MICROSURGERY VESUS GAMMA KNIFE

FOR MANAGEMENT OF SMALL SIZED ACOUSTIC NEUROMA

Systematic Review of Literature

Submitted for partial fulfillment of Master Degree in

OTORHINOLARYNGOLOGY

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Aim of the work

There is still debate about the best modality for treatment of VS. The aim of this work is to run a systematic review on small sized VSs (tumors up to **3** cm in size) to compare the outcome of microsurgery and gamma knife in order to reach the best treatment option in such patients**.**

Introduction

Vestibular Schwannoma (**VS**), also called acoustic neuroma is a benign intracranial neoplasm arising from Schwann cells, which myelinate the vestibular portion of the **8th** cranial nerve. Incidence of these tumors is approximately about **1** per **100,000** persons per year **(Propp et al., 2006).**

VSs are closely related to the cerebellopontine angle (**CPA**) which is a distinct V-shaped anatomical area with boundaries formed by the folding of the cerebellum around the pons and medulla and with the middle cerebellar peduncle as its floor **(Rhoton and Tedeschi, 1992).**

Although histologically benign, VS can lead to serious morbidity or even mortality from compression on cranial nerves or brain stem or from development of hydrocephalus **(Pollock et al.,1995).**

Gadolinium enhanced magnetic resonance imaging (**MRI**) of the internal acoustic meatus and CPA remains the standard method for diagnosing VS **(Propp et al., 2006)**.

There are **3** main treatment options available for patients with VS: watchful waiting, microsurgery and gamma knife radiosurgery (**GKS**) **(Kaylie and McMenomey, 2003)**.

There is no level **1** evidence (large randomized trials with clear-cut results and low risk for error) comparing microsurgery and GKS; the majority of data is level **3** or less, coming from retrospective studies (**Pollock, 2008).**

VS surgery has evolved tremendously since the first trial by Sir **Charles Balance** in **1894**. During the first half of the **20th** century, the mortality rate was greater than (**20%).** Since the early **1950s**, thanks to the advent of the operative microscope, cranial nerve monitoring and improved surgical and neuro-anathesiological techniques the rate has declined to less than **(1%**) (**Darrouz et al., 2004**).

The outcome after VS surgery is predominantly dependant on tumor size. Other factors include tumor biology, presenting symptoms, and surgical expertise. Although treatment of large tumors is clearly the domain of surgery, the present challenge is selection and timing of the best treatment for small tumors (**Gjuric and Rudic, 2008**).

The goal of surgery is complete removal of the tumor while goal of radiotherapy is tumor control which is defined as tumor regression or stable size (**Fayad and Brackman.,2005**).

Management of VS by radiation is through stereotactic radiosurgery (**SRS**) or stereotactic radiotherapy (**SRT**). SRS can be delivered by the gamma-knife (**GK**) or a dedicated linear accelerator (**LINAC**) (**Selch, 2009**).

In **1951**, **Leksell** introduced SRS which delivers a high dose of ionizing radiation to a stereotactically predetermined intracranial location in a single session (**Kaylie et al., 2000**). Radiosurgery has been proposed as an alternative treatment to microsurgery for tumors less than **3** cm in diameter (**Rosenberg et al., 2000**). Radiosurgery is rarely performed in patients with larger tumors, because the reduction in the dose necessary to minimize adverse effects on tissue decreases the efficacy of the procedure **(Kondoziolka et al., 1998).**

GK has been the most commonly reported type of radiotherapy for the treatment of acoustic tumours **(Fayad and Brackman., 2005).**

Tumor control is the principal aim of management of these tumors. However owing to the benign nature of most schwannomas, it requires the passage of long periods of time to ascertain whether this outcome measure has been achieved (**Rutherford and King , 2005**).

Facial nerve preservation is a critical measure of clinical outcome after VS management **(Yang et al., 2009).** Preservation of facial function varies according to tumor size and the surgeon's experience. In addition to tumor size, intraoperative electrophysiological facial nerve monitoring assists the surgeon in saving the nerve **(Lunsford et al., 2008).**

Given that very similar rates of tumor control and facial function may be achieved by the main treatment modalities of surgery and radiotherapy, new discriminators of better management have been sought. Hearing preservation has assumed this mantle as the objective that defines successful treatment (**Rutherford and King, 2005**).

Review of literature

Natural history of vestibular schwannoma

VS is a benign tumor of the **8th** cranial nerve that is found in the CPA and internal auditory canal (**IAC**) (**Shin et al., 2000**). The vestibular & cochlear components usually enter the meatus of the IAC as a single unit, and divide within the IAC (**Fig. 1**) (**O'Rourke and Wallace, 2004**).

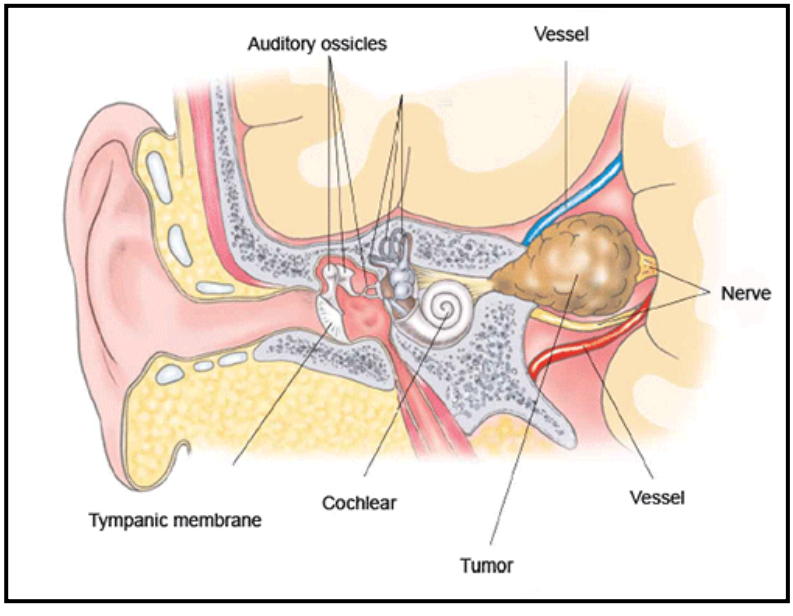

Fig.(1): An illustration for VS (**Matsumae, 2008**).

Most VSs originate at glial-Schwann cell junction (the junction between glial cells and myelin-forming Schwann cells), but they may occur anywhere along the nerve myelinated by Schwann cells (**O'Rourke and Wallace, 2004**).

Roosli et al., (2011) reported the origin of VS in **49** patients having **50** VSs. **12** tumors had their origin from the cochlear nerve and **38** tumors from the vestibular nerve. Among those originating from the vestibular nerve, **2** were intralabyrenthine, and **36** were in the IAC. Among these **36** tumors, **13** had their origin from the trunk, **13** from the superior vestibular nerve and **10** had their origin from the inferior vestibular nerve.

Histologically, VS contains Antoni A and B areas with nuclear atypia, whorls, scarring, chronic inflammation, sheets of macrophages, hyalinized and malformed vessels, recent and old thromboses and hemosiderin deposits (**Fig. 2**) (**Sobel, 1993).**

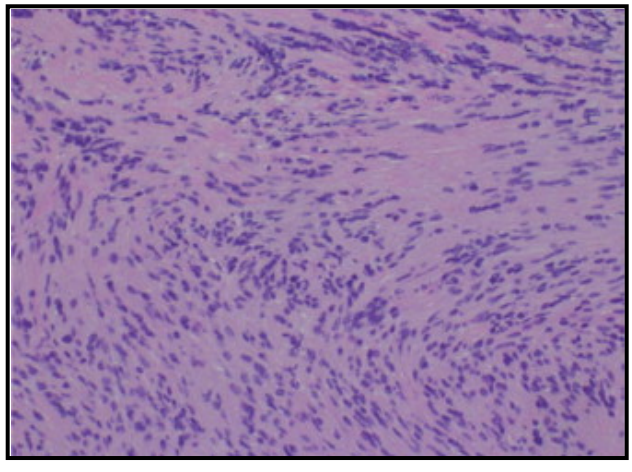

Fig.(2): Hematoxylin and eosin (H&E) stained VS demonstrating Antoni A and Antoni B cytoarchitecture (**Miller et al., 2012).**

As the VS grows, it gradually fills all the internal acoustic meatus and eventually protrudes out of the porus. It is not known how long this process takes; some tumors grow fast and others grow slowly **(Browning et al., 2008).**

Extra-meatal expansion of the tumour into the pontine cistern initially develops silently. Growth in this direction causes displacement and stretching of the **7th** and **8th** cranial nerves and the anterior inferior cerebellar artery (**AICA**). After further growth, the tumour expands sufficiently to touch and compress the cerebellum and the **5th** cranial nerve. Further growth and expansion causes compression and displacement

of the brainstem and the **4th** ventricle which leads gradually to hydrocephalus .The tumour may extend to the tentorium and can obstruct the cochlear aqueduct. Over time, the **5th** and **6th** nerves become stretched and thinned in much the same way as the **7th** nerve **(Browning et al., 2008).**

Reporting the size of VS has always been a great problem and there are at least **20** different classifications in use today. The main problem has been the inclusion and addition of the intrameatal element to the overall size of the tumour (**Kanzaki et al., 2003)**.

At the Consensus Meeting on Reporting Systems on Vestibular Schwannorna held at Keio University, Tokyo, in November **2001**, the following classification scheme was proposed and recommended for adoption (**table 1**). It was based on the size of the largest extrameatal diameter. It distinguishes between the intra- and extrameatal portions of the tumour. If the tumour is entirely within the meatus without any extension out of the porus, the term 'intrameatal' should be used. The extrameatal size of such a tumour is **zero**. The size of the intrameatal tumour can be recorded separately using such features as its length and width, for example **5 x 10** mm,

but the size of the intrameatal portion should never be added to the extrameatal tumour size **(Kanzaki, 2003)**.

Table (1) : Classification of VS according to size (**Kanzaki et al.,2003)**.

Grade	Extrameatal size	mm
1	Small	**1-10**
2	Medium	**11-20**
3	Moderately large	**21-30**
4	Large	**31-40**
5	Giant	**> 40**

Various ways of describing VS tumor growth have been proposed. The conventional method of assessing VS is by performing two-dimensional measurements. Some authors use the largest anteroposterior and/or mediolateral dimension, and sometimes combined with the craniocaudal dimension **(Saito et al., 2007).**

Nedzelski et al., (1992) and **Strasnick et al., (1994)** each followed **50** patients with VS for mean periods of **3.5** and **2.9** years, respectively and they reported that the mean tumor growth rate for VS is **1.1** mm/year.

The first Danish **'wait and scan'** prospective study followed a series of **123** patients with **127** VS over the period from **1973** to **1999**. It was updated three times (in June **1993**, June **1996** and June **1999).** In the third period from **1973** to **1999**, with a mean observation period of **4.2** years, the corresponding percentages were **(88 %)** showing positive growth, (**9 %**) no growth and **(6 %)** negative growth (**Fig.3) (Charabi et al., 2000)**.

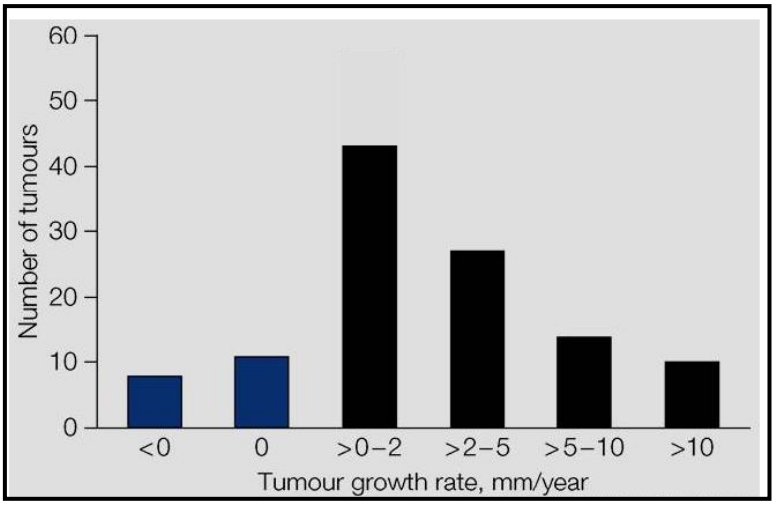

Fig.(3): Variation in growth rate of VS for mean observation period of **4.2** years in **127** VS. (**6 %)** of the tumours shrink, **(9 %**) show no growth, **45 %** grow from **0** to **2** mm per year, **(21 %)** from **2** to **5** mm per year, **(11%)** grow from **5** to **10** mm per year and (**8 %)** for more than **10** mm per year **(Charabi et al.,2000).**

All VSs have an unpredictable growth pattern. Some with no measurable growth initially can change and grow. Thus, patients allocated to a 'wait and scan' management policy must be followed indefinitely **(Charabi et al., 2000)**.

According to the meta-analysis conducted by **Smouha et al., (2005)**, they found that in **1345** VS patients, (**57%)** of tumors were nongrowing, whereas (**43%)** showed positive growth during a mean follow-up of **3.2** years**.**

Several studies, with longer follow-up periods ranging from **3** to **7** years, still report high nongrowing tumor rates **(Al Sanosiet al., 2006).**

The observed spontaneous involution of VS may be explained by tumor necrosis caused by intratumoral thrombosis and may be part of normal involution of tumors that have reached their maximum growing potential **(Luetje, 2000).**

According to **Leeuwen et al., (1995),** the presenting syptom in the majority of cases was unilateral hearing loss. Less frequent syptoms included tinnitus, vertigo, and facial paresthesia. Small tumors were associated with hearing loss and tinnitus, while vertigo and otalgia were infrequent. Large

tumors were associated with headache, facial paresthesia, and ataxia, while eye syptoms and dysarthria were less frequent

Evoked auditory brainstem response (**ABR**) has been a fundamental investigation in the differential diagnosis of perceptive hearing loss and other oto-vestibular disorders, as the sensitivity of ABR made it possible to exclude a retro-cochlear lesion, particularly VS **(Montaguti et al., 2007).**

When the tumour is completely confined to the IAC a definite diagnosis of VS becomes hard to achieve. It has been stated that VSs larger than **5** mm in diameter can be detected with a high degree of accuracy with high resolution CT **(Atilla et al., 1993).**

MRI is now the imaging method of choice for diagnosis of VS and allows for detection of tumors as small as **3** mm. Gadolinium-diethylenetriamine pentaacetic acid (**Gd-DTPA**) is a paramagnetic contrast agent that increases the intensity of VS on T1-weighted MRI scans **(Weber ,1992**).

Treatment of vestibular schwannoma

The treatment modalities for VS include conservative observation, microsurgery, stereotactic radiosurgery and fractionated radiotherapy **(Arthurs et al., 2011).**

A) Conservative management :

Progress in radiologic evaluation, especially Gd-DTPA MRI has resulted in the detection of small, less symptomatic VS. Some authors have proposed a "wait-and-scan" policy as a reasonable alternative to immediate surgical excision. The goal of this therapy, based on the usually slow progression of symptoms, is to minimize therapeutic risks and complications and to preserve an optimal quality of life in selected patients **(Shin et al., 2000).**

Conservative management has increasingly become a treatment option in appropriate cases **(Hoistad et al., 2001).** Observation may be recommended in elderly patients, those in poor health, those with small tumors but already poor hearing, or those unwilling to undergo microsurgery or radiotherapy **(Fayad and Brackmann, 2005**).