CASE SERIES

Temporal bone squamous cell carcinoma: A change in treatment

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SUMMARY

Temporal bone squamous cell carcinoma (TBSCC) is a rare head and neck malignancy with the incidence 0.8 –1.0 cases in 1 million population. We are reporting a case series on the TBSCC cases that were operated on at Sarawak General Hospital, Malaysia. Ten patients were identified and collected with the presentation and type of surgery performed. It has been challenging for us to manage with recorded 2 years surviving in 6 out of 10 patients operated within this period. An adequate management with proper surgical resection of tumour and radiotherapy can extend the life expectancy for TBSCC patients.

KEYWORDS:
Temporal bone, Squamous cell Carcinoma, Resection

INTRODUCTION

Malignant neoplasm of temporal bone is a rare disease with an incidence of 0.8 – 1.0 cases per million population per year.[1,2] Squamous cell carcinoma is one of the most common reported variants. Nonetheless, it only constitutes 0.2% of the total head and neck malignancy.[3] Other variants of this malignancy that have been reported include basal cell carcinoma, adenoid cystic, melanoma chondrosarcoma, Ewing's tumour and fibroxanthoma.[2] Temporal bone squamous cell carcinoma (TBSCC) has always been described for its aggressive behaviour with invasion to the surrounding structure via bony canals or intraosseous vessels.[4]

To date, to our best knowledge, there is no consensus on the management of this rare disease. There is still an ongoing debate with regards to the preferred staging method that should be used for TBSCC. In particular, Sarawak General Hospital (SGH), Malaysia is a tertiary healthcare centre in the state of Sarawak. SGH receives all the complicated surgery cases from all over the state. We aimed to review the outcome of the temporal bone surgeries in treating TBSCC in SGH, Malaysia.

MATERIALS AND METHODS

We retrospectively reviewed and audited 10 patients who were diagnosed with TBSCC from the years 2014 to 2019. Data was traced from the SGH record unit. All the patients were operated by the same surgeon. All patients went through regular Ear Nose Throat (ENT) examination. Patients who were confirmed with squamous cell carcinoma from the histopathological examination (HPE) of the external auditory mass were arranged for computed topography (CT) imaging of temporal bone as well as whole body staging. Modified Pittsburgh system was used to stage our patients.5 Operation was planned and introduced to all patients who were confirmed to have TBSCC from the biopsy of their external auditory canal tissue. During the review, we excluded those patients who were diagnosed with TBSCC but not fit for surgery after anaesthetist review, as well as patients who did not agree for any intervention. Surgical resection was deployed as the treatment for all the patients, followed by radiotherapy with or without chemotherapy. LTBR was performed on patients with T1 and T2 lesions, whereas STBR was performed in T3 and T4 lesions (Figure 1A).

RESULTS

The median age for this case series was aged 50 years with a male predominance. Of note, the youngest patient in our case series was only 13 years old. More than half of our patients presented with otorrhoea (n=7) and otalgia (n=6). Three patients reported hearing loss or reduced hearing, and external auditory canal mass was seen in two patients. Only one of our patients presented with facial nerve palsy. Most of our patients presented at stage III of the disease. One patient had a history of nasopharyngeal carcinoma with head and neck radiotherapy performed. Two patients had history of chronic otitis media for more than six months. Details of our patients is shown in Table I.

All the patients had lateral and subtotal temporal bone resection done as stated in Table I. A modified radical neck dissection was planned for patients who were noted to have cervical lymph node involvement in CT scan. Six of 10 patients had neck dissection performed, and one patient had HPE confirmed neck involvement. One of our patients did not go for radiotherapy as he had recently received radiotherapy for concurrent nasopharyngeal carcinoma.

DISCUSSION

Primary squamous cell carcinoma (SCC) of the temporal bone is an uncommon head and neck malignancy with aggressive behaviour. It was first described by Schwartze and

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Table I: Patients' data

No.	Age	Gender	Race	Comorbidities	Duration of presentation	Presentation	Side	Staging	HPE	Surgery	Parotid involvement from HPE	Radiotherapy	Status
1	13	M	Iban	Resolved otitis media	1 week	Otalgia	Right	T1N0M0	Moderately differentiated squamous cell carcinoma	Right lateral TBR, right Selective neck dissection	No	Yes	No recurrence for 2 years
2	46	M	Chinese	Diabetes mellitus, hypertension, end stage renal failure	1 month	Otalgia, otorrhea	Right	T1N0M0	Squamous cell carcinoma	Right lateral TBR	No	Yes	Death due to other disease after 1 year, i.e., Pneumonia
3	45	M	Iban	Nasopharyngeal carcinoma treated in year 2009	1 month	Otalgia, otorrhea, hearing loss	Right	T2N0M0	Moderately differentiated squamous cell carcinoma	Right lateral TBR	No	No	No recurrence for 5 years
4	53	M	Chinese	No comorbid	NA	Hearing loss	Left	T3N0M0	Invasive, moderately differentiated squamous cell carcinoma	Left subtotal TBR	No	Yes	No recurrence for 5 years
5	48	M	Bidayuh	Chronic otitis media for 6 years	3 months	Otorrhea, otalgia	Right	T3N0M0	Poorly differentiated squamous cell carcinoma	Right subtotal TBR	Yes	Yes	No recurrence for 3 years
6	68	F	Iban	No comorbid	6 months	Otalgia, otorrhea, facial weakness, vertigo	Left	T3N0M0	Well differentiated squamous cell carcinoma	Subtotal TBR, left parotidectomy, left MRND	No	Yes	Death due to disease
7	58	M	Chinese	No comorbid	2 months	Ear mass, otorrhea, postauricular fungating ulcer	Left	T3N0M0	Well differentiated squamous cell carcinoma	Right subtotal TBR, right total parotidectomy, right MRND	No	Yes	Death due to disease
8	62	M	Iban	No comorbid	3 months	Otorrhoea	Right	T3N0M0	Moderately differentiated squamous cell carcinoma	Right subtotal TBR, right parotidectomy and right MRND	No	Yes	No recurrence for 3 years
9	48	M	Malay	Diabetes mellitus, dyslipidaemia	10 years	Ear mass, bleeding, otalgia	Left	T3N0M0	Well-differentiated squamous cell carcinoma	Left subtotal TBR, left total parotidectomy, left MRND and pectoralis major free flap	Yes	Yes	No recurrence for 2 years
10	52	F	Iban	Hypertension	4 months	Ear mass, otorrhea, reduced hearing	Left	T4N1M0	Well differentiated SCC	Left subtotal TPR, total parotidectomy, left MRND	Yes	Yes	Death due to disease after 2 years

N.B.: MRND: Modified radical neck dissection, SCC: squamous cell carcinoma, TBR: Temporal bone, resection, SCM: sternocleidomastoid, NA: not available

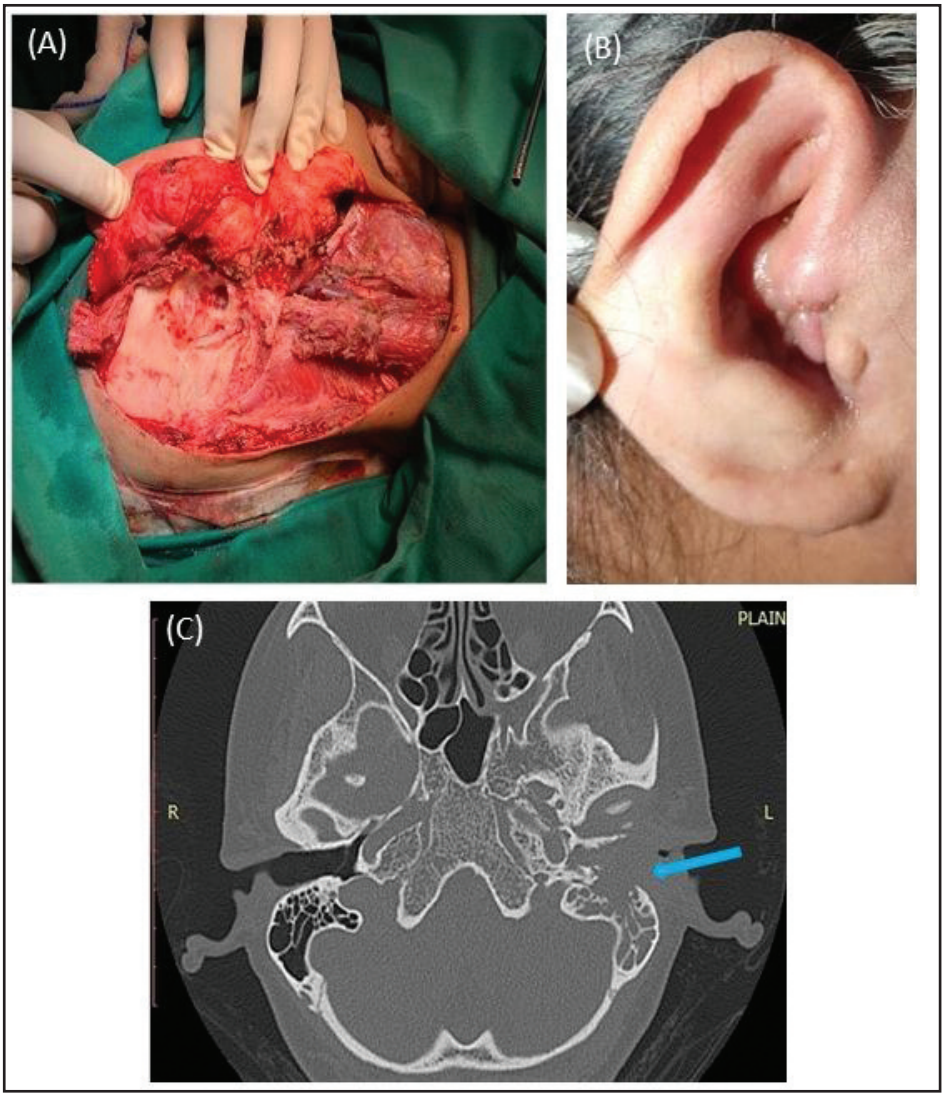

Fig. 1: Images of patients with temporal bone squamous cell carcinoma. (A) Intra-operative image of right subtotal temporal bone resection (B) External auditory mass (C) Computed tomography imaging at the base of skull in bone window, showing a mass in the external auditory canal of the left ear (blue arrow) that is invading into the left mastoid air cells.

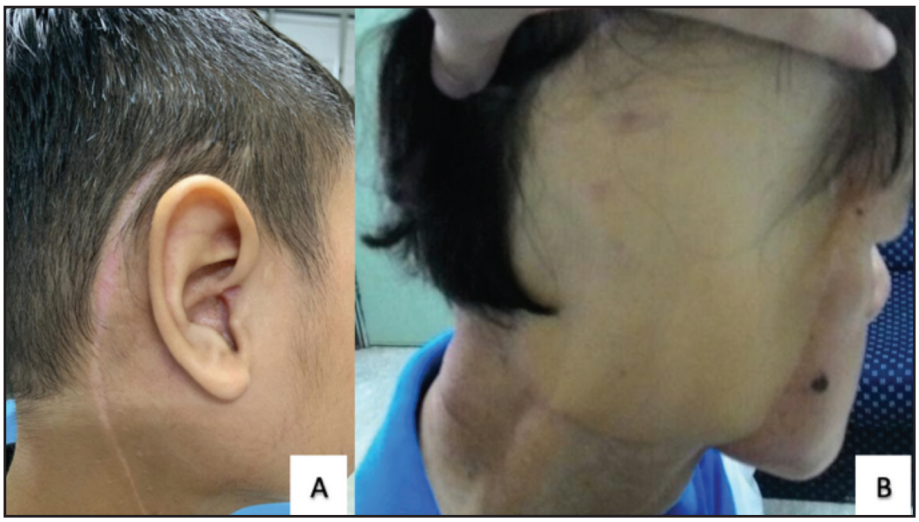

Fig. 2: Post-operative wound with (A) primary closure and (B) reconstruction with pectoralis major free flap.

Wide in 1775, but the first publication about TBSCC was only presented by Newhart in his case series in 1917.6 Due to the rarity of this condition, it remains a challenge to clinicians in diagnosing and managing the TBSCC. The incidence of TBSCC was reported to peak in the 6th decade of life with male predominance (male: female ratio of 1.7:1).[1,7] However, in our case series, the youngest patient was 13 years old and this raises the question of whether advanced age is truly a contributing risk factor for this rare malignancy.

The exact aetiology of TBSCC remains unknown. Whilst the use of alcohol and tobacco have always been related with SCC in head and neck, this cannot be applied to TBSCC.[8] More than half of our patients in this case series presented to us with otorrhoea and otalgia, with duration ranging from 1 months to as long as 6 months. One of them was treated as chronic otitis media for 6 years, and recently presented with bleeding from the ear with scanty foul-smelling discharge for 3 months with high suspicion of malignancy. Modified mastoidectomy was performed and revealed inflammatory granulation. Our findings correlate with previous studies, which show that chronic inflammatory process and chronic otorrhoea are risk factors for TBSCC.[9] It is postulated that chronic inflammation undergoes malignancy transformation when recovery fails.[10] Other symptoms such as facial nerve palsy, external auditory mass (Figure 1B), hearing loss with tinnitus are also found to be associated with TBSCC. History of radiation has been proposed as one of the causes of TBSCC.[11] Lending support to this, one patient in our case series had a history of nasopharyngeal carcinoma and completed radiotherapy was found to have TBSCC. Interestingly, human papillomavirus (HPV) exposure with detection of high-risk genotype HPV 16 and HPV 18 in chronic otitis media and cholesteatoma gave a hypothesis of relation between HPV with TBSCC.[12] The above mentioned presentations, however, are insidious presentations of temporal bone SCC that cause delays in seeking treatment.[13] The spread of TBSCC to cervical lymph nodes have also been reported. The pathway through which TBSCC spread is through the tegmen tympani, fissure of Santorini and foramen of Huschke into the middle cranial fossa or infratemporal fossa, thus causing headache, parotid swelling and ulceration in the surrounding skin.[1]

Imaging investigation helps clinician to evaluate extension of malignancy. High resolution computed tomography (HRCT) scan of the temporal bone provides useful information of the tumour and the surrounding bony erosion (Figure 1C).[13] However CT scan was reported to have limited role in identifying soft tissue involvement.[14,15] Magnetic resonance imaging (MRI) overcomes this limitation by giving information on perineural, parotid gland and any regional lymph node invasion as well as vascular and dural involvement.[15] Therefore, both HRCT and MRI are required for staging the disease. Biopsy is mandatory in order to establish the diagnosis of TBSCC.16 Additionally, imaging enables clinicians to precisely locate and obtain tissue biopsy for histopathological examination. The presence of inflammation and oedema may hinder the success of an accurate biopsy; thus, a deep tissue biopsy is often required to yield a positive result.[16] Further investigation with arterial or venous angiography can be arranged when there is vascular invasion seen on MRI. Imaging is also useful for disease staging and to guide the management or treatment planning for patients with TBSCC. Unfortunately, in SGH, MRI is usually approved for cases suspicious of neurological involvement only. CT scan is, thus far, sufficient in our setting to confirm the origin as well as the location of the tumour. Ninety percent of our operated patients were at disease stage ≤T3.

To date, the Modified Pittsburgh Staging system is widely used in staging TBSCC. American Joint Committee of Cancer (AJCC) staging for head and neck malignancies was used before the Pittsburgh system was introduced by Arriaga et al. in 1990 with accuracy of 98%.[14,17] Our centre is mainly operating on patient with tumour staging of T3 and below. Patient with stage T4 malignancies was reported to have 5 years survival rate < 30%.[7,18] The only stage 4 patient that had operation performed was noted to have T3 preoperative and patient succumbed within 1 year postoperatively.

Surgery is the mainstay treatment for TBSCC.[1] The primary consideration that a surgeon has to bear in mind is when and what kind of resection is adequate in treating the patient. In the current era of oncology surgery, a proper multidisciplinary discussion is required for patients before surgery, which involve otorhinolaryngology, oncology and plastic and reconstructive surgery. This helps in the designing and explaining the method of surgery and planned post-operative management for the understanding of patients towards their disease. The diminished hearing observed postoperative as well as the swallowing rehabilitation need to be explained to the patients, as this might give a huge impact to the quality of life of patients after the treatment.

The principle of *en bloc* resection is to maximise the negative surgical margin, at the same time preserving the function of unaffected structures.[4,10] Unlike cutaneous SCC, a definite margin of tumour could not be clearly resected during the burring.[19] Intraoperative frozen section maximise the benefit in assurance of negative margin during single operative setting. Depending on the anatomical extension of malignancy, options for temporal bone resection (TBR) that have been introduced, which include lateral temporal bone resection (LTBR), subtotal temporal bone resection (STBR) (Figure 1A). and total temporal bone resection (TTBR).[1] Wound closure post resection should also be included as part of the surgical planning. Flap reconstruction will be necessary if the resected wound is not able to close primarily (Figure 2). Patient should not only be counselled on the complications of wound breakdown and disease recurrence postoperatively, but also the cosmetic outcome post-surgery.[20]

The decision of parotidectomy during TBR is remains inconclusive.[21] Due to the vicinity of parotid gland and temporal bone, direct spread of this malignancy is of high index of suspicion. Although the percentage is reported < 50% in T1 and T2 lesions, it is advised not to take the risk to exclude parotidectomy.[18,21] Suggestions have been given to perform superficial parotidectomy in patients with T1 and T2 lesions, and total parotidectomy in T3 and T4 lesions during an extended TBR.[10,21] In our retrospective review, 6 of patients with T3 had total parotidectomy performed in which 4 had

positive malignancy seen (67%). We performed biopsy of parotid tissue for T1 and T2 lesions, with no parotid involvement. However, if CT imaging and intraoperative revealed parotid involvement, parotidectomy is performed intraoperatively. These patients also had neck dissection done under the same setting, and it was noted that none has positive lymph node involvement. There was only one patient who had lymph node involvement in advanced (T4) temporal bone SCC. As most patient might have problem in travelling to SGH, we had performed functional neck dissection under same general anaesthesia setting in our patients based on presence of enlarged cervical lymph node in both the CT and MRI scan, to prevent patient go for a second surgery.

Postoperative prognosis is said to be good in patient with early stage (T1 and T2). Most review show drastically higher mortality rate for T3 and T4. The 5-year survival is 100% in T1 and T2, but 69% in T3 and only 20% in T4.[10,18] Lymph node involvement is also an indicator of poor prognosis and higher recurrence rate.[8] In our series, adjuvant radiotherapy played an important role in the management as a clear margin is hard to identified during burring. Six out of 8 patients who went through radiotherapy showed no recurrence for at least 2 years. There was one patient with a T3 lesion who defaulted the planned radiotherapy and succumbed 6 months later. To date, there is no definite conclusion regarding the effectiveness of postoperative adjuvant radiotherapy in temporal bone SCC. Our patients, however, were further discussed with the oncology team post-operative and radiotherapy was given to most of our patients, as our HPE could not provide clear information on tumour free margin.

In SGH, surgical management was planned to patient with T1 – T3. As mentioned earlier, logistic issue is also a problem in Sarawak because the patients come from remote areas and often present to us at an advanced stage of disease. Hence, it becomes challenging to perform both advanced resection and regular radiotherapy in managing our patients. Nevertheless, the survival rate at >2 years in postoperative temporal bone SCC at SGH is 70%. Patients need to be briefed regarding the indication, complications, as well as the compliance with clinic review so that recurrence can be detected earlier. The aetiology of human papilloma virus should also be studied further in the future.

CONCLUSION

This case series does not represent a standard management in treating temporal bone squamous cell. However, it is aimed at sharing our surgical experience in treating this malignancy. Patients with severe painful non-traumatic, bleeding ear should always be alert to the possibility of malignancy, and an earlier intervention with well-designed resection could provide a better prognosis for the patient. Despite limited resources, the properly planned resection of tumour along with radiotherapy can help to extend the life expectancy of these patients. The challenging part in managing TBSCC is the experience of the surgeon in identifying a normal and tumour infiltrated margin intraoperatively for adequate resection. The compliance of the patients with the treatment and follow up is needed for regular monitor and keeping them disease-free in future.

CONFLICT OF INTEREST

None to declare.

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