Facial Nerve Schwannomas Mimicking as Vestibular Schwannomas

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Abstract

Objective The objective of this study was to identify preoperative and intraoperative findings that may aid in distinguishing facial nerve schwannomas (FNS) from vestibular schwannomas (VSs), particularly in cases limited to the internal auditory canal (IAC) and cerebellopontine angle (CPA).

Study Design This was a retrospective study.

Setting This study was set at a Tertiary Referral Center.

Patients Seventeen cases from October 2002 to July 2015 with an IAC/CPA mass presumed to be a VS who were found to have a FNS intraoperatively.

Main Outcome Measures The main outcome measures included preoperative presentation, intraoperative findings, and subsequent intervention.

Results Preoperative hearing loss and imbalance were seen in 70.5 and 64.7%, respectively. Suspicious intraoperative findings included: facial nerve incorporated intimately with the tumor capsule in 12 cases; spontaneous action potentials noted while drilling the bony IAC in 3 cases; and action potentials noted on stimulation of the entire tumor capsule in 10 cases. The mean long-term facial function was House–Brackmann grade II and the mean length of follow-up was 4.86 years.

Conclusion FNSs are rare and may be difficult to distinguish from VS preoperatively. Surgical findings that should raise concern include spontaneous action potentials during drilling the bony IAC, absence of a plane of dissection between the facial nerve and tumor, or stimulation of the tumor capsule.

Keywords

- facial nerve schwannoma
- electro-neuronography
- facial nerve monitoring

Introduction

Vestibular schwannomas (VSs) are the most common tumor of the internal auditory canal (IAC) and cerebellopontine angle (CPA), comprising 80 to 90% of the cases. The differential

diagnosis for masses of the IAC/CPA includes meningiomas, epidermoids, lipomas, metastases, and other unusual lesions, such as intra-axial tumors. Conversely, facial nerve schwannomas (FNSs) are very rare tumors of the temporal bone. By definition, they are encapsulated tumors originating from

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Schwann cells. These lesions are more commonly seen at the perigeniculate region, followed by tympanic and mastoid segments, although they may occur anywhere along the course of the facial nerve. These rare tumors account for approximately 1% of all CPA tumors.¹

A high index of suspicion is required in the diagnosis of a FNS. Although facial spasms and progressive or recurrent weakness are highly characteristic, many patients will not exhibit these classic symptoms. This is especially true for tumors limited to the IAC/CPA. In many cases, these tumors are clinically indistinguishable from VS, often presenting with unilateral hearing loss, tinnitus, and vertigo. Radiological imaging may provide little benefit in the differentiation of the two entities, especially if the fallopian canal is not involved or skip lesions are not present. Occasionally, the diagnosis is not made until surgical intervention is undertaken. This circumstance can create a dilemma for the surgeon and the patient. Traditional therapeutic recommendation included radical surgical resection with the placement of an interposition nerve graft. This would leave the patient with a severe facial paresis and irreversible and undesired cosmetic defect. In 2011, Wilkinson et al reported 79 FNS treated at the House Ear Institute over a 30-year time period, describing how the treatment algorithm has evolved.² FNS are slow growing, rarely invasive tumors. For this reason, surgical resection should be reserved for patients with facial paresis, House–Brackmann (HB)³ grade IV or worse, since patients without such compromise would likely lose greater facial nerve function and have poorer cosmesis after surgical resection. Alternatively, in patients with good preoperative facial nerve function, observation, bony decompression, and/or stereotactic radiosurgery may be considered.²

Although the literature describes greater than 600 cases of FNS, these are unusual lesions and their rarity poses a challenge in diagnosis and treatment.¹ Our aim is to add to the current literature and further elucidate preoperative findings that may aid in distinguishing FNS from VS, particularly in cases limited to the IAC/CPA. In addition, we describe surgical considerations, intraoperative findings, and postoperative outcomes.

Materials and Methods

Approval was obtained from the St. John Providence Health System's Institutional Review Board. A retrospective chart review was then conducted from January 2004 to July 2015 of all skull base cases. Patients with a presumed diagnosis of VS who were found to have a FNS at the time of surgery were included in our cohort. Patients younger than 18 years old were excluded. The following data were collected: demographic characteristics, clinical presentation, imaging, audiometry, electroneuronography (ENOG), treatment, surgical findings, and pre- and postoperative facial function as assessed by House–Brackmann scale. Descriptive statistics were used to characterize the patient population using Microsoft Excel.

Results

A total of 745 skull base cases were reviewed; 17 patients met inclusion criteria. The mean age at diagnosis was 47.5 years. The male-to-female ratio was 7:10. The ratio of right to left sidedness was fairly equivalent, with the left side involved in 52.9%. Preoperative symptoms are presented in **Table 1**. Preoperative hearing status was not available for four patients, and two patients were noted to have profound deafness on presentation. Sixteen patients (94%) underwent preoperative ENOG. Preoperative ENOG revealed a mean weakness of 17% (range, 0–85%). Four patients were noted to have a greater than 30% asymmetry (23.5%). Only three patients underwent ENOG testing postoperatively with a mean weakness of 68.3% (range, 31–100%).

All tumors were located in the IAC, 4 were limited to the IAC, while 13 (76.4%) had CPA extension. Preoperative imaging consisted of magnetic resonance imaging (MRI) scanning with and without gadolinium contrast, see **Figs. 1 and 2**. Preoperative imaging showed a mean tumor size of 1.7×1.6 cm. The overall mean time to treatment was 7.5 months. Surgical approach varied among patients based on tumor size and hearing status with a translabyrinthine approach for 14 patients, a retrosigmoid approach for 2 patients, and a middle fossa craniotomy approach for 1 patient. Suspicious intraoperative findings are listed in **Table 2**, and include facial nerve incorporated intimately with the tumor capsule in 12 cases; action potentials noted on stimulation of the entire tumor capsule in 10 cases; and spontaneous action potentials noted while drilling the bony IAC in 3 cases. Regarding tumor resection, three patients underwent complete resection with sacrifice of the facial nerve, while tumor debulking was performed in nine patients. Facial nerve decompression alone was performed in four patients. Decompression in conjunction with an incomplete tumor resection was performed in four patients. Decompression alone required no further treatment in two of the four patients; one patient was lost to follow-up and one patient required stereotactic radiosurgery 10 years later secondary to tumor growth. Of the three patients who underwent complete tumor resection, a greater auricular nerve graft was performed in one patient and cranial nerves VII to XII anastomoses in the remaining two patients. Long-term facial nerve function for grafted patients was HB grades II, IV, and VI, respectively.

The overall mean long-term facial function was HB grade II, and the mean length of follow-up was 4.86 years. The mean

Table 1 Preoperative symptoms

Symptom	N (%)
Facial weakness/spasm	1 (5.8)
Paresthesia	4 (23.5)
Hearing loss	12 (70.5)
Tinnitus	7 (41.1)
Imbalance	11 (64.7)

Fig. 1 (A) Axial T1-weighted, postcontrast image reveals a homogenous hyperintense lesion in the IAC extending into the CPA. (B) Coronal T1-weighted, postcontrast image depicting the same lesion. CPA, cerebellopontine angle; IAC, internal auditory canal.

tumor size at most recent follow-up was 4.27 mm. Regarding hearing function, the 14 patients who underwent translabyrinthine approach lost all hearing in the operated ear. Preoperative hearing was preserved in the two retrosigmoid patients. Preoperative and postoperative audiometric findings were not available for the middle cranial fossa patient.

Discussion

Widening of the fallopian canal on computed tomography or contrast enhancement on MRI of the facial nerve, particularly at the geniculate ganglion is concerning for a tumor of facial nerve origin.⁴⁻⁷ Conversely, the particularly unique and challenging FNS are those that are located within the IAC or CPA. Fagan et al described MRI findings of posterior displacement of the tumor relative to the IAC in four cases of FNS limited to the CPA.⁸ According to most other reports in the literature, there are no clear features to distinguish these

tumors from a VS radiologically when limited to the IAC or CPA.^{6,9}

Since the treatment algorithm and timing of surgery are quite different compared with that of a VS, the possibility of a FNS must always be considered. FNSs mimicking VSs have been reported to occur in 1 to 2% of VS.^{1,10-12} These patients may not present with the classic symptom of unilateral facial paresis/paralysis, instead they may complain of more classic VS symptoms such as unilateral tinnitus, hearing loss, and vertigo. In 2002, Sherman et al described five FNSs limited to the IAC/CPA, and only one such patient exhibited facial paresis.⁹ In this scenario, preoperative auditory brain stem response (ABR) and ENOG testing may be helpful in distinguishing these tumors from VS. A relatively abnormal ENOG with a normal ABR is suggestive of a FNS.^{4,5,7} Interestingly, in our cohort, preoperative ENOG revealed a mean weakness of 17%, which is neither particularly abnormal nor suggestive of a facial nerve lesion, although ENOG asymmetry was 30% or

Fig. 2 (A) Axial T1-weighted, postcontrast image depicting an enhancing lesion in the IAC with CPA extension. (B) Coronal T1-weighted, postcontrast image depicting the same lesion. CPA, cerebellopontine angle; IAC, internal auditory canal.

Table 2 Intraoperative findings of IAC/CPA facial nerve schwannomas

Surgical finding	N (%)
Lack of plane between facial nerve/tumor	12 (70.5%)
Action potential with stimulation of tumor capsule	10 (58.8%)
Spontaneous action potential with bony IAC drilling	3 (17.6%)

Abbreviations: CPA, cerebellopontine angle; IAC, internal auditory canal.

greater in four patients (23.5%) which may be of clinical relevance. Further studies of ENOG and FNS would be of interest, since it could provide potential insight into this rare diagnosis.

The traditional recommendation for the management of FNS was tumor resection and nerve grafting; however, it is now recognized that resection should be reserved for patients with already poor facial nerve function.² In the subset of patients with lesions in the IAC/CPA, mimicking a VS, surgical resection is often planned in cases of tumor growth or brain stem compression. In many patients with IAC/CPA FNSs, it is often not recognized as such until the time of surgery, due to the rarity of FNSs in the IAC/CPA region.¹³ Despite this, certain early intraoperative findings may assist the surgeon in confirming the diagnosis of a FNSs and may prompt the surgeon to redirect the treatment plan.

Sherman et al described an intraoperative finding of a diffusely thickened facial nerve in patients with IAC/CPA FNSs.⁹ Nadeau and Sataloff reported the finding of an inability to identify a plane of separation between the tumor and facial nerve.¹ Similarly, in our series of patients, the absence of a plane of dissection between the facial nerve and tumor was the most common intraoperative finding (70.5%).

Alternatively, our study describes two novel intraoperative findings that may suggest a FNS as opposed to the typical VS. These findings involve the use of facial nerve electromyographic monitoring during surgery. Improved postoperative facial nerve outcomes with the use of intraoperative monitoring has been repeatedly demonstrated.^{14–19} This has led to the routine use of monitoring during VS surgery over the past two decades and this has also been advised by the National Institutes of Health.²⁰ Consequently, we use facial nerve monitoring and Kartush nerve stimulating microdissecting instruments during surgeries that place the facial nerve at risk. With the use of these instruments, we noted facial nerve compound action potentials by simply touching the instrument to the edge of the tumor capsule in 10 patients (58.8%) in our cohort. Typically, with VS, stimulating the tumor capsule would not cause facial nerve stimulation—this difference may be the first clue that a FNS is encountered. Similarly, in three cases (17.6%), there were observed spontaneous action potentials during the drilling of the bony IAC. The presence of either of these intraoperative findings may spark concern for a FNS.

Conclusion

FNSs are rare tumors of the temporal bone, and when located in the IAC/CPA, they are often clinically indistinguishable from VSs. Since the incidence of VSs predominate as compared with FNSs, it is often difficult to identify and distinguish a FNS from a VS in patients without facial nerve paresis. Furthermore, the treatment algorithm and associated morbidity for VSs versus FNSs are significantly different, thus proper diagnosis is key in maximizing clinical outcomes. Surgical approach for tumor resection is not without morbidity, and the risks and benefits must be considered before surgery is warranted.

Our small sample size limited statistical analysis; however, our data suggest that preoperative ENOG and certain intraoperative findings may play a role in distinguishing FNSs. The facial paresis and paralysis that may result from tumor resection can be devastating to the patient and must be considered regarding discussion of therapeutic options and timing of surgery. Abnormal ENOG, unusually adherent tumor without a clear surgical plane between the facial nerve, or abnormal stimulation of the facial nerve through tumor capsule, or during bony IAC drilling may suggest a FNS. Since identification of an IAC/CPA tumor as a FNS is paramount, attention should be placed on the preoperative recognition of these tumors. Studies involving ENOG and IAC/CPA masses may help differentiate between these entities during the preoperative period and deserves further study.

Conflict of Interest

There is no financial disclosure or conflict of interest.

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