

## 유지 인대의 외부 고정을 통한 제1형 신경섬유종증 환자의 안면부 변형 교정

명유진 · 이윤호

서울대학교 의과대학 성형외과학교실

### External Fixation of Retaining Ligament in Correction of Facial Disfigurement in Type-1 Neurofibromatosis Patients

Yu Jin Myung, M.D., Yoon Ho Lee, M.D., Ph.D.

Department of Plastic and reconstructive Surgery, Seoul National University College of Medicine, Seoul, Korea

**Purpose:** In neurofibromatosis patients, complete surgical excision of the mass is almost impossible and surgical treatment usually consists of multiple serial excisions that only result in a debulking effect. Remnant tumor mass has a gravitational effect on facial soft tissues that leads to sagging of skin and soft tissue, and eventually, facial disfigurement and asymmetry. The purpose of our surgical method is to perform soft tissue lifting with longer lasting effect with less surgical risk of damaging facial nerve and vessels. With external fixation using K-wire or surgical screw, the procedure only called for a short incision length and had additional adhesive properties that enabled anchoring of soft tissue in a lifted position for a longer postoperative period.

**Methods:** A total of 5 neurofibromatosis patients (NF-1) visited our clinic for mass reduction and face lifting. The age of patients ranged from 13 to 42 (mean 28.8 years), and most patients had a long history of multiple excisions in the past. Face lifting was performed in 2 different areas, the periorbital area in 3 patients, and the midface in 2 patients. The materials used in fixation of retaining ligament were K-wire (n=3) and titanium screw (n=2).

**Results:** Follow up period was from 5 month to 3 years and 1 month (mean=2 years and 1 month). All patients conveyed satisfaction with the results and no major complications were reported. The lifting effect lasted for as long as 3 years, and there were no complaints of relapse of soft tissue depression or sagging within the operated area.

1 patient (M/13) needed secondary k-wire insertion and additional mass excision in 1 year and 10 months post-operatively due to tumor growth. In two patients with K-wire fixation, mild dimpling and tenderness were observed in the follow up period, but in about 2 months postoperatively, dimpling was relieved and there was no need for removal of fixing material.

**Conclusion:** Surgical lifting in neurofibromatosis patients can be challenging, for mass excision cannot be done completely and gravitational effect by residual mass can be persistent. External fixation of the retaining ligament in patients with neurofibromatosis can give satisfactory results-for incision length is relatively shorter, and the lifting effect can last longer compared to other various face lifting techniques.

**Key Words:** Neurofibroma, Retaining ligament

## I. INTRODUCTION

Type 1 neurofibromatosis (von Recklinghausen disease), the hereditary disease which neurofibroma involves the whole body including facial area, is the most common type of neurofibromatosis that consists of 85 to 90% of total cases.<sup>1</sup> Neurofibromas that arise on facial areas are especially difficult in total excision and have high rates of recurrence, so the patients tend to get repeated operations and the operations<sup>2</sup> aim for only debulking the tumor. Although the patients receive repeated mass excisions, noticeable size of tumor still remains in facial area, causing sagging of surrounding soft tissue by mass effect. Ultimately, the mass effect leads to facial disfigurement and asymmetry, the most common and stressful complaint of type 1 neurofibromatosis patients.

Although there were a few trials of performing face lifts and mass excisions simultaneously in order to correct the deformities, the surgical effect lasted for shorter time compared to the conventional face lift surgeries because of the tumor that remained or recurred. Additionally, the risk of damaging facial nerve or vessels were not ignorable because of the tumor was infiltrating on each surfaces of facial soft tissue.

Received December 1, 2010

Revised March 31, 2011

Accepted April 11, 2011

**Address Correspondence:** Yoon Ho Lee, M.D., Ph.D., Department of Plastic and Reconstructive Surgery, Seoul National University College of Medicine, 28 Yongon-dong, Chongno-gu, Seoul 110-744, Korea. Tel: 02) 2072-2374 / Fax: 02) 3675-7792 / E-mail: yhlee@snu.ac.kr

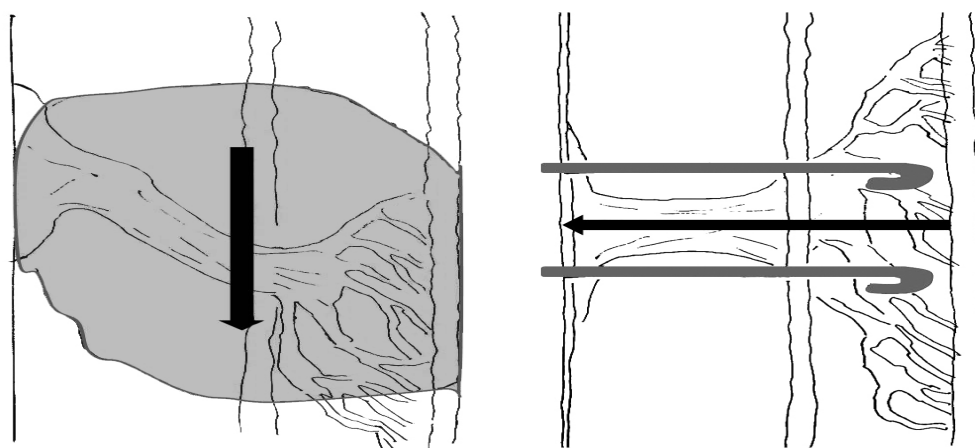
The authors performed a simple, minimally invasive method of external fixation method using K-wires (Zimmer, Inc. Indiana, USA) and non-absorbable titanium screws (Lorenz surgical Inc. Florida, USA) in order to strengthen the retaining ligaments of facial area and obtain longer-lasting face lifting effect. The results were highly satisfactory, with no major complications or recurrence of soft tissue sagging and disfigurement (Fig. 1).

## II. MATERIALS AND METHODS

Simultaneous mass excision and face lift with external fixation method were performed to 5 neurofibromatosis

type 1 patients that visited the clinic from 2006 to 2009. Patients' age ranged from 13 to 42 with the mean of 28.8 years, and the location of tumor were periorbital area in 3 patients, midface in 2 patients. 4 out of 5 patients had history of receiving mass excision on same location that the surgery was performed. Mean follow-up period was 25 months (Table I).

Preoperatively, extent of the tumor and amount of soft tissue sagging was assessed, and marking of the location was done where external fixation would be performed. After general anesthesia with orotracheal intubation, local solution (1% lidocaine with mixture of 1:200000 epinephrine) was infiltrated to incision line and around the tumor. For periorbital tumors suprabrow incision was



**Fig. 1.** Schematic drawings of effect of external retaining ligament fixation. (Left) Soft tissue of face is sagged downward by gravitational effect of infiltrating neurofibroma mass. Note the direction of gravitational force. (Right) After mass excision and fixation with two K-wires, the direction of force is now perpendicular to gravity and result in effective face-lifting.

**Table I.** Patient's Demographics

Patient	Gender / Age (year)	Mass location	Chief complaint	Operation technique (including mass excision)	Follow-up period (month)
1	M / 13	Lt. nasolabial fold	Lt. commissure sagging	#1. Retaining ligament fixation with K-wire to zygoma #2. Malar fat lifting	24
2	F / 38	Rt. periorbital area	Rt. eye ptosis	#1. Retaining ligament fixation with K-wire to upper lateral orbital rim #2. Lateral canthopexy, Rt. Eye	5
3	M / 24	Rt. periorbital area	Rt. eye ptosis	Retaining ligament fixation with titanium screw to upper lateral orbital rim	37
4	F / 42	Rt. periorbital area	Rt. eye ptosis	#1. D-E operation, Rt. eye #2. Retaining ligament fixation with titanium screw to upper lateral orbital rim	19
5	F / 27	Rt. temporal area	Rt. midface sagging	Retaining ligament fixation with K-wire to zygoma & upper lateral orbital rim	37



**Fig. 2.** A 10-year-old male patient with large neurofibroma on left cheek, causing sagging of left commissure. (Left) The patient underwent extensive surgical resection of tumor via intraoral and left alar rim approach, combined with retaining ligament fixation with k-wiring. (Right) Two k-wires indicated by red arrows fixed to his left zygoma can be confirmed by postoperative 3-D facial computed tomography.

done, and for midface tumors intraoral and external nasal alar fold incision was done (Fig. 2). Skin and subcutaneous soft tissue that were infiltrated by neurofibroma were excised, under extreme caution not to damage the facial nerve and vascular branches.

After excision of the tumor mass and wound closure, face lift operation with external fixation method was performed. In patients with midface tumor, the direction of the vector in face lifting was toward zygomatic arch from lateral commissure, and for the patients with periorbital tumor the lifting direction of forehead and periorbital soft tissue were headed from upper brow to the upper lateral border of frontal bone, where temporal adhesion is known to arise. After simulating the lifting effect with manual compression using operator's fingers, marking was done to the location where external fixation will be performed. The anatomical locations for fixation were anterior aspect of zygomatic arch, origin of zygomatic cutaneous retaining ligament, and upper lateral border of frontal bone, as described earlier. After 2 to 3 millimeters of slit incision, external fixation was done with K-wires or nonabsorbable titanium screws. K-wires were twisted 180 degrees like 'question mark' shape to avoid postoperative cutaneous exposure and tissue damage. Wires and screws were confirmed manually not to be exposed or damage the skin after closure of slit incisions.

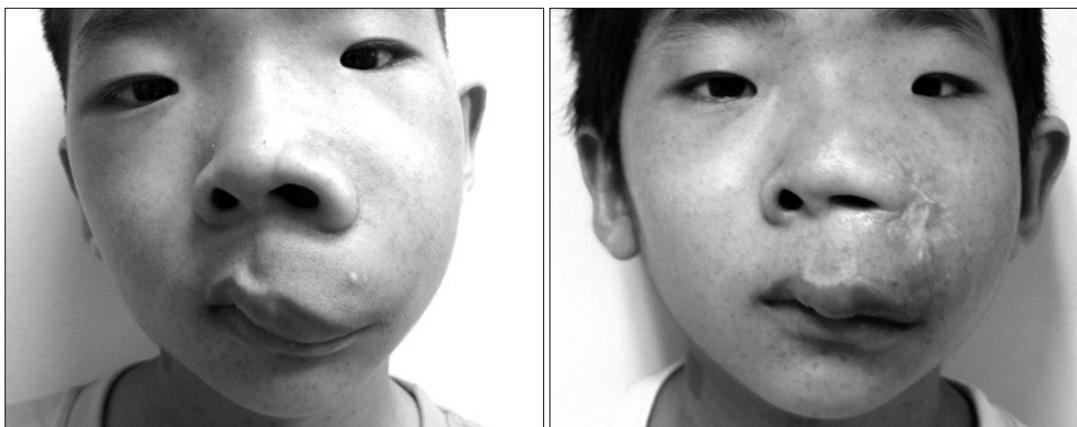
### III. RESULTS

Total of 6 operations were done to 5 patients. For every patient, there was noticeable improvement in soft tissue sagging, facial disfigurement, and asymmetry that patients complained about before the surgery (Fig. 3, 4). In four patients reoperation for recurrence of facial sagging was not needed, and in one patient additional mass excision and K-wire external fixation were done on postoperative 1 year and 10 months, for recurrence of primary neurofibroma mass was observed. There were no complications such as facial paralysis, infection on operated area or suture site dehiscence in follow-up period.

In two patients who had K-wire fixation complained about mild tenderness and dimpling on fixation site, but after about two months postoperatively the symptoms were cleared up and there were no need of removing the fixating material. In all of 5 patients, K-wires and screws were not removed and are persistent without causing any symptoms or complications.

### IV. DISCUSSION

Surgical lifting in neurofibromatosis patients can be challenging, for mass excision cannot be done completely and gravitational effect by residual mass can be persistent. For facial tissue sagging in type 1 neurofibromatosis patients, previous methods of surgical correction were



**Fig. 3.** Preoperative photo of the patient shown on Figure 1. (Left) Large mass on his left cheek is causing soft tissue sagging and depression to left side of commissure. (Right) Post-operative photo taken in 1 year and 3 month after the surgery. The effect of retaining ligament fixation is persistent, with minimal relapse of soft tissue sagging.



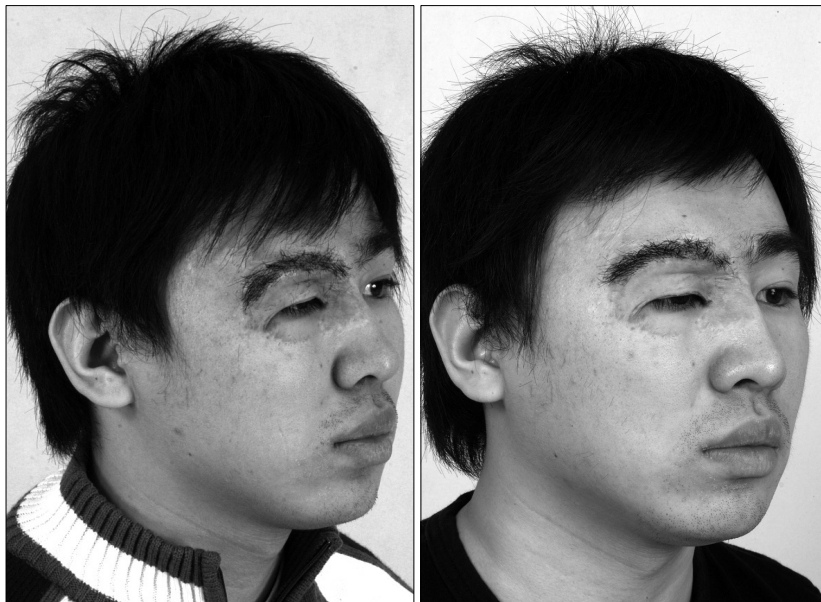
**Fig. 4.** (Left) A 42-year-old female patient with large neurofibroma on her right hemiface. Her primary complaint was sagging of her right upper eyelid with brow ptosis, and with tumor resection via upper blepharoplasty incision, retaining ligament fixation was done on right upper lateral orbital rim brow with additional incision on upper brow line. (Right) Postoperative photo taken in 1 year after the surgery. Her eye opening is significantly improved and her right brow is elevated.

either fixating with surgical suture<sup>3,4</sup> or performing lateral canthopexy.<sup>5</sup> Although these methods are very popular and time-honored, the surgical effects of these methods can be weakened and last for shorter period by gravitational effects of residual mass, compared to simple aging face patients without neurofibromatosis.

With anatomic and functional analysis of retaining ligaments, the fibrous supportive structure that holds soft tissue against gravity, Mendelson<sup>6</sup> emphasized fixation of weakened retaining ligaments in face lift procedures.

Retaining ligaments of facial area arise from periosteum or deep fascia to dermis layer superficially, creating a tree-like structure<sup>5</sup> to fix the facial soft tissue to normal anatomical location against gravity. They also fix all layers of soft tissue to facial bone or deep fascia, which give structural support to protect nerves and vessels against external shearing force.<sup>7</sup>

As normal anatomic structure can be interrupted by infiltrating neurofibroma, and by continuous overloading of retaining effect by gravity, the function of retaining



**Fig. 5.** (Left) A 24-year-old male patient with neurofibroma on his right upper eyelid and brow area. With mass excision, retaining ligament fixation was done with titanium screw fixation on right upper lateral orbital rim. (Right) Postoperative photo taken in 2 years after the surgery.

ligaments tends to grow weaker in neurofibromatosis patients. After Furnas<sup>8</sup> described about facial retaining ligaments there have been many reports about functional and anatomic importance of the ligaments, but there was no method trying to restore the function completely. External fixation can be an effective method of facelift surgery for neurofibromatosis patients, who need more retaining strength than normal patient with aging face.

Also by causing foreign body reaction the wires and screws can create soft tissue adhesions that can help retaining effect of the ligaments, and formation of subcutaneous scar tissue will have an additional effect of covering up the sharp sensation of wires or screws.

Two patients complained about mild tenderness or dimpling at the area of external fixation, but about 2 months after the operation the symptoms were not apparent, and there were no need of removing the fixating materials.

## V. CONCLUSION

Surgical correction of soft tissue sagging and facial disfigurement in type 1 neurofibromatosis patients is very difficult, for complete excision of facial neurofibroma is almost impossible and remnant mass gives continuous gravitational effect to facial soft tissue. To correct the deformity a few surgical methods with traditional facelift procedures have been introduced, but

shortcomings were present as the lifting effect lasted only for shorter period, and at the same time risk of damaging facial nerves and vessels is not ignorable.

External fixation in neurofibromatosis patients using K-wires or surgical screws can improve and restore the weight bearing capacity of facial retaining ligaments, preventing facial tissue from sagging downwards. And also with only slit incisions required, very short postoperative scarring can be yet another advantage. In neurofibromatosis patients that the results of traditional facelift surgery can easily be insufficient, external fixation method can be an excellent alternative as to avoid the side effect and still get satisfactory surgical results.

## REFERENCES

1. Neville BW, Hann J, Narang R, Garen P: Oral neurofibrosarcoma associated with neurofibromatosis type I. *Oral Surg Oral Med Oral Pathol* 72: 456, 1991
2. Jackson IT, Laws ER Jr, Martin RD: The surgical management of orbital neurofibromatosis. *Plast Reconstr Surg* 71: 751, 1983
3. Acarturk TO, Yigenoglu B, Pekedis O: Excision and "transcutaneous" lift in patients with neurofibromatosis of the fronto-temporo-orbital and auricular regions. *J Craniofac Surg* 20: 771, 2009
4. Lee YH, Noh JH, Choi SW: Correction of drooping mouth corner due to cheek mass by suspension with nasolabial dermal Flap. *J Korean Soc Plast Reconstr Surg* 29: 377, 2002
5. Marchac D, Britto JA: Remodelling the upper eyelid in the

- management of orbitopalpebral neurofibromatosis. *Br J Plast Surg* 58: 944, 2005
6. Mendelson BC: Extended sub-SMAS dissection and cheek elevation. *Clin Plast Surg* 22: 325, 1995
  7. Moss CJ, Mendelson BC, Taylor GI: Surgical anatomy of the ligamentous attachments in the temple and periorbital regions. *Plast Reconstr Surg* 105: 1475, 2000
  8. Furnas DW: The retaining ligaments of the Cheek. *Plast Reconstr Surg* 83: 11, 1989