnosis of a mass lesion in the ear canal includes benign lesions, such as adenoma or papilloma, and malignant lesions, such as squamous cell carcinoma, basal cell carcinoma, adenoid cystic carcinoma, mucoepidermoid carcinoma, adenocarcinoma,

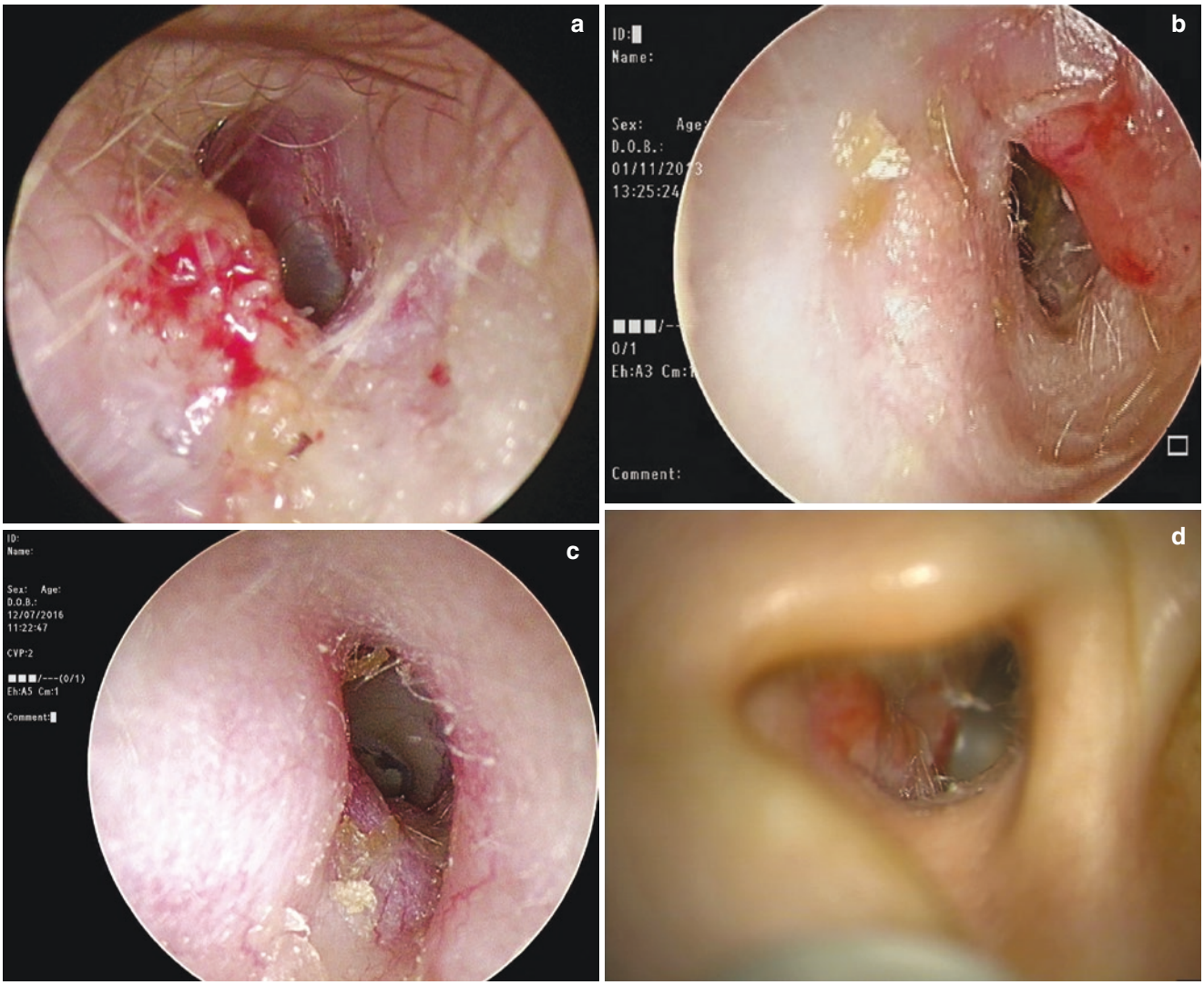

Fig. 7.2 Various cancers of the external auditory canal. (**a**) Squamous cell carcinoma of the left ear canal. (**b**) Basal cell carcinoma of the left ear canal. (**c**) Adenoid cystic carcinoma of the left ear canal. (**d**) Melanoma of the left ear canal

and melanoma. The malignant transformation of a benign papilloma to a squamous cell carcinoma in the ear canal has been described [75]. The temporal bone and ear canal are rare locations for metastatic cancers, usually from sites such as the breast, lung, prostate, or kidney [76–79].

The majority of patients with otitis externa or otitis media will respond to aural cleaning and eardrops or systemic antibiotics. Suspicion should arise when patients with these symptoms do not respond to standard therapy [72]. If patients with these symptoms do not respond to standard therapy, then any suspicious tissue should be sampled and sent for pathologic evaluation.

Only three cases of SCC isolated to the tympanic membrane have been reported in the English literature [80–82]. The classic clinical presentation is chronic drainage in an ear with granulation tissue on the tympanic membrane. Biopsy is essential but might only reveal dysplasia. In all three cases, definitive diagnosis was not established until surgical resection was performed.

Facial paralysis, when it occurs, is a very ominous sign and is linked with a poor prognosis [16, 46, 67]. The facial nerve generally becomes involved in one of the three ways: (1) extensive disease into the parotid to involve the nerve at the stylomastoid foramen, (2) erosion through the posterior canal wall into the mastoid or vertical segment of the nerve, and (3) erosion through the middle ear and along the middle fossa to involve the tympanic segment of the facial nerve [68]. All three modes indicate a large and aggressive tumor.

The incidence of facial nerve involvement ranges from 4% to 64% [5, 27, 45, 46, 69]. Gidley et al. reported a 15.5% incidence of facial nerve dysfunction at presentation in patients with SCC of the temporal bone which was present for an average of 6 months prior to presentation [5]. The incidence of facial paralysis is particularly high in patients with middle ear squamous cell carcinoma. Jia et al. found that patients with middle ear squamous cell carcinoma had a 55% incidence of facial paralysis [2].

Generally, right and left sides are equally affected. Gaudet et al. in a small study of only ten patients found that seven had left-sided tumors [83]. Gidley et al. in a study of 71 SCC of the temporal bone tumors found that 55% were on the left side and 45% on the right side [5]. In a study of 45 cases of SCC of the temporal bone, Bacciu identified a slight right-sided predominance, 57.8% right side versus 42.2% left side [46].

Thankfully, most cases of ear canal or middle ear SCC are unilateral. The earliest report of bilateral middle ear carcinoma was by Juby in 1957 [84]. Since then, there have been at least 23 case reports in the world literature of bilateral SCC of the ear canal and temporal bone [65, 85–105]. Most of these cases occur in patients with a prior history of chronic otitis media, and the tumors usually appear in a sequential and not simultaneous fashion.

Cervical Lymph Nodes

Cervical lymphadenopathy is a particularly poor prognostic sign associated with worse survival. The lymphatic drainage from the outer ear and ear canal is anteriorly to the parotid lymph nodes, posteriorly to mastoid lymph nodes, and inferiorly to levels II and III [108–112]. First echelon of nodes is the parotid gland and intraparotid lymph nodes [39, 113, 114]. Lymphatic drainage from the middle ear is poor [41] and runs to the mastoid and deep cervical nodes [82].

Level II is the most commonly involved neck nodes [5, 115]. Level III is a secondary drainage basin and usually does not become involved until level II has already been affected. Levels I, IV, and V are typically not involved from ear canal or temporal bone primaries [5, 114]. Level V nodes may become involved when tumors involve the postauricular skin. Submental lymph nodes are not involved.

Incidence of cervical lymph node metastasis has been reported to range between 10% and 23% [30, 45, 47, 113, 116–118]. In a large literature review, Rinaldo et al. reviewed 18 papers covering 491 patients and found an overall lymph node metastasis rate of 17.7% [114]. This finding refutes the claim that this tumor seldom metastasizes. Mazzoni et al. reported pathologically positive nodes 9 of 33 patients who underwent neck dissection. Their report highlights the fact that 5/29 clinically N0 necks had occult nodal metastasis [33].

Cervical nodal disease is an indicator of the aggressive nature of these tumors. Arriaga et al. reported 7/39 patients with positive lymph nodes [119]. In their series, patients with positive lymph nodes had only a 29% 2-year survival rate. In several series, patients with lymph node metastasis at presentation died of disease, usually within 24 months of diagnosis [39, 45, 120, 121]. Therefore, the presence of lymph node metastasis increases the staging so that T1-T3N1 is considered stage IV disease on the Pittsburgh staging system.

While death from temporal bone cancer is usually from local recurrence, cervical nodal disease is an important prognostic indicator of advanced and aggressive disease [114, 116]. Moffat et al. reported on a series of 39 patients with SCC of the temporal bone; 9 patients (23%) presented with lymph node metastasis, and all died within 27 months (mean 12.7 months). Of the remainder, 50% were alive and free of disease at a mean follow-up of 87 months [39]. It is important to note that patients with cervical nodal disease died of local recurrence and not regional or distant disease in their series.

Gillespie et al. reported that CT scan was typically adequate to detect nodal metastases; however, occult disease within the parotid gland was harder to detect [70, 122]. Choi et al. examined the rates of parotid and cervical lymph node metastasis and parotid invasion [123]. They studied 11 SCC and 10 adenoid cystic carcinomas of the external ear canal. They found that only two SCC (stages III and IV) involved parotid lymph nodes, and adenoid cystic carcinoma did not. SCC of the ear canal caused direct parotid invasion only in advanced-stage disease (stages III and IV), whereas 60% of patients with adenoid cystic carcinoma had direct parotid invasion, and it occurred with early-stage disease (stages I–IV) [123]. In their study, MRI missed direct parotid extension through the cartilaginous canal that was found on histologic study.

Diagnostic Imaging

Clinical examination is often limited by tumor filling the ear canal and obscuring the view of the tympanic membrane. Accurate imaging is necessary for staging and treatment planning. Computerized tomography (CT) and magnetic resonance imaging (MRI) each provides important information on tumor extent. Assessing tumor extent is essential since the main prognostic factor in temporal bone cancer is local extension [119]. Cross-sectional imaging (CT and MRI) are required for the staging of temporal bone cancers (Fig. 7.3). MRI and CT are often used in a complementary fashion [47, 69, 116, 124].

CT imaging has been correlated with pathologic findings in order to gauge the accuracy of the imaging. Arriaga et al. described that CT was accurate in 94 out of 96 comparisons [119]. They described one false negative, in which the CT did not diagnose soft tissue extension without bony erosion through the anterior canal wall, and one false positive, in which the CT predicted otic capsule involvement but mucosal biopsy was negative for tumor [119].

There are limitations of CT. CT cannot differentiate fluid and inflamed mucosa from tumor in the middle ear and mastoid [24, 68, 69]. Additionally, CT cannot distinguish inflammation from tumor when there is no adjacent bony erosion [68]. Anterior canal erosions less than 2 mm might be missed on CT [24, 125, 126]. However, bony canal defects greater than 2 mm are usually associated with tumor invasion through the bone and into anterior soft tissues [125]. CT scan

can miss microscopic extension of tumor from the anterior ear canal to the parotid gland [127]. When CT alone is used, radiographic underestimation and overestimation of disease has been reported. Leonetti et al. concluded that radiographic imaging underestimated disease in the middle ear and mastoid mucosa, tegmen tympani, middle fossa dura, and carotid canal [128]. Gillespie et al. found that CT underestimated disease anteriorly and felt that CT findings correlated best with advanced disease (T3 or T4) than with early-stage disease (T1 or T2) [70].

MRI has improved soft tissue detail and is essential for tumors that are producing nerve weakness or that invade the dura [70, 115, 124]. It has better resolution of disease affecting the parotid, temporomandibular joint, and petrous apex [70, 129].

PET/CT is important for patients with high-stage tumors (T3 or T4), since these patients are at higher risk of distant

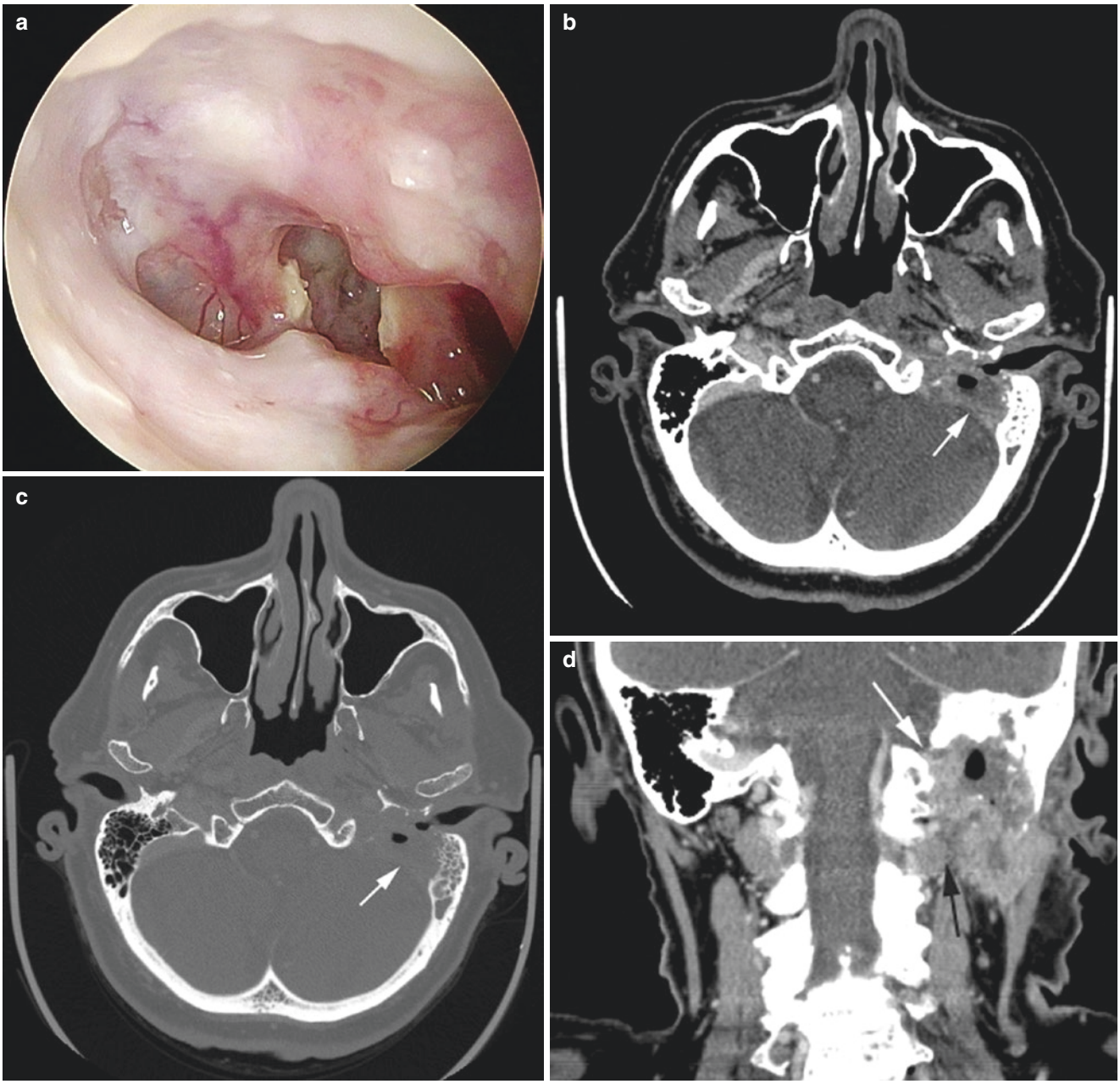

Fig. 7.3. Squamous cell carcinoma of the middle ear and temporal bone (stage IV) in an 82-year-old man. (**a**) Oto-endoscopic view. (**b**) Axial CT showing the mass filling the mastoid and middle ear. Note destruction of the posterior petrous bone (arrow). (**c**) Bone window CT showing the bony destruction from this tumor. (**d**) Coronal CT showing extension into the neck. White arrow marks hypoglossal canal. Black arrow marks disease at paraspinous muscles. (**e**) Contrast-enhanced axial T1 MRI at the same level as image (**b**). (**f**) Contrast-enhanced coronal T1 MRI showing no enhancement along the middle fossa dura at the same level as (**c**)

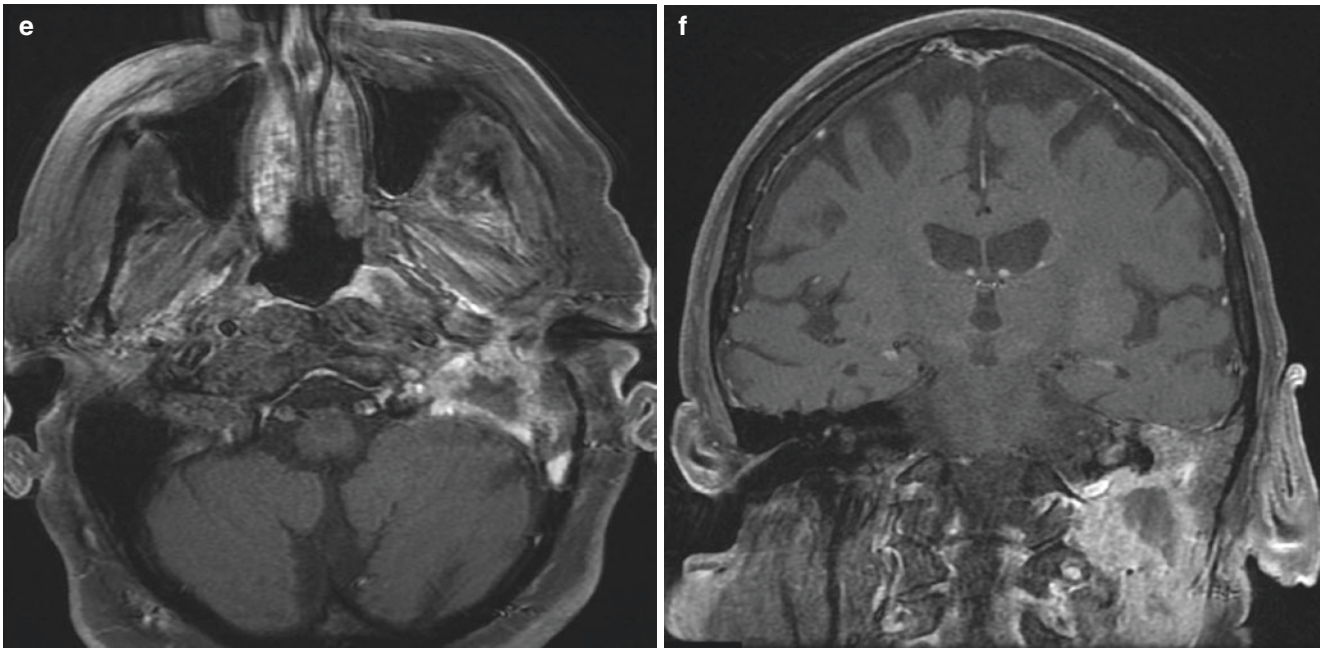

Fig. 7.3 (continued)

metastasis [69]. If distant disease was discovered on PET/CT, then primary surgery would not be offered. Instead, these patients are treated with neoadjuvant chemotherapy followed by concurrent chemoradiation therapy to the primary site.

Natural History and Directions of Spread

The bone of the ear canal and the eardrum is an early defense against the spread of SCC arising in the canal or outer ear. Once the ear canal and eardrum are penetrated, the tumor is able to spread anteriorly into the TMJ; inferiorly along the jugular vein, carotid artery, and lower cranial nerves; medially into the middle ear and mastoid; superiorly into the middle fossa and temporal lobe; and posteriorly into the sigmoid sinus and cerebellum. The numerous nerve and vascular foramina of the temporal bone allow spread of disease [33]. While the bone of the ear canal is relatively thick, the bone of the mastoid and tegmen is relatively thin or occasionally dehiscent, offering little resistance to invasion. The bone plates over many structures like the jugular bulb, carotid, tegmen, fallopian canal, and the labyrinth are thin and are vulnerable to tumor erosion [126].

The ear canal has preformed pathways like the cartilaginous fissures of Santorini, the petrosquamous suture line, and the bony foramen of Huschke that can facilitate spread of tumor anteriorly (Fig. 7.4) [126]. Embryologically, the ossification centers of the ear canal fuse to complete the tympanic ring. By age 5 years, the bone of the ear canal is usually completely fused. This foramen of Huschke is a sign of incomplete fusion of the ossification centers in the developing tympanic bone. The foramen of Huschke is a pathway from the anterior ear canal at the level of the tympanic membrane to the soft tissues anterior to the ear canal [125]. Additionally, Haversian canals and angiolymphatic pathways are additional pathways for the spread of cancer [113].

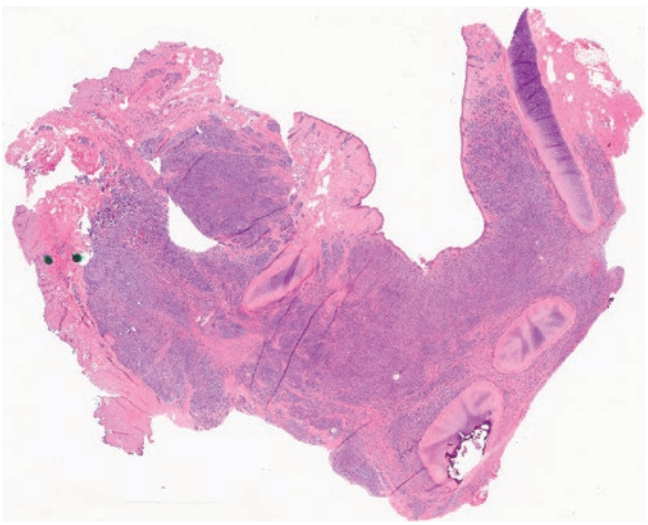

Fig. 7.4. Low-power photomicrograph of cancer spreading through the fissures of Santorini

The creation of canal wall down mastoidectomy cavities for cholesteatoma produces pathways for the spread of cancer. In the latter case, the fenestration procedure opens up the normally resistant inner ear and makes it more vulnerable to tumor invasion [35].

Leonetti et al. described the patterns of temporal bone invasions as (a) superiorly through the tegmen tympani into the middle cranial fossa, (b) anteriorly into the glenoid and infratemporal fossa, (c) inferiorly into the jugular foramen, (d) posteriorly into the mastoid air cells, and (e) medially into the middle ear, inner ear, and carotid canal [128]. In their study, CT and MRI underestimated disease in the mastoid mucosa, tegmen tympani, middle fossa dura, middle ear mucosa, and along the carotid canal. Underestimating disease preoperatively might lead to planning inadequate surgery. As a consequence, patients who later developed recurrence had radiographic underestimations in one or more of these locations.

Gidley et al. reported that extension anterior to the ear canal was present in 63% of patients [5]. They reported that tumor extended to involve the jugular foramen in 23%, the carotid artery in 11%, the infratemporal fossa in 11%, and the temporomandibular joint in 4% [5].

Histopathology

Squamous cell carcinoma can be graded based on level of differentiation. The degree of differentiation varies from study to study, and some authors have correlated degree of differentiation with survival. Well-differentiated tumors are found in 11–53%, moderately differentiated from 36% to 79%, and poorly differentiated from 4% to 36% [5, 39, 46, 48, 64, 129]. Gidley et al. reported perineural invasion in 7%, extracapsular extension in 2%, and vascular invasion in 4% [5].

Staging

The goal of staging is to sort patients with equivalent disease burden into the same group to allow a fair comparison with respect to treatment outcomes. Accurate staging is essential for treatment planning and to prognosticate survival. Staging is also necessary to counsel patients regarding treatment options and expected outcomes.

Accurate staging improves external validity and allows for analysis of the effects of specific disease characteristics, comorbidities, and treatment [67]. Staging must account for local disease extension, since this is the main determinant of survival [119]. Care must be exercised to avoid underestimating disease [70, 122]. Regional and distant disease must also be included. Given the complex anatomy of the temporal bone, clinical features alone are insufficient for accurate staging. Radiographic imaging and final pathologic findings are often necessary to complete staging [68, 130].

Several staging systems have been proposed over the years. Many of these early systems have just three stages, and they typically relied on physical examination findings. Prior to the development and refinement of cross-sectional, computerized imaging, surgeons relied on physical examination, plain film, and polytomography findings to assess extent of disease [131–135]. Goodwin and Jesse [88], Stell and McCormick [136], Pensak et al. [137], and Mandolis et al. [14] have all proposed different staging systems. These staging systems are reviewed in Chap. 2.

In 1990, Arriaga et al. published a landmark study in which CT radiographic findings were correlated with pathologic findings for squamous cell carcinoma of the external auditory meatus [119]. The authors excluded from consideration patients that had the ear canal involved secondarily from the external ear or parotid. They identified 12 anatomic areas to be assessed: external osseous meatus erosion anteriorly, posteriorly, inferiorly, and superiorly; infratemporal extension; middle ear involvement; otic capsule erosion; mastoid involvement; jugular fossa erosion; carotid canal erosion; tegmen erosion with middle fossa involvement; and posterior fossa involvement [119]. They reported a 98% concurrence between CT and pathologic extent of disease. This study highlights the fact that a careful and systematic review of CT scans is required for accurate prediction of disease extent. Their staging system ranks tumors by extent of local destruction (e.g., canal wall or soft tissue extension) and by involvement of medial structures (e.g., ear canal, middle ear/mastoid, inner ear involvement) and uses a TNM format for squamous cell carcinoma of the external auditory meatus. In their schema, any lymph node involvement was automatically considered a sign of advanced-stage disease; thus, Tl-3N+ and any T4 are considered stage IV [119]. This system has become known as the Pittsburgh staging system (PSS) and has undergone several amendments (Table 7.1).

Moody et al. modified the original PSS to move facial paresis or paralysis to T4, since a tumor that is affecting the facial nerve is either in the middle ear and eroding the medial wall (tympanic portion of the nerve), or has invaded the full thickness of the ear canal, or by involving tissue at the stylomastoid foramen (by definition >0.5 cm of soft tissue involvement) [68]. Facial nerve involvement, or its correlate facial paralysis, has been cited as an important prognostic feature in several papers [15, 113]; however, other papers have found that facial nerve involvement was not a significant factor [9, 116].

Higgins and Moody-Antonio performed a systematic review of the literature regarding outcomes for patients with or without facial paralysis in the setting of SCC of the ear canal and temporal bone [67]. Their study encompassed 21 studies covering 348 patients. They demonstrated that patient with facial paralysis had a significantly worse disease-specific survival and overall survival when compared to patients without facial paralysis. Furthermore, they showed that overall survival was worse for patients with facial

Table 7.1 Pittsburgh staging system, originally proposed by Arriaga et al. [119] with modification from Moody et al. [68]

T classification	
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 (<0.5 cm)
T3	Tumor eroding through the osseous EAC (full thickness) with limited soft tissue involvement (<0.5 cm) or tumor involvement in 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 with extensive soft tissue involvement (>0.5 cm), such as involvement of the TMJ or styloid process, or evidence of facial paresis
N classification	
N0	No regional nodes involved
N1	Single metastatic regional node <3 cm in size
N2a	Single ipsilateral metastatic node 3–6 cm in size
N2b	Multiple ipsilateral metastatic lymph nodes
N2c	Contralateral metastatic lymph node
N3	Metastatic lymph node >6 cm in size
Overall stage	
I	T1N0
II	T2N0
III	T3N0
IV	T4N0 and any T N+

Table 7.2 Proposed modification to the Pittsburgh staging system [33]

T classification	
T1	Tumor in the skin without bone involvement
T2	Tumor in the skin with bone/cartilage involvement, but not full thickness
T3a	Tumor extending <5 mm from cartilage to periauricular soft tissues or tumor strictly limited to the anterior bone wall and growing <5 mm into the parotid space
T3b	Same as for T3a, but extending >5 mm
T4a	Tumor growing into mastoid, without seventh paresis
T4b	Tumor growing into mastoid with facial paresis, or infratemporal space, or medial wall of tympanum, labyrinth, petrous bone (jugular foramen, internal carotid canal, petrous apex)

paralysis than those staged as Pittsburgh 2000 T3. There was no significant difference in overall survival between patients staged as Pittsburgh 2000 T4 and those with facial paralysis. From this analysis, they concluded that facial paralysis rightfully should be staged as T4 [67]. They also showed that the Pittsburgh 2000 staging was superior to the 1990 version and the Stell staging system by being able to discriminate overall survival in Stell T2 versus T3 disease and Pittsburgh T3 versus T4 disease.

Several studies have confirmed the correlation between Pittsburgh T stage and prognosis for squamous cell carcinoma [5, 8, 15, 39, 46, 113, 126, 130, 138], and this system has become the most frequently used for reporting results in contemporary literature [15]. A meta-analysis of the literature on squamous cell carcinoma of the temporal bone has shown that T classification is an independent factor for poor survival by Cox proportional hazard model (hazard ratio (HR) = 2.53, p = 0.002) [139]. Their multivariate analysis did not find that lymph node metastasis was a risk factor for survival (HR = 0.93, p = 0.939).

The Pittsburgh staging system has been applied to tumor histologies other than squamous cell carcinoma. Testa et al. applied the Pittsburgh staging system to describe 79 cancers of the external auditory canal, including SCC, basal cell carcinoma (BCC), and adenocarcinoma [118].

Gaudet et al. [83] used the Pittsburgh staging system for ten periauricular skin cancers. Their paper highlights the fact that large periauricular skin cancers might be staged as T4 by the prevailing AJCC system, but simultaneously, these tumors would be judged as T1 by the Pittsburgh staging system. Similarly, Essig and colleagues used the Pittsburgh staging system for large periauricular cancers that required lateral temporal bone resection [130].

Despite its popularity, problems with the Pittsburgh staging system have been identified, and changes have been suggested. Ito et al. state that extensive bone involvement correlates with prognosis, yet they do not define "extensive soft tissue involvement" [15]. Mazzoni et al. list four problems: "(1) The skin of the auditory canal. (2) Skin and bone and/or cartilage involvement, but not full thickness (the term 'full thickness' is appropriate for the anterior bone wall, while it needs to be defined for the other bone walls). (3) Anterior extension from anterior wall to parotid space, or from cartilage canal to periauricular soft tissues. (4) Extension from canal to mastoid and other sites of the temporal bone" [33]. They propose modifications to the PSS (Table 7.2).

Breau et al. proposed a modification to the Pittsburgh system for early-stage lesions based on the site of disease in the canal and less emphasis on the size of the primary tumor or degree of bony invasion [13].

Treatment Planning

Squamous cell carcinoma of the ear canal can require surgery, radiotherapy, and possibly chemotherapy. The complexity of treatment options emphasizes the role of the multidisciplinary team. Unfortunately, randomized trials do not exist to help guide treatment decisions. These patients should be seen by a number of consultants to arrive at the proper treatment. Consultations are typically sought from neurotology, head and neck surgery, radiation oncology, and medical oncology. Based on surgical requirements, consultations might be required from neurosurgery, oculoplastics, and plastic reconstructive surgery.

The Role of Surgery

Surgery has become a mainstay of treatment for cancers of the ear canal and temporal bone [46]. The extent of surgery has been a subject of debate and controversy. In a landmark study, Prasad and Janecka performed an early meta-analysis of 26 publications on temporal bone cancer covering 144 patients [140]. Their conclusion about surgical approach for temporal bone cancer was that patients who had tumors confined to the ear canal had similar survival rates whether they had mastoidectomy, lateral temporal bone resection, or subtotal temporal bone resection. This paper highlighted many of the deficiencies in the literature regarding temporal bone cancer and the surgical approaches to it. While some of its conclusions are no longer true, this paper launched better research and more rigorous outcome measures for this dreaded illness.

Up until the 1950s, radical mastoidectomy followed by radiotherapy was considered the treatment of choice for ear canal cancers. This approach is associated with high degree of recurrence and the potential for chronic drainage and osteoradionecrosis [141, 142]; however, Zhang et al. still promote this approach for SCC of the temporal bone [45]. Their philosophy is predicated on the belief that mastoidectomy causes less morbidity and mortality than temporal bone resection.

The goal of surgery is complete resection of the tumor with negative margins. Margin status is a clear prognostic indicator, and surgery must be conceived and designed in order to achieve a negative margin. Tumors that cannot be resected with negative margins are considered unresectable. Morris et al. reported close or positive margins in 38% of SCC ear canal. This number rose to 47% with T4 tumors [4]. Zhang et al. identified positive margins in 54% of early-stage (T1 and T2) tumors, and this finding prompted them to change from sleeve resection to lateral temporal bone resection for these early-stage tumors [127]. Rates of positive margins are quoted from 18% to 64% [5, 27, 30, 47, 68, 113, 130]. Clearly, achieving a negative margin is difficult in temporal bone cancer surgery.

Nomenclature of Surgical Procedures

The nomenclature surrounding surgical procedures has been confusing. Previous authors have named procedures such as wide local excision, sleeve resection, en bloc EAC resection, local canal resection, partial temporal bone resection, radical resection of the EAC, modified lateral temporal bone resection, and lateral temporal bone resection for procedures that excise lesions of the external auditory canal [6, 12, 70, 122, 138]. Medina et al. describe four types of lateral temporal bone resection [143].

Some authors have described "sleeve resection" where the skin of the ear canal and TM are removed and replaced with skin grafts [6]. The bone of the ear canal is left intact. The attractiveness of this approach is preservation of hearing and a lesser risk of neural or vascular injury.

The problems with this approach are a high rate of positive margins and a high recurrence rate. Goodwin and Jesse reported that sleeve resection for cancer of the external auditory meatus had a 41% recurrence rate [88]. Austin et al. reported that nine patients with T1 disease treated with local canal resection had a 5-year survival of 66%, and all failures were due to local recurrences [138]. Kunst et al. used local canal resection for T1 tumors, with a 25% recurrence rate [122]. Zhang et al. reported a recurrence rate of 46% and a positive margin rate of 54% with local canal resection [127].

A more recent study compared the results of local canal resection to lateral temporal bone resection for T1 and T2 temporal bone malignancies. This study showed a clear survival advantage and higher cure rate for patients who underwent LTBR with superficial parotidectomy over local canal resection [127].

Wide Local Excision

A clear distinction needs to be made about tumors located at the external auditory meatus versus tumors in the bony canal. Tumors that occupy the cartilaginous portions of the ear canal can be safely excised with surrounding soft tissues if the tumor does not extend medial to the bone-cartilage junction (Fig. 7.5). This procedure is more adequately termed "wide local excision" and is similar to other skin cancer excisions on the face and neck. Removing the underlying cartilage helps to ensure complete resection and to lower recurrence rate.

Tumors that extend medially into the canal past the bone-cartilage junction require lateral temporal bone resection for excision. The skin of the bony canal is quite thin, and an adequate margin of healthy skin is not present. Examining the ear with oto-endoscopes, in addition to the otomicroscope, is very helpful in making this determination.

Temporal Bone Resection

Lateral temporal bone resection is the workhorse of oncologic surgery for the temporal bone [4]. Lateral temporal bone resection (LTBR) is the en bloc removal of the ear canal, lateral to the facial nerve and stapes (Chap. 17). The procedure removes the bony ear canal, tympanic membrane, malleus, and incus. The stapes and inner ear are preserved. The facial nerve is generally preserved. LTBR can be combined with auriculectomy, parotidectomy, mandibulectomy, and neck dissection depending on the extent of disease.

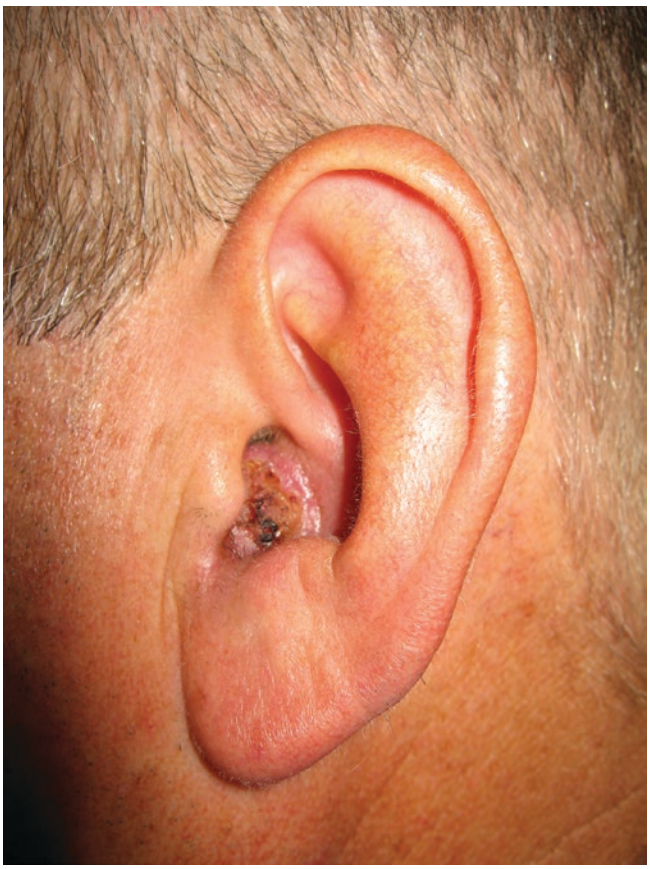

Fig. 7.5 Squamous cell carcinoma at the external auditory meatus, which was widely excised and reconstructed with a split thickness skin graft. The tumor did not extend into the bony ear canal and LTBR was not required

Limited dural resections can also be performed. Lateral temporal bone resection for Pittsburgh T1 is curative in 100% of patients [68].

Surgeons have devised procedures to remove the ear canal lateral to the tympanic membrane in an attempt to preserve hearing. Medina et al. first described this as "type I lateral temporal bone resection" [143]. Moody et al. describe this same procedure as "modified LTBR" [68].

On the other end of the spectrum, Moffat et al. recommend LTBR, total auriculectomy, mandibular condylectomy with superficial parotidectomy as a necessity for stage T1 and T2 tumors [39]. They justify this approach by citing the inability of radiographic studies to diagnose extent of disease into the TMJ and infratemporal fossa [39].

A subtotal temporal bone resection (STBR) is performed when disease extends past the tympanic membrane into the middle ear (Chap. 18). In this case, the labyrinth and cochlea are removed with a drill, and the disease is removed piecemeal. LTBR is usually a necessary first step to remove disease in the ear canal and to gain enough exposure for the middle ear, Eustachian tube, cochlea, and carotid artery. The margins of resection are the middle fossa dura superiorly, the carotid artery anteriorly, the jugular bulb inferiorly, and the sigmoid sinus and posterior fossa dura posteriorly. The internal auditory canal and anterior petrous apex are not removed. Dura is resected as dictated by frozen section and repaired with a dural graft. Microvascular free flap reconstruction is used for the majority of cases, since the surgical defect is too large for a temporalis flap. For tumors of the middle ear, Jia et al. showed that subtotal temporal bone resection had a higher survival rate than lateral temporal bone resection [2].

Total temporal bone resection (TTBR) expands the limits of the subtotal temporal bone resection to include the petrous apex and internal auditory canal. Again, dura resection is dictated by extent of disease as seen on frozen section pathology, and the dura is repaired with a dural graft. Microvascular free flap reconstruction is performed.

Both subtotal and total en bloc temporal bone resections have been described. Subtotal resection with piecemeal removal of disease beyond the limits of the subtotal temporal bone resection and postoperative radiotherapy is a reasonable option to total en bloc temporal bone resection [70]. Total en bloc temporal bone resection is fraught with increased morbidity and has not been shown to improve overall survival. In this setting of advanced disease, usually with carotid artery involvement, preoperative chemotherapy is emerging as likely the most efficacious approach. The interested reader is referred to Okada et al. for a description of subtotal and total en bloc temporal bone resection [144]. However, the introduction of chemotherapy for these large (T3 and T4) tumors has offered patients new hope without having to undergo such surgery.

Parotidectomy, Neck Dissection, Condylectomy, and Carotid Resection

The role of elective parotidectomy and neck dissection continues to be a source of controversy in the literature. Conley and Schuller in 1976 stated that the proclivity of ear canal cancers to metastasize to the parotid and cervical lymph nodes necessitates their inclusion in the treatment plan [43]. In 1977, Gacek and Goodman recommended that superficial parotidectomy be performed for all cases of en bloc temporal bone resection because of the proximity of the lymph nodes and to provide a more adequate margin of tissue anterior to the ear canal [145]. Parotidectomy is required where there is evidence of direct extension. Elective parotidectomy is recommended for accurate staging since many patients will have positive nodal metastases within the parotid. Chee et al. found direct extension to the parotid gland in only 1 of 14 cases of squamous cell carcinoma of the ear canal [24]. Morris et al. identified direct parotid invasion in 25% and parotid nodal metastasis in 43% of patients with SCC of the ear canal [4]. Parotid gland involvement has been noted as

high as 62% of patients with stage IV disease [107]. Moffat et al. considered parotidectomy as a routine part of treatment for squamous cell carcinoma of the temporal bone [39].

Performing parotidectomy for ear canal cancer is not universal. Madsen et al. reported on a series of 68 ear canal cancers, and parotidectomy and mandibular condylectomy were not performed in any of their patients [8]. In their series, primary neck dissections were performed in only two patients.

Neck dissection for the N+ neck is essential when surgical treatment is being planned. Elective neck dissection continues to be a controversial subject. The incidence of positive lymph nodes varies between 10% and 20%. Selective neck dissection to include levels II and III is performed for adequate staging and to select appropriate patients for adjuvant therapy [5]. The morbidity of adding selective neck dissection is minimal in this setting. Regional lymph node recurrences have been reported in SCC of the temporal bone [5].

Resection of the mandibular condyle is also controversial. Moffat et al. consider mandibular condylectomy with LTBR, auriculectomy, and superficial parotidectomy as a necessity for stage T1 and T2 tumors [39]. Hosokawa et al. recommend condylectomy and removal of the soft tissues anterior to the ear canal in cases where preoperative CT scan shows an anterior bony canal wall erosion of 2 mm or greater [125]. Their study of 15 patients with ear canal carcinoma of differing histologies found tumor in the anterior soft tissues in every case where the anterior canal wall had more than 2 mm of erosion. Clearly, extent of resection for the mandible is guided by preoperative imaging and intraoperative findings, as determined by frozen section pathology.

Carotid involvement is a particularly poor survival indicator. Masterson et al. showed that all patients who underwent surgical resection and who had carotid involvement died before 2 years [47]. Moffat et al. reported that 8 out of 39 patients had carotid involvement from SCCa; only one was alive at the time of the manuscript, and that patient was only 9 months from surgery [39]. Since carotid resection carries with it such a poor outcome and the potential for significant morbidity (i.e., stroke), carotid sacrifice is not performed [116]. Therefore, carotid invasion is considered a sign of unresectability [64].

Absolute contraindications to surgery include cavernous sinus involvement, massive intracranial involvement, unresectable neck disease, distant metastasis, and poor general health [115]. Relative contraindications to surgery include carotid artery involvement and lower cranial nerve involvement [146].

Reconstruction

In the days where radical mastoidectomy was the norm for resection of squamous cell carcinoma, split thickness skin grafts were used. Split thickness skin grafts have been used to re-epithelialize the cavity resulting from LTBR with successful grafting in 83% of non-irradiated patients [147].

In the setting of postoperative radiotherapy, patients with such open cavities have a high risk of developing chronic otorrhea and osteoradionecrosis [142]. Reconstruction and obliteration are used to avoid an open cavity, as in a canal wall down mastoidectomy. Obliteration of the cavity using either a temporalis muscle flap or microvascular free flap is advisable.

The temporalis muscle flap provides an excellent reconstructive option when the surgical defect results from removal of the ear canal only. Larger defects, especially those that involve auriculectomy, mandibulectomy, dural resection, or exposure of the great vessels of the neck require microvascular free flap reconstruction [148, 149]. Some authors have used pedicled pectoralis flaps and scalp rotational flaps in addition to temporalis flaps [66], but the disadvantage of these flaps is the length of the pedicle to reach the temporal bone. Pedicled regional flaps, such as trapezius or pectoralis major, are avoided since the temporal bone is at the very limit of the arc of rotation [149].

Microvascular free flaps are also favored in patients that have previously been treated with radiotherapy. The anterolateral thigh (ALT) flap is the most frequent donor site, although many different donor sites are available depending on the size of the defect and donor site anatomy. The ALT flap has several advantages including low donor site morbidity, permitting a two-team operation, and the ability to harvest fascia lata grafts, nerve grafts, and even vein grafts from the same donor site incision and earlier mobilization [149].

In patients with normal preoperative facial function, immediate reconstruction with nerve grafting is recommended. Facial nerve recovery is not affected by age of patient or preoperative facial function or the use of postoperative radiotherapy [149, 150]. Facial nerve grafts can also be placed and result in return of function even if the proximal nerve margin is positive [151]. Additionally, placement of gold weight and fascial slings are helpful at protecting the eye and reestablishing some facial symmetry.

Preoperative consultation with an anaplastologist is important for patients who will require auriculectomy. Auricular prosthesis can be attached with either adhesives or osseointegrated implants.

A maximum conductive hearing loss is an expected outcome of LTBR. Only a few papers have reported hearing results following LTBR. Cristalli et al. included hearing results on their report of 17 patients with SCC of the ear canal. Their graph indicates on average a 20 dB change at 3 kHz and 30 dB change at 4 kHz in the five patients that did not receive radiotherapy. Hearing at 6 kHz was significantly worse for the patients that received postoperative radiotherapy [66].

Hearing rehabilitation is an integral part of the surgical planning for patients who have temporal bone surgery. Placement of an osseointegrated implant for cochlear stimulation is a reasonable option. A transcutaneous abutment is not placed with the first surgery to avoid complications with anticipated postoperative radiotherapy. Since radiotherapy has an effect on osseointegration, placement of the transcutaneous abutment is delayed. Planned second stage for placement of the abutment takes place 3 months later if no radiotherapy is given and 6 months later if radiotherapy is required [152]. This secondary procedure is usually scheduled along with free flap revision, if needed.

Surgical Complications

Surgical complications include facial paralysis, wound complications, cerebrospinal fluid leak, meningitis, lower cranial nerve deficits, stroke from carotid sacrifice, postoperative hemorrhage, pulmonary embolus, or microvascular free flap failure [47, 149]. Perioperative mortality rate in surgical series has been quoted from 0% to 6% [14, 68, 107, 109, 119, 130, 149].

The Role of Radiotherapy

Radiotherapy has been used as definitive or adjunctive therapy for SCC of the temporal bone. The outcomes for primary radiotherapy have been reported in only a few papers.

Birzgalis et al. (1992) reported the results of primary radiotherapy on 60 patients with tumors involving the middle ear and temporal bone [153]. Treatment dosages ranged from 45 to 55 Gy. None of the patients in their series had radical resection of the primary tumor. They staged patients into two broad categories: early and late, where late indicated bony erosion, facial paralysis, multiple cranial nerve deficits, or nodal metastasis (Stell T2 and T3). They reported an overall 5-year survival of 32%.

Hashi et al. (2000) suggested that radiotherapy alone is sufficient for T1 tumors [154]. They treated five patients with Stell T1 tumors, and none of these patients experienced disease recurrence. Treatment doses were 65 Gy, and there were no long-term complications of radiotherapy. They also treated two Stell T2 tumors with radiotherapy alone (65 Gy and 75 Gy), and both patients developed disease recurrence and survived only 6 months [154].

Pemberton et al. (2006) reviewed the historic record of definitive radiotherapy for primary ear canal and middle ear squamous cell carcinoma in 123 patients performed at Christie Hospital, Manchester, UK. Twenty patients had radical mastoidectomy, but none had a radical resection of disease. Their paper used the Stell staging system. The clinical response to radiotherapy was complete resolution in 50%, partial resolution in 27%, and no response in 20% [117]. Recurrence was local in 18%, nodal in 15%, and distant in 5% [117]. DFS was 45% at 5 years; the mean time to recurrence was 1.6 years [117].

Generally, while a few papers have shown that definitive radiotherapy is adequate treatment for T1 tumors, most reports have indicated that radiotherapy is inadequate as primary therapy for SCC of the temporal bone [23, 138, 155]. The use of external beam radiotherapy as a postoperative adjunct to surgery was first put forth by Lederman and reinforced by Conley and Schuller [43, 86]. Most papers report using postoperative radiotherapy as part of multidisciplinary care. Surgery plus postoperative radiotherapy offers better survival than either one alone [30, 48, 138].

Indications for postoperative radiotherapy include positive margins, recurrent cancer, perineural invasion, bone invasion, lymph node metastasis, and advanced-stage disease [27, 68, 70, 116]. Doses between 60 and 66 Gy are typical for patients with negative margins. Dose is increased to between 68 and 72 Gy for patients with close or positive margins [156]. Recurrence in cervical lymph nodes has been reported in patients who did not receive postoperative radiotherapy [5].

Radiotherapy with doses around 60 Gy does show a survival benefit for tumors T2 tumors and higher [5, 16, 30, 68, 122, 157]. T1 tumors did not benefit in a statistically significant way with combined therapy [5]. In a meta-analysis, Moody et al. found that radiotherapy helped improve survival in patients with T3 and T4 tumors [68].

Postoperative radiotherapy can improve local control in patients with negative margins; however, Goodwin and Jesse found that radiotherapy was of no benefit when the tumor could not be completely excised [88]. Other authors have also commented that radiotherapy is not a substitute for achieving negative margins [119, 158].

Zhang et al. reported in 1999 their results for 33 patients with SCC of the temporal bone [45]. Eleven (11) patients with stage III and IV disease were treated with radiotherapy alone with a 5-year survival of 28.7%, but 20 patients with stage III and IV disease treated with combined surgery and radiotherapy at a 5-year survival of 59.6% [45].

Kitani et al. reported a 5-year survival rate of 51% using definitive radiotherapy with or without chemotherapy [159].

Ogawa et al. reviewed their experience treating 87 patients with radiotherapy and surgery and concluded that radiotherapy alone is reasonable for Stell stage T1 tumors, whereas combined surgery and radiotherapy was indicated for T2 and T3 tumors [155]. They found that patients with Stell T3 disease did significantly better with surgery and radiotherapy rather than radiotherapy alone [155]. Furthermore, they found that the timing of radiotherapy (preoperative, postoperative, or combined pre- and postoperative) when combined with surgery did not influence disease-free survival [155].

There is very little information on the results of brachytherapy for temporal bone cancer. In the 1930s and 1940s, Figi and Hempstead treated patients with radical mastoidectomy, electrodessication, and implantation of radium; their survival rate was around 32% [160]. Using a more modern approach, Mayer et al. used either low-dose-rate (LDR) or high-dose-rate (HDR) brachytherapy to treat seven patients postoperatively [161]. Four of these seven (57%) patients remained disease free 10 years after treatment [161].

Cristalli et al. employed intraoperative radiotherapy (IORT) in eight patients with SCC of the ear canal [66]. They delivered 12 Gy as the first dose of planned postoperative radiotherapy once frozen section margins were determined to be tumor free. They advocated its use on the basis of direct visualization of the tumor bed to minimize the risk of geographic miss.

Complications of radiotherapy include dermatitis, xerostomia, hearing loss, vestibular weakness, brain necrosis, and osteoradionecrosis [117, 156]. The mean time to bone or soft tissue necrosis is 4.6 years (range 0.8–13.7 s); the average dose was either 52.2 or 55 Gy [117].

Postoperative radiotherapy offers survival benefits to patients with T2 and higher-staged disease. Postoperative radiotherapy is indicated for patients with bone invasion, perineural spread, cervical lymphadenopathy, or recurrent disease. Definitive radiotherapy is an option for patients with limited, early-stage disease who either refuse surgery or are otherwise not candidates for surgery.

The Role of Chemotherapy

Chemotherapy has an increasingly important role in the management of ear canal and temporal bone cancer. While it is often reserved for large tumors (T3 and T4), the results of chemotherapy, with or without surgery and radiotherapy, have shown promise. Chemotherapy, as a systemic treatment, might also have implications for the rare patient with bilateral SCCa of the ear canal and temporal bone so that bilateral conductive hearing loss might be avoided.

In 2006, Nakagawa et al. published a retrospective review of 25 patients with primary SCCa of the external ear canal and middle ear. This early study of chemotherapy, combined with radiotherapy and surgery, used TS-1 (tegafur, gimeracil, and oteracil) and 5-FU or cisplatin (70 mg/m^2) and 5-FU (700 mg/m^2). Surgery (either LTBR or STBR) was performed on selected patients. T1 and T2 tumors were treated primarily with surgery, and these patients had 100% estimated 3-year survival. T3 and T4 patients were treated with surgery, radiotherapy, and chemotherapy. Several T4 patients were treated with chemoradiotherapy (CRT) alone. The 5-year estimated survival for T3 and T4 patients was 80% and 35%, respectively. They showed some effectiveness of concurrent CRT as stand-alone treatment for temporal bone SCCa; however, survival for T4 tumors was better with preoperative CRT and surgery than with CRT alone. They conjectured that preoperative CRT contributed to a higher rate of tumor-free margins in patients undergoing surgery for T4 tumors [129].

Shiga et al. reported the results of concurrent chemotherapy and radiotherapy in a group of 15 patients with SCC of the ear canal and temporal bone; 9 patients had stage IV disease [162]. Their protocol consisted of 5-FU continuous infusion 500–600 mg/M^2 on days 1–5, with docetaxel 50 mg/M^2 and cisplatin 60 mg/M^2 on day 2, and radiotherapy to total dose of 70 Gy. There were significant toxicities related to this therapy including grade 3 and 4 hematopoietic adverse events. Complete remission was achieved in eight of nine stage IV patients. They calculated a 67% 5-year disease-specific survival for patients with T4 disease.

In 2015, Shinomiya reported the results of CCRT in a group of ten patients treated with TPF protocol. Their 5-year overall survival rate was 60%. Even for T4 or unresectable tumors, the overall 5-year survival rate was 56% [163].

In 2015, Kitani et al. reported the results of combined chemotherapy and radiotherapy in a group of 13 patients with squamous cell carcinoma of the temporal bone [159]. Patients were staged according to the Pittsburgh system; tumors were stages II–IV. All patients were treated with definitive radiotherapy (2 Gy fractions to total dose of 70 Gy). Concurrent chemoradiotherapy (CRT) was given to patients under 75 years old and consisted of two cycles of 5-FU (1000 mg/M^2) and cisplatin (60 mg/M^2). Patients with renal disease received docetaxel (15 mg/M^2/week). Patients with unresectable disease received induction chemotherapy consisting of docetaxel 60 mg/m^2 on day 1, cisplatin 70 mg/m^2 on day 4, and 5-FU 1000 mg/m^2 on days 1–5 followed by concurrent chemoradiotherapy. Of note, three of five patients with stage II disease died of disease due to locoregional failure; two of these three patients received radiotherapy only. Using this protocol, patients with stage III disease fared much better with four out of five patients demonstrating long-term survival. None of the stage IV patients survived past 14 months, and all developed local recurrences. While the authors cite surgery as producing unfavorable quality of life due to facial paralysis and hearing loss, they do not report their patients' quality of life, hearing status, or facial nerve function following this combined chemoradiotherapy.

Takenaka et al. performed a meta-analysis of stage III and IV squamous cell carcinoma of the temporal bone and found that the 5-year overall survival rate between CRT and standard treatment (surgery with or without radiotherapy) equivalent (43.6% vs 53.3%, respectively, $p = 0.210$) [139]. Caution is required in interpreting these data since the CRT group contained patients that were surgically unresectable, and thus, these patients naturally had worse disease and prognosis.

When CRT and surgery were combined, the outcomes depended on the setting. Preoperative CRT patients (87.5%

5-year survival) had a slightly better outcome than patients who received CRT postoperatively (0% 5-year survival), but this difference was not statistically significant [139]. In a multivariate analysis using the Cox proportional hazard model, preoperative CRT was significantly associated with an improved overall survival (Hazard Radio (HR) 0.18, $p = 0.030$), while postoperative CRT did not affect survival (HR = 1.30, $p = 0.617$) [139]. They conclude that (1) preoperative CRT may improve survival in advanced external auditory canal SCC; (2) definitive CRT may be an effective alternative for surgical treatment; and (3) postoperative CRT may not be effective enough to salvage patients with worse prognoses [139].

In summary, chemotherapy has a role for advanced-stage disease (Pittsburgh III and IV) and surgically unresectable disease. Preoperative, multidrug protocols and radiotherapy seem to offer advanced-staged patients a better overall survival than conventional surgery and radiotherapy.

Intra-Arterial Chemotherapy

Theoretically, intra-arterial therapy has the advantage of delivering high-dose chemotherapy to the target tumor while avoiding systemic toxicity. In 1965, Tucker reported the original use of intra-arterial chemotherapy for six cases of ear canal and temporal bone cancer [21]. The role of superselective intra-arterial chemotherapy was first introduced for head and neck cancer by Robbins et al. [164]. A protocol for temporal bone SCC was piloted by Ueda et al. [165]. In their protocol, patients were given one dose of IV cisplatin (100 mg/patient) with sodium thiosulfate 1 week prior to radiotherapy. Two or three intra-arterial doses of cisplatin were given during radiotherapy (60 Gy at 2 Gy/day fractions). In this preliminary report of four patients, all patients developed complete remission of disease with mean follow-up of 30 months [165]. They reported only one patient with significant sensorineural hearing loss during the study period, and no patients had any thromboembolic complication from angiography [165].

More recently, Sugimoto et al. [166, 167] reported results in 12 patients. In their protocol, patients received conventional once daily external beam radiation (60–70 Gy) in 2 Gy doses, followed by intra-arterial cisplatin (200–600 mg in 2–6 portions). Surgical salvage was performed in patients with persistent disease. For the 12 patients (6 patients T3 and 6 patients T4) treated by this protocol, mean survival was 41.4 months in 9 patients. Two died of local recurrence and one died of distant disease. The application of this treatment requires skilled neurovascular interventionalist who can administer superselective chemotherapy, and this requirement limits the overall usage of this protocol.

Clinical Follow-Up

There are no current clinical guidelines dictating the frequency or extent of follow-up evaluations. Clinical evaluation of patients following temporal bone resection is limited given the fact that most patients with ear canal or temporal bone cancer will have a flap covering the operative site. The physical examination of these patients is limited to general factors such as weight, overall health, cranial nerve, and neck examination. For these reasons, cross-sectional imaging is indispensable and medically necessary for managing these patients.

Masterson et al. clinically evaluated patients every 2 months for the first 2 years and then every 6 months until 5 years following surgery [47]. Imaging is reserved for patients that have clinical signs of recurrence.

Moffat et al. describe seeing patients every 2 months for the first year, every 3 months for the second year, and then to every 4 months until 5 years after treatment [39]. Routine imaging was not performed unless clinically indicated. They reasoned that microscopic disease recurrence is difficult to discern from posttreatment changes on diagnostic imaging.

Mazzoni et al. describe seeing patients every 3 months for the year, every 6 months for 4 years, and then annually. Their imaging schedule includes CT and MRI every 6 months for the first year and then annually until 10 years [33].

Zanoletti et al. use clinical examination and contrast-enhanced head and neck MRI every 2 months in the first year, every 4 months in the second year, and every 6 months in the third to fifth years [1].

In our center, patients are seen every 3 months for the first 2 years following completion of treatment. Clinical follow-up is spaced to every 6 months for years 2–5. Contrast-enhanced cross-sectional imaging (either CT or MRI) of the head and neck is performed at each visit. An annual chest X-ray is performed for patients with early-stage disease. Patients with higher stage disease have an annual PET/CT performed. This schedule is similar to that described by Leong et al. [107].

Following the fifth year of follow-up, patients are transitioned into our Survivorship Clinic, where they are seen annually; and the focus of the visit moves from the primary tumor management to management of treatment complications and surveillance of second primary tumors (e.g., prostate, colon, or breast).

Recurrence and Survival

Recurrence is an ominous sign in this patient population, and recurrence is the major cause of death for temporal bone cancer. The pattern for recurrence is often the return of local disease [8, 33, 140, 165]. Local recurrence is especially common when surgical excision is incomplete, and a positive tissue margin is present [12, 122, 168].

Recurrence rates vary from 11% to 53% overall, and the recurrence rate can be as high as 72% in T3 and T4 tumors [4, 27, 45, 46, 69, 113]. For this reason, achieving local control is of paramount importance. Obtaining a negative margin is so important that even if surgery is extensive (i.e., total temporal bone resection) and a positive margin remains, the 2-year survival can be 0% [119].

Recurrence typically occurs within the first year of completing therapy [1, 5, 27, 46, 68, 113, 169], although there are instances of recurrences up to 3 years following treatment [5, 33, 68, 109]. Bacciu et al. described a series of 45 patients with SCC of the temporal bone; 13 patients (29%) developed recurrence within 1–30 months (mean 8.3 months) [46]. Once disease recurs, the average interval from local recurrence to death can be as short as 3.6 months [119]. Bacciu et al. reported a mean survival time after recurrence was 4.2 months (range 1–21 months) [46]. The median survival for patients with recurrence is approximately 16 months [1].

In a large, retrospective review of definitive radiotherapy for SCCa of the ear canal and temporal bone, Pemberton et al. reported local recurrence in 18%, nodal recurrence in 15%, and distant metastasis in 5% [117]. DFS was 45% at 5 years; the mean time to recurrence was 1.6 years [117]. Gidley et al. reported that 52.2% recurrences were local, 13.5% were to regional lymph nodes, 8.7% were both local recurrences and lymph node involvement, and only 4.3% patients had a solitary distant metastasis [5]. Lobo et al. reported local recurrences in 37% and distant metastasis in 16% [64]. Ogawa et al. reported recurrence in 38 out of 87 patients, 34 patients with local recurrence, 3 patients with regional lymph node recurrence, and 1 patient with distant metastasis (lung) [155].

Zanoletti et al. describe a series of 41 patients with SCCa of the ear canal or temporal bone, and they excluded any periauricular cancers [113]. All patients had negative surgical margins. Radiotherapy was administered to 23 patients. Eighteen patients (41%) developed recurrence within the study follow-up period. Their findings highlight several points: (1) the difficulty in assessing margins in temporal bone cancer, (2) the indications for adjuvant radiotherapy for SCCa in the temporal bone, and (3) the limitations of surgery in controlling disease in the temporal bone.

Factors associated with recurrence are positive margins [30, 68, 69], anterior canal involvement [13], dural involvement [33, 68, 113], facial nerve sacrifice [113], direction of spread in T4 tumors (anterior vs all other directions) [113], lymph node involvement [33], and pathologic tumor grade [113]. Factors not associated with recurrence include parotidectomy, neck dissection, and postoperative radiotherapy [113]. Zanoletti et al. in a review of 47 patients, including patients who presented with positive neck node, did not recur within the neck but instead recurred locally [170].

Surgical salvage for patients with local recurrence has a poor outcome, and these patients might be better served with palliative chemotherapy [1].

Survival

Overall survival for temporal bone squamous cell carcinoma has improved tremendously since Conley and Novack published their large series in 1960 [171]. They calculated an 18% 5-year survival rate. Fifteen years later, Lewis published his personal series of 100 cases of cancers of the ear and reported a 25% 5-year survival rate [172].

Overall Survival

Overall survival (OS) is defined as the time from a patient's first appointment until the last date of contact or death. Authors report various ranges for overall survival such as 2-year [6, 173], 3-year [129, 138], and 5-year [64]. Survival statistics are hard to compare since many studies have small numbers of patients or contain multiple histologic types or use various time intervals (2-year, 3-year, or 5-year) of survival.

Moody et al. report survival on a series of 32 patients with SCCa of the external auditory canal. They quote 2-year survival for stages T1 to T4 as 100%, 80%, 50%, and 7%, respectively [68].

More recent papers have seen increased survival in temporal bone cancer and used the standard 5-year survival statistic. Yin et al. reported an overall 5-year survival of 66.8% for their cohort of 95 patients. Stage I and II tumors had 100% 5-year survival, stage III tumors had 67.2%, and stage IV tumors had 29.5% 5-year survival [30].

Survival heavily depends on extent of disease. Gidley et al. found that overall survival was primarily affected by staging: Pittsburgh T stage (early T1 and T2 versus T3 and T4) [5]. Early-stage tumors (Pittsburgh stages 1 and 2) tend to have excellent 5-year survival. Late-stage tumors have much worse prognosis, but this has improved over time. The reported 5-year overall survival rate has risen from 57.5% to 72.5% for T3 cases and from 22.9% to 35.8% for T4 patients [174].

Prasad et al., in a literature review, found overall 5-year survival ranges from 80% to 100% for T1 tumors, 46–100% for T2 tumors, 17–100% for T3 tumors, and 14–54% for T4 tumors [126].

Takenaka et al. performed a meta-analysis of temporal bone squamous cell carcinoma [139]. Their study included 28 papers covering 752 cases. Their review showed an overall 5-year survival rate of 20–90% [139]. The weighted estimate of 5-year survival using a random effect model was 57% (95% confidence interval, 50–63%).

Their analysis showed that Pittsburgh T classification is an independent factor for poor survival by Cox proportional hazard model (hazard ratio (HR) = 2.53, $p = 0.002$) [139]. Their multivariate analysis did not find that lymph node metastasis was a risk factor for survival (HR = 0.93, $p = 0.939$) [139]. They report that patients with lymph node metastasis had worse survival compared to N0 patients (37.1 vs 47.8), but this was not statistically significant ($p = 0.372$) [139].

For patients with T3 or T4 disease, the overall 5-year survival was 72.5% and 35.8%, respectively, based on a subset analysis of 174 cases [139]. Their meta-analysis of these patients with T3 and T4 SCC of the EAC found that preoperative chemoradiation therapy followed by surgery improved overall 5-year survival rate to 85.7%, compared with surgery with or without radiation (53.5%), definitive CRT (43.6%), and postoperative CRT (0%) [139].

The survival rate for patients with positive margins is poor despite postoperative radiotherapy [68]. Takenaka et al. conjecture that these patients might fare better with preoperative CRT since postoperative treatment does not appear to improve overall survival [139].

Several studies have tried to tease out factors associated with overall survival. Masterson et al. in their multivariate analysis found that positive lymph nodes, poorly differentiated histology, and carotid involvement were indicators of poor prognosis [47]. Chi et al. found that survival was worse with poorly differentiated pathology [48]. Leong et al. found that patients with poorly differentiated tumors had significantly worse overall survival than patients with either well- or moderately differentiated tumors [107]. Others have not found histologic differentiation to affect overall survival.

For middle ear SCC, Gurgel et al. examined the SEER database and calculated a 23.9% 5-year observed survival rate for 135 reported cases [26]. For patients with localized middle ear disease, 5-year survival was slightly better for patients treated with surgery alone than with radiotherapy alone. For patients with regional disease, 5-year survival was better with surgery alone than with radiotherapy alone or combined surgery and radiotherapy [26].

Disease-Free Survival

Disease-free survival (DFS) measures the time from end of treatment until a locoregional recurrence is identified. In a large retrospective review of SCC of the temporal bone, Gidley et al. found that DFS was 60% [5]. Mazzoni et al. report an overall DFS of 49%. When divided into early- and late-stage tumors, they report a DFS for T1 and T2 tumors of 67%, and for T3 and T4 tumors the DFS is 41% [33].

Several factors have been found to affect DFS: positive margins [46], dura mater involvement [113], facial nerve sacrifice [16, 113], direction of spread in T4 tumors (anterior vs all other directions) [113], pathologic tumor grade [39, 113], Pittsburgh staging [5, 106, 130], and positive lymph nodes [64]. In a multivariate analysis, only dura mater involvement and nodal status were found to be independent factors affecting DFS [113].

Factors not associated with DFS include parotidectomy, neck dissection, and postoperative radiotherapy [113]. Essig et al. found that preoperative facial nerve palsy was not significantly correlated with disease-free survival [130]. Lobo et al. found that tumor histology did not make a significant difference in disease-free survival, but their study had only 19 patients [64].

Disease-Specific Survival

Disease-specific survival (DSS) is measured from the date of diagnosis until the last follow-up. The reported 5-year DSS for SCC of the temporal bone is 19–68% [5, 7, 8, 33, 46, 67, 175]. Mazzoni et al. further divided survival data based on early- and late-stage tumors. They found a DSS for T1 and T2 tumors of 92%, and for T3 and T4 tumors, the DSS is 48% [33]. Large tumors (Pittsburgh stage 3 and 4) have disease-specific survival ranging from 18% to 65% [69].

Several factors have been found to affect DSS: positive margins [46, 130], dura mater involvement [46, 113], facial nerve involvement or nerve sacrifice [46, 67, 113], direction of spread in T4 tumors (anterior vs all other directions) [113], pathologic tumor grade [113], Pittsburgh T stage [46, 67, 130], erosion of the medial wall of the middle ear [46], jugular bulb invasion [46], and TMJ invasion [46].

Higgins and Moody-Antonio performed a systemic review assessing 348 patients with temporal bone squamous cell carcinoma and found that DSS and overall survival were much worse for patients presenting with facial palsy compared to patients without facial weakness [67].

Factors not associated with DSS include parotidectomy, neck dissection, and postoperative radiotherapy [113]. Masterson et al. found that salvage surgery and brain involvement were not significant factors for disease-specific survival [47]. Essig et al. found that preoperative facial nerve palsy was not a significant correlation to disease-specific survival [130].

Bacciu et al. in a multivariate analysis found that only dural involvement was an independent predictor of DSS [46].

Zanoletti et al. found a significant difference in disease-free survival (DFS) based on the clinical and pathologic PSS by T stage, by lymph node involvement, and pathologic tumor grade [113].

Future Directions

Biomarkers

The family of signal transducers and activators of transcription (STAT) convey signals from extracellular stimuli and are important in inflammation, cell survival, differentiation, and proliferation. STAT3 has an important role in cancer development as an oncogene. STAT3 has been reported in head and neck cancers; however, STAT3 showed no significant correlation with survival in patients with temporal bone SCC [176].

Marioni et al. explored the expression of maspin in 29 archived squamous cell carcinomas of the temporal bone to determine patterns of recurrence and survival rates [106]. Cytoplasmic maspin (mammary serine protease inhibitor) is a tumor-suppressor protein. Their research found higher levels of maspin in patients who did not have a recurrence of SCCa of the temporal bone cancer. Additionally, they found that nuclear localization of maspin was also associated with significantly longer DFS [106].

Since bone invasion is an important prognostic factor for SCC of the temporal bone, Sugimoto et al. explored the factors that are responsible for epithelial-mesenchymal transition (EMT) [177]. EMT is determined based on histopathology. Their research showed an increase in vimentin expression in areas of bone invasion. They opined that increased EMT expression on biopsy might be a biomarker of propensity for bony invasion and thus a sign of poor prognosis.

Verrucous Carcinoma

Verrucous carcinoma is a variant of squamous cell carcinoma that is frequently seen in the oral cavity and larynx and only rarely has it been reported in the ear canal and temporal bone. To date, there have been only 18 cases reported in the English literature (Fig. 7.6) [178–182].

Unlike SCC, chronic irritation and smoking seem to be causally linked to the development of verrucous carcinoma [180]. Long-term, chronic (more than 60 years in one case [181]) otorrhea has been reported in a majority of cases.

Diagnosis can be delayed due to the benign appearance of biopsy material [180]. A superficial biopsy might only show a well-differentiated, extensively keratinizing squamous epithelium with blunt pushing margins and without cytological atypias microscopically [182]. Macroscopically, these tumors are superficially growing and fungating, papillomatous masses with well-demarcated borders. Multiple biopsies paired with clinically invasive behavior help to formulate the diagnosis [180, 182].

This tumor can be widely invasive throughout the temporal bone, including the petrous apex; however, the bony labyrinth, the facial canal, and the internal auditory canal can be spared from destruction [181]. This tumor type does not metastasize either to regional lymph nodes or distantly.

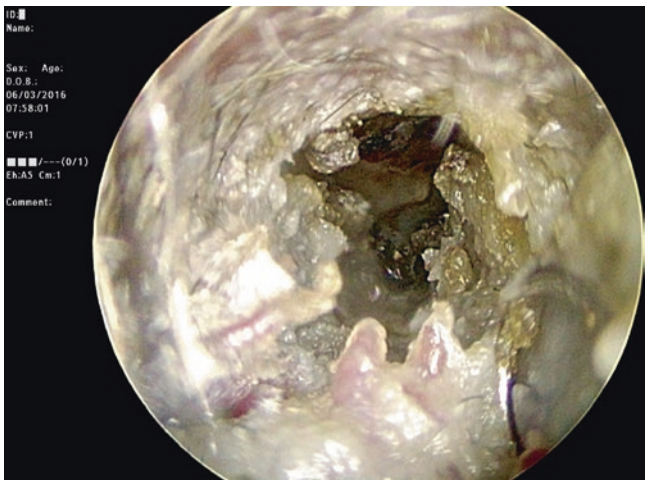

Fig. 7.6 Verrucous carcinoma of the ear canal. This tumor was staged as T1 and excised with lateral temporal bone resection. Postoperative radiotherapy was not indicated

Surgical excision is the primary treatment modality. The goal of achieving negative margins is emphasized for this tumor type, since adjuvant therapy has not improved outcomes.

The role of radiotherapy is controversial. Earlier reports indicated a potential dedifferentiation of tumor with radiation; however, more recent reports do not support this claim [182]. Radiotherapy was not helpful in preventing recurrences in patients with late-stage disease [178].

Radiation and chemotherapy have been linked with poorer prognosis, but this might be due to disease presenting at a later stage. For an inoperable tumor, chemotherapy consisting of vinblastine (2 mg/12 h IV, day 1), methotrexate (50 mg/8 h IV, day 2), and bleomycin (15 mg/6 h IV, days 2 and 3) as induction followed by three cycles with radiation (64 Gy in 2 Gy daily fractions) produced long-term survivor in one case [182].

Fully one-third of patients die within a year of diagnosis [180]. A few patients are reported to have long-term survival (4–10 years) [178]. These patients had complete resection of the tumor with negative margins and were treated with surgery alone. With such few cases reported, survival data is largely lacking.

Basal Cell Cancer

BCC of the ear canal and temporal bone is much rarer than SCC, but it represents the second most common tumor of the ear canal [8, 10, 12, 13, 118, 135, 137, 183, 184].

BCC also tends to have a better overall survival and prognosis than SCC [83, 88]. While the Pittsburgh staging system

was devised for squamous cell carcinoma of the temporal bone, it has been applied to other tumor histologies that affect this site [118]. Beyond stage 1 and 2, Testa et al. demonstrate that SCC has a significantly worse prognosis than similar staged BCC.

BCC has a propensity for recurrence, even with negative margins. Vandeweyer et al. reported results of six patients with BCCa of the external canal. Their patients presented with local recurrences even after radical surgery with negative margins; therefore, they considered BCCa an aggressive tumor with a poor prognosis [185].

Gaudet reports three patients with extensive periauricular BCCa [83]. Their study indicated that BCCa could be managed with surgery alone, but none of their patients had high-risk pathologic features.

In the MD Anderson series of temporal bone cancers, 42 patients had BCC involving the ear canal. The mean age was 67 years, and 83% were men. The most common presenting symptoms were hearing loss (36%) and otorrhea (26%). Recurrent lesions accounted for 60% of this patient population [186]. The tumor was confined to the cartilaginous canal in 52%. The bony ear canal was involved in 29%. Two patients (5%) had disease in the middle ear. Disease extended into the parenchyma of the parotid gland in 38%. Two patients developed regional nodal disease. Ten patients were able to be managed with wide local excision of the external auditory meatus and were reconstructed with a split thickness skin graft (Fig. 7.7). The remainder required lateral temporal bone resection, usually with total auriculectomy for advanced disease.

Parotidectomy was performed in 26 patients, and 62% of these patients had either direct tumor extension or metastatic disease within the gland (Fig. 7.8). Metastatic disease was only found in intraparotid and level II lymph nodes.

Fifty-five percent (55%) of patients received postoperative radiotherapy as indicated by recurrent tumor, perineural invasion, metastatic disease, or bony invasion.

The 5-year disease-specific survival was 100% with an overall survival of 84% and a 5-year disease-free survival of 77%.

Vismodegib, a hedgehog pathway inhibitor, can be used for locally advanced and metastatic basal cell carcinoma. This drug is especially useful for patients with basal cell nevus syndrome. This drug, however, does not produce durable disease remission. Surgical resection and radiotherapy are usually required for definitive treatment.

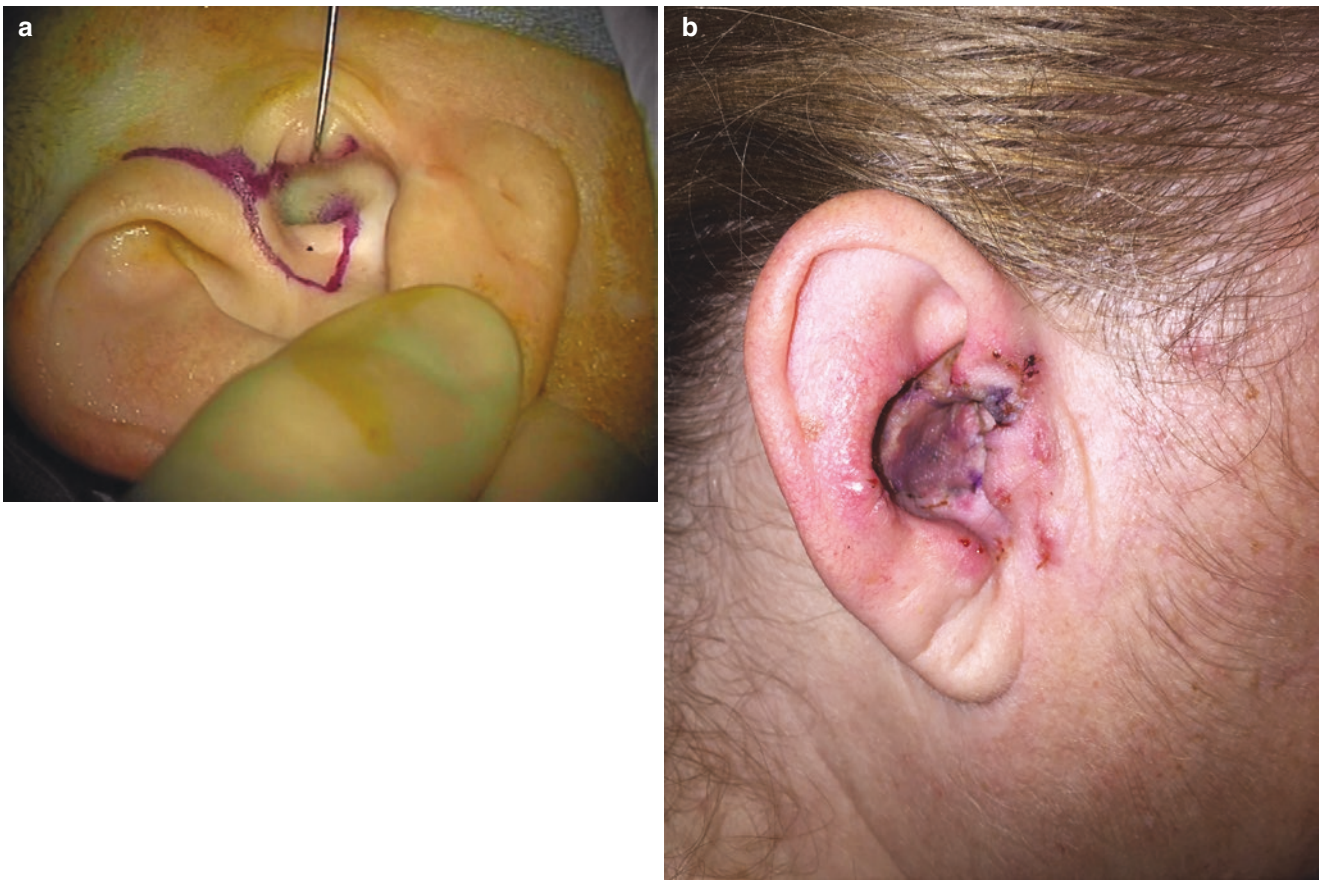

Fig. 7.7 Recurrent BCC of the right ear canal. (**a**) Endaural incision is performed to help with visualizing the ear canal portion. Additional tissues laterally and inferiorly were taken to achieve a final negative margin. (**b**) Nine days postoperative view of split thickness skin graft

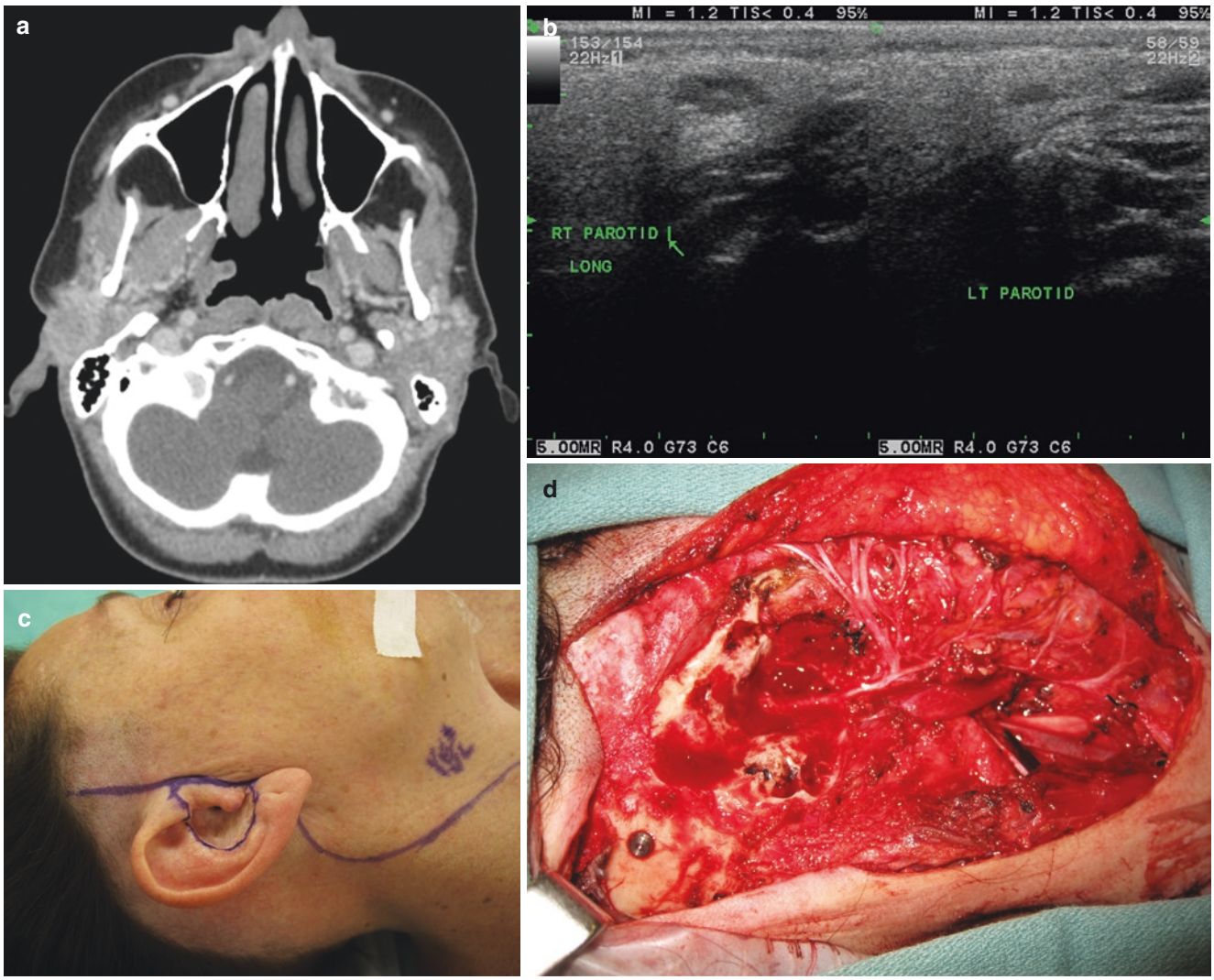

Fig. 7.8 Multiply recurrent BCC of the right ear. Lateral temporal bone resection was required. Final pathology revealed extensive perineural disease and metastatic lymph nodes in the parotid gland. (**a**) Preoperative CT showing disease within the right parotid gland. (**b**) Ultrasound-guided FNA confirmed BCC in the parotid. (**c**) Incisions marked. (**d**) Surgical defect following for lateral temporal bone resection, parotidectomy, and neck dissection. Osseointegrated implant is placed. Reconstruction performed with microvascular free flap

Conclusion

Squamous cell carcinoma is the most common malignant tumor of the ear canal, middle ear, and temporal bone. Basal cell carcinoma is the second most common tumor of the ear canal. Hearing loss, otalgia, and otorrhea are the classic triad of symptoms; however, this combination is seen in only 10% of cases. Squamous cell carcinoma of the ear canal is often mistaken for otitis externa or media until biopsy is performed, confirming the diagnosis. Cross-sectional imaging, either CT, MRI, or both, is required to evaluate the extent of disease. The Pittsburgh staging system is used, and it is a reliable predictor of survival. Surgery is the primary treatment for these tumors, especially for early-stage disease. Postoperative radiotherapy is indicated for tumors staged T2 and higher, recurrent lesions, and for patients with positive margins, perineural invasion, bone invasion, and lymph node metastasis. Preoperative chemotherapy and radiotherapy (CRT) has an emerging role for late-stage (T3 and T4) disease. The survival for temporal bone cancer has improved over the last 50 years, but overall survival still remains relatively low for advanced-stage disease.

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