Intratemporal Facial Nerve Schwannoma in a 5 Year Old Girl: A Therapeutic Dilemma

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Facial nerve schwannomas (FNS) are uncommon benign tumors that can arise from all segments of the facial nerve. Intratemporal schwannoma is a rare tumor in pediatric patients. Clinical manifestations may vary from none to facial paralysis, deafness, tinnitus, vertigo and an ear mass. We present the case of a child with an intratemporal FNS, and provide a detailed description of the diagnostic characteristics and the treatment dilemmas.

PATIENT DESCRIPTION

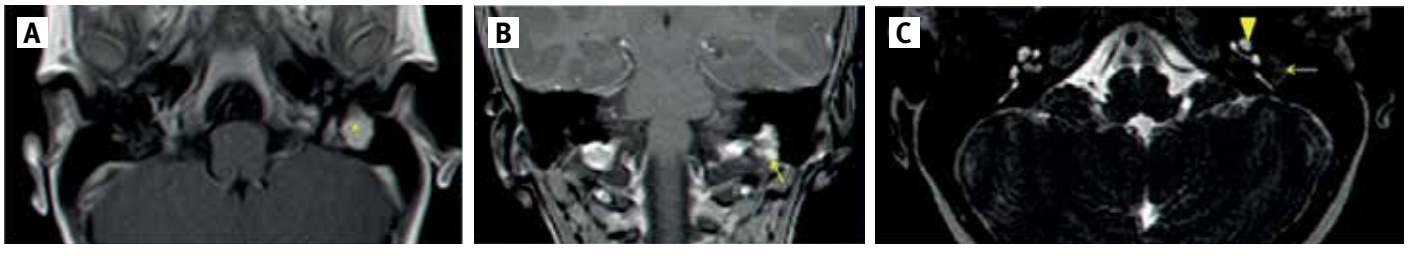
A 5 year old girl with a history of slowly progressive peripheral facial nerve palsy lasting for more than 2 years was referred for evaluation. The parents reported worsening of the paralysis in the months prior to presentation. Her medical and surgical history was otherwise unremarkable.

Physical examination revealed left peripheral facial nerve palsy, House-Brackmann (HB) grade 5. The rest of the physical examination was normal. An audiogram recorded normal hearing. Magnetic resonance imaging (MRI) and computed tomography (CT) scans demonstrated an enlarged fallopian canal with a gadolinium-avid lesion extending from the mid-tympanic segment to the stylomastiod foramen compatible with a schwannoma [Figure 1]. No stigmata of neurofibromatosis were present.

During surgery the tumor was found to extend from proximal to the cochleariform process to the neck just distal to the stylomastoid foraman. In its mastoid segment the tumor breached the fallopian canal towards the jugular bulb, and in its tympanic region reached the incus and pushed the stapes downwards towards the promontorium; the bulk of the tumor in this area prohibited preserving the posterior external auditory canal, necessitating creating a canal wall down cavity. The tumor was completely excised and nerve continuity reestablished with a sural nerve cable graft, supported by vascularized rotated temporalis muscle flaps. Frozen section microscopic studies established the health of the facial nerve edges. Hearing was reconstructed with a cartilage type 3 tympanoplasty. Facial nerve function returned to HB 4 but later deteriorated to HB 5. Therefore, at months 27 and 35 following surgery, she underwent a two-staged facial reanimation consisting of a cross-face nerve graft using a sural nerve followed by implantation of a gracillis muscle free flap.

Four years after surgery there is no evidence of a residual tumor. Although some asymmetry is noted in the resting state, dynamic facial nerve function is HB 2, and on audiometry a left conductive hearing loss with speech reception threshold is 35dB. Hearing on the right side is normal. Aeration of the residual middle ear and mastoid created a pseudo-posterior canal wall allowing exposure to water.

Figure 1. Images of an MRI scan of the temporal bones demonstrating the characteristics and extension of tumor. **[A]** T1-weighted gadolinium-enhanced axial section depicting a gadolinium-avid tumor in the fallopian canal. **[B]** T1-weighted gadolinium-enhanced coronal section. Tumor extended through the stylomastoid foramen to the neck. **[C]** T2-weighted axial cut, demonstrating the relationship of the tumor (arrow) at the second genu to the cochlea (arrowhead) and posterior semicircular canal



COMMENT

Although rare, FNS are part of the differential diagnosis of progressive peripheral nerve weakness in a child. The majority of FNS cases involve an intratemporal segment such as the geniculate ganglion, labyrinthine and internal acoustic canal [1]. However, tumor involving the intratemporal nerve in a pediatric patient is extremely rare. The impact of facial nerve palsy and hearing loss on social development and academic and language-related skills must be accounted for in the management of pediatric patients.

Diagnosis is based on history, physical examination and imaging. Application of facial nerve function tests in young children is difficult as they can be uncomfortable or painful. Most commonly the degree of facial nerve dysfunction will be established based solely on physical examination. The appearance of FNS in imaging is heterogeneous [1]. On CT, intratemporal FNS are seen by sharply defined bony enlargement of the fallopian canal, and in the middle ear and mastoid they may be seen as a multi-lobulated mass. On MRI, intratemporal FNS appear as a well-circumscribed fusiform, homogenously enhancing mass along the course of the intratemporal facial nerve. Most are isointense or hyperintense on T2-weighted series compared to brain tissue. Erosion of mastoidal air cells may mimic the MRI appearance of an invasive tumor. Progressive peripheral facial nerve palsy is best investigated with an MRI scan [1].

There are no accepted guidelines for the management of FNS. Recently, stereotactic radiosurgery was added as an option for treating FNS, but it is mostly reserved for small tumors with HB 1-3 [2]. Surgery is indicated once significant clinical manifestations are present, such as facial paralysis, or a large tumor compromising adjacent structures, such as brainstem compression or hydrocephalus [3]. However, since this is a slow-growing benign tumor with a slow-developing morbidity, determining the appropriate timing for surgical intervention in cases with minimal clinical manifestations can be difficult. In these cases, some authors advocate early intervention before greater nerve damage ensues and for better postoperative results. Others advocate expectant observation, with serial electroneuronography (ENoG) and MRI, and intervening once serial ENoG tests show a 50% decrease in the compound action potential [3], or before facial nerve function deteriorates to HB grade of 5 [4]. Expectant management is rationalized by the fact that best reported results of facial nerve function following tumor resection and nerve reconstruction are HB grade of 3-4 [4]. Delaying intervention until all nerve function is lost may result in facial muscle atrophy that will limit reconstruction options.

Once a decision for surgical intervention has been made, the location of the tumor and the hearing level determine the surgical approach. For patients with functional hearing a middle cranial fossa approach may be used and possibly aided by a mastoid approach for bony decompression of the facial nerve. A trans-labyrinthine approach may be used for patients with a tumor of the internal acoustic canal without functional hearing for better exposure during surgery [3]. The exact proximal reach of the tumor should be defined by imaging. Extracranial tumors can be managed with tympanomastoidectomy, most commonly canal wall up procedures.

A few reports support the possibility of preserving the facial nerve intact during extirpation with better postoperative results compared to facial nerve reconstruction [4]. In the case presented, during microsurgery and pathological study the tumor completely replaced the facial nerve, requiring complete transection and cable grafting.

Primary anastomosis of the cut nerve provides the best chance for good postoperative function. In the vast majority of cases the cut ends of nerves cannot be approximated without an intervening graft. The most commonly used grafts are the greater auricular nerve and the sural nerve. The graft is best supported by vascularized local flaps [5]. These flaps may compromise the mechanism of conduction of hearing. Harvesting a lengthy graft that can circumnavigate the middle ear and ossicles may prevent postoperative conductive hearing loss.

CONCLUSIONS

Intratemporal facial nerve schwannomas are rare in childhood but should be sought in a patient with progressive facial nerve palsy. Management requires consideration of the significant impact of hearing and facial nerve functions on social, academic and language development. Surgery should be timed according to the facial nerve function and potential damage to adjacent structures. Proper design and placement of a nerve graft for reanimation can reduce the risk of postoperative conductive hearing loss.

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"Pursue some path, however narrow and crooked, in which you can walk with love and reverence"

Henry David Thoreau (1817-1862), American essayist, poet, philosopher, abolitionist, naturalist, and historian