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. 4 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 Schwartz and
Table I: Patients’ data

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

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.
temporal bone SCC that cause delays in seeking treatment. More than half of our patients in this case series presented to us with otorrhoea and otalgia, with duration ranging from 1 month 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. It is postulated that chronic inflammation undergoes malignancy transformation when recovery fails. Other symptoms such as facial nerve palsy, external auditory mass (Figure 1B), chronic inflammation and oedema may hinder the success of an accurate biopsy; thus, a deep tissue biopsy is often required to yield a positive result. 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.

Surgery is the mainstay treatment for TBSCC. 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 involves otorhinolaryngology, oncology and plastic and reconstructive surgery. This helps in the designing and explaining the method of surgery and planned postoperative management for the understanding of patients towards their disease. The diminished hearing observed postoperatively 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. Unlike cutaneous SCC, a definite margin of tumour could not be clearly resected during the burring. 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). 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.

The decision of parotidectomy during TBR is remains inconclusive. 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. 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. 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 T4.14 Lymph node involvement is also an indicator of poor prognosis and higher recurrence rate.4 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.

**REFERENCES**

Case Series