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Initial Exploration on Temporal Branch of Facial Nerve Function Preservation in Plexiform Neurofibroma Resection

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Background: Large temporal plexiform neurofibroma (PNF) is an irritating problem that causes facial disfigurement. Surgical resection of PNF is the only effective way to remove the tumor as well as to improve the patient's facial appearance. However, temporal branch of the facial nerve (TBFN) in the tumor is prone to be destroyed during PNF removal. Thus, TBFN palsy is the inevitable complication after surgery and might induce other malformation and dysfunction. Therefore, the aim of this study is to reconstruct a nearly normal face contour while preserving the facial nerve function.

Purpose: Selective PNF removal technique was designed to protect TBFN during PNF lesions resection in our patients.

Methods: From May 2011 to June 2015, the authors had 10 patients who suffered from PNF in the temporal region with facial disfigurement and underwent selective PNF removal to correct the facial disfigurement while preserving TBFN as well.

Result: All patients obtained the improvement of facial appearance after surgery. The temporal PNF was removed and the TBFN function successfully maintained. Plexiform neurofibroma recurrence has not been relapsed during 6 to 49 months' follow-up.

Conclusions: In our initial exploration, TBFN function maintenance and facial appearance improvement can be achieved simultaneously by using PNF-selective removal surgery technique.

Key Words: Plexiform neurofibroma, temporal branch facial nerve

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Neurofibromatosis, a subfamily of neurocutaneous syndrome or phakomatosis, results from organ abnormality of ectoderm tissue. Its lesions are not only found in the brain, spinal cord, nerve, and skin, but also in the mesodermal and endodermal organs, such as heart, lung, bone, kidney, stomach, and intestine.[1–4] In 1987, the National Institutes of Health (NIH) proposed 2 different types of neurofibromatosis, namely Neurofibromatosis Type I (NF1) and Neurofibromatosis Type II (NF2). By 1997, Gutmann et al added Neurofibromatosis Type III (Schwannomatosis).

Neurofibromatosis 1, most common neurofibromatosis syndrome, also known as Von Recklinghausen disease or peripheral neurofibromatosis, is a rare disease with an incidence 1 of 4000. The distribution of NF1 epidemic is irrelevant to sex, population, or race. Neurofibromatosis type I is an autosomal-dominant genetic disorder caused by the genetic abnormality of the chromosome 17q11.2.[5] The average life expectancy of NF1 is 10 to 15 years shorter than that of the overall population.

Recent study shows 99% of neurofibroma is a clinical symptom of the neurofibromatosis patients, 90% of whom suffer from NF1. According to the pathological characteristics, NF can be divided into solitary, diffuse, and plexiform types. Plexiform neurofibroma (PNF) is one of the cardinal features of NF1. The incidence of PNF is 30% among NF1 patients. The probability of PNF occurring in the head and neck is 50%, which cause disfigurement and deformity. Tumor resection is the only effective method to restore cosmetic appearance and reconstruct function, but the facial PNF debulking is likely to damage the facial nerve, resulting in paralysis of the nerve after surgery.[6–8] The effective way to avoid facial nerve injury by neurofibroma resection has not been reported yet. To reduce intraoperative facial nerve injury and to preserve the temporal branch of the facial nerve (TBFN) function successfully, we creatively designed selective PNF removal approachment based on the anatomical distribution of facial nerve in the temporal region and the characteristic of PNF.

METHODS

Patients

Between November 2011 and May 2015 at Ninth People's Hospital Affiliated to Medical School of Shanghai Jiaotong University, 153 consecutive patients, age ranged between 5 and 41-year old, were diagnosed with neurofibroma. From 153 patients, 14 patients were diagnosed with temporal PNF and included in this study. The temporal PNF resection was performed among these 14 patients with facial disfigurement and deformity. Four patients underwent nonselective neurofibroma resection by using conventional surgical procedure. The remaining 10 patients (5 males and 5 females) adopted relevant facial nerve protective measures according to the anatomical features of the temporal branches in the PNF during the process of tumor resection (Table 1.). This study was

TABLE 1. Clinical Findings in Affected Patients

Patient No.	Sex	Age (Years Old)	PNF Type	Surgical Approachment	TBFN Injury in Operation	Relapse	Follow-Up (Mo)
1	M	41	Diffusely infiltration	Nonselective resection	+	–	49
2	M	19	Diffusely infiltration	Nonselective resection	+	–	40
3	F	22	Diffusely infiltration	Nonselective resection	+	–	45
4	M	36	Diffusely infiltration	Nonselective resection	+	–	48
5	F	5	Diffusely infiltration	Selective resection	–	–	6
6	F	6	Diffusely infiltration	Selective resection	–	–	6
7	M	8	nodular tumor	Selective resection	–	–	40
8	M	7	Diffusely infiltration	Selective resection	–	–	37
9	F	8	Diffusely infiltration	Selective resection	–	–	25
10	F	8	Diffusely infiltration	Selective resection	–	–	36
11	F	9	Diffusely Infiltration	Selective resection	–	–	36
12	M	11	Diffusely infiltration	Selective resection	–	–	37
13	M	19	Diffusely infiltration	Selective resection	–	–	6
14	F	29	Diffusely infiltration	Selective resection	–	–	22

TBFN, temporal branch of the facial nerve.

approved by the Institutional Review Board of the Shanghai 9th People's Hospital.

SURGICAL TECHNIQUES

Selective Plexiform Neurofibroma Resection With Temporal Branch of the Facial Nerve Protection

Nodular Plexiform Neurofibroma Resection

Localized resection was performed to PNF tumors with clear boundaries, and TBFN can be preserved.

The skin was incised from pretragal to temporal hairline. After skin flap had elevated, submuscular aponeurotic system (SMAS) was exposed. Then, TBFN was dissected carefully under SMAS in the vicinity region of the connection line of the pretragus and 1 cm lateral to the eyebrow. After TBFN was exposed, isolated, and preserved from the parotid-temporal fascia, the surrounding PNF entirely removed.

Diffuse Plexiform Neurofibroma Resection

This method also applied to diffusely infiltrative PNF lesions with obscured boundaries between the lesions and peripheral tissue, and TBFN could not precisely dissect and be isolated from the tumor. However, the fascia structure still could be able to identify according to the anatomical landmarks during operation.

The skin incision was made from pretragal to temporal hairline. This procedure covered 2 steps. The first step was PNF lesion removal under subcutaneous layer. After the skin flap had elevated, SMAS or superficial temporal fascia was exposed. PNF removal dissection was carried between the skin and the superficial temporal fascia. The second step was PNF lesion removal between superficial and deep layer of deep temporal fascia. We dissected the superficial layer of deep temporal fascia through superior border of lesions, exposed the lesion through superior rim of zygomatic arch then excised PNF lesion between superficial and deep layer of deep temporal fascia. The superficial layer of deep temporal fascia was closed with a suture superiorly of original position and removed loose tissue and skin. Lastly, the large part of PNF was removed, as well the tissues between superficial temporal fascia and superficial layer of deep temporal fascia, i.e. the parotid-temporal fascia was preserved during PNF dissection (Fig. 1.).

Nonselective Plexiform Neurofibroma Resection Without Temporal Branch of the Facial Nerve Protection

The skin incision was made from pretragal to temporal hairline. Skin flap was elevated, and PNF lesions between the subcutaneous and deep layer of the deep temporal fascia were completely removed without the preservation of TBFN.

RESULTS

Selective Plexiform Neurofibroma Resection With Temporal Branch of the Facial Nerve Protection

For one nodular PNF patient, TBFN could be isolated from tumor and kept intact during operation. After the temporal PNF was entirely removed, the cosmetic facial appearance had been restored (Fig. 2).

For nine patients treated with diffuse infiltrative PNF lesions, TBFN bound through the lesions, and the facial nerve barely

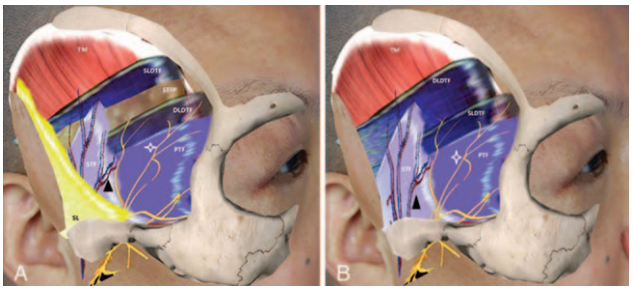

FIGURE 1. Surgical sketch for diffusely infiltrative PNF resection. (A) Preoperation. The normal temporal region soft tissue layers divided into 8 anatomical levels: dermis (D), subcutaneous layers (SL), superficial temporal fascia (STF), parotid-temporal fascia (PTF), superficial lamina of the deep temporal fascia (SLDTF), superficial temporal fat pad (STFP), deep lamina of the deep temporal fascia, and temporalis (DLDTF). Superficial temporal artery (black triangle) is in superficial temporal fascia. The TBFN (temporal branch of facial nerve, white star) is on the PTF. The NF tumor grows in all the soft tissue. SL and STFP NF compose of the major volume of the tumor. (B) Postoperation, subtotal NF tissue was removed; however, the tissue between the STF and SLDTF was preserved. The TBFN in PTF layer remained intact.

FIGURE 2. Patient, male, 8-year old, with PNF. Left, preoperative. PNF, obvious protruding to the left temporal region, facial asymmetry deformity can be observed. Middle, postoperative. Application of nodular PNF selective resection scheme, most of the temporal neurofibroma was removed. Right, postoperative. The function of TBFN was preserved, and the eyebrow can be raised freely after surgery.

isolated from the tumor. Temporal branch of the facial nerve was protected during entirely or sub-totally resection only by preserving the whole parotid-temporal fascia. After surgery, the cosmetic facial reconstruction was also achieved, and the function of the frontalis muscle movement of raising eyebrow controlled by TBFN was completely preserved (Fig. 3).

Nonselective Plexiform Neurofibroma Resection Without Temporal Branch of the Facial Nerve Protection

Although a good static facial appearance could be obtained after non-selective temporal PNF lesions entirely or sub-totally removal, the eyebrow motion was inevitably lost by TBFN resection during operation (Fig. 4). All PNF lesions did not relapse after surgery during 6 to 49 months' follow-up.

DISCUSSION

Plexiform neurofibroma is one of the cardinal clinical manifestations of NF1. Typical PNF will exist at birth, with 50% occurring in the head, neck, and throat. At birth, PNF may be only accompanied by slight symptoms, but the tumor will gradually grow and extend with the ages, and severe disfigurement and substantial loss of function will appear, which frequently leads to patient's social withdrawal.[9]

Histologically, PNFs consist of mixed tumor cells mainly containing Schwann cells, fibroblasts, perineurial cells, and mast cells. Schwann cells, with NF1 gene mutated, are the main composed cells in neurofibroma, which accounted for 40% to 80% of all the cells in tumor. Plexiform neurofibroma is a benign tumor, with malignant transformation to MPNST rate being 3% to 5%.

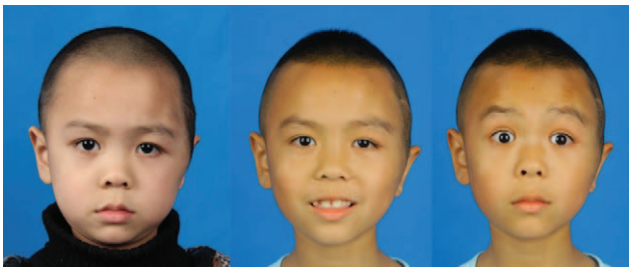

FIGURE 3. Patient, female, 5-year old, left preoperative. PNF, obvious prominent left temporal region, facial asymmetry deformity. Middle, postoperative. Application of diffuse infiltration PNF selective resection scheme, retained the tissue between the STF and the SLDTF, and removed subtotal tumor of temporal area. Right, postoperative. The function of TBFN was preserved after surgery, and the left brow was enabled to lift easily.

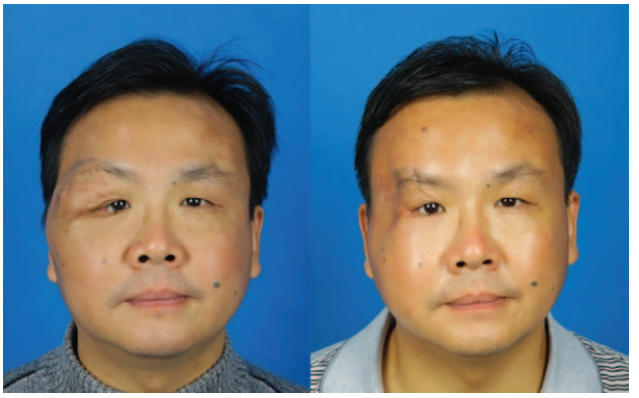

FIGURE 4. Patient, male, 49-year old, left preoperative. PNF, obvious prominent left temporal region, facial asymmetry deformity. Middle, postoperative. Application of nonselective PNF resection scheme, most of the temporal neurofibroma was removed. Right, postoperative. The function of TBFN was destroyed, and the eyebrow cannot be raised after surgery.

If PNF involves major nerves, irregular shape of nerve bundle will appear, hyperplastic neurilemma cell and collagenous fiber form curve cord, and the surrounding is a kind of amorphous matrix. Typical "target" sign will appear in T2 magnetic resonance imaging phase, namely the central nerve fiber cord present low signal, and the surrounding amorphous matrix is high signal.[10]

Plexiform neurofibroma is not sensitive to radiotherapy and tumor may continue to grow slowly after radiotherapy. Also, radiotherapy tends to increase the possibility of malignant transformation. Radiotherapy will therefore not be a first-line treatment for PNF. The recent research shows the effect of chemotherapy is still limited to shrink or diminish PNF. Tumor resection is the only effective way of PNF treatment.[11–13] The aim of surgical intervention for facial disfigurement involves tumor debulking, function restoring, and cosmetic facial appearance reconstruction.

Generally, PNF maintains an inconsistent, slowly growth. However, during specific periods, such as early childhood, puberty, or pregnancy, as well as suffering trauma, PNF is likely to grow rapidly. It has been reported that the proliferation of tumor was related to hormone level variation.[14,15] Although trauma is likely to stimulate neurofibroma growth, it remains unclear whether surgery related as a particular type of trauma that can promote the tumor's growth.

For the neurofibroma, subtotal resection removed more than 95% of the lesions during the surgery. The recurrence rate of subtotal excision is 45%, and total excision is less than 20%. For children, the postoperative recurrence rate after PNF resection reaches 40% to 60%. For children before 10-year old, the recurrence rate of neurofibroma after surgery is 60%, and for the children after 10-year old is 30%. The remains neurofibroma will grow with the age after surgery. The bigger the PNF is, the more extensively it will invade the surrounding tissues and thus will cause more severe bleeding during the surgery and acute tissue damage postoperatively, deformity tendency and possibility of relapse after the surgery. All these factors provide a strong indication for surgical intervention of prior diagnosis of PNF. Therefore, NIH suggests that early treatment should be performed to remove the lesion before it grows too large. This will help to reduce secondary deformity and dysfunction caused by tumor growing large.[16] Especially, facial PNF of the pediatric patients in this group, lesions were protruding in the temporal region, propagating, occupying, compressing, or invading the adjacent tissues, which could cause deformity of facial bone and soft tissue development. To reduce severe deformity

development due to tumor growth with the age, we tried to remove these lesions at the early stage.

Temporal branch of the facial nerve is one of the most important anatomical structures in temporal region. The primary function of TBFN is to control the frontal muscle, producing the eyebrow action.[17] However, temporal PNF lesions often involve TBFN or TBFN is coursing through temporal PNF, so the injury of temporal branches of facial nerve is inevitable when temporal neurofibroma need total or subtotal removal. Apparently, the injury to the temporal branch will result in the loss of the motor function of the frontal muscle. Consequently, the ipsilateral eyebrow cannot be raised, eyebrow will prolapse, and forehead wrinkles will disappear. As reported in this research, after nonselective PNF resection without TBFN protection, patients achieved a good facial static appearance improvement, but proper TBFN function undoubtedly lost as well.

To preserve the function of TBFN, we initially attempted and explored to design a selective PNF resection for 10 patients with temporal PNF. Clinically, PNF had 2 subtype growth patterns, nodular or diffuse infiltration.

For nodular PNF, the normal surrounding tissues are always pushed away by well-defined neurofibroma. The anatomic structure of the head and neck around localized PNF is still intact. Therefore, after the TBFN was isolated and safely protected from the tumor, the PNF could be entirely removed without damaging surrounding normal tissue, especially keeping the facial nerve integrity. At last, the TBFN function of localized PNF patients was preserved, and the aesthetic appearance achieved.

For diffuse infiltrative PNF, the abnormal tumor cells penetrate the surrounding tissues. Nerve fibers of the PNF lesions grow without their originally clear, regular, white, light, flexible boundaries. Afterward, the lesion shows irregular hyperplasia, thickening, even sheet extending. It is difficult to identify the anatomic structure, especially to make out TBFN from infiltratively growing neurofibroma in the temporal region. Also, tumor tissue texture is fragile, poor elasticity, and easy to bleed when touching, which greatly increases the difficulty of surgical dissection. It is nearly impossible to isolate TBFN from PNF without facial nerve injury.

The normal temporal region soft tissue layers divided into 9 anatomical levels: dermis, subcutaneous layers, superficial temporal fascia, parotid-temporal fascia, superficial layer of the deep temporal fascia, superficial temporal fat pad, deep layer of the deep temporal fascia, temporalis muscle, and pericarnium. Superficial temporal fascia is the continuation of SMAS fascia inferior of zygomatic arch. As we know, frontal and temporal branches of superficial temporal artery are the anatomic marker coursing through temporal superficial fascia. There are multiple layers of the mill-feuille structure in the parotid-temporal fascia, which is loose connective tissue located between the superficial temporal fascia and superficial layer of deep temporal fascia. Generally, 3 to 4 TBFN branches, parallel to each other, arise from the common facial nerve and parotid gland. The branches stretch vertically across the zygomatic arch, slightly protrude out, and then enter parotid-temporal fascia layer. Two to three anteromedia branches, which are oblique to forehead brow, control movement of forehead muscle and part orbicularis oculi muscle.[18]

It is hard to isolate and protect TBFN by fine dissection without normal anatomic nerve fiber structure and with excessive bleeding during PNF debulking. However, for diffusely infiltrative PNF, temporal 9 layers structure can be identified during the surgery. If we can completely preserve the parotid-temporal fascia, the TBFN could be avoided from injury because TBFN is located in the anatomic layer of the parotid-temporal fascia. During PNF debulking, the superficial temporal fascia with superficial temporal artery and the strong and shining superficial layer of deep temporal fascia were used as anatomic marker to indicate the position of parotid-temporal fascia. In the second selective neurofibroma debulking approachment for diffused PNF resection, the tissue from temporal superficial fascia to superficial layer of deep temporal fascia was preserved to assure the parotid-temporal fascia remains intact until the lesions maximally removed without facial nerve injury. After the operation, the appearances of patients not only showed improvement, but the function of the TBFN was maintained.

CONCLUSION

The approachment of neurofibroma resection with protection of facial nerve that we initially explored and creatively adopted is somehow complicated to achieve; it needs surgical skills and long-term experience of the surgeon. However, our novel surgical approachment resulted in both an excellent cosmetic appearance and maintaining facial animation function. The results showed that protection of facial nerve's function in entirely and subpartially PNF resection can be obtained, and the efforts to do so are very meaningful and improvement of the patient's social interaction and confidence can be observed.

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