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Review

Facial Nerve Tumors in Children: Two Clinical Cases and a Review of the Literature

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We provide an extensive review of clinical features, diagnosis, and treatment of primitive facial nerve tumors in children, and report 2 recent personal observations. We conducted a comprehensive literature search through PubMed, Medline, and ScienceDirect and collected information on patients' age, symptoms, tumor types and sites, diagnostic procedures, surgical approaches, and outcomes. Overall, we reviewed 26 pediatric cases from 20 papers. About 69.2% of children presented with some degree of facial palsy. Other symptoms included hearing loss, dizziness, and tinnitus. 84.6% of tumors were schwannomas, followed by meningiomas, epithelioid hemangioendothelioma, and germ cell tumors. The geniculate ganglion was the most commonly affected segment of the facial nerve. A total of 92.3% of children received surgery as complete or partial tumor resection. Facial nerve function improved in 26.9% of children. No tumor recurrence was reported. Facial nerve tumors are extremely rare in children but should be considered in the differential diagnosis of facial palsy, even in newborns. Audiometric and radiologic examinations are necessary; radiologic imaging allows to determine tumor localization, and the correct surgical approach surgery is suggested in almost all cases.

KEYWORDS: Children, neuroimaging, neurotology, otoneurology, temporal bone

INTRODUCTION

Primitive facial nerve tumors (FNTs) are benign, slow-growing neoplasms of the facial nerve (FN). Facial nerve tumors can be intrinsic, that is, arising from the nerve fibers themselves, or extrinsic, that is, from other tissues adjacent to the nerve.[1] Facial nerve tumors can originate anywhere along the course of the FN and are subdivided into intracranial, intratemporal, and extratemporal. Intracranial FNTs are located in the cerebellopontine angle (CPA) in close relationship with the brainstem; intratemporal FNTs grow inside the internal auditory canal (IAC) segment, the geniculate ganglion (GG) or the mastoid segment of the nerve; extratemporal FNTs occur after the stylomastoid foramen and involve the parotid branches of the FN. The most common symptoms of FNTs are facial palsy, hearing loss, tinnitus, and dizziness, although many tumors can be asymptomatic.[2-4]

Facial nerve tumors are very rare, with only 100 reports in the literature since 1930.[2,3] Facial nerve schwannomas, which are neoplasms born from the myelin-producing Schwann cells, are the most common FNTs accounting for about 5% of FN palsies.[5] In a study of 600 temporal bones, Saito and Baxter[6] found a 0.83% incidence of FN schwannomas. Because of their rarity and nonspecific symptoms, FNTs are often misdiagnosed. Most reviews focus only on specific FNT types, locations, or symptoms.[2,7-10] Individual reports of pediatric FNTs are rare, and no reviews are available. Therefore, our primary aim was to fill the gap in the literature by reviewing the clinical presentation, diagnostic evaluation, treatment, and outcomes of FNTs in children. Our secondary aim was to report 2 recent cases of pediatric FNTs from our practice.

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METHODS

Literature Review

We used the following key terms in our search: "facial nerve tumours," "paediatric facial schwannoma," "paediatric facial nerve paralysis," and "paediatric facial nerve tumours." The databases examined included PubMed, Medline, and ScienceDirect. We only included manuscripts written in English. Cases of neurofibromatosis and intra- and extratemporal lesions not involving the FN were excluded. Facial nerve tumors studies where adult and pediatric groups could not be differentiated were also excluded. Four hemangioma cases in children were not included because the FN appeared unaffected or was not the primary site of the tumor.[11-13] We selected 20 papers published between 1965 and 2019, of which 13 were case reports, 6 were retrospective studies of case series, and 1 was an overview of pediatric peripheral FN palsy. In these 20 papers, we identified 26 patients aged less than 18 years old who were affected by FNTs. We used the House–Brackmann (HB) facial nerve grading system to evaluate the degree of FN palsy. This system has 6 grades, spanning from grade I (normal function) to grade VI (total paralysis). We reviewed information on symptoms, FN status, types of treatment, and clinical outcomes. We could not perform a meta-analysis due to the small number of articles and patients.

Case Reports

Two patients were admitted to the ENT department of the Meyer Children's Hospital in Florence, Italy. We assessed the degree of hearing loss and the FN status along with neurological status. Diagnostic work-up always included gadolinium-enhancement magnetic resonance imaging (MRI) and computed tomography (CT). One patient received genetic testing for neurofibromatosis. Both patients underwent surgical treatment.

RESULTS

Epidemiology

The mean age of children diagnosed with an FNT was 7.3 years (2 months to 16 years). There were 11 females and 11 males. The gender and age of the remaining 4 children were not reported by Grinblat et al.[14] Twenty-two (84.6%) children had schwannomas (5 of them had a schwannoma in the parotid gland, 19.2%), 2 (7.7%) had meningiomas, 1 (3.8%) had epithelioid hemangioendothelioma, and 1 (3.8%) had a germ cell tumor (Table 1).

Clinical Presentation

Facial nerve palsy was the most common clinical presentation of an FNT in children (18/26, 69.2%). Six children (23.1%) had complete FN paralysis (HB grade VI), 4 (15.4%) had HB grade III facial palsy, 3 (11.5%) had HB grade V facial palsy, and 2 (7.7%) had HB grade IV facial palsy. Eight children (30.8%) had normal FN function. The FN function was not reported for 3 (11.5%) children. The age of palsy onset was reported for 16 children, averaging 5.1 years. In 2 patients reported by Kim et al[15] and Cushing et al,[16] studying old photographs and videos helped establish the true age at palsy onset, which was earlier than reported by parents. Facial nerve conduction was also evaluated in some patients in addition to visual examination and revealed abnormal response latencies and amplitudes.[15-19] Some degree of hearing loss was the second most common symptom (8/26, 30.8%) as assessed by audiometric tests. Three children (11.5%) had vestibular problems like dizziness, unsteady gait, or vomiting. One child (3.8%) had tinnitus, 1 had a tender mass in the external auditory canal (EAC), 1 had a middle ear cholesteatoma, and 1 had recurrent otitis media. One child with a germ cell tumor had bilateral sixth nerve palsy in addition to FN palsy. Five children (19.2%) with suspected parotid gland neoplasms had painless masses of various sizes at the level of the parotid gland.[20-24] Their FN function was normal. The symptoms above were not mutually exclusive and could co-occur.

Imaging

Most children underwent a combination of MRI and CT examinations (16/26, 61.5%). Four children (15.4%) underwent a CT examination only. Two children (7.7%) with parotid masses underwent MRI and ultrasound examinations. One child (3.8%) with a parotid mass had an ultrasound examination only. Computed tomography of some intratemporal FNTs revealed enlarged fallopian canal with some bone erosion (Table 2).[15,25,26] Magnetic resonance imaging usually revealed enlarged FN, with intratemporal schwannomas enhancing with contrast and appearing hypointense on T1-weighted images and hyper- or isointense on T2-weighted images.[25] Ultrasound revealed well-circumscribed homogenous intraparotid masses with hypo- and isoechoic patterns.[21,23] One meningioma enhanced with contrast and appeared isointense on T1-weighted MRI scans.[27] However, due to its limited spread to the CPA, it could not be distinguished from a schwannoma by a dural tail. The FN was asymmetrically enlarged on CT, but there was no characteristic petrous bone hyperostosis. Moskowitz et al[28] reported an epithelioid hemangioendothelioma with irregular edges and adjacent bone erosion on CT. The largest proportion of FNTs (11/26, 42.3%) involved the GG, either alone or with adjacent regions. Two FNTs (7.7%) were limited to the IAC. Five schwannomas (19.2%) were found on the extratemporal parotid branches of the FN. Thirteen FNTs (50.0%), including 4 schwannomas reported by Grinblat et al[14] involved multiple segments.

Treatment

Surgery was the most applied therapeutic approach (24/26, 92.3%). In 23 patients (88.5%), FNTs were resected completely, and partially in 1 patient (3.8%) with debulking of the EAC portion.[26] The retroauricular transmastoid approach extended with middle fossa craniotomy was the most common technique used in 10/23 (43.5%). Superficial parotidectomy was performed in all 5 intraparotid schwannomas (21.7%). A translabyrinthine approach was performed in 2 patients (8.7%). Other approaches were retrosigmoid, transotic, transotic-transparotid, transcervical, suboccipital, and subtotal petrosectomy. Depending on the segment of the FN resected, nerve continuity was restored via hypoglossal anastomosis or nerve graft. Grafting was not necessary for the intraparotid schwannomas, with nerve continuity remaining intact in at least 4 out of 5 children.[20-23] Watchful waiting approach with regular MRI with gadolinium and neurotologic examinations was applied in the FN schwannoma case reported by Cushing et al.[16] The treatment approach to the germ cell tumor in a 2-month-old child was not reported.[29]

Tissue Pathology

Fine-needle aspiration cytology (FNAC) was performed preoperatively in 5 intraparotid schwannomas,[18,20-24] providing a correct diagnosis only in one case reported by Coraglia et al.[22] Stromal characteristics of cells revealed by FNAC were suggestive of pleomorphic adenoma in 2 cases where a parotid mass was observed. Still, this diagnosis was changed to intraparotid schwannoma intra-operatively.[23,24] Transcanal biopsy was performed in a child with a mass in the EAC

Table 1. Facial Nerve Tumors in Children: Overview

Author, Year	Patient Age and Sex	Pathology	Site	Age of Palsy onset	Other Symptoms Apart from Facial Palsy	Treatment	Nerve Reconstruction	FN Function, HB Grade		Outcome and Follow-Up
								Preop	Postop	
Money and Halliday, 1965	7 y, M	Schwannoma	NA	6 y	Mild CHL, unsteady	TMA	VII-XII	NA	NA	"Nearly normal facial expression" in 1 year; no recurrence in 4 years
Gonzales-Pardo et al, 1980	16 y, F	Schwannoma	CPA	16 y	Dizziness, unsteady gait, vomiting	SC	VII-XII	NA	NA	HL, residual facial palsy, transient nerve V and VI impairment
Chinski et al, 1997	13 y, M	Schwannoma	GG-IAC	13 y	–	TMA-MFA	Auricular nerve graft	VI	VI	Mild CHL, no recurrence in 10 months
Van Den Abbeele et al 1999	2 m, M	Schwannoma	GG	2 m	–	MFA	Graft/ VII-XII	VI	IV	No recurrence in 4 years
	6 m, M	Schwannoma	GG-mastoid	6 m	Otitis media	MFA	Graft/ VII-XII	III	IV	No recurrence in 2 years
	15 y, M	Schwannoma	GG	14 y	CHL, chole	TMA	Graft	VI	IV	No recurrence in 2 years
Liu and Fagan, 2001	3 y, M	Schwannoma	GG-IAC	3 y	–	TLA	Auricular nerve graft	VI	III	HB grade III in 6 years
Kim et al, 2003	13 m, F	Schwannoma	GG-mastoid	3 m	Mild HL	TM-SLA	Sural nerve graft	III	IV	Worsened hearing, no stapedial reflex, no recurrence in 24 months
	17 m, M	Schwannoma	GG-mastoid	7 m	Mild HL	TM-SLA	Sural nerve graft	III	IV	Worsened hearing, no recurrence in 22 months
Ulku et al, 2004	6 y, F	Schwannoma	IAC	6 y	Severe SNHL, tinnitus, no reflex	RSA	VII-XII	IV	III	HB grade III in 5 years
Cushing et al, 2006	10 y, F	Schwannoma	GG-mastoid	8 m	–	Wait-and-scan	–	III	NA	NA
Alyono et al, 2014	11 y, M	Schwannoma	mastoid	–	Mild CHL, mass in the EAC	Debulking	–	I	II	Worsened FN function
Yafit et al, 2016	5 y, F	Schwannoma	GG-mastoid	3 y	–	TMA	Sural nerve graft	V	II	Mild CHL, no recurrence in 4 years
Grinblat et al, 2017	NA	Schwannoma	NA	NA	NA	TOA	Sural nerve graft	IV	IV	NA
	NA	Schwannoma	NA	NA	NA	TO-TPA	Sural nerve graft	VI	IV	NA
	NA	Schwannoma	NA	–	NA	TCA	NA	I	I	NA
	NA	Schwannoma	NA	–	NA	STP	NA	I	I	NA
Kumar et al, 1996	8 y, F	Schwannoma	Parotid	–	Parotid mass	SP	–	I	I	Transient postop FN weakness, resolved in 3 months
Kizil et al, 2008	7 y, M	Schwannoma	Parotid	–	Parotid mass	SP	–	I	I	No recurrence in 6 months
Coraglia et al, 2019	16 y, F	Schwannoma	Parotid	–	Parotid mass	SP	–	I	I	No recurrence in 5 years
Gumussoy and Ekmekci, 2019	9 y, M	Schwannoma	Parotid	–	Parotid mass	SP	–	I	I	NA
Khilnani et al, 2014	7 y, F	Schwannoma	Parotid	–	Parotid mass	SP	–	I	III	HB grade III in 3 months
Singh et al, 1975	14 y, M	Meningioma	IAC	10 y	Profound HL	MFA; TMA in 1 year	–	VI	NA	NA
Deep et al, 2017	4 y, F	Meningioma	CPA to mastoid	4 y	–	TLA	Sural nerve graft	V	NA	NA
Moskowitz et al, 2011	6 y, F	Epithelioid hemangioma	GG	5 y	Mild-to-moderate CHL, dizziness	TMA, MFA	NA	NA	NA	Transient dizziness, profound HL immediately postop
Ozkale et al, 2014	2 m, F	Germ cell tumor	NA	2 m	Bilateral nerve VI palsy	NA	NA	V	V	No improvement, bilateral nerve VI palsy

CHL, conductive hearing loss; Chole, cholesteatoma; CPA, cerebellopontine angle; EAC, external auditory canal; F, female; GG, geniculate ganglion; HB, House-Brackmann facial nerve grading system; HL, hearing loss; IAC, internal auditory canal; M, male; m, months; MFA, middle fossa approach; NA, not available; RSA, retrosigmoid approach; SC, suboccipital craniectomy; SNHL, sensorineural hearing loss; SP, superficial parotidectomy; STP, subtotal petrosectomy; TCA, transcervical approach; TLA, translabyrinthine approach; TMA, transmastoid approach; TM-SLA, transmastoid supralabyrinthine approach; TOA, transotic approach; TO-TPA, transotic-transparotid approach; VII–XII, nerve VII–XII anastomosis; y, years.

Table 2. Radiological Features of Facial Nerve Tumors

Tumor Type	Ultrasound	CT	MRI			
			T1-Weighted	T2-Weighted	Contrast Enhancement	Other Features
Schwannoma	–	Widened fallopian canal; asymmetric enlargement of FN	Hypoisointense	Hyperisointense	Yes	May be multifocal or have necrotic center
Intraparotid schwannoma	Well-defined hypoisoechoic homogeneous lesion	Well-defined lobulated homogenous lesion	Isointense	Hypointense	Yes	–
Meningioma	–	Asymmetric enlargement of FN,	Hypoisointense	Hyperintense	Yes	Dural tail
Hemangioma	–	irregular borders, bone erosion, calcification, bone spicules	Hypoisointense	Hyperintense	Yes	–

CT, computed tomography; FN, facial nerve; MRI, magnetic resonance imaging.

and revealed a schwannoma.[26] Frozen-section biopsy was performed intra-operatively in at least 19 patients to determine tumor type and confirm the resection margin. Facial nerve schwannomas appeared as well-circumscribed encapsulated bundles of spindle cells with nuclear palisading and no mitosis. They stained positive for the S100 protein.[20,23,30] Khilnani et al[24] reported brain tissue-like appearance of an intraparotid schwannoma. Two meningioma samples exhibited whorled patterns with psammoma bodies and some calcification.[19,27] They stained positive for the epithelial membrane antigen and negative for the S100 protein and CD34. Reticulin-stained epithelioid hemangioendothelioma contained many myofibroblasts and exhibited high vascularity.[28]

Outcomes

The main clinical outcomes included FN status, hearing status, and tumor recurrence. Facial nerve function improved in 7 children (26.9%) and remained the same in 9 children (34.6%) after tumor resection. Most notably, FN function improved from HB grade V to HB grade II in the child reported by Yafit et al[14] and remained normal in 2 children reported by Grinblat et al,[31] although the best expected postoperative outcome for intratemporal tumors is HB grade III. FN function worsened in 5 children (19.2%). FN functional outcome was not reported for 5 children (19.2%). All children with intraparotid schwannomas had normal FN function pre- and postoperatively except 1 child whose FN palsy reached HB grade III at 3 months postoperatively.[24] Hearing improved in 1 child (3.8%) and worsened in 6 children (23.1%). One child experienced mild transient impairment of the fifth and the sixth cranial nerves after surgery.[30] No tumor recurrence was observed in children whose tumors were resected.

Case Report 1

A 12-year-old male was admitted to our department in August 2020 for evaluation of head trauma after an accidental bicycle accident. He suffered progressive hearing loss and tinnitus in the right ear, which had started a few months earlier, and right FN palsy, which had become obvious a year earlier. Clinical examination revealed HB grade III FN palsy. Audiometric assessment revealed moderate sensorineural hearing loss above medium frequencies on the right side (Figure 1). Eye examination was normal. Genetic testing excluded neurofibromatosis type 2. Computed tomography scan revealed an enlarged right IAC and a mass involving the GG and the tympanic

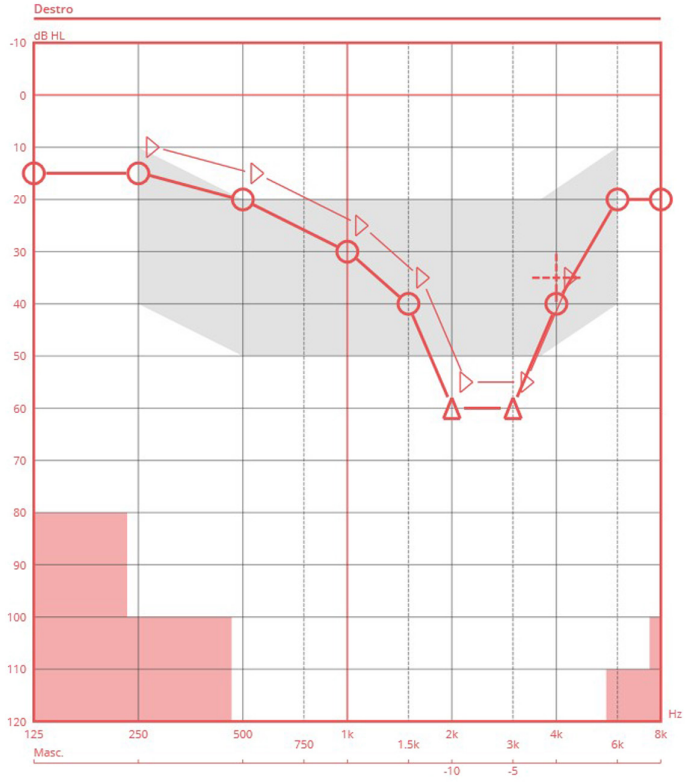

Figure 1. Right ear: pure tone audiometry.

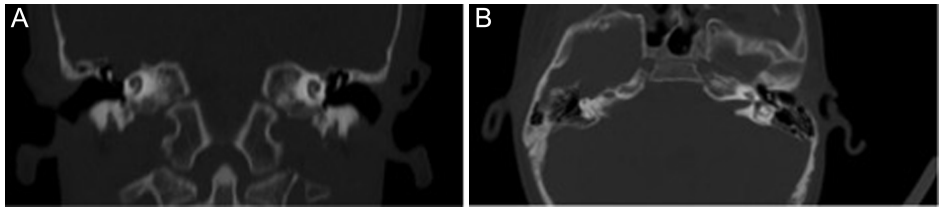

Figure 2. CT scan shows the mass on the right side involving the GG and the tympanic segment on the FN, in coronal (A) and axial (B) view. CT, computed tomography; GG, geniculate ganglion.

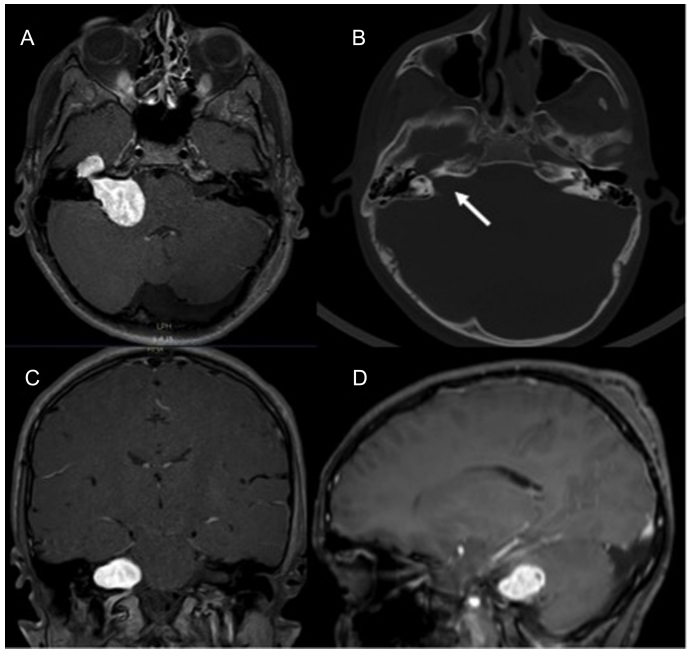

Figure 3. A: axial contrast-enhanced 1-weighted MRI shows enhancement of the right side mass. B: CT scan showing the tumor erosion of the right internal acoustic canal (white arrow). C and D: Coronal and sagittal MRI showing the mass effect of the tumor compression and dislocating the brainstem. CT, computed tomography; MRI, magnetic resonance imaging.

segment of the FN (Figures 2 and 3). T1-weighted MRI showed enhancement of the right side mass (Figure 3). The initial diagnosis based on clinical and radiological findings was FN schwannoma. In October 2020, the patient underwent surgical treatment. The tumor covering a wide area of the tympanic segment, GG, intralabyrinthine segment, and the IAC was completely removed using a modified transcochlear approach type A. A wide portion of the FN affected by the tumor was sacrificed. Histologic evaluation of the mass confirmed the diagnosis of FN schwannoma. The patient was discharged 1 week after surgery. Postoperative FN function was HB grade VI. Six months later, the patient underwent FN anastomosis between the FN nerve and the masseteric nerve. Magnetic resonance imaging confirmed complete tumor removal.

Case Report 2

An 18-month-old girl was admitted to our department in March 2021 with left FN palsy of HB grade V, which was noticed soon after birth. Clinical history was unremarkable for known pathologies. Conditioned orientation response audiometry revealed normal hearing on both sides. CT showed a left FN enlargement at the GG level (Figure 4), and MRI revealed mild gadolinium enhancement (Figure 5). The patient underwent surgical treatment. A retroauricular transmastoid approach was performed, and the incus was removed. The tympanic segment of the FN until the GG was edematous, and no tumor was found. The FN segment from the digastric crest to the GG was decompressed (Figure 6). The patient had no complications and was discharged one week after surgery. Postoperative FN function was HB grade V. The first follow-up MRI is planned 6-8 months after the surgery.

This study was approved by Ethics Committee of Florence University. According to local ethical review board guidelines, all patients sign an informed consent on admission to hospital for their inclusion in observational studies with anonymised data extraction.

DISCUSSION

According to our review of FNTs in children, histologies included schwannomas, meningiomas, one hemangioma, and one germ cell tumor. We could identify only 26 cases in the literature published between 1965 and 2019, which shows how exceedingly rare these pathologies are. As reported in adults, facial palsy was the most common clinical presentation of an FNT in children.[2,4] The severity of the palsy ranged from mild facial weakness to complete paralysis. Hearing loss was also common, followed by vestibular problems. Tumors were generally slow growing, with symptoms gradually developing over the course of months or years. About 42.3% of the tumors involved the GG region, which also agrees with the literature.[1,2] Females were as likely to develop FNTs as males.

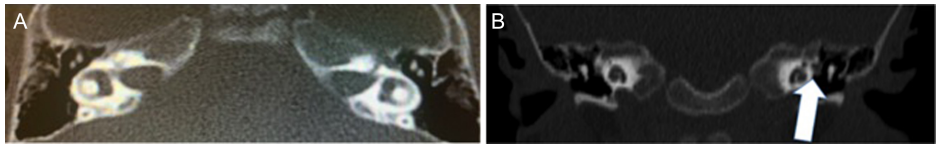

Figure 4. CT scan shows an enlargement of the left FN at the level of the GG in the axial view, white matter arrow (A) and in the coronal view, white matter (B). CT, computed tomography; FN, facial nerve; GG, geniculate ganglion.

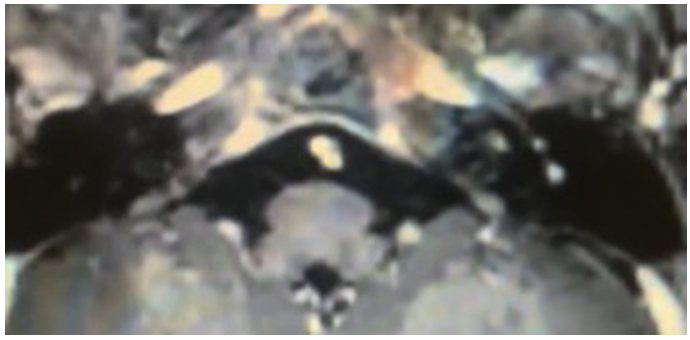

Figure 5. T1-MRI shows mild gadolinium enhancement at the GG level on the left side. GG, geniculate ganglion; MRI, magnetic resonance imaging.

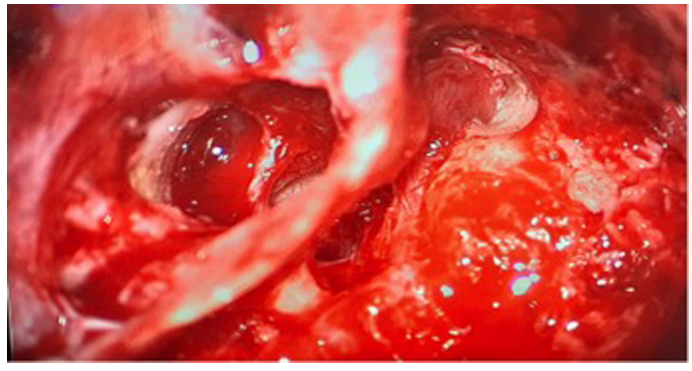

Figure 6. Surgery imaging of the edematous FN. FN, facial nerve.

Diagnosis

Most children underwent neurotologic (like audiometry and electromyography) and radiologic examinations. Although MRI and CT are the most effective noninvasive diagnostic tools for FNTs, they can still lead to an incorrect diagnosis.[15,24,27,32] To prevent accidental nerve damage, a preoperative biopsy should only be performed after imaging.[26] However, preoperative biopsies are likely to be inconclusive or even misleading.[18,20,23,24] An intra-operative frozen-section biopsy is the most robust way to establish the correct diagnosis and rule out malignancy.[23]

Although extremely rare, FNTs should be included in the differential diagnosis of facial palsy in children. Gradual onset of palsy in the first weeks or months after birth indicates an FNT, while congenital palsy is present immediately after birth, and idiopathic Bell's palsies have a sudden onset.[25] Old photographs and videos of the patient can help establish the true age of facial palsy onset.[15,16] Fallopian canal enlargement on CT and contrast enhancement on MRI provide further evidence in favor of an FNT.[15]

Facial nerve tumors located in the CPA or the IAC are often confused preoperatively with vestibular schwannomas.[7,32,33] Normal hearing, as identified by audiometric tests, can help rule out a vestibular schwannoma, like in the cases reported by Gonzales-Pardo et al[27] and Deep et al.[30] It is especially important to perform audiometric tests in small children because hearing loss can be underestimated in a younger population.[14] Facial nerve tumors are also more likely than vestibular schwannomas to cause facial palsy and extend to the fallopian canal and the labyrinthine segment, which is visible on MRI.[1]

Intraparotid schwannomas are often misdiagnosed as pleomorphic adenomas because FN schwannomas comprise only about 1.4% of intraparotid neoplasms, whereas pleomorphic adenomas are much more common.[34] Both types present as painless masses with no facial palsy. Two patients reviewed here were misdiagnosed as pleomorphic adenomas after imaging and FNAC, with the correct diagnosis established intra-operatively.[23,24]

Facial nerve schwannomas are also less common in children than neurofibromas.[25] Unlike schwannomas, neurofibromas arise from endoneural connective tissue, are more intermixed with the nerve fibers, and are more likely to become malignant.[3,20,35,36] They can be differentiated immunohistochemically using calretinin and CD34.[22,37]

Additionally, FNTs should be differentiated from granular cell tumors, glomus tumors, and congenital cholesteatoma.[7,10,32] Unlike FNTs, cholesteatomas do not enhance with contrast on MRI.[3]

Facial nerve tumors should also be distinguished from each other. Meningiomas arise from arachnoid cells of the meninges and rarely occur extracranially.[27] They may have a dural tail on MRI and calcification on CT.[27] Their more aggressive nature can cause a sudden worsening of symptoms and requires more generous resection.[19,27] Psammoma bodies are the distinguishing histological feature of meningiomas. Hemangiomas are vascular neoplasms that redirect the blood flow. Therefore, small hemangiomas can cause disproportionately strong FN palsy due to ischemia and not due to compression, as is the case with schwannomas.[7] On MRI and CT, hemangiomas have irregular borders and may contain bone spicules, while schwannomas are more likely to be multifocal.[7]

Genetic evaluation was not performed in any of the reviewed cases of sporadic FN schwannoma. According to literature, patients with solitary tumors are not routinely tested.[38] However, young patients may have multiple tumors due to a genetic condition. Other authors sustain that 14% of children with isolated meningiomas and 13% with schwannomas later fulfill the diagnostic criteria for neurofibromatosis type 2.[39]

Treatment

Facial nerve tumor treatment options include radiological observation, bone decompression, tumor debulking, complete tumor resection, and radiotherapy. The choice of treatment depends on the size and type of the tumor, the duration and degree of facial palsy, the hearing status, and patient's consent.

Complete tumor resection was the most common treatment (88.5%), but the timing of intervention was controversial. Generally, FN function after resection was not better than HB grade III, with the notable exceptions reported by Grinblat et al[14] and Yafit et al.[31] Therefore, the consensus is to monitor tumor growth radiologically if the FN function is normal or below HB grade III and to resect the tumor if the FN function is HB grade III or above.[32] Liu and Fagan[32] also argued that tumor resection is indicated when it compresses the CPA. Some authors promote earlier intervention because, firstly, children have better regeneration capacity and, secondly, muscle deterioration and nerve fiber infiltration are not advanced yet.[15,29,31] This is thought to lead to easier surgery and a better outcome.[31]

Residual/intact hearing should be preserved whenever possible. According to Van Den Abbeele et al,[25] surgery should be delayed for some children until hearing preservation methods can be used. When hearing is lost, translabyrinthine and transotic approaches can be used.[18] Grinblat et al[14] argued that complete resection of an extensive tumor is more important than hearing and FN preservation. Kim et al[15] also emphasized that the FN should be entirely exposed during surgery, and the resection margin must be confirmed by frozen-section biopsy.

More conservative alternatives, for example, partial tumor debulking and bone decompression, prolong normal FN function and are now being used more often.[40,41,42] Alyono et al,[26] however, emphasize that the FN may still be damaged during debulking.[26]

Generally, adult surgery techniques are safe for children, but children's bones are thinner and need less drilling, and the mastoid process is absent or not fully developed in children under 3 years old, which makes the FN more vulnerable during surgery.[14]

Meningiomas and hemangiomas should be resected more generously because they spread more aggressively and are more likely to become malignant than schwannomas.[19,27,28] Radiotherapy is not recommended in children due to the risk of malignant transformation and other long-term complications.[26]

Outcomes

Facial nerve function improved or remained the same in 16/26 (61.5%) of the reviewed cases. The best functional outcomes were

achieved with intraparotid schwannomas, which were easier to strip free of the nerve without damaging it.[20] All children with an intraparotid schwannoma maintained normal FN function after surgery except one, although the follow-up period in his/her case was only 3 months, and he/she might have recovered later.[24] Outcomes are mainly affected by the age of palsy onset, the duration of palsy before surgery, and the degree of FN function, although Van Den Abbeele et al[25] did not find such relationships in their case studies.[1,5,32,40] There is less risk of immediate and complete facial paralysis with conservative treatment, whereas radical tumor resection almost inevitably results in HB grade III palsy.[32] Grafting type does not seem to affect the outcome.[15] Prognosis is usually better in children than in adults, although Ozkale et al noticed poorer outcomes in younger children than in older children.[29,43]

Case Reports

We also reported 2 recent observations that were initially classified as FNTs. In the first patient, the diagnosis of FN schwannoma was confirmed. The tumor and the associated segment of the FN were removed, resulting in postoperative HB grade VI palsy. In the second patient, the diagnosis was changed intra-operatively to FN edema. The affected segment of the FN was decompressed, resulting in postoperative HB grade V palsy.

Limitations

This review considered primarily case series and single case reports. Large studies are unfeasible due to the extreme rarity of FNTs in children. Furthermore, data obtained from the scattered reports available in the literature are unavoidably heterogeneous and incomplete, precluding robust meta-analysis.

CONCLUSION

FNTs are extremely rare in children but should still be included in the differential diagnosis of facial palsy along with vestibular schwannomas, neurofibromas, pleomorphic adenomas, cholesteatomas, and other pathologies. Hearing status should be assessed with audiometric tests. Radiologic imaging is necessary to determine tumor localization and a surgical approach but is not always sufficient to distinguish between all tumor types. Biopsy (e.g., FNAC) can be attempted after imaging, but in most cases, it remains inconclusive. Common adult surgical techniques can be used in children, despite some anatomical differences, but after tumor resection FN function is likely to become HB grade III or worse.

Ethics Committee Approval: This study was approved by Ethics Committee of Florence University.

Informed Consent: Verbal and written informed consent was obtained from the patients who agreed to take part in the study.

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