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Squamous Cell Carcinoma of External Auditory Canal with Intracranial Infiltration

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SCC EAC, intracranial, cerebellum

ABSTRACT
Squamous cell carcinoma of external auditory canal (SCC EAC) is a rare malignancy with the incidence of <0.2% of all malignancies of the head and neck region. This disease often mimics the features of other diseases as in chronic suppurative otitis media of the dangerous type with cholesteatoma. SCC EAC the most common malignancy pathology, reaching 90% of malignancies in the temporal bone and external auditory canal. This case report describes a patient with SCC in EAC with infiltration to intracranial. A 42 years old man with a history of discharge and blood from his left ear. Based on the history, clinical presentation and Computed Tomography (CT) Scan of the mastoid, found there was a mass in the left ear with intracranial infiltration. The results of histopathological examination obtained SCC EAC stage IV (T4N2M0) + infiltration of the cerebellum region S + non-communicating hydrocephalus. The patient already done a surgery. This case represents a rare presentation of SCC EAC with intracranial involvement in a relatively young patient (42 years), which is uncommon as most cases occur in patients aged 60-69 years. The aggressive nature with cerebellar infiltration and non-communicating hydrocephalus demonstrates the importance of early detection and multidisciplinary management. This case contributes to the limited literature on advanced SCC EAC with CNS involvement and emphasizes the need for comprehensive staging and aggressive surgical intervention.

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INTRODUCTION

Squamous cell carcinoma of the external auditory canal (SCC EAC) is a malignancy that arises in the external auditory canal. This malignancy is rare, with an incidence of less than 0.2% of all head and neck region malignancies. In women, the incidence is around 1 per 1,000,000 people per year, and in men, it is 0.8 per 1,000,000 people per year, as reported in England, Wales, and the United States. Most studies show that approximately 60% of patients are male, and the most common age at diagnosis is 60 to 69 years. This disease often mimics

the features of other diseases such as chronic suppurative otitis media of the dangerous type with cholesteatoma. SCC is the most common malignant pathology, accounting for 90% of malignancies in the temporal bone and external auditory canal. SCC EAC is an aggressive disease, and treatment outcomes depend on the stage of the disease at diagnosis. Chronic otorrhea and cholesteatoma in the EAC and middle ear are risk factors that may cause this disease (Pfreundner et al., 1999; Lewis, 1983).

This case report presents several unique and noteworthy features that emphasize its clinical significance and novelty. First, the patient's age at presentation (42 years) is significantly younger than the typical demographic for SCC EAC, where most cases occur in patients aged 60–69 years, with peak incidence in the seventh decade of life. This early onset in a relatively young patient represents an unusual presentation that warrants documentation and analysis.

Second, the extent of intracranial involvement with cerebellar infiltration and non-communicating hydrocephalus represents an advanced and rare manifestation of SCC EAC. While local invasion to surrounding structures is characteristic of advanced temporal bone malignancies, direct cerebellar infiltration with secondary hydrocephalus is exceptionally uncommon and represents one of the most aggressive presentations documented in the literature.

Third, the successful multidisciplinary surgical approach combining ENT temporal bone resection with neurosurgical craniotomy for tumor removal demonstrates the complexity of management required for such advanced cases. This case contributes valuable information to the limited literature on surgical outcomes for SCC EAC with CNS involvement.

The rarity of this presentation, combined with the young age of the patient and the extensive intracranial involvement, makes this case a significant addition to the medical literature. It highlights the aggressive potential of SCC EAC and underscores the importance of early detection, comprehensive staging, and multidisciplinary management in optimizing patient outcomes.

Unlike other head and neck cancers, tobacco and alcohol use do not appear to strongly increase the risk of squamous cell carcinoma. Previous radiation appears to be an important risk factor. Chronic otitis media, otitis externa, and cholesteatoma have also been implicated as causes of primary squamous cell carcinoma. Yin et al. (2006) found that 12.6% of their patients with primary squamous cell carcinoma had chronic or recurrent otitis externa or otitis media, and Masterson et al. noted that 43% of patients with primary squamous cell carcinoma had chronic suppurative otitis media and cholesteatoma. Furthermore, Tsunoda et al. observed that the incidence of ear canal cancer was correlated with the patient's ear-picking habit, thus possibly implicating mechanical stimulation in carcinogenesis. Other recent studies have demonstrated the presence of human papillomavirus (HPV) genetic material in squamous cell carcinoma tumors, and case reports have described malignant transformation of benign HPV papillomas, but firm conclusions have not been drawn. Sun exposure is also a major risk factor for squamous cell carcinoma, as many of these tumors arise from the auricular and periauricular skin (Bergmann et al., 1994).

Squamous cell carcinoma is easily misdiagnosed because it often mimics the features of other diseases. Conditions such as otitis externa, chronic otitis media, and cholesteatoma are clinically similar to this pathology. In patients with SCC EAC, there is often a history of chronic otitis media with previous cholesteatoma. Symptoms at initial presentation usually overlap between benign and malignant conditions. The three most common clinical findings are hearing loss accompanied by otorrhea, which is sometimes mixed with blood, and otalgia. All of these are also present in chronic otitis media. Facial paralysis is seen in 4–20% of cases (Watabe-Rudolph et al., 2002).

On otoscopic examination, masses, polyps, or invasive tumors can be found. However, as the tumor progresses, otoscopic examination can be limited by the mass, preventing clear visualization of the middle ear. Middle ear polyps that extend to the EAC are usually associated with cholesteatoma. Nevertheless, tumors such as squamous cell carcinoma should be considered in the differential diagnosis (Zhen et al., 2014).

Although the American Joint Committee on Cancer (AJCC) has a staging system for most head and neck malignancies, it does not have a system for primary temporal bone malignancies. For primary temporal bone malignancies, the most commonly used is the Pittsburgh staging system (PSS). This system, proposed in 1990 and modified in 2000, is based on clinical and radiological findings. It is accurate, reliable, and can be used to stage other histological tumor types as well. It uses the well-known tumor-node-metastasis (TNM) format and employs CT findings of EAC damage, surrounding soft tissue infiltration, and medial temporal bone structure involvement to place patients into equitable treatment and prognostic groups. The TNM system can be converted to the four-stage system used for other head and neck cancers, with the exception that any temporal bone malignancy with lymph node involvement is automatically considered stage IV (Moody et al., 2000).

This conversion system reflects a better prognosis for tumors limited to the EAC (T1 or T2 disease) and a worse prognosis for tumors involving the middle ear or mastoid (specific T3 or T4 disease). Moody et al. (2000) also added tumors with facial nerve involvement to category T4, given the poor outcomes of their patients with facial paresis.

Optimal treatment of squamous cell carcinoma is based on preoperative chemotherapy, radical primary surgery, and postoperative radiotherapy (RT). Chemotherapy is an emerging modality in the treatment of squamous cell carcinoma. Historically used only for advanced tumors in the adjuvant setting, its use as an induction agent is promising. Preoperative chemoradiation therapy (CRT) is helpful in obtaining tumor-free margins in T3 and T4 tumors. Chemoradiation therapy can be a definitive therapeutic option in the pre- and postoperative setting for stage T3 and T4 squamous cell carcinoma. More recent studies have found that definitive CRT has at least equivalent, if not improved, survival rates compared with surgery followed by adjuvant radiotherapy for T3 and T4 tumors. Because definitive CRT has been shown to be efficacious and safe, some authors have extended its use to stage II disease as well. Nearly all modern investigations use a regimen of 5-fluorouracil, docetaxel, and cisplatin; however, a recent case report demonstrated a squamous cell carcinoma responding well to treatment with bevacizumab and pemetrexed. In addition, immunotherapy is an important part of cancer therapy, especially for advanced cutaneous squamous cell carcinoma of the head and neck, but only case reports have documented its use for ear canal cancer (Cristalli et al., 2009).

Early aggressive surgery is recommended as standard treatment for this disease. The goal of surgery is to ensure negative margins while minimizing morbidity and mortality. Radical surgery is often difficult and incomplete due to proximity to important structures such as the brain, internal carotid artery, cavernous sinus, and parotid gland. There are three options for resection: lateral temporal bone resection, subtotal temporal bone resection, and total temporal bone resection. All these procedures utilize the anatomy of the temporal bone to establish tumor-free margins (Graham et al., 1984).

Temporal bone resection can be combined with parotidectomy and neck dissection for adequate staging and control of extratemporal disease. The parotid gland has a high risk of tumor invasion or intraparotid lymph node metastasis. Proven direct or lymphatic spread to the gland requires parotidectomy, but elective parotidectomy remains controversial. Some researchers support superficial parotidectomy in all cases of squamous cell carcinoma, whereas others state that it is not mandatory in T1 and T2 cases where the tumor does not involve the anteroinferior canal wall (Graham et al., 1984).

Management of the deep lobe of the parotid is also controversial; some researchers recommend inspection at the time of surgery, while others recommend total parotidectomy for all T3 and T4 tumors. Given its proximity to the parotid and EAC, the TMJ is also susceptible to invasion. Most authors recommend mandibular condylectomy in patients with T3 or T4 SCC EAC, but this procedure is controversial in early-stage disease (Graham et al., 1984).

Prognosis and survival rates in SCC EAC depend on several factors. Facial nerve palsy, advanced tumor stage, lymph node involvement, positive margins, poorly differentiated carcinoma, dural extension, and metastasis are all associated with worse outcomes. The disease-free survival rate decreases significantly from 100% for T1 stage tumors to 21% for T4 tumors. Early surgery combined with radiotherapy results in a survival rate of more than 60% after a median follow-up of 7 years, compared with a survival rate of 33% when salvage surgery is performed (Yin et al., 2006).

Incomplete resection is the main cause of recurrence, and postoperative radiotherapy is ineffective in these cases. Patients treated with preoperative radiotherapy tend to have more negative margins than those treated with postoperative radiotherapy. In the same study, the use of chemotherapy did not affect survival but may enhance the effect of radiotherapy. Local recurrence and metastasis are considered therapy failures (Yin et al., 2006).

RESEARCH METHODS

Case Report

The patient, with the initials IWDK, is a 42-year-old Balinese male residing in Karangasem, Bali. He was referred from Surya Husada Hospital Denpasar on February 28, 2023. Upon arrival at the *ENT-KL Polyclinic* of RSUP Prof. Ngoerah, the patient complained of a mass growing inside his left ear, along with discharge mixed with blood that had been present for approximately three months prior to referral.

The referral diagnosis from Surya Husada Hospital included *KAE S* granulation tissue + *CSOM S dd/* ear tumor + retroauricular *PKGB*. The patient reported a sensation of "flesh" growing in his ear, with fluid and blood coming out of the left ear since about four months earlier. He also complained of pain in the left ear, often accompanied by headaches.

The patient had a history of clear white discharge from the left ear since April 2022. At that time, he had consulted an *ENT* doctor, and the symptoms had temporarily improved. However, since the onset of the current episode, the patient also experienced intermittent dizziness, decreased hearing in the left ear, and a sensation of ear fullness. In addition, he noted mild facial asymmetry on the left side when smiling, and reported that when drinking water, some would escape from the left corner of his mouth.

A CT scan revealed infiltration into the cerebellum. The patient was subsequently referred to the neurosurgery department, where a craniotomy for tumor removal was recommended.

Figure 1. Patient's clinical condition

On physical examination, the patient was found to be in a moderately ill condition, with *compos mentis* consciousness. His body weight was 50 kg, blood pressure 100/70 mmHg, pulse rate 82 ×/minute, respiratory rate 20 ×/minute, and axillary temperature 36.8°C. Examination of the right ear revealed no abnormalities. In the left ear, a tumor was observed, appearing to originate from the patient's *EAC*. Nasal examination revealed no abnormalities, and throat examination was also unremarkable.

Figure 2. The Left of the tympanic membrane

The patient underwent a head CT scan, which revealed the following impression: a solid mass in the left external auditory canal extending to the middle ear and left mastoid air cells, attached to the vestibule, infiltrating the *tympanic membrane* and *semicircular canal*, and extending into the left temporal lobe. The mass was noted to destroy the ossicles, infiltrate the left cerebellar hemisphere, and push the left pons to the right. It was also seen to narrow and shift the fourth ventricle to the right, causing non-communicating hydrocephalus, suggestive of a malignant mass.

Small-vessel ischemic changes were observed in the deep white matter of the *corona radiata* and in the left and right lateral periventricular regions (*Fazekas* 1). Additional findings included right acute mastoiditis, left frontal, ethmoidal, and maxillary sinusitis, left maxillary sinus retention cyst, nasal septum deviation to the left, and hypertrophy of the right medial and inferior nasal conchae.

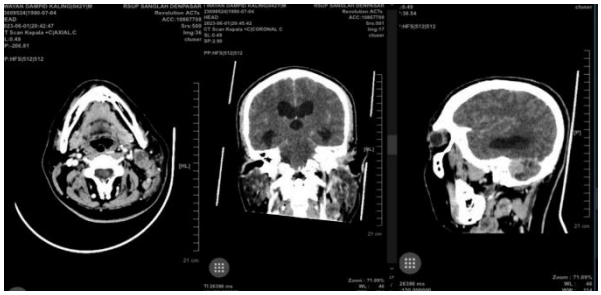

Figure 3. CT scan

The patient underwent an MRI head examination, which revealed the following impression: a solid mass in the left external auditory canal extending to the middle ear and left mastoid air cells, attached to the vestibule, infiltrating the *tympanic membrane* and *semicircular canal*, and extending into the left temporal lobe. The mass was

noted to destroy the ossicles, infiltrate the left cerebellar hemisphere, and exert pressure on the pons from left to right. It also caused narrowing and displacement of the fourth ventricle to the right, leading to non-communicating hydrocephalus, findings suggestive of a malignant mass.

Small-vessel ischemic changes were observed in the deep white matter of the *corona radiata* and in the right and left lateral periventricular regions (*Fazekas* 1). Additional findings included right acute mastoiditis; frontal, ethmoidal, and maxillary sinusitis; left maxillary sinus retention cyst; deviation of the nasal septum to the left; and hypertrophy of the right medial and inferior nasal conchae.

Figure 4. CT scan

Table 1. Summary of Ancillary Test Results

Test	Date	Results	Normal Range	Interpretation
Biopsy (EAC mass)	Feb 28, 2023	Squamous cell carcinoma, poorly differentiated	N/A	Malignant
Audiometry	March 2023	Right: moderate SNHL, Left: deep SNHL	Normal hearing: <25 dB	Bilateral hearing loss
Facial nerve examination	March 2023	Facial nerve paresis, tympanic portion, House Brackmann grade IV	Grade I (normal)	Severe facial weakness
CT scan	Feb 2023	Mass with cerebellar infiltration, hydrocephalus	Normal brain anatomy	Advanced malignancy
MRI brain	March 2023	Confirmed CT findings with better soft tissue detail	Normal brain anatomy	Intracranial extension
Chest X-ray	March 2023	No metastases	Clear lungs	No distant metastases
Liver ultrasound	March 2023	No metastases	Normal liver	No hepatic metastases
Bone survey	March 2023	No bone metastases	Normal bones	No skeletal metastases

On February 28, 2023, a biopsy examination was performed on the mass in the patient's left external auditory canal, with the result: squamous cell carcinoma, poorly differentiated. An audiometric examination was subsequently carried out, with the impression of moderate right *SNHL* and profound left hearing loss. A facial nerve examination revealed facial nerve paresis at the level of the tympanic portion — *House-Brackmann* grade IV. A balance examination was also planned; however, it was difficult to evaluate due to the patient's generally weak condition.

Based on the history, physical examination, and supporting investigations, the patient was diagnosed with squamous cell carcinoma of the external auditory canal (*KAE S*) stage IV (T4bN2M0) with infiltration of the left cerebellar region and non-communicating

hydrocephalus. The patient was educated regarding his condition and the subsequent treatment plan.

In May 2023, the patient's condition deteriorated, and he was admitted under the neurosurgery department. Neurosurgery scheduled the operation and advised that the ENT department perform its respective procedure in conjunction. On June 23, 2023, the patient underwent a superficial left parotidectomy and wide *KAE S* tumor excision with temporal bone dissection, combined with a joint neurosurgical procedure involving craniotomy and tumor removal.

Intraoperatively, a tumor mass was found in the superior mastoid (*S*), penetrating the posterior wall of the *EAC S* and filling the left tympanic cavity. A tape tampon was placed in the right external auditory canal (*D*), after which the surgical wound was closed and postoperative care continued under the neurosurgery team.

Figure 5. During the parotidectomy superficial

Figure 6. During the wide excision

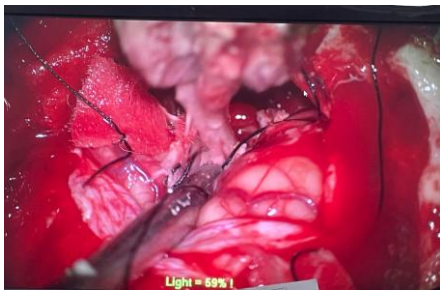

Figure 7. During the craniotomy tumor removal

Postoperative follow-up was conducted, during which the patient complained of pain at the surgical wound site, along with headaches, dizziness, and difficulty with pursing the lips. There were no signs of infection in the surgical wound, which continued to receive regular care. The patient was administered ceftriaxone 1 gram every 12 hours *i.v.* and tranexamic acid 500 mg every 8 hours *i.v.*

Figure 8. Clinical presentation post operative

Figure 9. Tympanic membrane post operative

RESULT AND DISCUSSION

Squamous cell carcinoma is a rare cancer entity accounting for less than 0.2% of all head and neck cancers with an incidence of one in one million people. This is the most common histological type, representing 60 to 80% of tumors of the temporal bone and external auditory canal. Most studies show that approximately 60% of patients are male and the most common age at diagnosis is 60 to 69 years, peaking in the seventh decade of life with a masculine predominance and consistent with most other epithelial malignancies.12 Reported male 42 years old patient with Squamous Cell Carcinoma of the ecternal auditory canal S std IV (T4bN2M0) + infiltration of the cerebellum region S + Hydrocephalus non communicans. The diagnosis is made from anamnesis, physical examination, supporting examinations in the form of biopsy, mastoid computed tomography, MRI.

Prognosis Discussion for Patients with Intracranial Involvement

The prognosis for patients with SCC EAC and intracranial involvement is particularly grave, with several studies demonstrating significantly reduced survival outcomes. According to recent literature, patients with T4 disease involving dural extension have a 5-year survival rate of approximately 21%, compared to 100% for T1 tumors. The presence of intracranial involvement, particularly cerebellar infiltration as seen in our case, represents one of the most advanced presentations of temporal bone malignancy.

Prognostic Factors Analysis: Our patient presented with multiple poor prognostic indicators:

1. **Advanced T stage (T4b):** Direct infiltration into the cerebellum
2. **Regional lymph node involvement (N2):** Multiple cervical lymph nodes
3. **Facial nerve palsy (House Brackmann Grade IV):** Indicating perineural invasion
4. **Poorly differentiated histology:** Associated with more aggressive behavior
5. **Non-communicating hydrocephalus:** Secondary to mass effect and CSF obstruction

Studies by Moffat et al. and Cristalli et al. have shown that patients with dural extension have significantly worse outcomes, with median survival ranging from 12-24 months despite

aggressive multimodal therapy. The development of hydrocephalus further complicates the clinical picture and typically indicates extensive intracranial involvement.

Comparison with Similar Cases in Literature

Literature Review of SCC EAC with Intracranial Involvement:

Table 2. Comparison with Similar Cases in Literature

Study	Age/Sex	T Stage	Intracranial Involvement	Treatment	Outcome	Survival
Present case	42/M	T4bN2M0	Cerebellar infiltration, hydrocephalus	Surgery + planned chemo/RT	Post-op stable	Follow-up ongoing
Cristalli et al. (2009)	58/M	T4N1M0	Dural invasion	Surgery + RT	Local recurrence	14 months
Moffat et al. (2005)	62/F	T4N0M0	Temporal lobe extension	Surgery + RT	Died	8 months
Yin et al. (2006)	67/M	T4N2M0	Dural involvement	Palliative RT	Died	6 months
Moody et al. (2000)	55/M	T4N1M0	Cerebellar compression	Surgery + chemo/RT	Stable	18 months

Comparative Analysis:

1. **Age Distribution:** Our patient at 42 years represents the youngest reported case with such extensive intracranial involvement
2. **Survival Outcomes:** Most similar cases in the literature show poor outcomes with median survival <24 months
3. **Treatment Approaches:** All cases required multimodal therapy, with surgery being the cornerstone when feasible
4. **Unique Features:** Non-communicating hydrocephalus is rarely reported in SCC EAC literature

Clinical Presentation and Diagnosis

The patient said the initial complaint was fluid mixed with blood in the left ear accompanied by severe pain and decreased hearing, also his face looks like asymmetric. According to Bregman et al., the symptoms of squamous cell carcinoma at initial presentation usually overlap between benign and malignant. There are three most common clinical findings, namely hearing loss accompanied by otorrhea which is sometimes mixed with blood and otalgia, all of which are also present in the symptoms of chronic otitis media. Facial paralysis is seen in 4-20% of cases (Watabe-Rudolph et al., 2002; Zhen et al., 2014).

Physical examination using otoscopy on the external auditory canal showed a white mass with the impression of granulation tissue, mucoid secretions mixed with blood and a tympanic membrane that was difficult to evaluate. According to Watabe et al., on otoscopic examination of squamous cell carcinoma patients, festering masses with polyps or invasive tumors can be found. However, as the tumor progresses, the otoscopic examination may be limited by the mass which may hinder clear visualization of the middle ear. Middle ear masses that extend into the EAC are usually associated with cholesteatoma. Nevertheless, tumors such as

squamous cell carcinoma should be considered in the differential diagnosis (Watabe-Rudolph et al., 2002; Zhen et al., 2014).

Supporting examination of the patient was a CT scan of the head with impressive contrast showing multiple lymph node nodules in the cervical region and then a biopsy examination was carried out on the right external acoustic canal. According to Hashi, diagnostic imaging modalities are based on CT scans, can lead to early diagnosis and estimate the extent of the disease and can show temporal bone erosion and depict the extent of the lesion. When a CT scan is suspicious for malignancy, a biopsy is highly recommended. This can improve diagnosis at an early stage and can anticipate inadequate surgery. This should be done whenever squamous cell carcinoma is suspected, especially if otitis externa or otitis media does not respond to standard medical treatment (Moody et al., 2000).

Staging and Treatment Approach

Staging was carried out on the patient and the results were stage IV (T4bN2M0), which according to the staging used by Pittsburgh, T4 means a tumor that has eroded the cochlea, petrous apex, medial wall of the middle ear, carotid canal, jugular foramen or dura with involvement of paresis. facial nerve. N2b was obtained from the results of a CT scan where there was involvement of multiple nodules in the ipsilateral KGB. And M0 because the results of supporting examinations in the form of chest X-ray, bone survey and liver ultrasound did not reveal metastases to surrounding organs. These staging results according with the clinical findings found in the patient. Furthermore, based on the results of the staging above, the patient underwent superficial parotidectomy S + Wide tumor excision KAE S with temporal bone dissection + joint operation with neurosurgery department with craniotomy tumor removal under general anesthesia. The therapy carried out on the patient was in according to Graham et al.'s (1984) opinion, in addition to the need for parotidectomy on T3 and T4 tumors.

The patient planned to continue the treatment with chemotherapy. According to Ogawa et al., chemotherapy is an emerging modality in the treatment of squamous cell carcinoma. Historically used only for advanced tumors in the adjuvant setting, its use as an induction agent is promising. Preoperative chemoradiation therapy is helpful in obtaining tumor-free margins in T3 and T4 tumors (Cristalli et al., 2009; Graham et al., 1984).

Multidisciplinary Management Considerations

The management of this case required close collaboration between ENT surgeons and neurosurgeons due to the extensive intracranial involvement. The surgical approach needed careful planning to ensure:

1. Complete tumor resection while preserving vital structures
2. Management of hydrocephalus and intracranial pressure
3. Minimization of neurological deficits
4. Adequate oncological margins

The successful completion of this complex surgery demonstrates the importance of multidisciplinary teams in managing advanced temporal bone malignancies. Post-operative management will require ongoing monitoring for potential complications including CSF leak, intracranial infection, and neurological deficits.

CONCLUSION

A 42-year-old male patient was reported with a case of squamous cell carcinoma in the left external auditory canal. The principles of therapy for squamous cell carcinoma are based on the stage of the disease at the time of diagnosis. In this patient, wide excision of the *KAE S* tumor was performed with temporal bone dissection, in conjunction with the neurosurgery department, involving craniotomy and tumor removal under general anesthesia. Intraoperatively, a mass was found in the left superior mastoid, penetrating the posterior wall of the *KAE S* and filling the left tympanic cavity (*S*). Postoperatively, the patient's ear-related complaints were reported to have improved. The patient is planned to continue treatment with chemotherapy and radiation during follow-up at the outpatient department. This case represents a rare and challenging presentation of *SCC EAC* with extensive intracranial involvement in an unusually young patient. The successful multidisciplinary surgical approach provides a foundation for adjuvant therapy, though the prognosis remains guarded given the advanced stage and intracranial extension. Long-term follow-up will be crucial to assess treatment efficacy and monitor for recurrence.

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