

# Temporal Bone Basaloid Squamous Cell Carcinoma: A Rare Case Report

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## ABSTRACT

External Auditory Canal (EAC) carcinoma is uncommon tumor of head and neck malignancies and the commonest histology is Squamous Cell Carcinoma (SCC). The variant of SCC known as Basaloid Squamous Cell Carcinoma (BSCC) is extremely rare in this anatomical site. BSCC may present along with squamous dysplasia of the surface mucosa or typical SCC. It shows advanced age group propensity and some studies shown tobacco and virus may be contributor factors. BSCC may presents with symptoms and signs mimicking other benign otologic lesions such as otitis externa and chronic suppurative otitis media. Thus, the final diagnosis is established late. Since it is rare neoplasm, not much studies reported and the gold standard of management is still controversial. Pittsburgh Staging System (PSS) was used to facilitate the best management for patient's quality of life. Survival rate is better in earlier stage. Whereas an advance stage with positive surgical margin has poorer outcome. We present a 63-year old gentleman with chronic blood stain ear discharge which the final diagnosis was BSCC. The option of treatment and literature review will be discussed.

**Keywords:** Basaloid squamous cell carcinoma, Squamous cell carcinoma, Temporal bone carcinoma, External auditory canal, Treatment, Prognosis.

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## INTRODUCTION

Temporal Bone Carcinoma (TBC) is reports as less than 0.2% of all tumours of the head and neck which infrequent and reveals one to six cases per one million people annually<sup>1</sup>. Histologically, SCC is commonly reported in temporal bone malignancy and usually affected those in the fifth and sixth decades of life<sup>2,3</sup>. Besides, SCC of EAC also four times more common than basal carcinoma<sup>2</sup>. The typical presentation is otorrhea, otalgia, hearing loss and bleeding. This presentation may lead to delay in diagnosis<sup>2,4</sup>.

BSCC is the variant of SCC and it is extremely rare in EAC. Wain et al, contriver for variant type of SCC known as BSCC in 1986 based on unusual histological finding<sup>5</sup>. BSCC is high grade neoplasm of SCC variant described as rounded nests of cytologically highly atypical, mitotically active basaloid epithelial cells with high nuclear/cytoplasmic ratios and hyperchromatic nuclei<sup>6</sup>. The differential diagnoses of BSCC is Basal Cell Carcinoma (BCC). Morphologically, BCC exhibit retraction artefacts and palisading at the periphery as well as presence of stromal mucin. Further immunohistochemistry test may aid in making the diagnosis, in which BCC will show positivity to BerEp4 while Epithelial Membrane Antigen (EMA)-positivity is seen in SCC<sup>7</sup>. As a variant of SCC, BSCC express EMA, nevertheless, it must be remembered that BerEp4 may also be positive in BSCC<sup>8</sup>.

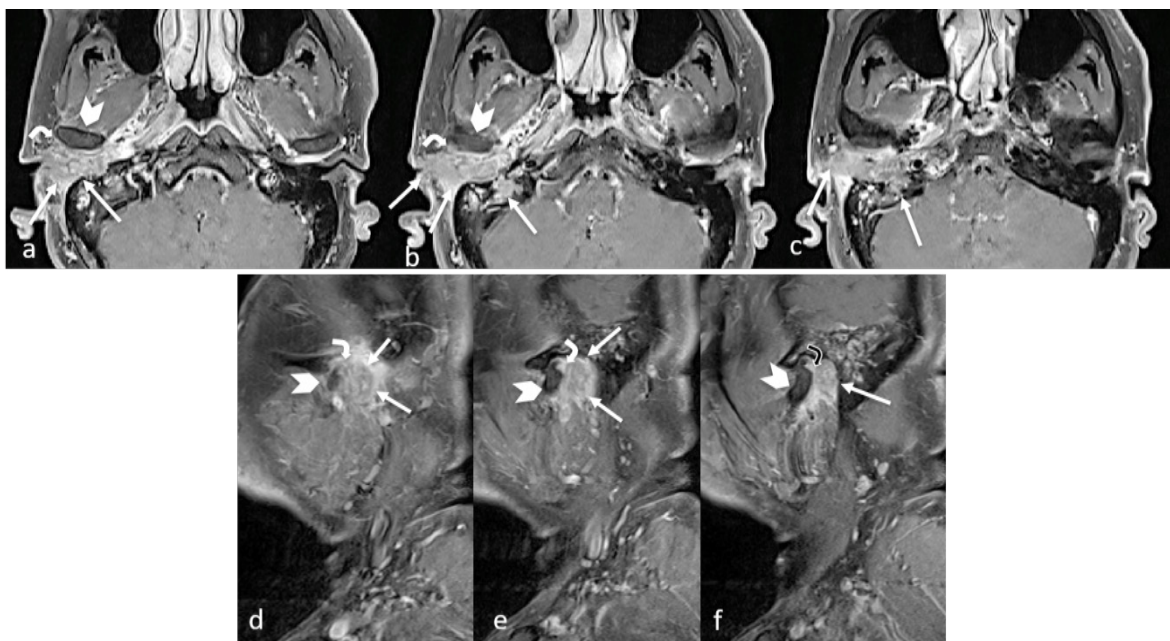
There is no standardizing staging for the TBC<sup>7</sup>. Even, American Joint Committee on Cancer (AJCC) has an inadequate staging system for primary temporal bone malignancy. Thus, Pittsburgh modified TNM staging

system was applicable in various previous studies<sup>9</sup>. The survival rates of BSCC patient related to origin of neoplasm shown statistically insignificant<sup>10</sup>. The management and outcome of BSCC remain controversial since this neoplasm is extremely rare and limited case studies published. Here we discussed the treatment option for BSCC and reviewed the literatures.

## CASE REPORTS

A 63-year-old man with underlying hypertension on treatment presented with chronic blood stain foul smelling discharge of the right ear for three months prior to admission. It was associated with otalgia and right ear reduced hearing. He also had tinnitus which he describes as buzzing sound and persistent. Otherwise, he had no symptom of facial asymmetry, vertigo or increase intracranial pressure. On examination, his facial was nerve intact. Otoscopic examination revealed fungating mass at right external auditory canal with bloodstain. Hearing assessment performed noted right ear profound hearing loss while left side normal at lower frequency and down sloping sensorineural hearing loss at high frequency likely presbycusis. The previous biopsy of ear canal mass from other private medical centre showed severe dysplasia.

Magnetic Resonance Imaging (MRI) neck performed and noted right external auditory canal mass with mastoid and right Temporomandibular Joint (TMJ) extension as well as suspicious involvement of deep lobe of the right parotid gland. Bilateral lymph nodes noted less than 1cm size at bilateral level Ib, II, IV and IV and the largest left level IV measuring 0.7cm in short axis diameter (Figure 1). He underwent right modified lateral temporal bone resection



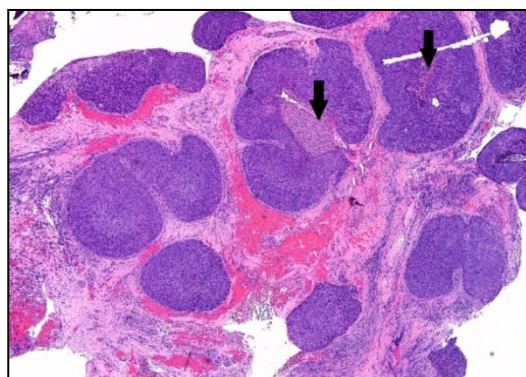
**Figure 1:** MRI T1 fat saturation post contrast in axial from inferior to superior (a, b, c) and in sagittal from lateral to medial (d, e, f). The mass occupies the external auditory canal extending to the middle ear cavity (white arrow). Anteriorly, extension into the right Temporomandibular Joint (TMJ) is suspected (curve block arrow). The tumor extension is limited only to the lateral part of the TMJ as medially the tumor margin is clearly seen to be separated from the TMJ (curve black arrow in (f)). The mandibular condyle shows normal enhancement without any cortical break suggesting non-involvement of the condyle (arrowhead).

for right temporal bone carcinoma. Intraoperatively, incision made three finger breath from right ear posterior sulcus, skin incised until muscle and temporalis fascia harvested, mastoid air cells drilled exposing the sigmoid sinus posteriorly, anteriorly until the posterior wall of EAC and zygomatic root. Inferiorly until mastoid tip and superiorly until tegmen. Korner's flap raised and antrum identified. Operative finding noted right ear fungating mass at anterior cartilaginous EAC with contact bleeding and occupied the EAC. Thus, unable to visualize tympanic membrane.

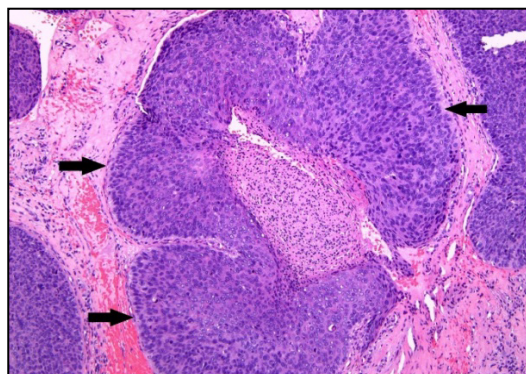
Tumour identified within entire mastoid air cells which occupying antrum, epitympanum, mesotympanum (wrapping around ossicles), hypotympanum and eustachian tube. Posterior wall, floor and anterior EAC eroded. Annulus and tympanic membrane also affected

by tumour. Malleus intact however long process of incus eroded. Right lateral semicircular canal identified with overlying tumour however no erosion seen. Facial nerve was anatomically and physiologically identified at tympanic and mastoid segments. The tumour including cartilaginous EAC, tympanic membrane, malleus and incus removed. Posterior wall of EAC was brought down. Eustachian tube plugged with surgicel and lined with bone wax.

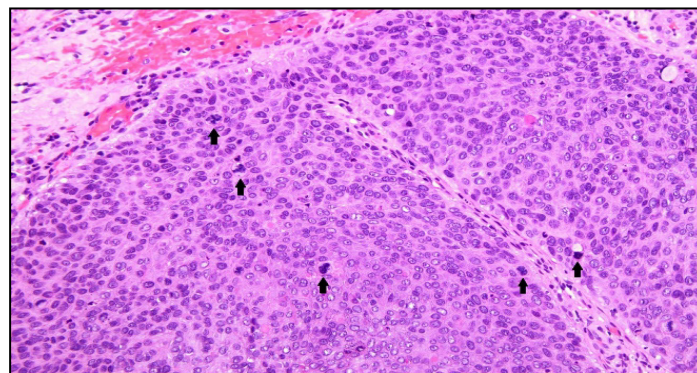
Histopathology Examination (HPE) was reported BSCC in the background of high-grade squamous dysplasia (Figure 2-5). The right tympanic membrane and floor of EAC showed high grade squamous dysplasia with areas suspicious for invasion. Otherwise, tissues taken from right sinodural angle and right retro labyrinthine air cells are negative for malignancy. He had an uneventful



**Figure 2:** The tumour is composed of malignant basaloid cells arranged in nests with central comedo-necrosis (arrow). (H&E, x40).

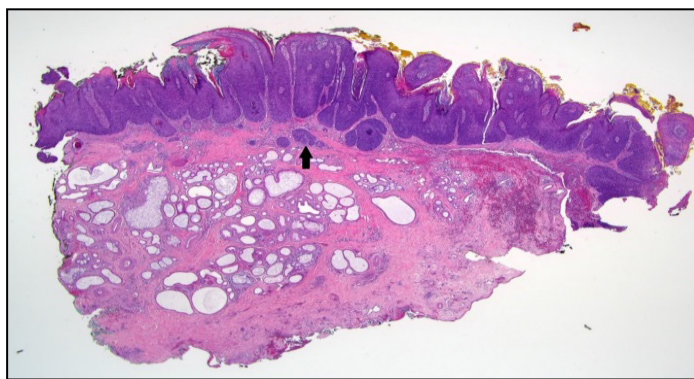


**Figure 3:** Higher magnification view highlights peripheral palisading of the tumour cells (arrow). (H&E, x100).



**Figure 4:** Malignant basaloid cells displaying round to oval hyperchromatic nuclei with inconspicuous nucleoli. Abnormal mitoses are brisk (arrow). (H&E, x200).





**Figure 5:** Surface epithelium showing high grade dysplasia with foci of invasion arranged in basaloid nests (arrow). (H&E, x12.5).

recovery and was discharge after completed 5 days of intravenous ceftriaxone 1 gm daily. There was no facial nerve palsy and any cerebrospinal fluid leak rhinorrhoea.

Based on Pittsburgh modified TNM staging system, this case is stage IV (T4N0M0).

## DISCUSSION

BSCC is composed of basaloid and squamous components, forming nests (Figure 5), islands and lobular pattern. Dysplastic changes are seen in adjacent intact epithelium<sup>6</sup>. The basaloid cells display variable pleomorphic round to oval hyperchromatic nuclei with occasional conspicuous nucleoli (Figure 2 & Figure 4)<sup>10</sup>. Features of nuclear palisading (Figure 3), comedonecrosis (Figure 2), mucin production, high mitotic activity and surrounding desmoplasia can be identified<sup>2,6</sup>. Wierzbicka et al conducted study based on data of 40 million population, primary cancer of temporal bone carcinoma detected predominant in EAC or middle ear which contribute about 76.4% of total patient post-surgery for temporal bone malignancy<sup>4</sup>.

The role of imaging is primarily for an accurate description of the anatomic spread of tumor as well as the structures involved since it is crucial in the surgical planning. All skull imaging for temporal bone malignancy ideally should have thin section in two different planes imaging with Computed Tomography (CT) and MRI<sup>11</sup>. CT is superior in the delineating bone whilst MRI is superior to visualize the soft tissue and bone marrow involvement. Post gadolinium sequence allows solid tumor delineation and extension<sup>11</sup>. In our case, MRI neck revealed the extension of tumor to adjacent structures and positive radiological lymph nodes noted.

He underwent right modify lateral temporal bone resection after considering the anatomical involvement of neoplasm. BSCC known as poor prognosis, some study recommended CT scan thorax as lung is favourable site for BSCC metastasis<sup>10</sup>. Winzenburg et al supported the evident of BSCC in previously treated at early stage with no local recurrence subsequently passed away due to lung metastasis<sup>12</sup>. Therefore, we have to schedule patient for close follow up and high index of suspicious to look for insidious sign of lung metastasis.

Pittsburgh staging universally applied in different centres to standardize the studies of temporal bone SCC besides to evaluate treatment and survival outcome<sup>13</sup>. Most of published literature supported that surgery should be considered as first line management after considering patient's outcome and staging status<sup>13-16</sup>.

Globally, few types of resections were introduced such as Lateral Temporal Bone Resection (LTBR) or also known as lateral petrosectomy, Subtotal Temporal Bone Resection (STBR) or subtotal petrosectomy and Total Temporal Bone Resection (TTBR) or total petrosectomy<sup>4,17</sup>. These surgery types can be performed either via en bloc or piecemeal method. The aim of surgery is to achieve complete resection with the maximize tumour free margin<sup>17</sup>. BSCC with no evidence of metastasis and operable case should be treated primarily with salvage surgery, subsequent reconstruction<sup>15</sup>. The distant free flaps if indicated for reconstruction and the locoregional control are same for SCC and BSCC<sup>15</sup>. Other operative procedures need to be considered are parotidectomy and neck dissection based on extension of disease invasion<sup>17</sup>.

Primary tumour removal with negative tumour margin is the key of patient survival factor<sup>18</sup>. Arriaga et al reported increase survival rate following partial or STBR with negative margin as 79%<sup>9</sup>. En bloc LTBR, elective parotidectomy and level II lymph node dissection for tumour T1 and T2, while for T3 tumour treated with piecemeal STBR, parotidectomy and level II and III neck dissection<sup>17</sup>. TTBR is treatment of choice when involving petrous apex in T4 tumour. Any tumours invading nearby structures will be considered mandibulectomies, zygoma resections or dura resections as indicated<sup>17</sup>.

Medina et al reported tumour less than T2 with no node involvement will be treated with LTBR while those with advanced stage will be considered either LTBR or an extended temporal bone resection may be total or subtotal<sup>19</sup>. Bacciu et al did en bloc complete resection with removal of suspicious soft tissue margin in tumours T1, T2 and selected T3 cases such as tumour confined to EAC. Lobo et al did parotidectomy in cases of parotid node metastasis, parotid gland involvement either clinically or radiologically or when anterior wall of EAC affected by tumour subsequently the temporalis is used

muscle flap to cover the wound. They also proceed with neck dissection in those clinically had neck finding. The most common muscle flap used for reconstruction is temporalis flap however not exclude other options such as skin grafts, parascapular free flap and sternocleidomastoid flap<sup>2</sup>.

Those BSCC with higher-grade tumour preferably proceed with lymph node dissection<sup>20</sup>. However in our case study, we are not proceeding with neck dissection but he underwent chemoradiotherapy post operatively. Complete removal of tumour with free margin and subsequently completed with postoperative radiotherapy was recommended<sup>2</sup>. Thus, result in survival rates of 5-years between 40% and 70% then drastically down 20% in advanced stages<sup>2</sup>. However, another study proved that preoperative Chemoradiotherapy (CRT) would result better outcome to achieved negative margin at the operation<sup>16</sup>. Combination of complete en bloc operation with tumour-free surgical margin and preoperative CRT increase the 5-year estimated survival for T3 and T4 patient to 80% and 40% respectively compare to previously reported as 20% in T4 patients<sup>16</sup>.

In certain circumferences, the surgeon will change the procedure from STBR to LTBR in considering patient's quality of life in advanced disease. Thus, the risk for positive margin is high<sup>16</sup>. Patient will benefit better outcome in considering CRT prior operation however in metastasis case it is insignificant. In order to achieve clear margin intraoperatively, frozen section was introduced<sup>9</sup>. In our case, the tumours were removed and facial nerve was preserved as it was not affected by tumour.

Few factors will contribute to poor prognosis such as distant metastasis, positive surgical margins, involvement of dura, facial or cranial nerves or first visit with excruciating pain<sup>21</sup>. In some cases, it will be considered inoperable when the neoplasm invading the carotid or with evident middle or posterior fossa involvement<sup>2</sup>.

Comparison between BSCC with neck pathology and poorly differentiated squamous cell carcinoma with neck pathology revealed overall survival are 16 months and 32 months respectively<sup>12</sup>. Combination of cetuximab and radiotherapy will prolong duration and survival in locoregional disease control of advanced head and neck malignancies<sup>22</sup>. Few authors agreed that post-operative adjuvant CRT will decrease loco-regional recurrence of head and neck SCC indirectly increase patient survival rates in any tumour stages or poorly histopathological entity<sup>23,24</sup>. Cooper et al considered those underwent simultaneous postoperative combined therapy (chemotherapy and radiotherapy) shown better in local and regional rate control of two years compare to radiotherapy alone, however as general survivor rate still poor<sup>25</sup>. Postoperative radiation patient with positive margin reported as low survival rate<sup>9</sup>. Thariat et al reported that BSCC patient has high regional control rate compare to other SCC subtypes.

The different anatomical origins of BSCC shown statistically insignificant difference compare to survival rate of patients<sup>10</sup>. Thus, we can conclude that the survival rate of temporal

bone BSCC patient may result similar with others BSCC origin. Yet, the nature and prognosis of head and neck BSCC still controversial compare to primary SCC due to limited studies and small retrospective series<sup>10</sup>. In this case study, modified LTBR performed, preserving facial nerve and patient underwent postoperative chemoradiotherapy.

## CONCLUSION

Temporal bone carcinoma frequently presented at an advanced stage due to late diagnosis. BSCC about five percent of SCC which is aggressive, mostly presented a high stage, favourable to lymph node and systemic metastases especially lung. High index of suspicious by attending physician might help to diagnose malignancy earlier. There is no well-established definitive treatment due to limited retrospective studies and rarity of neoplasm. Surgery is the best option of treatment for BSCC and chemotherapy or radiotherapy will be performed as indicated. In the bright side, BSCC is good locoregional control and sensitive to chemo-radiotherapy treatment.

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