12 Management of Vestibular Schwannomas

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12.1 Introduction

The modern management of vestibular schwannomas represents a triumph of surgery and medicine. What was once a fatal disease with an almost equally fatal treatment is now curable with relatively little morbidity or mortality.

12.1.1 Historical Background

The earliest recorded attempt to remove an acoustic neuroma was by Charles McBurney in 1891. After removing the occipital skull with a mallet and chisel, he was unable to remove any tumor. Unfortunately, this initial effort proved fatal for the patient. Later that decade, a British surgeon named Charles Ballance successfully removed a vestibular schwannoma by a lateral suboccipital route (Stone 1999). Dr. Ballance performed the surgery in a staged manner with the craniotomy on one day and the tumor resection at a later date. He used blunt finger dissection to free the tumor from the pons and cranial nerves. Reportedly, the patient lived 18 years after the surgery, albeit with persistent facial and trigeminal nerve palsies (Stone 1999). The lateral suboccipital craniotomy that Charles Ballance employed became the standard approach for the early pioneers of vestibular schwannoma surgery.

The early attempts at surgical treatment of vestibular schwannomas carried substantial mortality, up to 78% by 1913 (Jackler 1994). Harvey Cushing, one of the great pioneers of neurosurgery, was successful in reducing the mortality rate to 20% by 1917 (Cushing 1921). Dr. Cushing emphasized the need to be gentle with the structures of the central nervous system and associated cranial nerves, arteries, and veins. In addition, he obtained meticulous hemostasis with the use of bone wax, silver clips, and electrocautery. Walter Dandy, a pupil of Dr. Cushing, further advanced the field by advocating complete tumor removal by gently stripping the capsule of the tumor away from the pons and cerebellum. Cushing, in contrast, chose to leave a small cap of tumor behind to avoid injury to the brain stem, arteries, and cranial nerves. The technical contributions of both surgeons were instrumental in reducing the mortality rate to 10% by 1931 (Jackler 1994). Despite this reduction in mortality, these pioneering surgeons were unable to remove tumors without critically injuring the facial nerve. The incidence of facial nerve impairment in the early and mid-20th century was significant, with the incidence of facial nerve impairment ranging from 43% to 80% (Cushing 1921, Jackler 1994).
palsy after removal of an acoustic neuroma was on the order of 60% by 1960. William House introduced the surgical techniques that were necessary to completely remove acoustic neuromas while preserving the pons, facial nerve, and anterior inferior cerebellar artery (House 1964).

12.1.2 Contemporary Surgery

Dr. House employed the middle fossa approach to the internal auditory canal, and he refined translabyrinthine craniotomy in the early 1960s (House 1964). The middle fossa approach provided exposure of the internal auditory canal, which was difficult to access by the more popular suboccipital approach. Translabyrinthine craniotomy (translab) provided more direct access to the cerebellopontine angle than suboccipital craniotomy, with less injurious cerebellar retraction than the suboccipital route. Dr. House was able to successfully utilize these approaches by employing the operating microscope and the high-speed drill in these surgeries. The operating microscope allowed for precise dissection of the tumor away from the facial nerve, brainstem, cerebellum, and cerebellar arteries. The high-speed drill enabled the surgeon to precisely remove the bony coverings of the auditory canal and cerebellopontine angle without injuring the adjacent neurovascular structures. These were impossible to achieve with the older techniques utilizing a mallet and chisel. These early attempts at translabyrinthine craniotomy were met with considerable resistance. Cushing felt that, “there is no possible route more dangerous or difficult” (House and House 1964). By utilizing microscope magnification and a high-speed drill, Dr. House was able to overcome such hurdles and remove small tumors with very little risk of facial nerve injury and very low mortality for the time (House and Hitselberger 1964). In 1964, Dr. House reported 5% mortality amongst all of his surgical patients, and partial or complete preservation of facial nerve function in all 41 patients who had translabyrinthine surgery for acoustic neuroma removal (House and Hitselberger 1964).

Neurosurgeons later adopted the operating microscope and high-speed drill for suboccipital craniotomy as well. Large tumors were eventually resected by the suboccipital route with little injury to adjacent neurovascular structures and low mortality. Di Tullio et al. (1978) reported 91% success in complete tumor removal with only 12% facial nerve palsy and 3.7% mortality. More recent series report even better outcomes with 99% incidence of complete tumor removal and less than 1% mortality with the suboccipital, retrosigmoid approach (Ebersold 1992; Gormley 1997).

12.1.3 Stereotactic Radiosurgery

A major revolution in the management of vestibular schwannomas occurred with the development of the gamma knife stereotactic radiosurgical unit. In 1968, a pioneering neurosurgeon named Lars Leksell designed the first stereotactic unit to deliver gamma radiation to treat arteriovenous malformations and tumors of the brain (Leksell 1983). Stereotactic radiosurgery allows the neurosurgeon to treat intracranial lesions without the attendant risks of craniotomy including cerebellar contusion, cerebrospinal fluid (CSF) leak, and meningitis. The major disadvantage of this treatment modality is that radiosurgery fails to eradicate the tumor; rather, it only ceases the tumor growth.

In the early 1980s, Noren et al. (1983), presented some of the earliest cases of vestibular schwannomas treated with stereotactic radiosurgery. The authors initially reported reduction of the tumor size among 30% of their patients and arrest of growth among 60%. These data were viewed with considerable skepticism at the time, since radiosurgeries did not result in tumor removal, only in the control of tumor growth. To confirm that the treatment truly controls tumor growth, 10–20 years of follow-up is necessary. These long-term follow-up data are now beginning to emerge. As a result, stereotactic radiosurgery now has an important role in the management of vestibular schwannomas (Leksell 1983; Linskey et al. 1990; Lunsford et al. 1990, 1992; Flickinger et al. 1991, 1993, 1996, 1998; Lunsford and Linskey 1992; Kondziolka and Lunsford 1993; Kondziolka et al. 1998; Flickinger 1999).

As we enter a new millennium, the neurosurgeon and neurotologist have four important tools to treat vestibular schwannomas. A judicious combination of the suboccipital craniotomy, translabyrinthine craniotomy, middle fossa approach, and stereotactic radiosurgery represents the complete armamentarium in the modern management of vestibular schwannomas.