Sporadic unilateral vestibular schwannoma in the pediatric population

Clinical article

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Object. Vestibular schwannomas (VSs) are rare in the pediatric population. Most often, these lesions manifest as a bilateral disease process in the setting of neurofibromatosis Type 2. Even in the absence of additional clinical diagnostic criteria, the presentation of a unilateral VS in a young patient may be a harbinger of future penetrance for this hereditary tumor syndrome.

Methods. The authors retrospectively reviewed the charts of a cohort of 7 patients who presented with apparently sporadic, unilateral VSs. These patients had previously undergone surgery via translabyrinthine, retrosigmoid, or combined approaches. Clinical outcomes were reviewed with emphasis on facial nerve function and follow-up for signs and symptoms of a heritable disorder.

Results. All patients underwent microsurgical resection in a multidisciplinary effort by the senior authors. The average tumor size was 4.57 cm, with an average duration of symptoms prior to definitive diagnosis of 31.2 months. The tumor size at the time of presentation followed a trend different from reports in adults, while the duration of symptoms did not. At a follow-up average of 6.3 years (range 1–12 years), 100% of patients demonstrated good facial function (House-Brackmann Grade I or II). No patient in this cohort demonstrated symptoms, objective signs, or genetic analysis indicating the presence of neurofibromatosis Type 2.

Conclusions. Diagnosis and management of sporadic, unilateral VSs in children is complicated by clinical presentations and surgical challenges unique from their adult counterparts. Careful consideration should be given to a heritable genetic basis for sporadic unilateral VS in the pediatric population. Results of genetic testing do not preclude the necessity for long-term follow-up and systemic investigation. In patients who present with large tumors, preliminary experience leads the authors to suggest that a combined retrosigmoid-translabyrinthine approach offers the greatest opportunity for preservation of facial nerve function. (DOI: 10.3171/2009.3.PEDS08434)

Key Words • facial nerve • neurofibromatosis • pediatric neurosurgery • retrosigmoid approach • translabyrinthine approach • vestibular schwannoma
tosomal dominant condition NF2, a retrospective analysis indicates that 10–25% of VSs are initially unilateral at presentation. From an alternative vantage point, the risk of NF2 in patients with an apparently sporadic unilateral VS is extremely small except for those in the pediatric age group. The possibility of NF2 must therefore be taken into consideration in the evaluation and treatment of unilateral tumors in children, as the chance that a contralateral tumor could develop may influence the management of the primary side. In the present study, we reviewed the presentation, surgical approach, and outcome in a consecutive series of pediatric patients harboring apparently sporadic, unilateral VSs. Special attention was given to analysis of facial nerve preservation in addition to follow-up for signs and symptoms of NF2.

Methods

Patient Population and Symptoms

Of all the cranial base operations performed by the senior authors at Loyola University Medical Center between April 1996 and May 2007, 7 children were identified with apparently sporadic, unilateral VSs. Five patients were girls and 2 were boys. The mean age of the patients was 15.1 years, ranging from 9 to 18 years. The average duration of symptoms was 31.2 months. The most common presenting symptom was hearing loss, which was reported in all but 1 patient. Six patients (86%) presented with balance or gait instability, and 3 (43%) presented with tinnitus. Facial numbness and paresthesias were noted in 2 patients (29%). Nausea and vomiting occurred in only 1 patient. Of the 7 patients, 1 reported overt difficulty swallowing and abnormal phonation, indicative of lower cranial nerve dysfunction. Objective presenting signs included papilledema, nystagmus, partial abducens nerve palsy, abnormal corneal reflex, facial weakness, and cerebellar ataxia.

Magnetic resonance imaging provided for preoperative determination of tumor dimensions and impingement on surrounding cranial structures (Fig. 1). Maximal tumor diameter ranged in size from 1 to 7 cm, with a mean size of 4.57 cm. Six of the 7 tumor measurements were > 4 cm. Five patients in this group were identified with preoperative radiographic evidence of hydrocephalus.

Determination of Surgical Approach

Three different surgical techniques were used for tumor resection: the translabyrinthine, the retrosigmoid, and the combined retrosigmoid-translabyrinthine approach. The selection criteria used to determine the surgical approach was based on several anatomical factors including the size of the tumor, extension of the tumor into the IAM, and the distance between the jugular bulb and the superior petrosal sinus. Additionally, the surgical plan was influenced by preoperative hearing functionality as assessed with audiometric studies. In patients with smaller tumors extending into the fundus of the IAM or if the patient had a wide-angled petrous apex (a normal or enlarged space between the jugular bulb and the superior petrosal sinus), a translabyrinthine approach was considered. A purely retrosigmoid approach was considered for medially based tumors. For the majority of the patients in this cohort, it was recognized that very large tumor sizes (most commonly > 4 cm) necessitated a combined approach to overcome the inherent individual limitations of each.

Surgical Technique

One translabyrinthine, 1 retrosigmoid, and 5 combined translabyrinthine-retrosigmoid approaches were used in the surgical management of 8 tumors in the same number of children. The multidisciplinary cranial base team composed of a pediatric neurosurgeon, a neurotologist, a neuroanesthesiologist, and intraoperative electrophysiological monitoring personnel was assembled in much the same fashion as for the adult skull base tumors routinely treated at this tertiary care academic medical center.

The combined translabyrinthine-retrosigmoid approach, uniquely developed by the lead authors, seeks to incorporate the strengths of the 2 separate approaches to maximize the likelihood of facial nerve preservation when performing gross-total removal of very large tumors. The workbench was completed first, allowing for both aspects of the 2 individual approaches. Sequentially, the translabyrinthine isolation of the IAM and the lateral aspect of the facial nerve were performed prior to durotomys for the continuation of the retrosigmoid portion of the exposure. Ultrasonic aspiration and debulking of the tumor mass were performed, initially defining and later maintaining the surgical plane between the tumor and the cerebellum. Perseverance eventually led to the establishment of a similar plane between the lesion and both the brainstem and cranial nerves. Paramount to the success of the dissection was the early identification of the facial nerve medially to facilitate a medial to lateral dissection of the tumor away from the facial nerve and other neural structures. After tumor removal, attention was solely focused on the facial nerve, which was stimulated at the most proximal region of the brainstem root exit zone. Anatomical preservation of the facial nerve was achieved in all cases and further documented by a response to this stimulus. If no response was initially obtained, the current was increased in 0.1-mA increments until a response was achieved. Finally, the wound closure was performed in the usual, routine fashion.
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Outcome and Follow-Up

Several end points were observed with an emphasis on facial nerve function in addition to long-term follow-up for signs and symptoms of a heritable disorder. Relating to this, the completeness of tumor resection, intraoperative electrophysiological facial nerve activity, facial nerve response at the finale of the procedure, clinical facial nerve function using the House-Brackmann facial nerve grading scale (Table 1), complications, and genetic studies were reviewed. Additionally, review of a routine postoperative questionnaire was used to help summarize patient perception of facial function and long-term outcome. Uniform follow-up averaged 6.3 years, ranging from 1 to 12 years.

Results

Operative Procedure

The average duration of the procedure was 12.5 hours, ranging from 4 to 17 hours. In 4 patients (57%) CSF diversion procedures were necessary prior to definitive resection. Gross-total resection was achieved in all patients as determined on postoperative MR imaging studies (Fig. 2). In 1 patient, apparent tumor recurrence was treated with stereotactic radiosurgery; this patient is currently undergoing follow-up with serial imaging. Pathological analysis revealed the diagnosis of schwannoma and the absence of atypia in all specimens.

The average duration of hospital stay was 7 days, with a range of 4–20 days. The longest hospital stay reflected the complexity associated with a 2-stage operation performed over the interval of 1 week. There were no operative deaths in this cohort of patients.

Facial Nerve

In 2 patients, facial nerve weakness was noted preoperatively (House-Brackmann Grade III and VI). These patients were assumed to have had the deficit due to the direct effect of the tumor on the nerve. Routine follow-up included objective assessment in the immediate postoperative period, with questionnaire forms, and at routine clinic visits. Ultimately, all 7 patients (100%) achieved good facial function over time (House-Brackmann Grade I or II).

Postoperative Complications

There was no incidence of CSF leaks, wound infections, or hematomas in our patients, which are the common perioperative complications associated with this procedure. Two patients experienced brief postoperative episodes of psychosis attributed to corticosteroid therapy, a known adverse effect that has been demonstrated to be more variable in children. One patient experienced postoperative cerebellar and brainstem swelling, resulting in a transient ipsilateral palsy of cranial nerve IX and X. These symptoms resolved by the time of discharge and were attributed to venous congestion.

Genetic Analysis

One patient underwent a full genetic evaluation at the time of admission in light of the highest concern for NF2 in this cohort given a familial history of hearing loss and a personal history of congenital cataracts. Molecular analysis including single-strand conformation polymorphism studies and fluorescence in situ hybridization with probes directed at the 22q11 region failed to reveal any abnormality.

Discussion

The risk of unilateral tumors presenting as the first manifestation of NF2 is related to the age of the patient. It has been determined that individuals < 30 years of age are at the highest risk of developing a contralateral tumor and thereby fulfilling the diagnostic criteria necessary for NF2. According to the NIH consensus statement, these criteria are as follows: 1) the presence of a bilateral cranial nerve VIII mass visible on Gd-enhanced MR images; or 2) a first-degree relative with NF2 and either a unilateral cranial nerve VIII mass or 2 of the following: neuro-
fibroma, meningioma, glioma, schwannoma, or posterior capsular cataract or opacity at a young age. Evans et al. also reported that ~ 6% of all individuals with an apparently isolated VS are mosaic for an NF2 gene mutation, and may therefore go on to develop various features of NF2 despite an initial negative genetic analysis.

The dilemmas inherent to the treatment of VSs in patients with NF2 are well-known and are highlighted by facial nerve paralysis, in addition to a multitude of other tumors and complications. These considerations are magnified in children and can be extrapolated to the apparently sporadic VS patient population given the implications for impairment of childhood development and lifelong disability in the setting of possible future contralateral tumors. Clinical management, including the selection of operative approach and execution of surgical technique, must be aimed at minimization and deferment of these complications, as in all patients.

Although significant controversy remains regarding the optimal treatment, timing, and selection of a surgical approach, tumor size and the presence of serviceable hearing remain important, common, and consistent factors in this decision process. By exposing the areas both anterior and posterior to the sigmoid sinus, a prominent vertical and lateral exposure of the tumor is established. It was found that the retrosigmoid portion of the exposure was particularly helpful in instances of a high jugular bulb, as a solely translabyrinthine approach limits vertical exposure. In conjunction, the translabyrinthine approach allowed for early identification of the facial nerve distally, enabling its visualization at the porous acousticus and within the posterior fossa. The translabyrinthine approach also allows the most lateral portion of very large tumors to be reached, maximizing the potential for GTR. The results of facial nerve preservation may be related to the combined approach that allows for early identification of the facial nerve proximally, facilitating its immediate and frequent monitoring. Preservation of facial nerve function is paramount, especially in children, as it is often regarded as the single most important determinant of quality of life in the postoperative setting.

A combined translabyrinthine-retrosigmoid approach was most advantageous for tumors that extended to the fundus of the IAM and were very large, as was almost universally found in our cohort of pediatric patients with apparently sporadic unilateral VSs.

In a meta-analysis of 1345 adult patients, the average size of adult VS was reported as 11.8 mm. In our cohort, the average size tumor size was 4.57 cm, substantially larger than that reported in the adult literature. A similar average size tumor size was 4.57 cm, substantially larger than that reported in the adult literature. A similar

Recurrence of VS after GTR is rare. However, a single patient in our study was found to have a 1-cm recurrence at the time of a routine, 6-month postoperative surveillance MR imaging study. She then underwent stereotactic radiosurgery. Currently, attainment of tumor control in this patient remains to be elucidated given the short interval since the time of her treatment. The clinical behavior of recurrent VSs, such as in this child, remains unclear. Microscopic fragments of the tumor may remain viable after GTR, and if given proper vascularization, propagate. The role of stereotactic radiosurgery after microsurgery has been explored and shown to be an effective measure in the treatment of both tumor regrowth and for use after subtotal resections. Although the goal of microsurgery is ultimately total tumor removal, the benefit of stereotactic radiosurgery in children with very large tumors may rest in the planned combination of dual modality treatment.

Conclusions

Although children typically present with larger VSs than adults, it is possible to preserve good long-term facial nerve function. A combined translabyrinthine-retrosigmoid approach provides excellent results in terms of GTR and ability to meticulously preserve the facial nerve. Follow-up for a heritable genetic basis should be undertaken in all cases of apparently sporadic unilateral VS in the pediatric population.

Disclaimer

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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