Aggresive Mass Excision Through Nasolabial Fold Area in the Treatment of Facial Neurofibromatosis : Case Report

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Background : Neurofibromatosis type 1 is a rare disease which can manifest itself by the development of plexiform neurofibromatosis, with craniofacial deformities. In this paper, we propose special consideration in excision nasolabial fold mass to help lifting procedures in neurofibromatosis patients.
Methods : We are reporting two case, both presented with von Recklinghausen's disease, a 21 years old male patient with neurofibromatosis in his right hemifacial for which he had been operated on five times previously at other center, and a 24 years old male with neurofibromatosis in his left hemifacial.
Result : After several stages of reconstruction which started with nasolabial fold mass excision, the result was satisfactory, by using the multiple stage repair, it was done until the end result is similar to anatomical form
Conclusion : The mass debulking procedures for these patients, which started with Nasolabial Fold mass excisison are continued with durable lifting procedures. This method gives results a satisfactory lifting procedure.
Keyword : *Neurofibromatosis type I, nasolabial fold mass excision, Von Recklinghausen disease*

Latar Belakang: Neurofibromatosis type 1 adalah penyakit langka dengan manifestasinya berupa perkembangan neurofibromatosis plexiformis, dengan cacat kraniofasial. Dalam makalah ini kami mengusulkan pertimbangan khusus untuk melakukan eksisi massa lekuk nasolabial untuk membantu proses pengangkatan pada pasien neurofibromatosis.
Metodologi : Kami melaporkan dua kasus, keduanya dengan penyakit Von Recklinghausen's, pasien laki-laki berusia 21 tahun dengan neurofibromatosis di daerah hemifasial kanan dimana dia telah menjalani operasi sebanyak lima kali pada rumah sakit pusat lain, dan seorang pasien laki-laki 24 tahun dengan neurofibrosis di daerah hemifasial kiri.
Hasil : Setelah beberapa tahap rekonstruksi yang dimulai dengan eksisi massa lekuk nasolabial, hasilnya cukup memuaskan, dengan menggunakan beberapa tahapan perbaikan, hal itu dilakukan sampai hasil akhir menyerupai bentuk anatomis yang normal.
Kesimpulan : Prosedur pembedahan untuk mengurangi massa pada pasien ini, yang dimulai dengan eksisi pada lekuk nasolabial dilanjutkan dengan prosedur pengangkatan lama. Metode ini memberikan hasil prosedur pengangkatan yang memuaskan.
Kata kunci : *Neurofibromatosis type I, eksisi massa lekuk nasolabial, Von Recklinghausen.*

Neurofibromatosis type 1 (von Recklinghausen disease) was first described by von Recklinghausen and Festscher in 1882,[1] referred to as peripheral neurofibro-matosis, is a rare disease which can manifest itself by the development of plexiform neuro-fibromatosis, with craniofacial deformities as the result of alterations of the bone and in-filtration of soft-tissue. The neurofibromatosis type 1 mutation may affect the structure of connective tissues, explaining the distinctive softness and smoothness of the skin [2].Skin rheologic profiles of neurofibromatosis type 1 patients demonstrated hyperextensibility above neurofibromas and a lack of elasticity in general, including areas unaffected by tumors[3]. Neurofibromatosis type 1 patients with lesions involving the face or their whole body usually undergo many surgical procedures during their lifetime, as their

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Presented in The Fifteenth Annual Scientific Meeting of Indonesian Association of Plastic Surgeon. Semarang, Central Java. Indonesia.

Disclosure: *The authors have no financial interest to disclose.*

skin properties lead to an inexorable deterioration of functional and cosmetic results [4].

Surgical treatment for these deformities is difficult, especially in craniofacial area and needs aggressive procedures. Many surgeon prefer multiple serial excisions, as it is almost impossible to remove the entire tumor in one stage of reconstruction. However, because mass debulking are usually not accompanied by durable lift procedures, the skin sags again because of the gravitational effect on the remaining tumor.

We are reporting two case, both presented with von Recklinghausen's disease, a 21 years old male patient with neurofibromatosis in his right hemifacial for which he had been operated on five times previously at other center, and a 24 years old male with neurofibromatosis in his left hemifacial. In treated neurofibromatosis patient we use multiple stage reconstruction, with special consideration excision on nasolabial fold area which we believe will be very helpful in decreasing mass of the tumor so the skin will not sags again by the time we perform lifting procedure.

Case 1

A 21 year old male patient was admitted to Plastic Surgery Department of Cipto Mangunkusumo Hospital with a diagnosis of Von Recklinghausen's disease, NF type 1 in his right hemifacial region with history of 5 debulking surgery in the past at other center. (See Fig 1 to 6)

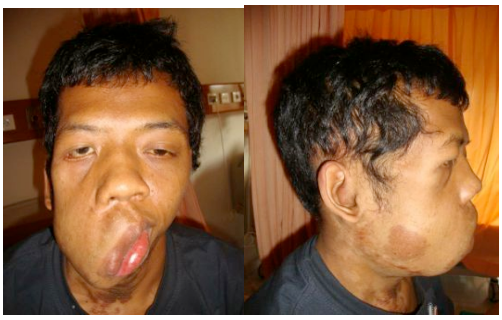

Fig 1. Preoperative appearance

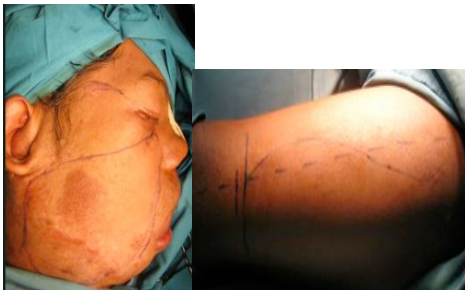

Fig. 2 Left: incision design on the tumor. Right : tensor fascia lata design.

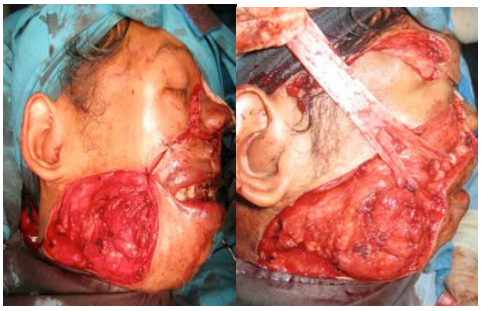

Fig 3. Left: nasolabial fold area excision. Right: the tensor fascia lata flap was crossed to right facial, one side was sewn to upper and lower right oris commisure, the other side was anchored to temporalis fascia, with anchoring on the zygoma arch in between

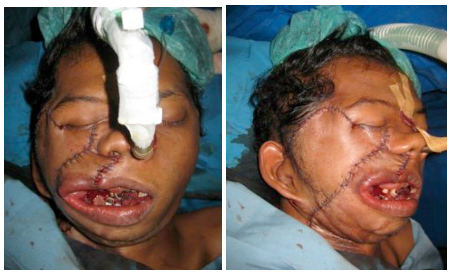

Fig 4. Intraoperative appearance

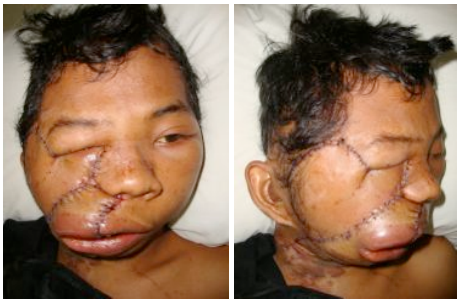

Fig 5. Result 3 days after the procedure

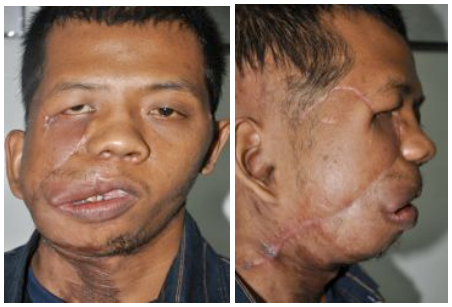

Fig 6. Result 2 month after the procedure

Case 2

A 24 year old Indonesian male was admitted to Plastic Surgery Department of Cipto Mangunkusumo Hospital with a diagnosis of NF type 1 of the left facial region. The massive and enormous tumour involving the inferior region of his left forehead, left eyebrow, left orbit, left temporal region, left auricle, left midfacial region (cheek), left nose, left side of the mouth and lips, left mandibular and submandibular region extending to his chin is confirmed and with an appearance like melting wax from a candle.

Multiple surgeries were necessary in order to remove the patient's tumor from his body. Each procedure was performed under general anesthesia.

First stage operation (30/06/09)

An incision was made at the left neck region alongside with the sternocleidomastoid muscle through cutis and subcutis. Sternocleidomastoid muscle was separated. Identification of commune carotid artery was made and preserved to the proximal, identification of external carotid artery was made, preserved and controlled with "lasso" technique. (see fig 7-9)

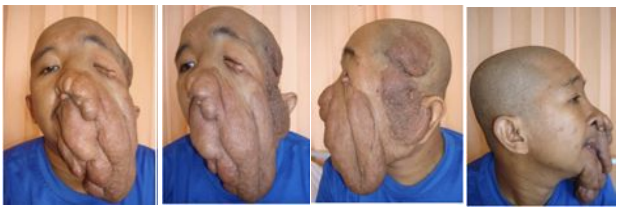

Fig 7 .Preoperative appearance

Fig 8. Incision design on the tumor.

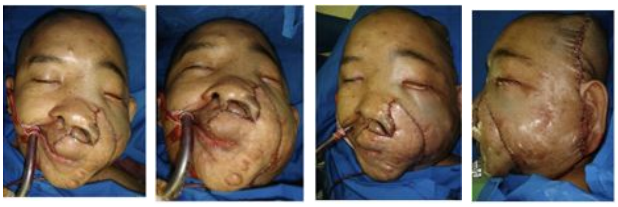

Fig 9 Intraoperative appearance

Second stage operation (15/07/09)

Excision of tumor mass. The left ear is anchored alongside the lateral canthus. The lateral canthus is then anchored to the superolateral periosteum. Sub-cutis was sewn with 2.0 cutting prolene. Intra dermal was sewn with 4.0 cutting prolene, dermal was sewn with 5.0 cutting prolene.(see fig 10)

Fig 10. Preoperative and intraoperative appearance.

Third stage operation (31/07/09)

An incision of the left orbital region was made into a flap and the tumor was extirpated. An incision to the superior orbital rim was made and dissected until naso orbital junction was visualized. An anchoring was made on the nasoorbital junction to the left lateral canthus with screw and wire. An incision according to the a' design was made into a flap and the tumor was excised with skin sparring. On the left nasal region a Cinh suture was made. The left oris commisure was anchored to the zygoma arch with 6.0 cutting prolene. A left eyebrow lifting was done with Treit technique

with O prolene. 3 incision was made at the scalp region, a number 16 abocath was inserted to insert a number 0 prolene, and the eyebrow was then anchored to the periosteum.(see Fig 11)

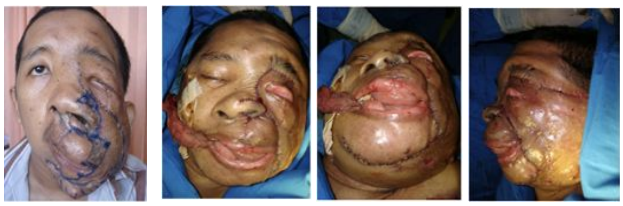

Fig 11. Incision design preoperatively and intraoperative appearance.

Fourth stage operation (14/08/09)

Reduction of mass at the parotid, retroauricula, and left buccal region. Intradermal was sewn with 2.0 vicryl dan 3.0 prolene, cutis was stitched with 4.0 prolene. A mass reduction was made at the left supra auricula, there is an attachment of the loose superior helix cartilage, then tied to the periosteum with 1.0 prolene, a skin sparring was done. Intra dermal was sewn with 4.0 vicryl and 4.0 prolene. Cutis was sewn with 4.0 prolene. Mass reduction was made at the left buccal region, 3.0 cromic was used for inner stitching, mucose is stitched with 3.0 cromic. The subtle but noticeable diffrence in mass size is shown in the serial photo taken after each operations. (see Fig 12)

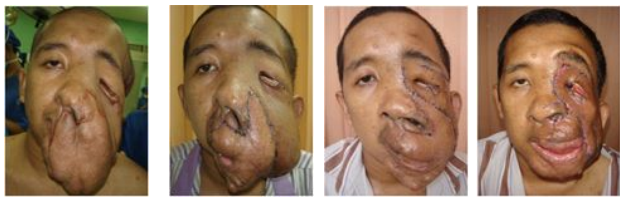

Fig 12. Post operative appearance after each operation.

DISCUSSION

Neurofibromas (NF) are benign peripheral nerve sheath tumors composed mainly of Schwann cells and fibroblasts[5] arising from any type of nerve[6] anywhere except in brain or spinal cord,[7] insinuating in normal tissue.[8] Two main types of NF: neurofibromatosis 1 (NF-1) and neurofibromatosis 2 (NF-2). NF-1 in the population (90% of cases), affecting 1 in 4,000 individuals. Onset occurs as early as childhood, and people of all sex, race, and ethnicity are affected equally. The NF-1 gene on chromosome 17, characterized by tumor formations, usually manifest themselves within the peripheral nervous system. Symptoms include Lisch nodules, cafe´-au-lait spots, axillary freckling, numerous fibromas, multiple developmental problems, macrocephaly, optic glioma, hype-tension, short stature, seizures, and distinctive osseus lesions. The severity of each NF-1 case is impossible to predict, and a patient might exhibit all or just a small number of these symptoms [1 2].

Plexiform neurofibromas, pathognomonic for NF type 1, is a slow-growing tumor with an unpredictable growth pattern occurs in a variety of locations including skin and subcutaneous tissue, craniofacial region, paraspinous structure, mediastinum, viscera, and retroperitoneum [9].Rapid growth may occur during early childhood, puberty, or pregnancy, but spontaneous regression does not occur. The incidence ranges from 24.1 % (in the National Neurofibromatosis Foundation International Database[10]) to 32 percent of the patients for Huson et al [11].Facial plexiform neurofibromas usually occur unilaterally and extend from nerves such as trigeminal, facial, or glossopharyngeal ones,[8] insinuate deeply and extensively, making their total removal impossible without sacrificing nontumoral tissues [2]. Surgical treatment is not a complete tumor resection, but a reduction to acceptable cosmetic and functional results [20].

These patients comes to us with neurofibromatosis type 1. In treated these patients we use multiple stage reconstruction, with special consideration excision on nasolabial fold area which we believe will be very helpful in decreasing mass of the tumor so the skin will not sags again by the time we perform lifting procedure. We adapt this technique based on systematic multiple stage surgical approach using Nagata's technique in which he modified and utilized the static suspension method with the tensor fascia lata [12]. The surgical approach we use was by doing excision on the corner of the mouth or nasolabial fold area to achieve

sufficiently wide coverage and large mount of mass because the tumor were extensive hemi-facial lesion. To achieve facial symmetry, tensor fasciae latae sling of the mouth corner or nasolabial fold area were performed. By doing this, we hope the tensor fascia lata could minimized further deformity caused by regrowth of the tumor after resection by being a substitute layer of the skin.

The same technique was done in netting operation where the netting of neurofibroma using Teflon mesh as a substitute for the sub-cutaneous layer and a barrier against drooping and minimizing further deformity caused by regrowth after resection, to maintain facial contour and function and to prevent progressive disfigurement.[14] The Teflon mesh is still very expensive and hard to get for most of these patients, so we have not use this technique.

Other approach such as the facial aesthetic unit remodeling monobloc translesional resection technique offered increased predictability in functional and cosmetic results, and allowed us to operate on extensive hemifacial lesions with a lower transfusion risk[4] is still an optional technique that we consider to use in the future.

CONCLUSION

Many procedures of operation with multiple stages reconstruction had to be done for Neurofibromatosis patient during their lifetime. The mass debulking procedures for these patients, which started with Nasolabial Fold mass excisison are continued with durable lifting procedures. This method gives results a satisfactory lifting procedure.

Acknowledgement

We gratefully thank Ishandono Dachlan, M.D. for his assisstance in reviewing this article and Nandita M. Putri, M.D. for basic editing and proof-reading.

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