

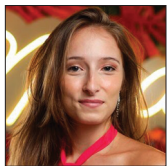
Case Report

Temporal bone squamous cell carcinoma: Aggressive behavior coursing with cerebellar invasion and hydrocephalus

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ABSTRACT

Background: Temporal bone squamous cell carcinoma (TBSCC) is a very rare condition. The prognosis is dismal for advanced tumors. Due to its rarity, information in the literature is scarce. Here, we report a unique case of TBSCC with cerebellar invasion and hydrocephalus.

Case Description: A 46-year-old reported right-sided hearing loss and a painful right retroauricular mass for 4 months. Magnetic resonance imaging revealed a 8.7 × 7.6 × 6.4 cm mass invading the right temporal and occipital bones. After a biopsy and 3 surgical procedures over 6 months, the diagnosis of TBSCC was obtained. Due to invasion of the cerebellar tissue and obstructive hydrocephalus, a ventriculoperitoneal shunt was performed. The patient was referred for adjuvant radiotherapy. However, palliative care was initiated due to tumor progression.

Conclusion: We report a case of advanced TBSCC with poor prognosis despite surgical treatment and radiotherapy. More data are necessary to provide new and better treatment to these patients.

Keywords: Cerebellum, Hydrocephalus, Squamous cell carcinoma, Temporal bone

INTRODUCTION

Temporal bone squamous cell carcinoma (TBSCC) is a rare head and neck tumor with aggressive behavior.^[8] Squamous cell carcinoma (SCC) represents around 40% of all temporal bone tumors and accounts for 0.2% of the total head and neck malignancy.^[5,18] The worldwide annual incidence is estimated at approximately 1–6 cases per million.^[14] Late stage at presentation is common, resulting in delayed diagnosis and poor prognosis.^[8,11] The periauricular soft tissues, parotid gland, temporomandibular joint, and mastoid are often affected by tumor progression. In advanced stages, the carotid canal, jugular foramen, dura mater, middle, and posterior cranial fossae can be invaded.^[2,7] The incidence of cerebellar invasion is not known, and after careful

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literature review, there were no cases reported. Our aim is to report a challenging and unique case of aggressive TBSCC presenting with cerebellar tissue invasion and hydrocephalus.

CASE DESCRIPTION

A 46-year-old longshoreman went to a University Hospital reporting right-sided hearing loss and a painful right retroauricular mass, which was noticed 4 months earlier after being unable to wear his safety helmet. He also reported rapid weight loss of 4 kg in 1 month. His only previous comorbidity was bilateral occupational hypoacusis. Clinical examination revealed a right retroauricular mass, in addition to right anacusis and left hypoacusis. On careful inspection, there was no evidence of skin lesions other than diffuse milia [Figure 1]. He was admitted to the hospital and a brain magnetic resonance imaging (MRI) showed an expansive mass measuring $8.7 \times 7.6 \times 6.4$ cm with heterogeneous enhancement, areas of hemorrhage, and calcification. The lesion invaded the mastoid, petrous part of the right temporal bone, and squamous part of the occipital bone with no evidence of involvement of the cerebellar tissue [Figure 2]. Chest, abdomen, and pelvis computed tomography (CT) did not reveal any other mass, tumor, or nodal involvement.

A tumor biopsy was performed and the histopathological findings were negative for malignancy and infection screening (GeneXpert – tuberculosis, Gram staining, and mycological culture). After that, due to the rapid tumor growth, he underwent another 2 surgical partial resections with positive margins over a period of 5 months. The tumor was not completely resected due to invasion of petrous bone and the risk of injury to the facial nerve. In the first

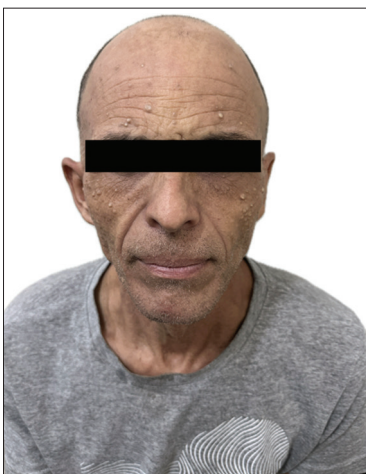


Figure 1: The patient exhibited multiple subepidermal keratin cysts that appeared as small, firm white papules in varying numbers, consistent with milia.

surgical approaches, samples from the temporal bone tumor were predominantly represented by keratin sheets with parakeratosis, sometimes forming horny pearls, with extensive degenerated areas. Foci of preserved squamous epithelium were observed, showing areas with enlarged nuclei, but without signs of stromal invasion and criteria for malignancy. The sample was also subjected to an immunohistochemical study with p16, which was negative.

Two months after the last surgery, a new brain MRI revealed local recurrence and invasion of the cerebellar tissue [Figure 3]. Another partial resection was performed [Figure 4]. The specimen exhibited fragments of squamous epithelium displaying cells with moderate to marked pleomorphism, with an enlarged and irregular nucleus, and frequent atypical mitotic figures, superficially infiltrating the cerebellar cortex and fibroconnective tissue. The rest of the material was similar to that observed in the first surgeries. In view of all these findings, it was considered to be squamous carcinoma. Pathology samples are collected in Figure 5. The Modified Pittsburgh Moody Scale was compatible with T4N0M0 (grade IV) TBSCC.

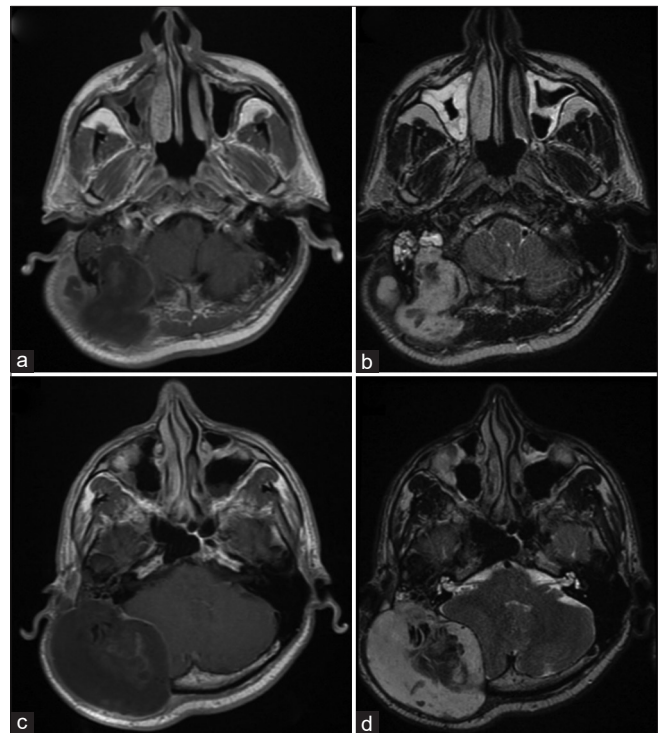


Figure 2: First magnetic resonance imaging of the brain showing a huge lesion in the temporal bone. (a) Axial T1-weighted with contrast exhibiting temporal bone mass with peripheral contrast enhancement – right side. (b) Axial T2-weighted image shows hyperintense signal abnormality in the mastoid part of the temporal bone. The occipital bone was invaded. (c) Contrast-enhanced axial T1-weighted showing a large temporal bone tumor on the right side. (d) The mass was predominantly hyperintense on the T2-weighted image.

One month after the last procedure, the patient exhibited a recurrent pattern of bulging deformity with a new holocranial headache. He did not present focal deficit symptoms. The new MRI revealed infiltration of the mass into the cerebellar region, accompanied by compression of the fourth ventricle and obstructive hydrocephalus [Figure 6]. Therefore, he underwent a left parietal ventriculoperitoneal shunt (VPS). Radiotherapy was started 6 weeks after VPS. At present, his surgical wound is open [Figure 7] and he is being monitored by palliative care.

DISCUSSION

Temporal bone malignancies are rare, constituting only 0.2% of all head and neck malignancies. The global annual incidence is estimated to be 1.3 cases per million, and it is detected in approximately one in every 5000–20000 patients presenting with otological complaints.^[4,14] SCC is responsible for even fewer cases, despite representing the most prevalent histological type among temporal bone tumors.^[9] TBSCC constitutes 60–80% of primary tumors in the temporal bone; yet, it comprises only about 40% of all temporal bone tumors. Across studies on TBSCC, around 60% of patients are men,

with the most frequent age of diagnosis falling between 60 and 69 years.^[5,17]

In contrast to other head and neck cancers, the risk of primary TBSCC does not appear to be significantly elevated by tobacco and alcohol consumption. Instead, prior radiation emerges as a notable risk factor. In addition, chronic otitis media, otitis externa, and cholesteatoma have been identified as potential contributors to primary TBSCC.^[1,12] Recent studies have indicated the existence of human papillomavirus (HPV) genetic material in TBSCC tumors. Case reports have even detailed the malignant transformation of benign HPV papillomas. However, definitive conclusions are still pending. To date, in our patient samples, the expression of biomarker p16 (highly correlated with HPV infection in head and neck SCC) was negative. Sun exposure emerges as a significant risk factor for TBSCC, especially since many of these tumors originate from the skin around the ear. Nonetheless, the precise cause for most cases remains elusive.^[10]

The principal clinical indicators of temporal bone malignancies include otalgia, otorrhea, and hearing loss. Gradually, signs indicating more serious conditions, such as facial weakness or the presence of a parotid or neck mass, may manifest.^[5] Establishing a diagnosis of TBSCC requires a mandatory biopsy. The presence of inflammation or edema can pose a challenge to obtain an accurate biopsy. Consequently, a deep tissue biopsy is often essential to ensure a conclusive and positive outcome.^[13] In our case, it was difficult to obtain a definitive diagnosis due to pathological samples with large amounts of keratin and differentiated tissue. The diagnosis was only confirmed after the last resection including tumoral samples invading the cerebellar tissue.

Given that a substantial portion of the temporal bone is typically not directly visible, imaging studies play a crucial role in both staging and management. CT scan and MRI provide complementary information about the extent of the tumor.^[8] Accurately assessing tumor spread is essential, as the degree of local tumor extension stands out as a key prognostic factor for TBSCC.^[3]

The main staging system used for temporal bone malignancies is the Pittsburgh Staging System (PSS). The PSS adopts the familiar tumor-node-metastasis (TNM) format, relying on CT findings such as bone external

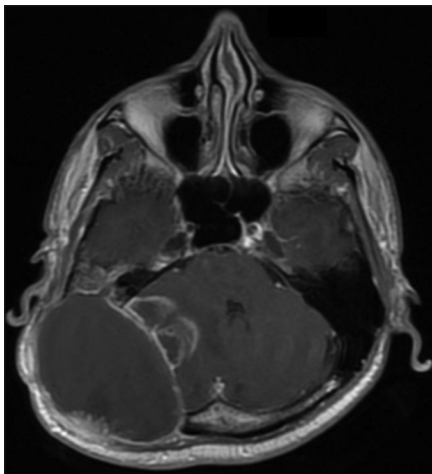


Figure 3: Contrast-enhanced axial T1-weighted brain magnetic resonance imaging shows temporal bone tumor invading the cerebellar tissue with dislocation of the IV ventricle.

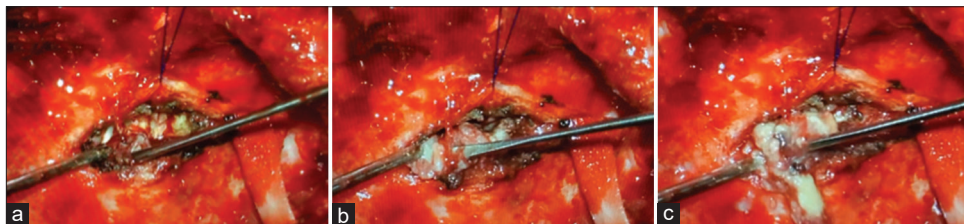


Figure 4: Microsurgical resection of the tumor invading the cerebellar tissue. (a) The tumor was identified after the dural opening. (b) Circumferential dissection between the tumor and normal cerebellum parenchyma. (c) Tumor removal using a microdissector.

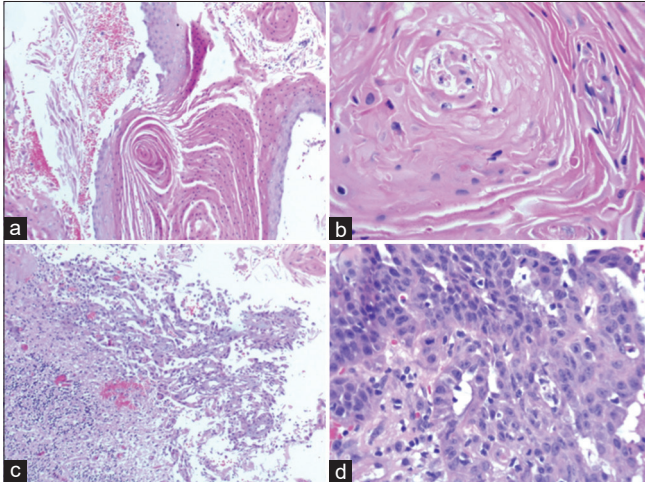


Figure 5: Temporal bone tumor samples: (a) Hematoxylin and Eosin (H&E) stain ($\times 200$). Squamous epithelium with some maturation, keratin sheets with parakeratosis, sometimes forming horny pearls. (b) H&E stain ($\times 400$). Foci of preserved squamous epithelium showing areas with enlarged nuclei, but no signs of stromal invasion. (c) H&E stain ($\times 100$). Sample from the cerebellar region. Squamous epithelium presenting poorly cohesive cells with moderate to marked pleomorphism, superficially infiltrating the cerebellar cortex. (d) H&E stain ($\times 400$). Sample from the dural region. Squamous epithelium with an enlarged and irregular nucleus, and frequent atypical mitotic figures infiltrating fibroconnective tissue.

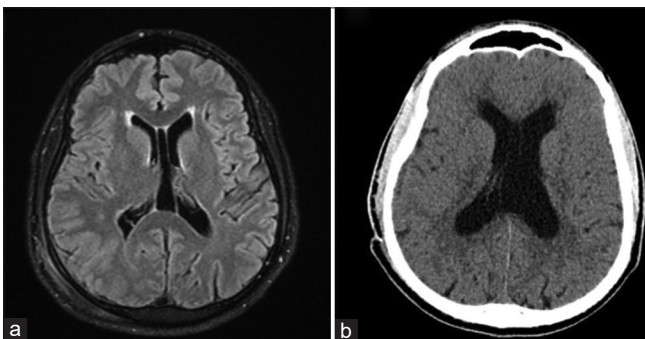


Figure 6: Comparison between the first brain magnetic resonance imaging (MRI) (fluid-attenuated inversion recovery [FLAIR] sequence) and head computed tomography (CT) after symptoms of hydrocephalus. (a) Preoperative MRI-FLAIR shows a normal lateral ventricle without radiological signs of hydrocephalus. Normal anatomical variation is observed: Cavum septum pellucidum. (b) Head CT showing enlargement of the lateral ventricles and transependymal edema: Low-density change on CT around the margins of the ventricles.

auditory canal (EAC) destruction, surrounding soft-tissue infiltration, and involvement of medial bony temporal structures. This approach allows the categorization of patients into treatment and prognostic groups. TNM can be transformed into the standard four-stage system used for other head and neck cancers, with the exception that any temporal bone malignancy with lymph node involvement



Figure 7: Final appearance of the wound after four attempts at tumor resection and adjuvant radiotherapy. The surgical wound is open and the patient was referred for palliative care.

is automatically classified as stage IV.^[3] This conversion reflects the more favorable prognosis for tumors limited to the EAC (T1 or T2 disease) and the worse prognosis for tumors affecting the middle ear or mastoid (specific T3 or T4 disease).^[9]

Due to the rare occurrence of TBSCC, there is a paucity of adequate randomized trials, leading to wide variation in the extent of resection among different authors and institutions. In general, indicators of low disease-specific survival rates in patients with TBSCC include nodal involvement, poorly differentiated histology, carotid involvement, positive margins, stage T4, dural invasion, and temporomandibular joint invasion.^[10] The standard approach for the oncological management of TBSCC is surgical intervention. Three resection options are available: Lateral temporal bone resection, subtotal temporal bone resection, and total temporal bone resection. Each of these procedures takes advantage of the anatomy of the temporal bone to establish tumor-free margins and can be executed either en bloc or in a piecemeal manner.^[8]

Regarding surgical treatment, we considered the significant morbidity associated with total resection of the temporal bone and the absence of proven survival benefit.^[16] Most radical procedures result in the loss of sensorineural hearing and often compromise facial nerve function.^[11] Furthermore, removing the ear canal and even the auricular pavilion results in significant cosmetic impact.^[15,16] In our case, the decision of a piecemeal partial resection was previously shared with the patient that agreed with the non-radical procedure, especially preserving his facial nerve function.

Radiotherapy for TBSCC is typically administered in the adjuvant postoperative period. Some indications include

lymph node metastasis, perineural invasion, positive margins, recurrent tumors, and bone invasion.^[14] Numerous studies have also demonstrated better survival rates with adjuvant radiotherapy compared to surgery alone, especially for T2 and higher tumors. On the other hand, there are limited benefits seen with postoperative radiotherapy for patients with completely resected T1 tumors.^[6]

CONCLUSION

TBSCC is a very aggressive tumor associated with poor prognosis. We reported a unique case coursing with cerebellar invasion and hydrocephalus despite multiple surgical resections. There was also tumor progression after radiotherapy. More studies on this topic are necessary to provide new and better treatment to these patients.

Ethical approval

The Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

REFERENCES

- Allanson BM, Low TH, Clark JR, Gupta R. Squamous cell carcinoma of the external auditory canal and temporal bone: An update. *Head Neck Pathol* 2018;12:407-18.
- Arena S, Keen M. Carcinoma of the middle ear and temporal bone. *Am J Otol* 1998;9:351-6.
- Arriaga M, Curtin H, Takahashi H, Hirsch BE, Kamerer DB.

- Staging proposal for external auditory meatus carcinoma based on preoperative clinical examination and computed tomography findings. *Ann Otol Rhinol Laryngol* 1990;99:714-21.
- Conley JJ. Cancer of the middle ear and temporal bone. *N Y State J Med* 1974;74:1575-9.
 - Gidley PW, DeMonte F. Temporal bone malignancies. *Neurosurg Clin N Am* 2013;24:97-110.
 - Kunst H, Lavielle JP, Marres H. Squamous cell carcinoma of the temporal bone: Results and management. *Otol Neurotol* 2008;29:549-52.
 - Leonetti JP, Smith PG, Kletzker GR, Izquierdo R. Invasion patterns of advanced temporal bone malignancies. *Am J Otol* 2000;122:882-6.
 - Lovin BD, Gidley PW. Squamous cell carcinoma of the temporal bone: A current review. *Laryngoscope Investig Otolaryngol* 2019;4:684-92.
 - Madsen AR, Gundgaard MG, Hoff CM, Maare C, Holmboe P, Knap M, *et al.* Cancer of the external auditory canal and middle ear in Denmark from 1992 to 2001. *Head Neck* 2008;30:1332-8.
 - Masterson L, Winder D, Marker A, Sterling JC, Sudhoff HH, Moffat DA, *et al.* Investigating the role of human papillomavirus in squamous cell carcinoma of the temporal bone. *Head Neck Oncol* 2013;5:22-9.
 - Mazzoni A, Danesi G, Zanoletti E. Primary squamous cell carcinoma of the external auditory canal: Surgical treatment and long-term outcomes. *Acta Otorhinolaryngol Ital* 2014;34:129-37.
 - McRackan TR, Fang TY, Pelosi S, Rivas A, Dietrich MS, Wanna GB, *et al.* Factors associated with recurrence of squamous cell carcinoma involving the temporal bone. *Ann Otol Rhinol Laryngol* 2014;123:235-9.
 - Moffat DA, Wagstaff SA. Squamous cell carcinoma of the temporal bone. *Curr Opin Otolaryngol Head Neck Surg* 2003;11:107-11.
 - Moody SA, Hirsch BE, Myers EN. Squamous cell carcinoma of the external auditory canal: An evaluation of a staging system. *Am J Otol* 2000;21:582-8.
 - Ngu CY, Mohd Saad MS, Tang IP. Temporal bone squamous cell carcinoma: A change in treatment. *Med J Malaysia* 2021;76:725-30.
 - Prasad S, Janecka IP. Efficacy of surgical treatment for squamous cell carcinoma of the temporal bone: A literature review. *Otolaryngol Head Neck Surg* 1994;110:270-80.
 - Yin M, Ishikawa K, Honda K, Arakawa T, Harabuchi Y, Nagabashi T, *et al.* Analysis of 95 cases of squamous cell carcinoma of the external and middle ear. *Auris Nasus Larynx* 2006;33:251-7.
 - Zanoletti E, Marioni G, Stritoni P, Lionello M, Giacomelli L, Martini A, *et al.* Temporal bone squamous cell carcinoma: Analyzing prognosis with univariate and multivariate models. *Laryngoscope* 2014;124:1192-8.

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