



Review Article

Middle Cranial Fossa Surgery for Hearing Preservation in Vestibular Shwannomas: A Comparative Review of Contemporary Findings

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Abstract

Vestibular Schwannomas (Vs) are slow growing, benign tumors that derive from the eighth cranial nerve and the neural crest of the inner ear. Historically, they have been considered benign because of their slow growth rate and asymptomatic nature. However, case studies show that when left untreated, they can grow into large tumors that may cause sensorimotor disturbances, hydrocephalus, and untimely death. Common symptoms of Vestibular Schwannomas are hearing loss, tinnitus, headaches, imbalance, dizziness, vertigo and paresthesia.

A specific protocol for pre-operative and post-operative test batteries has not yet been established and there are currently no reliable predictors for tumor growth. Diagnosis of Vs relies on neuroimaging techniques: Computerized Tomography and Magnetic Resonance Imaging.

Several procedures are presently in use for the treatment of Vestibular Schwannomas: Middle Cranial Fossa, Retrosigmoid, and Translabyrinth Approach, Conventional External Beam Radiation Therapy and Stereotactic Radiosurgery (SRS). Although there have been significant advances in current technologies and surgical procedures, all treatments are associated with risk of hearing loss and other complications. The outcomes and complications of treatments are discussed in this paper, with the middle cranial fossa as the preferred method of treatment for hearing preservation.

Keywords: Vestibular Schwannomas, Active Surveillance; Wait and Scan; Surgical Resection; Middle Cranial Fossa; Retrosigmoid; Translabyrinth Approach; Conventional External Beam Radiation Therapy; and Stereotactic Radiosurgery (SRS)

Objective

The objective of this review is to draw a comparison between the hearing outcomes of traditional surgical procedures and radiation treatments for vestibular schwannomas, with the middle cranial fossa approach as the preferred method of treatment to preserve integrity of hearing.

Method

Cross reference findings gathered from publications about

three surgical procedures and two radiation therapies have been presented and examined in this paper, along with a detailed interpretation of the findings associated with detection, diagnosis, treatment, and post-operative hearing results of small to large Vestibular Schwannomas. The procedures discussed are the middle fossa, retrosigmoid, translabyrinthine approach; radiotherapy, and fractionated stereotactic radiotherapy.

Hearing loss has been defined as 61-80 decibels (db). Tumor size has been defined as 2 cm for small to medium size tumors; 4 cm for large tumors [1].

The information gathered for this review was obtained from online sources, journal publications, original research papers, clinical studies, and review articles, gathered from the National Library of

Medicine and the National Center for Biotechnology Information. Outside sources have been referenced, accordingly. Terms searched were: Vestibular Schwannomas, Active Surveillance, Wait and Scan, Surgical Resection, Middle Cranial Fossa, Retrosigmoid, Translabyrinth Approach, Conventional External Beam Radiation Therapy, and Stereotactic Radiosurgery (SRS). Inclusion was limited to publications pertaining to hearing loss from Vestibular Schwannomas and their respective treatments.

Introduction

Vestibular Schwannomas (Vs) are a benign tumor, stemming from Schwann cells anywhere along the path of the eighth cranial nerve, the neural crest, and the internal acoustic meatus. Vs account for most brain tumors, with 80% to 90% located in the cerebellopontine angle [2].

Historically, Vs have been considered benign because of their slow growth rate and asymptomatic nature. However, case studies show that when left untreated, they can grow into middle-sized to large tumors that can cause sensorimotor disturbances to patients) [2]. Large schwannomas proximity to the brain stem and cerebellopontine angle can also make them especially dangerous to patients: causing hydrocephalus, a condition that leads to compression or invasion of cerebrospinal fluid to neurovascular structures [3], resulting in death [1].

The most common symptoms of Vestibular Schwannomas are unilateral sensorineural hearing loss (94% of patients) and tinnitus (83% of patients). Other common symptoms are headaches, imbalance, dizziness, vertigo and paresthesia [2].

Presently, there is no genetic profile for patients with Vs Fujita. The only consistency is a genetic alteration in the inactivation of the neurofibromatosis type 2 (NF2), merlin: a cytoskeletal protein encoded by the NF2 gene on chromosome 22q [1].

Vs cases are categorized as either sporadic and unilateral: occurring randomly rather than by a genetic condition; and, bilateral: neurofibromatosis type 2 (NF2) [1].

Incidence rate of Vs is 1 in 100,000 per year [4]. Prevalence rate is 1 in 2000, and 1 in 500 in patients 70 and older [1].

Several treatments are presently in use for Schwannomas: Active Surveillance (Wait and Scan), Surgical Resection (Middle Cranial Fossa, Retrosigmoid, and Translabyrinth Approach), Conventional External Beam Radiation Therapy and Stereotactic Radiosurgery (SRS).

Pre-Operative Considerations

A specific protocol for test batteries has not yet been established to measure indicators such as tumor size and tumor growth; though identifying tumors appears to be central to management options and affects treatment of choice [5]. This might be due to individual

differences in the inner ear and individual differences in tumors: while some tumors remain stable over time, others progress to cause life-threatening symptoms in a very short time [3]. Magnetic Resonance Imaging (MRI) is currently the method of choice for detection, diagnosis, tumor characterization, operative planning, and post-operative treatment evaluation [6].

In recent years, sporadic diagnosis for Vestibular Schwannomas has increased and the use of traditional microsurgeries to treat Vs has decreased, due to the adoption of serial imaging as the most common initial evaluation and treatment strategy, especially for small sized Schwannomas [1].

Neuropathology: A Brief History

Vestibular Testing

The first identification of breakthrough patterns in Schwannomas occurred in the 1920s, by Nils [5]. In the 1960's, advanced audiometric testing led to increased sensitivity in tests for retrocochlear lesions and for earlier diagnosis and detection of VS Radparvar. Jerger utilized the automatic audiometry for cochlear and retrocochlear lesions, allowing to differentiate between different types of hearing loss. Testing was expected to show unilateral sensorineural high frequency loss [5].

Robert Barany work on vestibular testing involving ice water testing for the integrity of vestibulo ocular reflex, and caloric procedure, earned him a Nobel prize [5].

Wait and scan

Observation without therapy have become a popular approach for the recognition of Vestibular Schwannomas. Findings justify preliminary observations that wait and scan is a viable approach for managing small tumors. As the use of Radiological Imaging has increased, management has shifted to wait and scan, and tumor size at time of diagnosis has decreased. Earlier detection has also meant treatment for larger tumors is more readily available [5].

Radiological Imaging

The microscope was introduced to neurosurgery by Theodore Kurze, who used the device for Vestibular Schwannoma resection in 1957 [5]. 1963, Harvey Gass, described the opaque cisternography procedure of early lesions in the CPA [5]. In 1964, Robert Scanlon, improved the procedure and advocated for early diagnosis of small intracranial tumors [5].

CT Scans

In 1971, Computerized Tomography (CT) scans replaced all tests for evaluation of tumors. Metrizamide cisternography using nonionic water soluble subarachnoid contrast, reported by the Arne Grepe in 1974, and gas CT cisternography using intrathecal air or filtered carbon dioxide as a subarachnoid contrast material by Ove Sortland in 1979 [5].

Magnetic Resonance Imaging

Diagnosis for Vestibular Schwannomas was revolutionized in the 1980s with the development of the Magnetic Resonance Imaging machine by Raymond Damadian, Paul Lauterbur and Peter Manfeld [5]. Magnetic Resonance Imaging(MRI) is currently the method of choice for detection, diagnosis, tumor characterization, operative planning, and post-operative treatment evaluation [6].

Surgical Procedures

The procedures presently in use for the treatment of Vestibular Schwannomas are the Middle Cranial Fossa, Retrosigmoid, and Translabrynth Approach, Conventional External Beam Radiation Therapy and Stereotactic Radiosurgery (SRS). Of the three surgical procedures, the Middle Fossa and Retrosigmoid approaches, are the preferred method of resection for small to medium sized schwannomas and are associated with integrity of hearing; while the translabyrinth approach is utilized for larger tumors and associated with complete hearing loss.

Middle Fossa

The middle fossa is a type of surgical procedure for the removal of small to medium schwannomas that allows for hearing integrity. The extradural approach that was first utilized more than 130 years ago. It was originally described by Parry in 1904 and utilized in the 1960's by House [7, 8].

The procedure has become an effective treatment for pathological conditions in the temporal region while minimizing trauma and hearing loss. The approach gives access to challenging regions like the cerebellopontine angle (CPA), petroclivial area, basilar artery, and cavernous sinus [7].

The middle fossa approach is typically utilized for tumors 2 cm and smaller in size. MCF surgery usually involves elevating the temporal lobe with a retractor to access the petrous bone. Required elevation of the brain is often associated with irreversible damage to the temporal lobe, cerebrospinal fluid leak, and gliosis [8].

Surgery of the middle cranial fossa requires specialized understanding of extreme variability in temporal lobe anatomy, and different surgical landmarks. Consequently, the middle cranial fossa and its variations, are considered one of the most difficult surgical procedures [7].

With further advances, the fossa approach has become one of the primary surgical procedures for resection of schwannomas and accessing challenging regions of the brain [7]. Reported success of hearing preservation is 55% to 70%.

Retrosigmoid

In 1925, Dandy, advocated for Intracapsular Denuncleation: a procedure that removes tumors by separating them from their

capsule without affecting the nerve sheath, followed by a capsular dissection from the brainstem with the unilateral suboccipital surgical approach. After further refinement, this procedure became known as the Retrosigmoid Approach [5].

The retrosigmoid approach is the most popular approach for surgical resection of vestibular schwannomas, being one of two resection procedures utilized for hearing preservation [5]. This procedure is suitable for patients with serviceable hearing and is often used for small to medium sized schwannomas [9].

Retrosigmoid surgery involves dissecting the transverse sinus and sigmoid sinus. After an incision to the dura, the cerebellum is retracted, the cerebellomedullary cistern is opened, cerebrospinal fluid released, and the tumor separated from the cochlear nerve [6].

Translabrynth

The translabyrinth approach, utilized for medium to large schwannoma resection, is considered the best approach for patients with unserviceable hearing. The procedure requires minimal brain retraction to access the internal auditory canal (IAC), minimizing the risk involved in parenchymal injury [10].

Translabrynth surgery involves creating a surgical incision behind the ear lobe, creating a cavity to expose the dura and inner ear, removing bone for access to the internal auditory canal and removing the tumor. Because the translabyrinth approach involves chiseling out the mastoid and removing the translabyrinth entirely, complete hearing loss is unavoidable [9].

Radiosurgery

Modern refinements to traditional surgical procedures began in the 1960s with advancements in the operating microscope, audiometric testing, radiological imaging, electromyography, and stereotactic radiosurgery [5].

Conventional Beam Therapy

Beam therapy is an effective treatment for Vestibular Schwannomas, that involves a radiation beam delivered precisely to the tumor. The types of beam therapy are Stereotactic radiosurgery, hypofractionated, fractionated, photon and proton beam therapy. Radiation therapy does not remove the tumor but inactivates Schwann cells, reducing the size of the tumor. The effect of radiation on hearing is largely dependant on dose to the cochlea: independent of radiation treatment modality. Hearing preservation is associated with smaller tumor size.

Stereotactic Radiosurgery (SRS)

In 1993, the first tumors were treated with SRS [5]. SRS is considered the dominant treatment for small to large Vestibular Schwannomas due to its safety and control rates [11]. SRS can be delivered with the gamma knife (elektra ab), cyberknife (accuray,

inc), zap-x (zap surgical systems, inc), other linear accelerator-based techniques, and protons frameless/ frame based [11].

In SRS, high-energy gamma rays, align the beam of radiation toward the isocenter of the tumor. This practice offers a noninvasive alternative to surgery [9]. SRS has been shown to be effective in tumors 3 cm and smaller. This form of treatment is not efficacious for tumors larger than 3 cm, as large tumors often require decompression. Failed gamma-knife radiosurgery is associated with complications and more difficulty in correcting surgical injury than other forms of treatments because it may result in severe fibrosis [6].

Findings

A multivariate comparison of surgical and radiation procedures and their outcomes indicated that there is a correlation between early onset detection, treatment, and prevention of hearing loss in Vestibular Schwannomas.

Although there have been significant advances in current technologies and surgical procedures, all treatments are associated with risk of hearing loss and other complications. Tumor size does not correlate with degree of hearing loss, as small tumors limited to the auditory canal result in hearing loss [1]. Phenotypical considerations should be considered when choosing a surgical approach.

Traditional surgical procedures, middle cranial fossa and retrosigmoid microsurgery, have been viewed more favourably for small to medium sized tumors. The middle cranial fossa approach has been associated with irreversible damage to the temporal lobe and 70% temporal gliosis [8].

Retrospective studies show that patients who underwent primary microsurgery for Vs from 2002 to 2012, where preoperative hearing did not differ between approaches, concluded that a decline in hearing was greater in the retrosigmoid approach (55.5 dB and 45.6 discrimination) than in the middle fossa (38.9 dB and 31.7) ($p < 0.011$ and 0.033 , respectively). The observed effects remained after controlling for tumor size.

Small to large vs, delivered platforms, including the gamma knife (elektra ab), cyberknife (accuray, inc), zap-x (zap surgical systems, inc), other linear accelerator- based techniques, and protons frameless and frame based [11].

There is a higher incidence of recurrence of vestibular shwannomas in patients treated with radiation therapy: hearing loss observed in post operative followups in comparison to other treatments is significantly higher in patients treated with radiation therapy. Radiation therapy damage to the inner cochlea can be attributed to varied doses in radiation therapy.

Oncological findings report poorer than expected results for hearing preservation in Stereotactic patients than less invasive

microsurgical and endoscopic techniques, debate surgical resection vs treatment. To date, no randomized control trials are available for patients of Stereotactic Surgery [5].

The outcomes and complications of treatment are comparable between procedures and mostly dependent on the expertise of the surgeon. Currently, neuroendoscopic procedures are utilized to complement conventional surgeries [8].

Discussion

Post-operative Test Battery

A specific protocol and test battery has not established correlation between clinical outcome and tests before and after surgery. To this day, there are no reliable predictors for tumor growth. This might be due to differences in tumor growth rate: while some tumors remain stable over time, others progress to cause life-threatening symptoms in a very short time [5]. Magnetic Resonance Imaging (MRI) is currently the method of choice for detection and diagnosis of Vestibular Schwannomas [3]. Sporadic diagnosis for Vestibular Schwannomas has increased due to the adoption of neuroimaging techniques (MRI) [1].

Conclusion

After a thorough assessment of the diagnostic tools, treatments presently available, the author has concluded that the implementation of surgical techniques in the early onset of Vestibular Schwannoma Tumors can help to prevent hearing loss [7].

A specific protocol and test battery has not established correlation between clinical outcome and tests before and after surgery, Though identifying tumors appears to be central to management options and affects operating view, proper testing is not yet in place. There are currently no reliable predictors for tumor growth. This might be due to differences in tumor growth rate: while some tumors remain stable over time, others progress to cause life-threatening symptoms in a very short time [5]. Magnetic Resonance Imaging (MRI) is currently the method of choice for detection and diagnosis of Vestibular Schwannomas [3].

The approaches utilized for hearing preservation are the retrosigmoid craniotomy and the middle cranial fossa, the retrosigmoid approach being the most popularized approach. Loss of hearing was greater with RS approach than the MF, even when accounting for tumor size and other variables. Findings of hearing preservation are consistent in small to medium sized tumors [6].

Tumor size is an important parameter that affects nerve localization, extent of resection, postoperative outcomes and complications [6].

Small to large vs, delivered platforms, including the gamma knife (elektra ab), cyberknife (accuray, inc), zap-x (zap surgical systems,

inc), other linear accelerator- based techniques, and protons frameless and frame based Excellent safety and control rates [11-14]. Gamma-knife radiosurgery is associated with severe fibrosis [6].

Recurrence of vestibular schwannomas after radiation therapy: the overall ratio of hearing loss observed post operative followups in comparison to other treatments is higher. Radiation therapy Damage to the inner cochlea or patients cochlear implants, this observation can be attributed to varied doses in radiation therapy.

Optimal operation depends on the tumor characteristics, patients hearing status, surgeon comfort, objective of operation. Intraoperative electrophysiological techniques combined with neuronavigation may be helpful to improve resection [6].

VS microsurgery has been successful at achieving tumor removal with minimal morbidity and mortality. Although a general consensus for the treatment of vestibular schwannomas has not yet been established, the middle cranial fossa remains a viable alternative treatment for hearing preservation of tumors at an advanced stage.

Ethics

The present paper utilizes qualitative and retrospective methods to describe its findings. Patients' personal data were omitted from this paper. As this was a review of published research, no requirement of ethical approval needed to be fulfilled.

Availability of data and material

None.

Competing interests

The author declares that there were no conflicts of interest with regard to the content in this review.

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Authors' contributions

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