OTOLOGY, NEUROTOLOGY, AND SKULL BASE SURGERY

CLINICAL REFERENCE GUIDE

Theodore R. McRackan, MD
Derald E. Brackmann, MD

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FOREWORD

This remarkable text, edited by Drs. McRackan and Brackmann, is a significant addition to our armamentarium of references in the field of otology, neurotology, and skull base surgery. The chapters are well-written and encompass the scope of the subspecialty. Each author is a well-recognized expert in their field and the format of the chapters by outline is extremely easy to follow. It is an outstanding reference for residents, fellows, and practicing otolaryngologists as well as neurotologists. The text is simplistic and easily addresses any question that may arise in the mind of the reader. This is clearly a reference guide. No attempts have been made to increase the prose to describe the topics, but that is not to say that the key issues are hard to assess using the format. The editors are to be commended in formulating a text that is extremely user-friendly to the reader. In these days where the majority of information we seek is available on the Internet, this particular book will be a completely different offering that will be a welcome addition to anyone's library. I look forward to receiving my copy!!!

—Harold C. Pillsbury, MD
Professor and Chief
Otolaryngology-Head and Neck Surgery
The University of North Carolina at Chapel Hill

CHAPTER

29

Vestibular Schwannoma

Theodore R. McRackan and Derald E. Brackmann

NOMENCLATURE

- Virchow named lesion neuroma due to parallel fibers on histology appear to be axons
- Initially thought to be cochlear origin due to associated hearing loss
- Murrey and Stout discovered origin cell to be Schwann cell
- Preferred name is currently vestibular schwannoma (VS) although acoustic neuroma is still widely used

EPIDEMIOLOGY

- Represent approximately 6% of all intracranial tumors
- Prospective Denmark database estimate approximately 20 VS per 1 million population
- Incidence increasing due to awareness and improved imaging access and quality
  1. 7.8 VS/million 1976 to peak 23 VS/million year 2004
- Tumor size at time of diagnosis has decreased from 30 mm in mid-1970s to current 10 mm
- Mean age at diagnosis has also decreased from 58 years to 49 years

PATHOPHYSIOLOGY

- Arise from vestibular division of eighth cranial nerve
  1. More commonly the inferior vestibular nerve
- VS do not likely arise from Obersteiner-Redlich transition zone (glial-schwannian junction) as initially thought
  1. Most tumors form lateral to the Obersteiner-Redlich transition zone; oligodendrocytes produce myelin medial to junction and Schwann cells lateral
  2. Greatest density of Schwann cells lies at **Scarpa's ganglion**; VS likely arise from Schwann cell population associated with the Scarpa's (vestibular) ganglion; like VS, Scarpa's ganglion can lie near junction but also at various locations
- Understanding of genetic origins of sporadic VS is still uncertain; tumor biology is mostly understood through studying Neurofibromatosis type II (see Chapter 31)
- There are two histological patterns; Antoni A are densely packed cells with an organized whirled appearance (*Verocay* body); Antoni B are more loosely packed collection of vacuolated pleomorphic cells; Antoni B may occur more frequently in large tumors
  1. Stain positive for S-100

PRESENTATION

- Hearing loss is most common presenting symptom and present in 95% of patients
  1. Typically gradual onset unilateral sensorineural hearing loss
  2. During clinical course up to 26% of patients report sudden onset SNHL
  3. Normal hearing patients are more likely to have tumors <1 cm
- AAO-HNS has standardized hearing reporting in VS (Table 29–1)
- Vertigo/dysequilibrium is most common presentation in patients with normal hearing
  1. Occurs in 20–50% of patients
  2. Vertigo typically occurs relatively early in symptomology due to peripheral nerve disturbance but can spontaneously resolve
  3. Dysequilibrium typically occurs late in course due to cerebellar involvement
  4. Due to slow rate of tumor growth, majority of patients compensate
- Tinnitus reported in 53–70% of patients
  1. Variable in nature
  2. Asymmetric tinnitus without hearing loss should warrant investigation for retrocochlear pathology
- Trigeminal nerve symptoms occur in approximately 20% of patients
  1. Most commonly malar hypesthesia or parasthesia
     a. Can result in absent or decreased corneal reflexes
     b. Very rare in tumors <1 cm, but occurs in 20% of patients with 1–3 cm tumors and 50% in tumors larger than 3 cm
  2. Given high prevalence of headache in general popular, hard to establish association with acoustic neuromas; vestibular schwannoma-related headaches are likely dull with mild to moderate intensity

TABLE 29–1. American Academy of Otolaryngology-Head and Neck Surgery Hearing Classification after VS Surgery

Hearing Class	PTA	SDS
A	≤30	≥70%
B	31–50	50–71%
C	>50	>50%
D	Any	<50%

Note. *PTA* = pure-tone average; *SDS* = speech discrimination score.

- Facial nerve (FN) dysfunction is relatively uncommon (<10% of patients)
  1. Can present as facial twitching or paralysis/paresis
  2. Either symptom should raise concern for primary facial nerve tumor
- Late symptoms are much less common today due to early detection
  1. Increased intracranial pressure from hydrocephalus can result in nausea/emesis, diplopia, papilledema, and lethargy to obtundation
  2. Lower cranial nerve symptoms of hoarseness, dysphagia, and dysarthria are rare in modern era

DIAGNOSIS AND EVALUATION

- Audiologic evaluation
  1. Pure-tone audiogram (PTA) and speech discrimination scores (SDS) show wide variability from normal to no response
  2. Rollover (worsened SDS with increasingly loud presentation) and acoustic reflex decay (acoustic reflex fades during prolonged presentation of signal) were once thought to be good markers for retrocochlear pathology; over time these have been found to be poorly reliable
- Auditory brainstem response (ABR) testing
  1. Attractive as screening tool because it is noninvasive, inexpensive, and rapid
  2. The interaural wave I to V latency difference (IT5) is most reliable screening measure for VS; latency of 0.2 ms or more is considered abnormal
  3. Sensitivity ranges from 63–95%
     a. Highest sensitivity with tumors larger than 2 cm (100%)
     b. Decreased to 60% with tumors less than 1 cm
  4. Stacked ABR increases sensitivity to 95% for all tumors
- Electronsytagmography
  1. Abnormal results in 98% of superior vestibular tumors and 60% inferior vestibular nerve tumors
  2. Reduced ipsilateral caloric response and spontaneous nystagmus to contralateral side are common findings
  3. Wide variety of findings in larger tumors including: abnormal saccadic pursuit, bilateral horizontal gaze nystagmus, and failed fixation suppression
  4. Can be used to help identify vestibular nerve of origin in planning for hearing preservation surgery for middle cranial fossa surgery (discussed later)

- Magnetic Resonance Imaging
  1. Isotense or hypotense on T1 and T2, but enhance with gadolinium
  2. VS are centered on IAC, have spherical shape without dural attachment or hyperostotic adjacent bone
  3. Up to 100% sensitivity and specificity in modern era with gadolinium
  4. Able to accurately display size, location, depth of penetrance into IAC (discussed later for hearing preservation surgery)
  5. Heavily T2 weighted fast spin-echo series can help identify location of facial nerve (FN) nerve with regard to the VS
     a. This series can also identify VS with 98–100% sensitivity and 99.5–100% specificity without gadolinium

NATURAL HISTORY

- Determination of tumor growth varies based on study; some define intracanalicular tumor growth as growth to extrameatal tumor whereas others simply define tumor growth as growth in any dimension by 2 mm; most precise gauge is volumetric analysis
- Difficult to determine true VS growth rates as larger tumors are intervened upon and not included in natural history studies; therefore, there is an inherent bias toward decreased growth rates
- Collected data in meta-analyses show approximately 50% of VS grow (data ranges from 6–73% in literature)
- Mean growth rates in all VS 1–2 mm/year; this rate increases to 2–4 mm/year in those tumors with proven growth
- Majority of studies show no relationship between gender, age, duration of symptoms, initial hearing level, and laterality and likelihood of VS growth
  1. Multiple studies have shown that VS larger than 2 cm have increased propensity to grow
  2. Some evidence exists for increased growth rates of extrameatal compared to intrameatal tumors

TREATMENT

- Overall goal is to maintain patient quality of life and intervene when the decrease in quality of life from treatment outweighs the effects of further tumor growth