

NEUROFIBROMA TYPE 1 WITH INTRACRANIAL INFILTRATION

A CASE REPORT

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ABSTRACT

Neurofibromatosis (NF) is an AD dysplasi of mesoderm and ectoderm characterized by numerous neurofibromas and cafe-au-lait spot. It can develop on cranial and cavum orbita. Additional abnormalities such as episode of seizure, learning disabilities, and speech difficultie may present. Presented a rare case that has underwent surgery by multidisciplinary team consist of neuro surgeon, plastic surgeon, and ophthalmologist. A 31 years old female patient admitted to Dr. Soetomo Hospital with mass at left face since she was 3 years old. Clinical examination shows a large plexiform mass at left hemifacial, distopia left oculi, and facial deformity. Radiological examination shows a soft tissue mass destructing the left temporal and maxillary bone, a temporal lobe herniation to frontobasal, and a suppression on left optical nerve. In the present time, the patient is satisfied with her performance and vision. The result of surgery is satisfying. (QM 2017;01:44-47)

Keyword : Neurofibroma type 1, intracranial neurofibroma

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ABSTRAK

Neurofibromatosis (NF) adalah dysplasi AD dari mesoderm dan ektoderm ditandai dengan sejumlah Neurofibroma dan bercak cafe-au-lait. Ia dapat tumbuh pada tulang tengkorak dan cavum orbita. Kelainan tambahan seperti episode kejang, ketidakmampuan belajar, dan kesulitan bicara dapat ditemukan. Sebuah kasus langka yang telah dilakukan operasi oleh tim multidisiplin yang terdiri dari neuro bedah, bedah plastik, dan dokter mata., yaitu seorang pasien wanita berusia 31 tahun dirawat di Rumah Sakit Dr. Soetomo dengan massa di wajah sebelah kiri sejak ia berusia 3 tahun. pemeriksaan klinis menunjukkan massa plexiform besar pada hemifacial kiri, distopia meninggalkan oculi, dan deformitas wajah. Pemeriksaan radiologi menunjukkan massa jaringan lunak merusak diri tulang kiri temporal dan maksila, lobus herniasi temporal frontobasal, dan penekanan pada saraf optik kiri. Pada saat ini, pasien puas dengan kinerja dan visi. Hasil operasi adalah memuaskan. (QM 2017;01:44-47)

Kata kunci : neurofibroma tipe I, neurofibroma intrakranial

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INTRODUCTION

Plexiform neurofibroma occurred in the head and neck region can be disfiguring, causing cosmetic embarrassment to the affected patients. Surgical management is the mainstay of therapy. It is challenging because of the vascularity, the risk of primary hemorrhage, and infiltration of facial soft tissues as well as the need to preserve functions and maintain aesthesis

Neurofibromatosis is an autosomal dominant disorder involving both the central and peripheral nervous system. Its clinical mark is the development of multiple cutaneous and subcutaneous nodular tumor (Shin, J., Persing, JA., Throne, CH 2007, ,Kapur, S., Michael, BL 2013).

The hallmark of the NF-1 disease is tumors along the nerve sheath from the dorsal root ganglion to the terminal nerve branches. Neurofibromas divided into five main types: localized cutaneous neurofibromas, diffuse cutaneous neurofibromas, localized intraneural neurofibromas, massive soft-tissue neurofibroma, and plexiform neurofibromas (Shin, J., Persing, JA., Throne, CH 2007).

Plexiform neurofibromas are practically unique as NF-1 due to its composition of nerve sheath cells that proliferate along the length of nerve. Plexiform neurofibromas are frequently associated with hypertrophy of the soft tissue and hyperpigmentation or hypertrichosis of the overlying skin. Their growth possibly causing

destruction or compression of local tissue and furthermore causing significant morbidity. Plexiform lesions occur in 16% to 40% patients with NF-1, and are found on the trunk in 43% to 44% patients, the extremities in 15% to 38% patients and in the head and neck in 18% to 42% of patients (Creange, A. et all 1999, Kapur, S., Michael, BL 2013).

A CASE REPORT

This retrospective study was performed in Plastic Reconstructive and Aesthetic Surgery Department in Dr. Soetomo Hospital Surabaya reporting a patient with intracranial infiltrated Neurofibroma type 1. This patient underwent surgery involving multidiciplinary team consist of plastic surgeon, neuro surgeon and ophthalmologist..

A 31 years old female patient admitted to Dr. Soetomo Hospital with plexiform mass on her face and multiple nodules on her upper and lower extremity since she was 3 years old. She complained about the deformity on her face and blurred vision on her left eye and intermittent headache



Figure.1. ed left temporal bone, lateral wall of the left orbit, maxillary and mandible bones, also left temporal lobe herniation to left orbital cavity^(Rahmania D, 2010)

The patient underwent surgery with coronal approach extend to facelift incision

(Figure 2). During surgery, a mass on subgaleal layer infiltrated to left temporal fascia is found. The mass moreover destruct the left temporal, left fronto zygoma and left zygoma arch bone. The left temporal lobe herniation to left orbital cavity led to a suppression onto left oculi (Figure 3). A neurofibroma reduction and left lateral cantopexy has been performed to maintain simetrical position of the left orbit. The mass was successfully removed then leaving a defect on the left temporal side with duramater and herniation temporal lobe in left orbital cavity as its base. In this condition, the exploration of left orbital cavity and positioning the temporal lobe to the normal position can't be performed since the increase of intra cranial pressure might occur. The surgical site closed after drain was inserted (Figure 4).



Figure 2. Desain Incision pre operative (Rahmania D, 2010)



Figure 3. Temporal defect (Rahmania D, 2010)



Figure 4. Post Operative : Top : after reductive excision of neurofibroma, skin excess was left. Bottom : after left lateral cantopexy and reducing skin excess. (Rahmania D, 2010)

3 days after surgery, significant changing of of the patient appearance was gained. The function of left N.VII was decrease, but contraction response of maxilla and mandibular nerve branches was present.

RESULT AND DISCUSSION

From the descriptive study above, the management of Neurofibromas with intracranial infiltration should be performed by multidisciplinary team. Reconstruction for the destructed bone will be performed later. Generally the cosmetic appearance of this patient is very satisfactory.

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