

Reduction Surgery of Giant Hemifacial Neurofibromas: A Case Report

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Abstract

Neurofibromatosis type 1 (NF-1), first described by Von Recklinghausen (1882), is an autosomal dominant disease caused by a spectrum of mutations in NF-1 gene. We reported a case of giant facial neurofibromas in 26 years old male patient without family history of neurofibromatosis type 1. Features typical of neurofibroma, including an enlarged nerve fascicle composed of elongated nuclei and scant cytoplasmic cells, were identified. Excision was performed with preservation of parotid duct. The mass weight was about 5,5 kg. There were some challenges during the procedure, such as bleeding, hypovolemia, and preservation of vital structure.

Key words: Giant facial neurofibromas, benign tumour, neurofibromatosis type 1, surgical reduction

Introduction

Giant facial neurofibromas cause functional and neurological damage and deficits. Surgical resection is the mainstay of treatment and is a major challenge for surgeons with regard to restoration of defects after tumor resection. We presented a case of surgical reduction of facial neurofibroma of an enormous size and difficulties that posed a rare challenge for the surgeons. In this case report, the reduction was

performed in an elliptical design, on the outer edge of the lateral canthus of the right orbital.

Case Presentation

We reported a case of 2016, a giant facial neurofibromas in 26 years old male patient without family history of neurofibromatosis type 1. Written informed consent was obtained from the patient for publication of this case report and any accompanying images. Family history did not reveal any similar complaints in immediate or distant relatives. The patient was from a low social economic family, graduated from junior high school, unemployed.

He was found by health workers during a social service. Due to a lack of information, he had never sought medical opinion before. Even so, he had high

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hopes for mass reduction surgery. From the results of the examination, the function of the right facial nerve was reduced. The ophthalmologist said the visual acuity in the right eye was within normal limits, except for the ptosis of the palpebra. There was café au lait all over his back and in the axillary region. General health conditions were within normal limits.



Figure 1. Frontal view before operation

Pre-operation

We had provided an explanation beforehand and the patient had understood the possible reduced function of the right facial nerve as a result of the surgery. We had designed a reduction incision near the lateral canthal area of the right eye. We had made the design by considering the possible bleeding that could occur.



Figure 2. Designing the reduction

During Operation

The anesthetist had prepared 3 bags of Whole Blood (WB) and 3 bags of Pack Red Cell (PRC). The blood pressure was 153/78 mmHg and pulse 92x/m. Prolene 1.0 was inserted to the orifice of parotid duct, but we had trouble for further insertion. The reduction was performed in 4 hours by 3 operators, 2 assistants, 1 anesthetist, 2 anesthetist assistants, and 1 circulating nurse. During surgery, enlarged nerve fascicles, dilated blood vessels and brownish patches all over the tissue were found. The fluid was entered through 2 IV lines. Total blood loss was 3500 ml, post-operative blood pressure was 121/64, and the pulse was 88x/m. The administered fluid were 2500 ml Ringer Lactate + 1000 ml normal Saline, 1000 ml Voluven, 700 ml WB, and 230 ml PRC. Then, the hemovac drain was inserted.



Figure 3. The reduction mass, weighing 5.5 kg.



Figure 4. After suturing

Post Operation

On day 1, the hemoglobin level was about 8, the patient was administered with four bags of PRC. On day 4, the hemoglobin level was about 10, and the hemovac drain was taken out. The patient was allowed to discharge from hospital on day 5.



Figure 5. After 1 year

Discussion

Neurofibromatosis type 1 (NF1), affecting approximately 1 in 3000 individuals, is one of the most common inherited genetic conditions¹. NF1 is principally associated with cutaneous, neurologic, and orthopedic manifestations² that can cause damaging functional and cosmetic effects. The diagnosis of NF1 has been relied primarily on clinical criteria due to its high degree of accuracy and the absence of reliable molecular tests. Genetic testing can be particularly helpful for patients who present with an unusual phenotype or an incomplete clinical picture³.

It is said that NF1 is caused by a mutation in NF1 gene that encodes tumor-suppressing neurofibromin

protein, a Ras inactivator. It can be classified according to anatomical location: cutaneous, subcutaneous, intraneural, and plexiform. Plexiform neurofibromas (PN), histologically benign tumors of peripheral nerves which arise from Schwann cells, occur in 20%–50% of all patients with NF1⁴. The nerves are transformed into a thick convoluted mass, which is likened to a bag of worms. Treatment of PN mainly consists of symptom management and / or surgical resection⁵.

However, these tumors involve nerves, blood vessels, or other internal organs, thus complicating surgery with often incomplete resections followed by tumor regrowth, or morbidity. The challenges during the operation was how to keep hemodynamics stable and how to perform all actions as measured and as quickly as possible. Although there have been preclinical and clinical studies of several molecularly targeted compounds, no medical or therapeutic treatment has been approved to date for PN⁶. Continuous care of the patient is the main management of NF1 by monitoring clinical manifestations according to age, with the aim of early recognition and treatment of symptomatic complications².

Thus has been reported a case report of giant hemifacial neurofibromas, with an enormous size. The reduction has been done by considering the speed of surgery, also the possible amount of blood loss, so that the patient could recover early with optimal results. However it may still needed further reduction, but the amount of mass removed will be much smaller. In conclusion, this case report can be used as a reference in other cases of reduction of facial neurofibroma.

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Ethical Clearance: Taken from Bina Sehat Hospital committee

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