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Case Report

Intratemporal facial nerve neurofibroma: a case report and review of literature

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ABSTRACT

Facial nerve neurofibromas (FNN) are a rare benign tumor of facial nerve can arise from anywhere along the course of the facial nerve from the cerebellopontine angle (CPA) to the extracranial branches within the parotid. They are most commonly located in the parotid gland, and FNN in temporal bone are rarely reported. Intratemporal facial neurofibroma arise from facial nerve in internal auditory canal or in Fallopian canal. It usually manifests as progressive facial palsy, but also can present as sudden or repetitive facial palsy mimicking Bell's palsy. We reported a case of intratemporal facial neurofibroma arising mainly from mastoid segment in 17-year-old female who presented with progressive facial palsy for 3 years. We briefly reviewed previous cases of intratemporal facial neurofibroma reported in the literature to provide a comprehensive understanding of this rare entity.

Keywords: Neurofibromas, Facial nerve, Facial palsy, Bell's palsy

INTRODUCTION

The primary tumours of facial nerve are rare. They may develop from Schwann cells, the endoneurial fibroblast support cells, the perineurial cells that resemble epithelial cells, or the blood vessel.[1] The two forms of neurogenic neoplasms of the facial nerve-schwannomas and neurofibromas are derived from Schwann cells.[2] FNN are rare benign tumors, which originate along the facial nerve and are most commonly located in the parotid gland, and FNN in temporal bone are rarely reported.[3] The majority of neurofibromas occur sporadically, although about 10% ultimately prove to be associated with neurofibromatosis type1 (NF1).[4]

We described the extremely rare occurrence of a histologically verified neurofibroma primarily arising in the mastoid segment of the facial nerve in a patient not fulfilling diagnostic criteria for neurofibromatosis. Tumor was completely removed through trans-mastoid approach and facial nerve grafting done using great auricular nerve. Histopathological report confirmed the diagnosis of facial nerve neurofibroma. This case was misdiagnosed as idiopathic bell's palsy at few other centers and therefore, we also performed a brief review of the literature to evaluate the clinical features of intratemporal facial neurofibroma to increase the awareness of clinicians on this rare entity and minimize misdiagnoses in the future.

CASE REPORT

A 17-year-old female was presented with history of the left-sided lower motor neuron facial nerve palsy for 3 years without facial spasm, hearing loss, or other cranial nerve palsies and no family history of neurofibromatosis. The onset of facial palsy was insidious and progressed to compete facial palsy over a period of 6 months. Facial palsy accompanied by otalgia during the early stage of facial palsy. Examination revealed a complete peripheral facial nerve palsy (HB grade VI) on the left side with negative Schirmer's test. There was no parotid mass or

stigmata of neurofibromatosis, and other examinations were otherwise unremarkable.

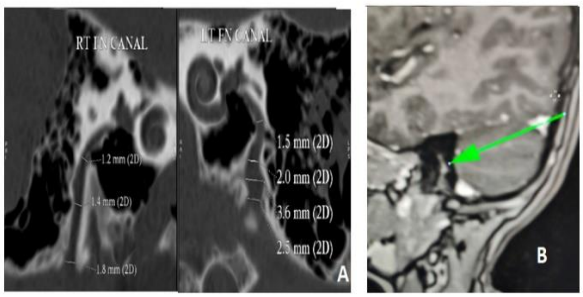

Figure 1: (A) Constructed HRCT scanning revealed smooth expansion of left mastoid segment of fallopian canal in comparison with right side; (B) the coronal view of T1-wighted MRI revealed thickening of mastoid segment of facial nerve and enhancing after gadolinium administration.

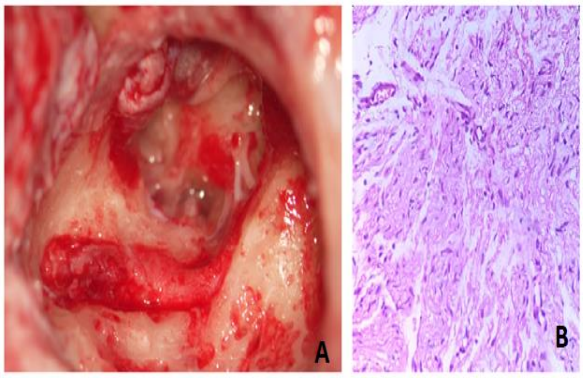

Figure 2: (A) Intraoperative macroscopic specimen; (B) representative histopathological image; intraneural proliferation of wavy spindle cells; cross sections of axons observed throughout; hematoxylin and eosin.

As it was long standing facial nerve palsy, she was subjected to radiological imaging. HRCT temporal bone and MRI with gadolinium (Gd) contrast were done. On HRCT, smooth bony expansion was noted in left vertical Fallopian canal (Figure 1A). On MRI, there was thickening of mastoid segment of left facial nerve for about 9-10 mm and measures about 4.3×5 mm (TR×AP) on axial plane, and the same area enhanced after intravenous administration of gadolinium (Gd) (Figure 1B). Hence possibility of facial nerve tumor was suspected and surgery was performed.

A transmastoid approach was performed. Vertical segment of facial nerve was exposed. Vertical segment was wider. Epineurium was normal. Epineurium was incised, perineurium was wider and bulbous (Figure 2A). Affected segment was excised and sent for HPE. Grafting was done using great auricular nerve. Histopathology reported as tumor formed by spindle cells oriented in fascicles with spindled to wavy nuclei with bipolar cytoplasmic processes (Figure 2B). Facial nerve palsy was recovered to grade IV at the end of 3 months and patient is still under follow up.

DISCUSSION

FNN can develop anywhere along the course of the facial nerve from the CPA to the extracranial branches within the parotid. The intratemporal facial neurofibromas arise from facial nerve in internal auditory canal or in Fallopian canal. Although the incidence of intratemporal facial nerve neurofibromas remained unclear, they are substantially less frequent than schwannomas.[5] Neurofibromas can be divided into three categories: localized, diffuse, and plexiform. The majority of neurofibromas are sporadic, localized and have a very low risk of malignant transformation. On other hand, the plexiform type is pathognomonic for neurofibromatosis type 1 and is more likely to progress to malignancy.[6]

As schwannomas and neurofibromas share common imaging characteristics including multi-segmental and multifocal occurrence, the diagnosis could not be made based on clinical and imaging findings alone.[3,5,7] The differential diagnosis between the two tumors dependent on pathological examination. Schwannomas are composed exclusively of neoplastic cells that simulate the appearance of differentiated Schwann cells, while neurofibromas display a mixture of cell types, including Schwann cells, perineurial cells, and endoneurial fibroblasts. Schwannomas typically are well circumscribed and composed of spindle cells organized as cellular areas with nuclear palisading (Antoni A) and pauci-cellular areas (Antoni B). Neurofibromas are less cellular and not as circumscribed as schwannomas and contain considerable extracellular myxoid material, wavy collagen fibers, and occasional neurites.[8]

After screening, a total of histopathologically verified 31 cases were included in the analysis (Table 1). Facial palsy was the first presentation of intratemporal facial neurofibroma in majority of reported cases 90% (28/31). In cases presenting with facial palsy, the typical manifestation of progressive onset was the most common (60%), the remaining cases presenting with sudden onset mimicking Bell's palsy. Repetitive facial palsy with vertigo observed in one case. Auditory symptoms were the second most common symptoms and included otalgia with the onset of facial palsy, hearing loss, and pulsatile tinnitus.[1,3,5,7,10] In our case, patient presented with progressive facial palsy with otalgia accompanied the palsy at the onset.

Table 1: Presentation.

Authors	Total cases	Presenting symptoms
Dai et al[5]	11	10 cases with FP 4 cases with otalgia accompanied FP One case with repetitive FP with vertigo
Liu et al[1]	10	All cases with FP 3 cases with otalgia accompanied FP One case with SNHL accompanied FP
Liu et al[3]	6	3 cases with FP 2 cases with conductive hearing loss One case with pulsatile tinnitus
Yamamoto et al[10]	2	All cases with FP
McMonagle et al[12]	1	FP
Our case	1	FP with otalgia

Facial nerve tumors which could either grow along the facial nerve and presenting with facial palsy or invaded the middle ear directly and presenting with auditory symptoms like hearing loss and pulsatile tinnitus. It seemed that facial palsy was the predominant manifestation of intratemporal FNN and hearing loss or vertigo was rare. This may be explained by the fact that intratemporal facial nerve typically grew along facial canal other than intruding into middle ear cavity. Contrarily, intratemporal facial nerve schwannomas had a substantially higher incidence of hearing loss or vertigo, with rates of 61% and 39%, respectively.[11]

Similar to schwannomas, intratemporal FNN usually affected more than one segment of facial nerve (Table 2). In Liangfa et al report, two or more segments of facial nerve were affected in all the cases with FNN (all 6 cases with FNN).[1] In Liu L et al study, multi-segment involvement of the facial nerve was found in 50% of the cases (5 of 10 cases with FNN) and usually containing geniculate ganglion or tympanic segment.[3] In Dai et al report, two or more segments of facial nerve were affected in 54% of cases (6 of 11 cases with FNN).[5] Yamamoto el at report, two or more segments affected in all cases (2 of 2 cases).[10] In our case, the mastoid segment was only the affected segment.

Table 2: Affected segments.

References	Affected segments	
Dai et al[5]	Case (N)	Segments affected
	1	GG
	2	LS, GG
	3	IAC, LS, GG, TS, MS
	4	GG, TS, MS
	5	MS
	6	MS
	7	MS
	8	LS, GG, TS
	9	PS
	10	TS, MS
	11	PS, MS
Liu et al[1]	12	GG, LS
	13	IAC, LS, GG, TS, MS
	14	GG
	15	MS
	16	LS, GG, TS
Yamamoto et al[10]	17	GG
	18	TS, MS,
	19	TS, MS,
	20	TS
	21	GG
	22	TS, MS
	23	GG, TS
McMonagle et al[12]	24	LS, GG, TS

Continued.

References	Affected segments	
Liu et al[3]	25	GG, TS
	26	GG, TS
	27	GG, TS
	28	TS, MS
	29	TS, MS
	30	GG, TS, MS
Our case	31	MS

IAC, internal auditory canal; LS, labyrinthine segment; GG, geniculate ganglion; TS, tympanic segment; PS, pyramid segment; MS, mastoid segment.

It's worth mentioning that there were no skip lesions (discontinuous involvement of facial nerve segments) in FNN, as reported in Liu et al and Dai et al reports.[3,5] But skip lesions were found in 20%–29.2% cases with schwannomas.[9] In other words, skip lesions may be an important exclusive point of facial nerve neurofibromas from schwannomas.

Complete tumor removal is the standard choice, and nerve grafting is required if there is nerve deficit. The preoperative facial nerve function influences on the discission of preservation of facial nerve integrity if possible or not. In those cases, with favorable facial nerve level (grade IV or better), preservation of facial nerve integrity is still recommended if possible since preservation of nerve integrity may harvest better outcomes than nerve grafting. For those cases with poor preoperative facial nerve function (grade V or worse), complete tumor removal with severely degenerated fibers followed by nerve grafting would be better than preservation of nerve integrity.[3,5]

The choice of surgical approach for inratemporal FNN varies depending mainly on tumour location, but also levels of hearing and surgeon preference are taken into account. The intratemporal tumor can be removed completely by either transmastoid approach or middle cranial fossa approach or middle cranial fossa combined with transmastoid approach.[3]

Regarding to nerve graft, greater auricular nerve is most commonly used, since it is convenient to harvest and has similar diameter of facial nerve. If the required nerve graft is longer, sural nerve or the lateral femoral cutaneous nerve may be a good choice.[5]

In our case, the tumor was removed completely with by transmastoid approach. Patients underwent greater auricular nerve graft, since preoperative facial nerve function was grade VI.

CONCLUSION

In conclusion, facial nerve tumors are often misdiagnosed as Bell's palsy initially. Imaging is not routinely obtained for Bell's palsy patients, but it is highly recommended in the case of progressive paralysis beyond 3 weeks, complete paralysis with sensorineural hearing loss on the affected side, recurrent ipsilateral facial paralysis, polyneuropathy, absence of any clinical recovery 4 to 6 months after the onset of paralysis and the presence of facial twitching.

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