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CASE REPORT

Case Series: A 5-year-experience ofTranslabyrinthine Approach in Managing Cerebellopontine Angle Tumour in Ministry of Health Malaysia

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ABSTRAK

'Vestibular schwannomas' (VS) juga dikenali sebagai neuromas akustik, adalah tumor jinak pada cabang vestibular saraf kranial kelapan dan berasal daripada sel Schwann yang melapisi cawangan vestibular. Tumor ini menyumbang kepada 6-10% daripada semua tumor intrakranial dan 60-90% daripada tumor sudut 'cerebellopontine' (CPA). Artikel ini menerangkan pengalaman kami dalam mengendalikan tumor CPA melalui pendekatan 'translabyrinthine' dalam konteks Kementerian Kesihatan Malaysia, dengan penglibatan pasukan pelbagai disiplin. Kami membincangkan demografi, persembahan klinikal biasa, histopatologi tumor CPA, penginapan di hospital dan komplikasi. Terdapat 26 pembedahan telah dijalankan dari tahun 2018 hingga 2022, di mana seramai 18 (69.2%) pesakit adalah wanita, dan 8 (30.8%) adalah lelaki. Purata umur pesakit ialah 49.35 ± 13.39. Daripada 26 pesakit, 19 (69.2%) mempunyai fungsi saraf muka selepas pembedahan yang normal. Walau bagaimanapun, seramai 7 pesakit (30.8%) mengalami kelumpuhan saraf muka kekal dari House-Brackmann gred II hingga gred IV. Data kami menunjukkan bahawa empat (15.4%) pesakit mengalami kebocoran cecarir serebrospinal. Tempoh penginapan hospital adalah antara 5 ke 51 hari, dengan median 8.5 dan julat antara kuartil (IQR) 5.75. Pembedahan neuroma akustik melalui pendekatan 'translabyrinthine' ialah rawatan yang selamat dan berkesan. Konsep pembedahan saraf/ENT antara disiplin disarankan kerana setiap langkah operasi memerlukan kepakaran yang khusus.

Kata kunci: Interdisiplin; 'schwannoma vestibular'; 'translabyrinthine'

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ABSTRACT

Vestibular schwannomas (VS) are benign tumours of vestibular branch of vestibulocochlear nerve, originated from the glial cells that form the myelin sheaths. The tumours account for 6% to 10% of all intracranial tumours and 60% to 90% of cerebellopontine angle (CPA) tumours. This article described our experience in managing CPA tumour via *translabyrinthine* approach in the setting of Ministry of Health in Malaysia, with multidisciplinary team involvement. We discussed the demographics, common clinical presentation, histopathology of the CPA tumour, hospital stays and complications. There were 26 surgeries performed from 2018 until 2022, whereby a total of 18 (69.2%) patients were female, and 8(30.8%) were male. The mean age of the patients was 49.35 $\pm$ 13.39. From a total of 26 patients, 19 (69.2%) had a normal post-operative facial nerve function. However, a total of 7 patients (30.8%) suffered from permanent facial nerve palsy ranging from *House-Brackmann* grade II to grade IV. Our data revealed that four (15.4%) patients sustained cerebrospinal leak. The total duration of hospital stays ranged from 5 days to 51 days with median of 8.5 and interquartile range (IQR) of 5.75. Acoustic neuroma surgery via *translabyrinthine* approach is a safe and compelling treatment. The interdisciplinary concept is recommended as each step of the operation requires specialist expertise.

Keywords: Interdisciplinary; translabyrinthine; vestibular schwannoma

INTRODUCTION

Vestibular schwannomas (VS) are benign tumours of vestibular branch of vestibulocochlear nerve, originated from the glial cells that form the myelin sheaths (Swartz 2004; Thapa et al. 2019). The tumours account for 6-10% of all intracranial tumours and 60-90% of cerebellopontine angle (CPA) tumours (Swartz 2004). It is slow growing but can cause local erosion of the internal auditory canal (IAC) and compression of the proximal nerves commonly 5th and 7th cranial nerves (Swartz 2004; Thapa et al. 2019). The incidence of the tumor is 1 in 100,000, and it usually affects individuals between their 30s and 50s (Rosahl et al. 2017). The aim of managing patients with VS is to improve the quality of life (Rosahl et al. 2017). The factors influencing the management of VS includes patients' age, surgical morbidity of different approaches, tumour size and hearing level preoperatively with the aim or goal of surgery to excise the tumour with a low morbidity and preserving facial nerve function (Zanoletti et al. 2016). In a study by Tonn (2000), the author found the interdisciplinary Neurosurgery/ Ear, Nose and Throat team concept deemed necessary as each step of the operation is performed with their respective expertise. The core of managing VS is variable, thus makes it hard to create a standard treatment pathway for this disease. Therefore, it is more important to establish a continuous interdisciplinary meeting than to establish a common standard (Rosahl et al. 2017). Previous study mentioned three established general

approaches, which are middle cranial fossa approach, *translabyrinthine* approach and retrosigmoid approach. All the approaches possessed their own indication and specific risk and complications (Rosahl et al. 2017). In the following, this article described our experience in managing CPA tumour via *translabyrinthine* approach in the setting of Ministry of Health in Malaysia, with multidisciplinary team involvement. We discussed the demographics, common clinical presentation, histopathology of the CPA tumour, duration of intensive care unit (ICU) and hospital stays and complications. This paper is very important because it is first to describe the Ministry of Health's experience treating CPA tumours.

MATERIALS AND METHODS

Data collected from Hospital Tuanku Jaafar (HTJ), Hospital Sultan Ismail (HSI) and Hospital Sultanah Bahiyyah (HSB). All patients who had been diagnosed with CPA tumour between from March 2018 until December 2022 recruited and undergone *translabyrinthine* approach of surgery and were managed by multidisciplinary team of healthcare professionals that includes Otorhinolaryngologists, Neurosurgeons, Audiologist, Rehabilitation specialist and Psychiatrist. A total of 26 surgeries were performed. *Translabyrinthine* approach is defined by approach to CPA by dissection of the internal auditory meatus, mastoidectomy as well as labyrinthectomy (Tonn 2000). Those selected for this approach have non-functional hearing based on the guideline by American Academy of Otolaryngology – Head and Neck Surgery (AAO-HNO), which was divided into 4 classes of hearing ability (Rosahl et al. 2017). Non-functional or non-serviceable hearing is defined by hearing threshold >50 dB and word recognition score 50% (Rosahl et al. 2017). The dimension of the tumour is graded according to the New and Modified Reporting Systems from the Consensus Meeting on Systems for Reporting Results in Vestibular Schwannoma. It is graded into five grades (Visagan et al. 2018). The grades are grade 1 (size 1-10 mm), grade 2 (size 11-20 mm), grade 3 (size 21-30 mm), grade 4 (size 31-40 mm), grade 5 (size >40 mm). Facial nerve function assessed using the House-Brackmann grading system, based on a six-point grading system, where grade 1 represents normal facial function and grade 6 represents complete paralysis based on gross appearance and motion (Thapa et al. 2019).

The analysis was done using Statistical Package for the Social Science (SPSS) version 27 and Windows Excel Windows 11. Data were presented as median (interquartile range),if the data was not normally distributed (Shapiro-Wilk test of normality $p < 0.05$) and as mean with standard deviation if the data was normally distributed (Shapiro-Wilk test of normality $p > 0.05$).

RESULTS

Pre-operative Data

A total of 61.5% of the tumour arise from the right CPA and 38.5% from the left side. The mean age of the patients was 49.35 $\pm$ 13.39. (Table 1).

There were 26 surgeries performed; a total of 18 (69.2%) patients were female, and 8(30.8%) were male. Malay race

constituted the majority (85%), followed by Indian (7.7%) and Chinese (7.7%) (Table 2).

Patients were then categorised based on the tumour laterality, tumour size, pre-operative and post-operative facial nerve status, duration of surgery, intensive care unit stays, and total hospital stays. Patients were diagnosed with severe to profound hearing loss, with a total of 73% of patients suffered from tinnitus, followed by imbalance or unsteadiness (46%). Facial paraesthesia accounted for eight patients (30.8%) (Table 3).

The tumour was categorised based on the size. A total of 9 (34.5%) patients had tumour size 31-40 mm. This was followed by 11-20 mm and 21-30 mm, with a total of 6 participants respectively (Table 4).

Post-operative Data

Post-operatively, data on duration of surgery, facial nerve function, complication of CSF leak, intensive care unit (ICU) stay, and total hospital stay

TABLE 1: Data on patients' age, tumor laterality and presenting symptoms

Patient No.	Age	Tumour laterality	Symptoms
1	28	Left	Left ear tinnitus and reduced hearing with facial paraesthesia
2	30	Left	Left facial palsy with imbalance
3	64	Right	Right sided reduced hearing with tinnitus and gait disturbance
4	50	Right	Gait disturbance with right tinnitus and reduced hearing and facial paraesthesia
5	46	Left	Left ear tinnitus and hearing loss, left facial nerve palsy and paresthesia
6	62	Right	Right sided hearing loss with tinnitus and vertigo and unsteady gait
7	45	Right	Unilateral tinnitus
8	50	Right	Right sided hearing loss and imbalance with facial paraesthesia
9	52	Left	Left sided hearing loss with tinnitus and unsteady gait with headache
10	16	Left	Left unilateral hearing loss with tinnitus, occasional imbalance and giddiness
11	74	Right	Right sided reduced hearing with tinnitus
12	45	Left	Left sided reduced hearing with tinnitus and headache
13	27	Right	Right sided reduced hearing with facial paraesthesia
14	38	Left	Left unilateral hearing loss
15	63	Right	Right sided reduced hearing with unsteady gait and diplopia
16	59	Right	Right sided reduced hearing with tinnitus and imbalance
17	63	Left	Left sided reduced hearing with unsteady gait
18	42	Right	Right sided reduced hearing with tinnitus
19	52	Right	Right sided reduced hearing with tinnitus and imbalance and facial paraesthesia
20	63	Right	Right ear tinnitus and facial pain
21	53	Right	Right ear tinnitus and reduced hearing
22	52	Right	Right sided reduced hearing with tinnitus and imbalance and giddiness with facial paraesthesia
23	58	Left	Left sided hearing loss
24	44	Right	Right ear tinnitus with reduced hearing
25	53	Left	Vertigo
26	54	Right	Right sided hearing loss with tinnitus and facial paraesthesia

TABLE 2: Patients' demographics information (N=26)

Demographics	No. of participants (n)	Percentages (%)
Gender		
Male	18	69.2
Females	8	30.8
Races		
Malays	22	85
Chinese	2	7.7
Indians	2	7.7

TABLE 3: The common presenting symptoms among the patients

Symptoms	n (%)
Severe to profound unilateral hearing loss	26 (100%)
Tinnitus	19 (73%)
Unsteadiness/ imbalance	12 (46%)
Facial paresthesia	8 (30.8%)
Headache	3 (11.5 %)

TABLE 4: Tumour size

Tumour size group*	n (%)
0-10mm	0
11-20mm	6 (23.1%)
21- 30mm	6 (23.1%)
31- 40mm	9 (34.6%)
>40mm	5 (19.2%)

*New and Modified Reporting Systems from the Consensus Meeting on Systems for Reporting Results in Vestibular Schwannoma classification

were obtained. Preoperatively, 23 (88.5%) had House-Brackmann (HB) grade I facial nerve grading, two patients (7.7%) had HB grade II and one patient had HB grade III. From a total of 26 patients, 19 (69.2%) had a normal post-operative facial nerve function. However, a total of seven patients (30.8%) suffered from permanent facial nerve palsy ranging from *House-Brackmann* grade II to grade IV (Table 5).

Our data revealed that four (15.4%) patients sustained cerebrospinal leak, which resolved after insertion of lumbar drain for a week. Histopathology of the excised tumour via the *translabyrinthine* approach showed that 88.5% were VS, whereby others included meningioma and endolymphatic sac tumour which accounted for 11.5% (Table 6).

Duration of surgery for all patients ranged from 10 to 20 hours, with mean of 13 hours. Patient number 13 underwent *translabyrinthine* surgery with concurrent cochlear implant surgery, hence the duration of surgery was longer. The duration of ICU stays ranged from 10 hours to 432 hours, with median of 24 and IQR 18. In addition, the total duration of hospital stays ranged from 5 days to 51 days, with median of 8.5 and IQR 5.75 (Table 7).

TABLE 5: Pre- and post-operative facial nerve grading

House-Brackmann grading	Pre-operative facial nerve grading [n (%)]	Post-operative facial nerve grading [n (%)]*
I	23(88.5%)	19 (69.2%)
II	2 (7.7%)	1 (3.8%)
III	1 (3.8%)	1 (3.8%)
IV	0	3 (11.5%)
V	0	2 (7.7%)
VI	0	0

*12-months post-operative

TABLE 6: Post-operative diagnosis of cerebellopontine angle tumours

Post-operative diagnosis	n (%)
Vestibular schwannoma	23 (88.5%)
Others	3 (11.5%)

DISCUSSION

VS is a benign tumour involving the IAC, CPA, cochlear and labyrinth that can lead to displacement and compression of the brainstem. Therefore, causing hearing loss, tinnitus, vertigo, dizziness, headache, hypoesthesia and cranial nerve palsies (Rosahl et al. 2017; Zanoletti et al. 2016).

Over time, acoustic neuroma surgery

TABLE 7: Duration of surgery, Intensive care unit (ICU) stays and total hospital stays

Patient number	Duration of surgery (hours)	Duration of ICU stays (hours)	Duration of total hospital stays (days)
1	17	30	51
2	13	42	19
3	10	17	6
4	13	30	7
5	11	432	60
6	14	72	8
7	11	48	11
8	12	12	10
9	11	15	23
10	12	10	8
11	12	22	9
12	12	24	14
13	20	48	13
14	14	24	13
15	16	24	14
16	12	24	8
17	12	16	7
18	13	12	9
19	13	12	8
20	15	12	8
21	15	12	8
22	13	12	8
23	13	24	5
24	14	24	20
25	12	12	7
26	10	48	5

has advanced and produced amazing results. The goal of surgery is to completely remove the tumour while maintaining hearing and facial nerve function (Zanoletti et al. 2016).

As surgical techniques have advanced, other disciplines have emerged to provide patients with excellent, holistic care. The interdisciplinary team comprised of otorhinolaryngologist, neurosurgeons, audiologists, anesthesiologists and physical therapists (Sweeney et al. 2014).

There are many approaches to CPA tumor described in previous literatures. The *translabyrinthine*, middle cranial fossa and *retrosigmoid* craniotomies remain the most utilised surgeries (Darrouzet et al. 2004; Sweeney et al. 2014). The surgical approach depends on the tumour size and site, preoperative hearing threshold, functional status and surgeon preference (Darrouzet et al. 2004; Rosahl et al. 2017).

The *translabyrinthine* approach is preferable for patients with non-serviceable hearing. The advantageous are good exposure of the lateral IAC with no cerebral retraction (Darrouzet et al. 2004; Rosahl et al. 2017; Sweeney et al. 2014). Most importantly, this approach provides safe exposition of the facial nerve (Rosahl et al. 2017). The facial nerve is identified from the mastoid segment until the internal auditory meatus and CPA (Azmi et. al. 2006). If the facial nerve is severed or injured during the tumour removal, *translabyrinthine* offers direct approach for immediate, intraoperative nerve repair (Azmi et al. 2006).

Other approaches as commonly mentioned in other literatures are the *retrosigmoid* and middle cranial fossa approach. *Retrosigmoid* approach offered a bird's-eye view of the posterior fossa, thus best for large tumour. It is considered as hearing preservation surgery for smaller tumours. Whereby, the middle fossa craniotomy is best the approach for small tumours in the IAC with good hearing (Darrouzet et al. 2004; Rosahl et al. 2017).

Our data centred on the *translabyrinthine* approach of surgery. As evidenced in our study, the low morbidity rate is also supported by previous study (Darrouzet et al. 2004; Sweeney et al. 2014; Zanoletti et al. 2016). *Translabyrinthine* surgery reportedly has an operative mortality rate ranging from 0-2% (Zanoletti et al. 2016). In a case series of patients undergone *translabyrinthine* surgery, the complications included subdural hematoma and CPA hematoma with incidence rate of 0.4% and 0.6% respectively. Other less common complications are cerebellar oedema, brainstem hematoma transient aphasia and lower cranial nerve palsies (Zanoletti et al. 2016).

Our data showed low rate of complications, namely post operative facial nerve palsy and CSF leak. A total of 19 patients (69.2%) had a normal post-operative facial nerve function. There are factors that may attributed to the post-operative facial nerve function as mentioned by Zanoletti et al. (2016). The author correlated the size of tumour with post-operative facial nerve function. The rate of normal facial nerve function with tumour less than 1 cm, is more than 96%. However, this rate drops further in larger tumour size (Zanoletti et al. 2016). But bear in mind that paresis of the facial nerve may inevitably occur due to compromised vascularity to the nerve during tumour dissection (Azmi et al. 2006).

The cases of cerebrospinal fluid (CSF) leakage ranged from 2.8-14% from

previous studies (Zanoletti et al. 2016). Additionally, as reported by Azmi et al. (2006), CSF leak was found in four from 15 cases (26.7%). Our data showed that four from 26 patients had CSF leak. The leak can be at the skin incision, otorrhoea if the tympanic membrane was perforated or rhinorrhoea. In our cases, the CSF leaked from the incisional wound. Specifically in *translabyrinthine* surgery, three pathways exist for CSF to reach the temporal bone air cells i.e. the dural opening into the posterior cerebral fossa, the exposed air cell tracts proximal to the IAC and the CSF tracking via the breach of the IAC fundus to the vestibule (Azmi et al. 2006). Intraoperatively, cautious closure of mastoid cavity with musculocutaneous flap and the use of fibrin glue are the hallmark of our current technique (Darrouzet et al. 2004). In the present study, we advocate lumbar drain as part of the conservative management in CSF leak, which resolved after a week. Our case series did not encounter post-operative meningitis complication. The present study showed that the total duration of hospital stays ranged from 5 to 51 days, which we do not stratify according to tumour size, as previously reported (Visagan et al. 2018). In the mentioned study, the length of hospital is rather similar in both *translabyrinthine* approach and retrosigmoid approach, but the difference is eminent with tumour size 31-40 mm for both approaches, whereby retrosigmoid approach have a longer stay. Length of stay is a complex parameter that requires further study to gauge as there are many confounding factors. According to our experience, approximately 60% of the tumour were typically debulked, and a follow-up imaging study of Magnetic Resonance Imaging is usually conducted six months after surgery to evaluate tumour growth.

CONCLUSION

Acoustic neuroma surgery via *translabyrinthine* approach is a safe and compelling treatment. The complications rate is low with acceptable facial nerve outcome. The interdisciplinary concept is recommended as each step of the operation requires specialist expertise.

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