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거대 층상 신경 섬유종 절제 후 전외측 대퇴부 유리피판술을 이용한 재건

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Surgical Correction of Disfiguring Plexiform Neurofibroma Using an Anterolateral Thigh Free Flap

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Purpose: Neurofibromas of neuroectodermal origin are commonly found in Von Recklinghausens disease or neurofibormatosis type 1. It is an autosomal dominant disease caused by mutation of the long arm of chromosome 17. It can present from small nodules to disfiguring giant tumor. Plexiform neurofibroma is benign in most cases, but it could be transformed into malignant tumor, which requires surgical excision. To cover the defects after the excision, a number of surgical correction methods are available. This study is to report a surgical correction of disfiguring plexiform neurofibroma using anterolateral thigh free flap for extensive defects after surgical excision of neurofibrona.

Methods: Data of five neurofibroma patients with an average age of 39 including medical history, physical examination, computed tomography, and magnetic resonance imaging were checked. No disease other than neurofibroma were detected. Biopsy on the excised tissues was performed. The follow-up period was 7 to 27 months.

Results: The average size of defects after complete excision of neurofibroma was 13 × 10~25 × 15 cm. Defects were covered by anterolateral thigh free flap, while donor sites were covered by local flap, split thickness skin graft and regional flap. Throughout follow-up, there were no complication, relapse, or any abnormalities.

Conclusion: Despite various surgical correction methods are applicable to defects after excision on disfiguring plexiform neurofibroma, coverage of massive defects is still challenging in plastic and reconstructive surgeon. We have made five successful cases of surgical correction of disfiguring plexiform neurofibroma using anterolateral thigh free flap.

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I. INTRODUCTION

Neurofibroma is a characteristic clinical symptom of neurofibromatosis, which is also characterized by other symptoms, such as, cafe-au-lait spots and Lisch nodules. Neurofibromas of neuroectodermal origin are commonly found in Von Recklinghausen's disease or neurofibromatosis type 1 (NF-l). They are manifestations of an autosomal dominant disease caused by mutation of the long arm of chromosome 17. The prevalence of neurofibroma is about one per 3000 in the general population.[1,2]

Clinically and macroscopically, neurofibroma can be classified as localized neurofibroma derived from a single fascicle or as plexiform neurofibroma derived from multiple nerves and fascicles.[3] Neurofibroma sizes range from small to disfiguring giant tumors, which can cause functional disabilities. Neurofibroma may present abnormalities of skin, soft tissue, the nervous system, the vascular system, or bones.

Indications for surgical excision include continuous growth, functional abnormalities, clinical symptoms, such as, pain, or malignant transformation.[3] Generally, a local flap is used for coverage after the excision of small nodules, but for large masses various surgical correction methods are available and the issue is still being actively researched. Furthermore, most neurofibromas are multiple, which limits donor sites and makes surgical correction difficult. Here, we introduce a method of covering wide defects after the surgical excision of disfiguring plexiform neurofibroma based on the use of an anterolateral thigh free flap.

II. CASE

From January 2008 to March 2010, we treat five patients

with plexiform neurofibroma (Table I). All required surgical excision because of a disfigured appearance and functional limitation.

No clinical symptoms of neurofibromatosis, except for cafe-au-lait spots, were found, and although neurofibroma is dominantly inherited, only one patient had a familial history. No patient had any other underlying disease, or a surgical history. Two patients had disfiguring neurofibroma on face and three had multiple neurofibroma, which manifested at various body locations. Preoperative evaluations were conducted using computed tomography (CT) and magnetic resonance imaging (MRI) to determine lesion sizes, and non-invasive computed tomographic angiography was used to determine blood vessel conditions.

After surgical excision, defects were covered using an anterolateral thigh free flap and donor sites were closed using local flap, skin graft, or island flap. Average follow-up period was 17.8 months (7~27 months).

Case 1

A 35-year-old woman presented with huge disfiguring neurofibroma on her left cheek and showing ectropion, and a mastication problem due to the size of the mass. She had become socially withdrawn because of her appearance. On the CT scan, the mass appeared to spread along the skin and soft tissues on her left upper, lower eyelid, cheek, malar area, nose and upper lip. No bones or other organs were involved, and nothing other than neurofibroma was found in her medical history. The size of the defect left after excision was 14 × 12 cm. It was reconstructed using an anterolateral thigh free flap. The donor site was covered using a local flap and a split thickness skin graft. Neurofibroma was confirmed histologically. During the 27-month follow-up period, no relapse or abnormality occurred. The patient exhibited improvements of drooping and ectropion symptoms caused by mass, and the limitation of faical expression improved to some degree over time as the flap is bulky, but the recommended debulking procedure was not carried out, because the patient did not want it. The patient was also satisfied with the improvement of focal bluish color and dimpling skin lesion (Fig. 1).

Table I. Summary of Cases

Case	Age	Sex	Follow-up (month)	Site	Defect size (cm)	Donor site coverage	Functional outcome	Complication
1	35	F	27	Left cheek	14 × 12	Local flap and STSG	Good	No abnormality
2	24	F	18	Right temple, cheek and neck	13 × 10	Local flap	Good	Infection
3	39	F	7	Right lower leg and ankle	25 × 15	TRAM	Excellent	Donor site; partial necrosis
4	52	M	15	Right forearm	15 × 8	Local flap	Good	No abnormality
5	45	M	22	Left ankle and foot	20 × 17	STSG	Good	No abnormality

STSG, Split thickness skin graft; TRAM, Transverse rectus abdominis musculocutaneous island flap.

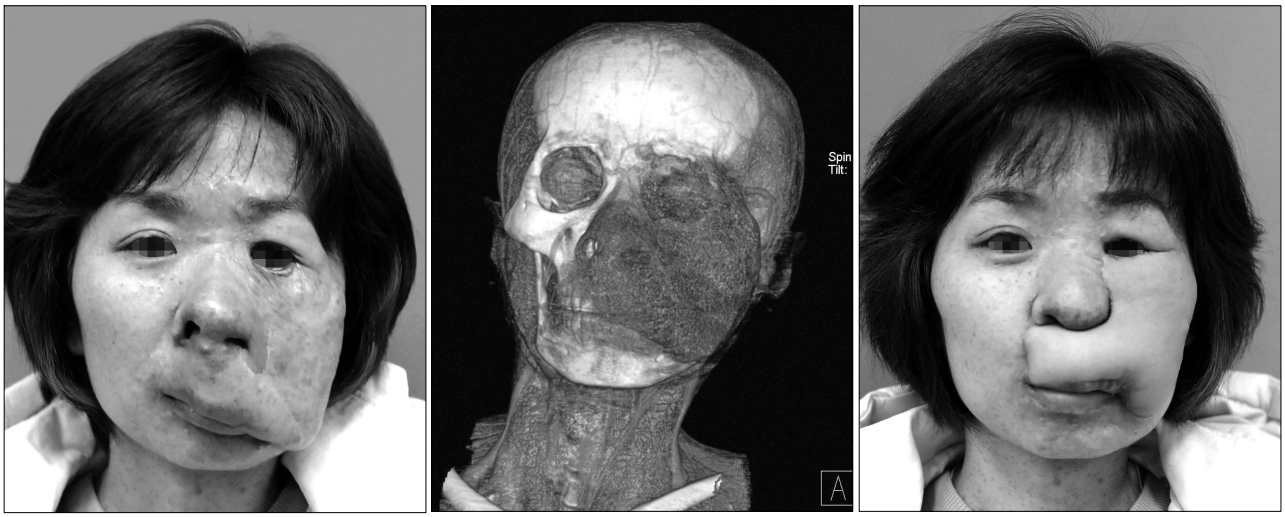

Fig. 1. (Left) A 35-year-old woman with neurofibroma on the left upper, lower eyelid, cheek, malar area, nose and upper lip. (Center) 3-D facial CT with enhancement. (Right) Postoperative view in 2 years after the surgery.

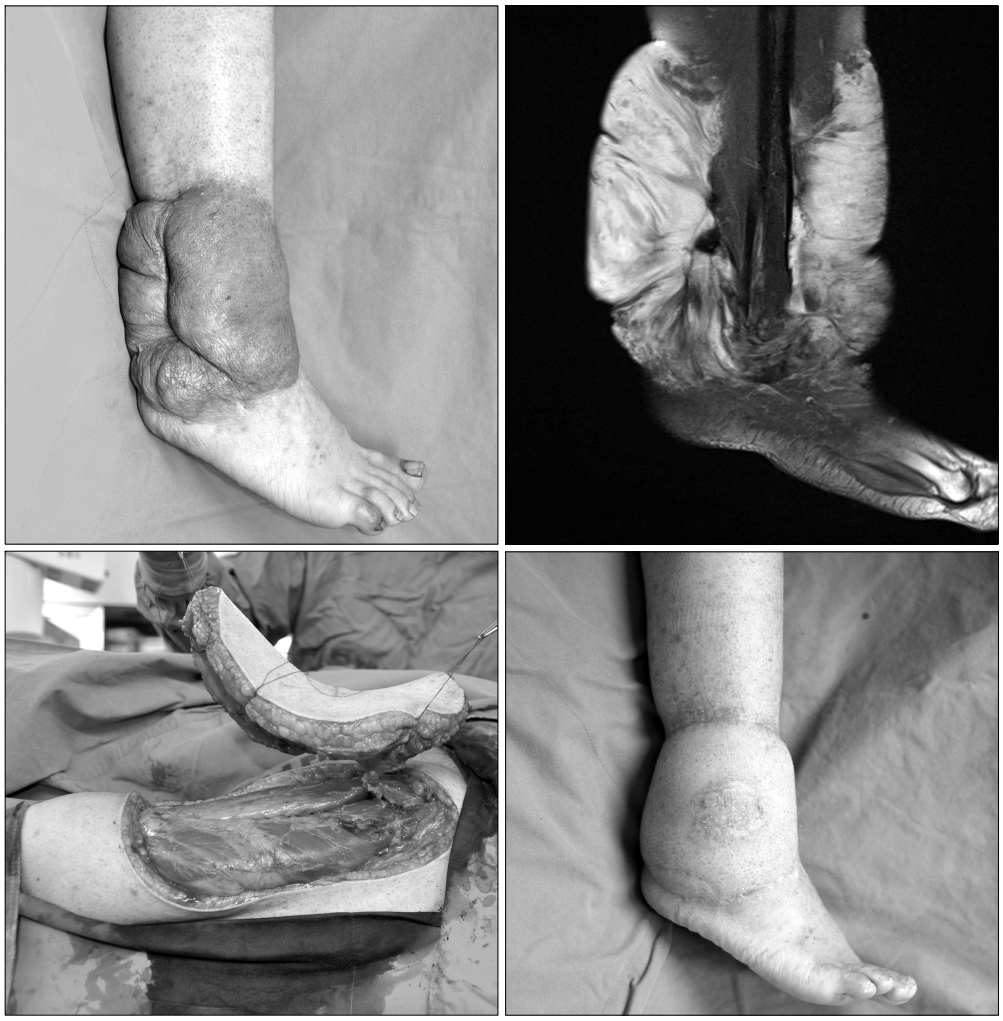

Fig. 2. (Above, left) A 39-year-old woman with neurofibroma on the right lower leg and ankle. (Above, right) Preoperative enhanced ankle MRI showed high signal intensity in the right lower leg and ankle. (Below, left) Elevation of the anterolateral thigh flap. (Below, right) Photograph taken in 6 months after the surgery.

Case 2

A 39-year-old woman presented with a large mass on the right lower leg and ankle. She was unable to dorsiflex the right foot because of the mass. In addition, she had variously sized multiple neurofibroma lesions and cafe-au-lait spots, but no other symptoms were evident. Furthermore, there was no evidence of invasion of bones or other organs by CT scan and MRI. The 25 × 15 cm defect remaining after excision was covered using an anterolateral thigh free flap, but it was not possible to cover the donor site with a local flap. Moreover, skin graft cannot be used because multiple lesions interfere harvesting. Instead, the defect was covered using a transverse rectus abdominis musculocutaneous island flap. At 6 weeks after the operation, walking and weight-bearing rehabilitation were started. No other abnormalities were observed during the 7-month follow-up, and the patient was pleased with the reconstruction using proper autoplasty following the removal of tumors for cosmetic purposes despite some degree of bulkiness, establishment of joint due to tumors was also improved by rehabilitation (Fig. 2).

The size of defect after surgical excision was 13 × 10~25 × 15 cm, and all were reconstructed using an anterolateral thigh free flap. Donor sites were covered using a local flap, a split thickness skin graft, and regional flap. Biopsy results showed no abnormalities other than positivity for neurofibroma, and all flaps survived well. During the average 17.8-month follow-up period, one split thickness skin graft procedure was performed due to partial necrosis of the donor site, and reconstruction was conducted because of infection by external auditory canal obstruction. No relapses or other

abnormalities were encountered. Patients began facial muscle and lower limb rehabilitation at an average of six weeks after operation. Results were functionally successful and aesthetically satisfactory by 5-point Likert scale in all five cases (Table I).

III. DISCUSSION

Neurofibroma can develop solely, but is more frequently associated with neurofibromatosis. Local or multiple masses of various sizes are found on those affected. Neurofibroma is usually benign, but it can transformed to be malignant, and it can cause movement disorders and other complications depending on its location and size.

Plexiform neurofibroma is asymmetric, and may cause deformation and functional disorders by overgrowing surrounding tissues. Complete surgical excision is the treatment of choice, although it is often difficult due to the invasive nature of the disease. Furthermore, plexiform neurofibroma has a relapse rate as high as 50%.[3]

Because of its overgrowing tendency, surgical correction of neurofibroma after excision is challenging. Small masses can be covered using local advancement flaps, but wider defects must be covered using skin grafts, regional flaps, or other procedures. Furthermore, because most neurofibromas are of the multiple form, donor sites are restricted, which limits surgical possibilities and presents obstacles to successful results. For reconstruction of large defect after excision of neurofibroma, it can be used for free flap such as scapular flap, rectus abdominis flap, radial forearm free flap, latissimus dorsi flap. Scapular flap and latissimus dorsi flap require harvesting in the lateral position and skin on the back is among the thickest in the body. Rectus abdominis flap is so bulky nature and radial forearm free flap is so thin. Therefore, free flap mentioned above is not suitable to treat giant neurofibroma in areas such as including head and neck. The author also observed less neurofibroma in thigh, used as a good donor site for the reconstruction of neurofibroma. As there are not sufficient cases of this, however, studies are needed for a larger number of cases.

The anterolateral thigh free flap is widely used for reconstruction of head, neck and other site. The advantage of anterolateral thigh free flap is that tissues of the thigh allow size adjustment. It can be harvested with a skin flap as large as that of abdominal perforator flaps. The vascular pattern allows the use of a more versatile design with double skin paddles based on multiple perforators. Including musculocutaneous flap in surrounding areas, large or 3-dimensional reconstruction like chmeric flap is possible. The flap pedicle is relatively long. Donor sites morbidity is low and most donor sites can be closed directly. Flap thicknesses are easily modified; that is, it is possible to make a flap as thin as radial forearm free flap. Furthermore, simultaneous operation by two teams is possible, which can reduce operation time. Therefore it has advantages over scapular, parascapular and latissimus dorsi flaps.[4,5]

Reconstructions based on the use of an anterolateral thigh free flap have high flap survival rates as high as 95.68% due to the use of advanced microsurgical technique.[6]

A number of surgical correction methods are available for covering the defects caused by excision of a disfiguring plexiform neurofibroma. Due to the advantages offered by anterolateral thigh free flaps, we used it to cover these defects, and we achieved successful results. However, our study was limited by the small number of cases, applied area, and a relatively short follow-up period, and references concerning neurofibroma. We hope that this report encourages the publication of more case reports and research on this surgical procedure.

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