Serial Excisions of Three Big Tumors on the Upper Right Side of Nose, Left Nostril, and Right Side of Chin of NF 1 under Tumescent Surgical Anesthesia in a Woman

Sri Lestari, Fesdia Sari, Nadya Hasriningrum, Elsi Kemala Putri, Yulia Eka Suryani
Dermato-Venereology Department of Dr. M Djamil Hospital/ Medical Faculty of Andalas University, Padang, Indonesia

Keywords: Neurofibromatosis type-I, excision, flaps

Abstract: Background: Neurofibromatosis type-1 (NF1) is autosomal dominant and multi-system disorders. An incidence 1 in 3000 live births. There are three big tumors on the face that could disturb function organs around the tumors and she wanted to remove that tumors. Case report: A case of 32-year-old woman with chief complaint there were brownish patches and bumps that painless and not itchy on the most part of the body since 20 years ago. At the age of 10 year-old there were appeared multiple skin colour bumps and growing bigger all over the body. On physical examination there were multiple skin colour tumors on most of the body, three big tumors on upper right side of nose, left nostril, and right side of chin, caféau-lait spots, generalized distribution, disseminated. We consulted to Ophtalmologic Departement there were Lish's nodules on iris of her eyes, on Neurology Departement no neurologycal focal defisits were found. Histopathological examination is neurofibroma. We excised three big tumors on the upper right side of nose, left nostril, and right side of chin with eliptical, subcutaneous tissue pedicle island flap and long inferiorly melolabial transposition flap designs under tumescent surgical anesthesia and there were good cosmetic result. Discussion: NF-1 is best cared for within a multidisciplinary approach, which has access to a wide range of subspecialists. We excised three big tumors on upper right side of nose, left nostril, and right side of chin every two-weeks to maintanance function of organ with good cosmetic result.

SCIENCE AND TECHNOLOGY PUBLICATIONS

1 INTRODUCTION

Neurofibromatosis (NF) is a term that has been applied to a variety of related syndromes, characterized by neuroectodermal tumors arising within multiple organs and autosomal-dominant inheritance. At least 8 different clinical phenotypes of neurofibromatosis have been identified and are linked to at least two genetic disorders. Neuro-fibromatosis type I (NF-1) is the most common type of the disease accounting 90% of the cases, and is characterized by multiple café-au-lait spots and the occurrence of neurofibromas along peripheral nerves (Burton et al., 2012; Dimitrova et al., 2008). Von Recklinghausen's neurofibromatosis (NF-1) is inherited in an autosomal-dominant and has a prevalence 1 per 3000 and 1 per 5000 live births (Dimitrova et al., 2008).

The diagnosis NF-1 was made according to the presence of four of the seven diagnostic criteria of the National Institute of Health Consensus Development Conference at least two of the following criteria must be present to make the diagnosis of NF-1 (Burton et al., 2012; Dimitrova et al., 2008). Five or more cafe-au- lait spots larger than 5 mm in diameter in prepubertal patients; six or more cafe-au-lait spots larger than 15 mm in diameter in postpubertal patients, two or more neurofibromas of any type, or one plexiform neurofibroma, axillary or inguinal freckling, optic glioma, two or more Lisch's nodules, a distinctive osseous lesion (pseudoarthrosis of the tibia or sphenoid wing dysplasia), a first-degree relative diagnosed with NF-1 in accordance with the above criteria (Dimitrova et al., 2008; Moraes et al., 2013; Ghalayani et al., 2012; Goldberg & Alam, 2004).

There is no medical treatment for NF-1 at this time. Neurofibroma therapy is not required and commonly unsuccessful with high rate of recurrences. Discrete cutaneous neurofibromas may be removed surgically to improve cosmetic or to prevent local irritation (e.g., from brushing for lesions in the hairline or from rubbing against the shoe for those on the foot). Deeper neurofibromas may require surgical removal when they push on vital structures, such as a

Lestari, S., Sari, F., Hasriningrum, N., Putri, E. and Suryani, Y.
Serial Excisions of Three Big Tumors on the Upper Right Side of Nose, Left Nostril, and Right Side of Chin of NF 1 under Tumescent Surgical Anesthesia in a Woman.
DOI: 10.5220/0008158604110414
In *Proceedings of the 23rd Regional Conference of Dermatology (RCD 2018)*, pages 411-414
ISBN: 978-989-758-494-7
Copyright © 2021 by SCITEPRESS – Science and Technology Publications, Lda. All rights reserved

dorsal root neurofibroma that infiltrates the neural foramen and compresses the spinal cord. Complications of surgery include regrowth of the original tumor and nerve damage (Burton et al., 2012; Dimitrova et al., 2008).

We reported three big tumors of NF1 on the upper right side of nose, left nostril, and right side of chin of 32 year-old woman, and she wanted to remove that tumors because of blocking of the right eye sight, push the left of nostril and disturb the right side of lower lip.

2 CASE

A 32 year-old woman, came to out-patient Department of Dermato-Venereology on November 28th 2017 with there were skin colour bumps that painless and not itchy on the most part of the body since twenty years ago. There were three big skin colour bumps on the face: on the upper right side of nose, left nostril, and right side of chin. There was positive family history, her daughter was 6 years old complained skin colour bumps on the back. On physical examination we found multiple skin colour tumors, the largest amount being on the upper right side of nose, left nostril, and right side of chin ranging from 1-4 cm, caféau-lait spots. On Ophtalmology Departement there were lish nodules on her eyes and there were no optic glioma. On Neurology Departement there was no neurologycal focal defisits were found. We excised three tumors on the upper right side of nose, left nostril, and right side of chin with eliptical, subcutaneous tissue pedicle island flap and long inferiorly melolabial transposition flap designs under tumescent surgical anesthesia to maintanance the function of organ around the tumors with good cosmetic result. Histopatological examination is neurofibmotasosis.

2.1 Procedural Operation

First time we operated the tumor on the upper right side of nose with eliptical design. From the area between eyebrows, we injected local anesthesia Pehacain® then incised with blade no.15. Using infiltrator cannula 3 mm diameter, we delivered surgical tumescent solution anesthesia 35 cc under the tumor and to subcutaneous tissue around the nose and the skin became bulging. We waited 20 minutes and after that we injected with Pehacain® (lidocaine HCl 2% 20 mg and epinephrine 12,5ug) superficially along the incision lines. Excised the tumor and anastomosed the wounds in subcutaneus space with 4-0 chromic gut and epidermis with 5-0 Prolene suture®.

Two weeks later we excised the tumor on the left nostril with subcutaneous tissue pedicle island flap design. From the area between eyebrows, we injected local anesthesia Pehacain® then incised with blade no.15. Using infiltrator cannula 3 mm diameter, we delivered surgical tumescent solution anesthesia 40 cc under the tumor and to subcutaneous tissue around the left nasolabial fold and left cheek and the skin became bulging. We waited 20 minutes and we injected local anesthesia Pehacain® superficially along the incision lines. Excised the tumor and anastomosed the wounds in subcutaneus space with 4-0 chromic gut and epidermis with 5-0 Prolene suture®. Two weeks later we excised the tumor on the right side of chin with long inferiorly melolabial transposition flap design. From the mid right mandibula, we injected local anesthesia Pehacain® then incised with blade no.15. Using infiltrator cannula 3 mm diameter, we delivered surgical tumescent solution anesthesia 80 cc under the tumor and to subcutaneous tissue around the chin and right cheek, until the skin became bulging. We waited 20 minutes and we injected local anesthesia Pehacain® superficially along the incision lines. Excised the tumor and anastomosed the wounds in subcutaneus space with 4-0 chromic gut and epidermis with 5-0 Prolene suture®. There were good cosmetic result after surgery and the function of the organs became normal.

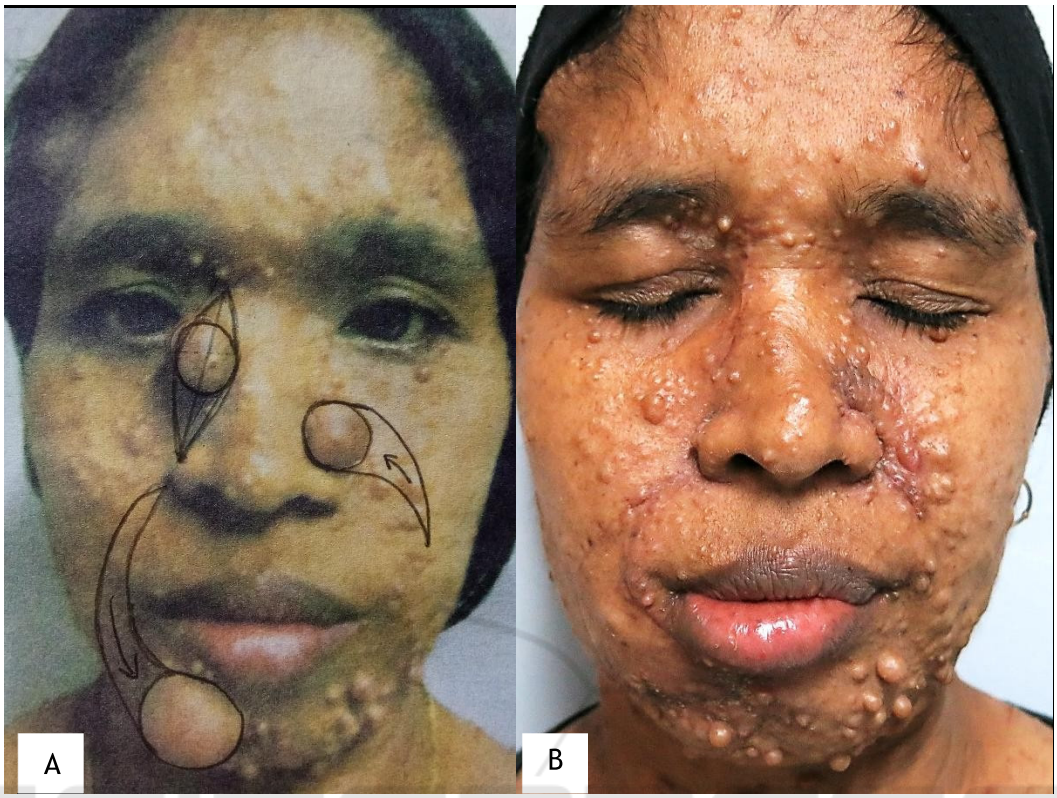

Figure 1. Design operations (A) eliptical on the upper right side of nose, subcutaneous tissue pedicle island fiap on left nostril, and long inferiorly melolabial transposition flap on right side of chin (B) four weeks after the third operation of the tumors.

SCITEPRESS SCIENCE AND TECHNOLOGY PUBLICATIONS

3 DISCUSSION

The patient has done serial exicions of three big tumors on the upper side of right nose, left nostril and right side of chin. We excised the tumor on the upper right side of nose with eliptical design. The elliptical excision remains an adaptable and essential surgical strategy. Elliptical excision are easily designed and can be adapted to many situations. The classic ellipse is formed by tracing 2 arcs of a circle on the skin. The arcs, which are symmetrical with respect to the midline axis separating them, intersect at their ends to form a convex shape. Commonly used curvature is variable, but typically leads to a 1:3 to 1:4 width-length ratio between the short and long axes of the formed ellipse. Intersection of the arcs an elliptical angle of 30° has been traditionally assumed at the ends (Goldberg & Alam, 2012).

The tumor on the left nostril was excised with subcutaneous tissue pedicle island flap design from the left cheek to the left side of the nose. The two most commonly used local flaps for repair of cutaneous defects of the nose and in which the donor sites of the flaps are confined to the nose are the subcutaneous tissue pedicle island advancement flap and the bilobe flap. The island flap is used for repair of defects located at the anterior aspect of the alar groove. The bilobe flap is used to repair small cutaneous defects of the nasal tip and caudal dorsum. It is based on subcutaneous tissue and portion of the transverse nasalis muscle. A triangular shaped flap with its base making up the cephalic border of the defect is designed with the apex of the flap positioned laterally. The posterior border of the flap rests in the alar groove. The anterior border extends cephalically and slighly medially from the defect and is designed to recruit skin of the nasal side wall. The anterior border then arcs laterally to meet the posterior border in the alar facial sulcus. The nasal skin is undermined widely and the proximal and distal one-third of the flap in undermined in the subcutaneous plane. The central one-third of the flap remains pedicled on the subcutaneous tissue. The flap in undermined only to the degree that there is sufficient mobility to allow the flap to be advanced into the recipient site. The flap is advanced and secured at the recipient site first and

then the donor site is closed to the repair (Baker, 2007).

The tumor on the right side of chin was excised with long inferiorly melolabial transposition flap design. The location and size of defect prevented repairing the wound with a single unipedicle advancement flap because of the inelasticity of the chin skin. A long inferiorly melolabial transposition flap was selected. The flap was design to recruit skin from melolabial fold. It was slightly curved in its linear axis to parallel the melolabial crease. This facilitated placement of the flap donor site scar directly with in the melolabial crease. Because the flap was long relative to the width of the base, the standing cutaneous deformity that formed on transposition of the flap was not excised for fear of compromising the vascularity of the flap (Baker, 2007).

4 CONCLUSION

NF-1 demonstrates a true proliferative process of neuroectodermal tissue and it is need multidisciplinary approach. There were no definitive therapy and surgical therapy only for cosmetic and maintain the function of organ around the tumors. We have done operated of three tumors of NF1 of 32 year-old woman every two weeks on the upper right side of nose, left nostril, and right side of chin with eliptical, subcutaneous tissue pedicle island flap and long inferiorly melolabial transposition flap designs under tumescent surgical anesthesia to maintain the function of organs around the tumors with good cosmetic result.

REFERENCES

Baker, S.R., 2007. Flap classification and design. In: Baker SR, editor. Baker local flap in facial reconstruction. Second editon. Philadelphia; Elsevier Inc, pp. 71-107.

Baker, S.R., 2007. Transposition flaps. In: Baker SR, editor. Baker local flap in facial reconstruction. Second editon. Philadelphia; Elsevier Inc, pp. 133-157.

Burton, S.C., Burkhart, C.N., Goldsmith, L.A., 2012. *Neurofibromatous*. In: Freedberg IM, Eisen AZ, Wolff AK, Austen KF, Goldsmith LA, Katz SI, editors. Dermatology in general medicine. 8th ed. New York: Mc.Graw-Hill, pp. 1679-1686.

Dimitrova, V., Yordanova, V., Pavlova, V., Valtchev, V., Gospodinov, D., Parashkevova, B., & Balabanov., C., 2008. A case of neurofibromatosis type 1. Pleven, Bulgaria. *Journal of IMAB - Annual Proceeding (Scientific Papers), 14*, pp. 1-5.

Ghalayani, P., Saberi, Z., & Sardari, F., 2012. Neurofibromatosis type I (von Recklinghausen's disease): A family case report and literature review. *Dental research journal*, *9*(4), pp. 483-488.

Goldberg, L. H., & Alam, M., 2004. Elliptical excisions: variations and the eccentric parallelogram. *Archives of dermatology, 140*(2), pp. 176-180.

Moraes, F. S., Santos, W. E. D. M., & Salomão, G. H., 2013. Neurofibromatosis type I. *Revista Brasileira de Oftalmologia*, *72*(2), pp. 128-131.

SCITEPRESS SCIENCE AND TECHNOLOGY PUBLICATIONS