Esthetic subunit approach in massive facial plexiform neurofibroma: a case report

M. de la Torre Macías, B. Berenguer Frohner, C. Lorca García, E. de Tomás Palacios

Pediatric Plastic Surgery Unit. Pediatric Surgery Department. Gregorio Marañón General University Hospital. Madrid (Spain).

ABSTRACT

Plexiform neurofibroma is a presentation of neurofibromatosis type 1 (NF1) which can cause great facial deformities. Treatment rarely has a healing effect, so the surgical approach is aimed at improving esthetics and function. It requires a cross-disciplinary approach and typically needs multi-stage surgery.

This is the case of a 16-year-old male patient with NF1 presenting with left periorbital and malar facial plexiform neurofibroma with slow-growth intraconal and extraconal invasion.

He presented at the plastic surgery consultation for facial soft tissue deformity correction. Removal was performed using an esthetic subunit approach, with canthopexy and orbital cavity reconstruction, resulting in facial region symmetrization. This allowed for remarkable esthetic and functional improvement, facilitating ocular prosthesis adaptation. The subsequent use of selumetinib allowed the lesion to be stabilized.

KEY WORDS: Neurofibromatosis type 1; Plexiform neurofibroma; Esthetic subunit approach; Ocular prosthesis.

ABORDAJE POR SUBUNIDADES ESTÉTICAS EN EL NEUROFIBROMA PLEXIFORME FACIAL MASIVO: A PROPÓSITO DE UN CASO

RESUMEN

Los neurofibromas plexiformes son una forma de presentación de la neurofibromatosis tipo 1 (NF1) que pueden originar grandes deformaciones faciales. El tratamiento de estas tumoraciones casi nunca es curativo, el abordaje quirúrgico tiene por objetivo mejorar la estética y la función. Requiere un abordaje multidisciplinar y suele necesitar cirugía por etapas.

Se presenta el caso de un paciente varón con NF1 que presenta un neurofibroma plexiforme facial periorbitario y malar izquierdo con invasión intra y extraconal de crecimiento lento.

Acude con 16 años a la consulta de cirugía plástica para corrección de las deformidades faciales de partes blandas. Se realiza exéresis mediante abordaje por subunidades estéticas, realizando cantopexia y reconstrucción de la cavidad orbitaria, resultando en una simetrización de la región facial. Con ello se obtiene una notable mejoría estética y funcional, facilitando la adaptación de la prótesis ocular. El uso posterior de selumetinib ha permitido estabilizar la lesión.

PALABRAS CLAVE: Neurofibromatosis tipo 1; Neurofibroma plexiforme; Abordaje por subunidades estéticas; Prótesis ocular.

INTRODUCTION

Neurofibromatosis is a genetic condition characterized by the occurrence of cutaneous neurofibroma, with two variants: type 1, when located at the level of the peripheral nerves, and type 2, when located at the level of the central nervous system[1].

Plexiform neurofibroma is a benign hamartomatous tumor. At the craniofacial level, it is typically unilateral, extending from the branches of the trigeminal, facial, or glossopharyngeal nerves[2].

We present the case of a 16-year-old male patient with massive facial plexiform neurofibroma (MFPN) and the therapeutic approach used.

CLINICAL CASE

Male patient with multiple café au lait spots and left optic glioma at birth associated with congenital glaucoma and left buphthalmos. He was diagnosed with NF1.

During follow-up in another healthcare facility, and given the ocular involvement present, he required various goniotomies and trabeculectomies. When he was two and a half years old, full left eye enucleation and scalp neurofibroma removal were carried out, which required subsequent surgical revisions.

Correspondencia: Dr. Manuel de la Torre Macías.
E-mail: manueldltm@gmail.com

Date of submission: September 2019 *Date of acceptance:* March 2020

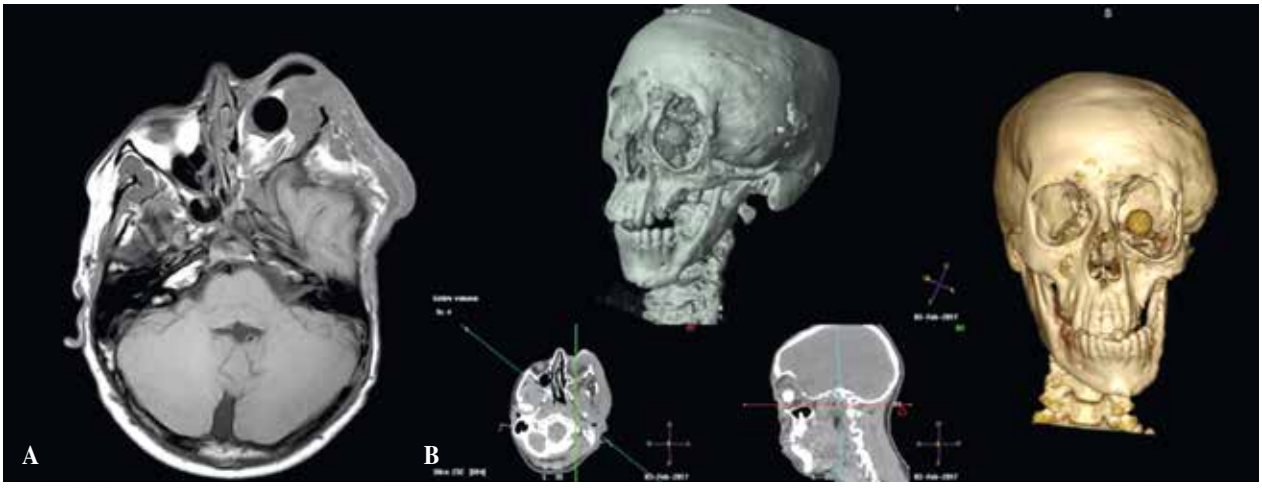

Figure 1. Preoperative images: A) MRI coronal cut, demonstrating a large left hemifacial tumor. B) Bone deformity 3D reconstruction.

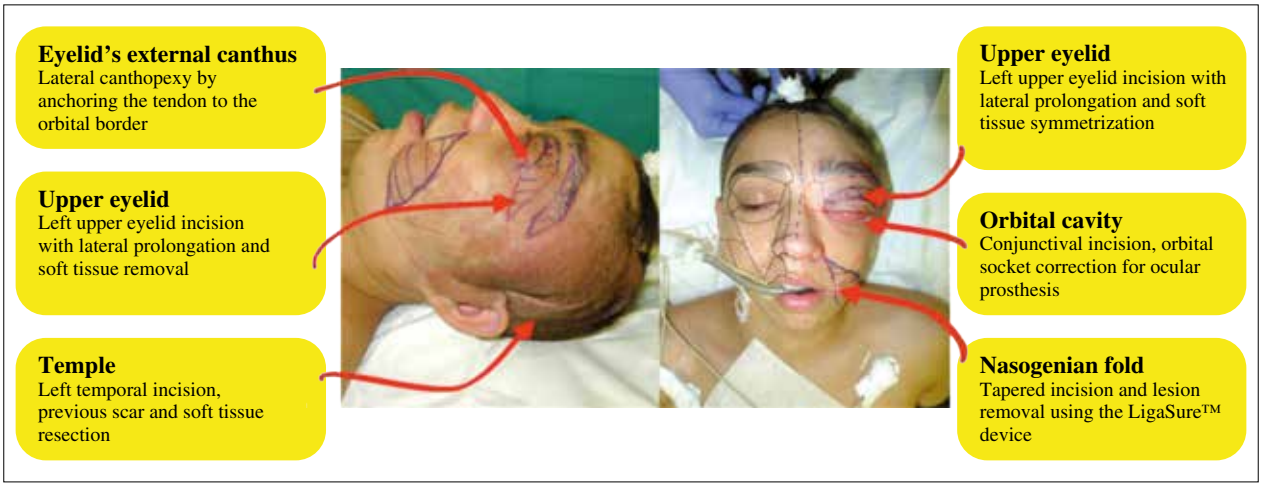

Figure 2. Preoperative surgical marking: esthetic subunit approach. Delimitation of cutaneous resection areas.

A soft tissue increase was recorded at the left malar and periocular level, which caused great deformity. When he was 16, he was referred to our consultation for assessment purposes. A radiologic study using CT-scan and MRI (Fig. 1) was performed. It showed left cervico-facial multi-compartmental plexiform neurofibroma with orbital, malar, parotid, and pre-auricular involvement of the mastication and parapharyngeal space, with intracranial extension and involvement of the orbital branches of the trigeminal nerve (V1, V2, and V3). No vessels eligible for embolization were found. The case was presented at a cross-disciplinary session, including Pediatric Neurology, Dermatology, Genetics, Oncohematology, Radiology, Maxillofacial Surgery, and Pediatric Plastic Surgery departments. A first surgery was jointly decided upon to improve facial deformity.

Preoperative surgical marking of facial esthetic subunits was key in the procedure. Comparing the facial units from the diseased side with those from the healthy side helped decide which tissue should be removed and which incisions should be carried out (Fig. 2). The objective was to reduce such great hypertrophy, increase symmetry, and hide incisions in the natural expression lines.

Surgery was performed under general anesthesia using orotracheal intubation. Antibiotic prophylaxis (amoxicillin-clavulanic acid) was administered. For analgesic and bleeding control purposes, a tumescent infiltration with 1% lidocaine and 1 mg/L adrenalin was carried out using a subcutaneous needle. Resection was conducted using the LigaSure™ device (Valleylab, Tyco International Healthcare, Boulder, CO)[3].

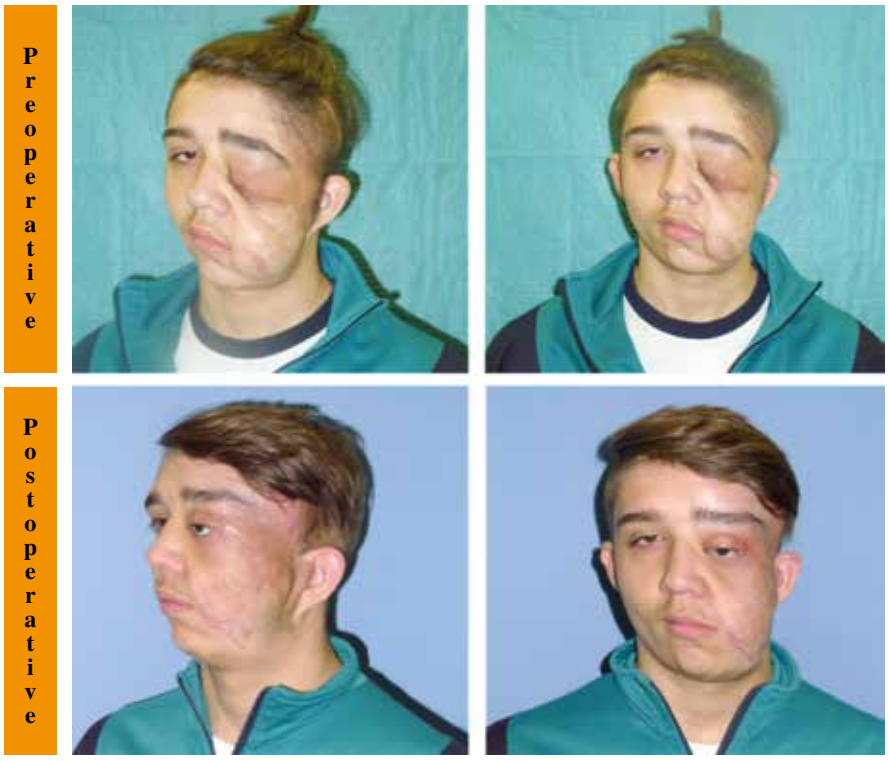

Figure 3. Comparison between preoperative and postoperative appearance.

The ocular region was approached through upper eyelid blepharoplasty, with a lateral prolongation until the temple. A tapered excision of the eyebrow deformity was carried out, and an extensive lateral canthopexy was performed while anchoring the canthal tendon to the orbital border with 4/0 non-absorbable monofilament suture to symmetrize the axis of both eyelid incisions and improve prosthesis position.

The soft tissues were resected using a transconjunctival approach to normalize the orbital cavity, while in situ verifying that the prosthesis was well adapted. The tapered incision at the level of the left nasogenian fold allowed the malar area to be approached. At the left temporal level, the previous scar was resected, and the volume of the frontal area, the zygomatic area, and the temple was reduced. Careful hemostasis and suture by planes were carried out using 4/0, 5/0, and 6/0 absorbable monofilament sutures. For the skin, 5/0 absorbable monofilament intradermal continuous suture was used. The scalp area was stapled and reinforced using 3/0 interrupted monofilament stiches.

The patient did not require blood product transfusion, and he was discharged after 48 hours, with oral analgesic and antibiotic treatment for one week.

Pathological examination confirmed the resection of tissue borders compatible with MFPN, without malignity signs.

Stitches were removed one week following surgery, and staples were removed 10 days postoperatively. The incisions were left covered with Steri-Strip™ and Micropore™ (3M, St. Paul, MN) paper tape for one month.

The patient had no postoperative complications, with a favorable mid-term situation (Fig. 3). He claimed to be highly satisfied with the result. During follow-up, Selumetinib was administered for lesion control and stabilization purposes. After a 3-year follow-up, no re-interventions have been required so far, and ocular prosthesis adaptation remains correct.

DISCUSSION

Neurofibromatosis type 1 or Von Recklinghaussen's syndrome is a dominant autosomal disorder with variable penetration caused by mutations in NFT1, which is located in chromosome 17q11.2[1]. These mutations bring about benign hamartomatous tumors at the level of the peripheral nerves called neurofibromas[1,4]. Although they are typically located in the skin, they can also be present in other tissues. They start to occur at puberty, with endocrine influence playing a role in neurofibroma development[5].

NF1 expresses in various forms, from small neurofibromas to large deformities caused by plexiform neurofibroma (PN)[1]. PN is a subtype of neurofibroma with the following characteristics: it is multiple (as it involves various thickened nerves), it is recurrent, and it consists of a mix of fibroblasts, Schwann cells, axons, and numer-

ous collagen fibers. 1% of NF1 patients are estimated to have facial involvement in the form of MFPN, defined as a >25% involvement of the facial surface[6].

MFPN typically arises in the orbital-temporal region, involving the socket, the eyes, the eyelids, the lips, the oral commissure, the facial muscles, the nerves, and the bones. It has high malignization potential (3.5%) in adults. In children, it grows faster and is associated with higher malignization risk (up to 20%)[7].

Treatment requires multiple specialists: neurologists, pediatricians, dermatologists, and surgeons from different specialties, according to the level of involvement. The objective is to achieve esthetic, functional, and preventive improvement, and occasionally healing[8,9]. In general, deformity is the most worrisome occurrence for NF1 patients[10].

Preoperative MFPN planning is typically carried out through CT-scan and MRI to determine bone and soft tissue structure involvement. These tests also prove useful during follow-up[1].

In pediatric patients, surgery tends to be deferred until the lesions have been stabilized in order to reduce the number of procedures. In case of malignization suspicion or facial structure compromise, it can be rescheduled to an earlier date[9].

Given its benign character, complete resection with oncologic margins is only justified in case of confirmed malignity. Therefore, MFPN surgery is typically intralesional and should respect facial innervation and functional structures[11].

Surgical alternatives based on "lifting" or facial suspension procedures are not indicated owing to the lack of elasticity, excess of extensibility, and alteration of the microvasculature of the skin involved. They provide with insufficient surgical results, they increase the number of procedures, and they are associated with higher risk of bleeding[1], since wide dissections are required.

Consequently, some authors advocate the use of previous lesion embolization[12]. However, given that neurofibroma is not always connected with a single arterial branch, embolization is sometimes unfeasible. Therefore, the risk of bleeding should be considered, blood products should be made readily available, and intraoperative bleeding should be reduced by planning ahead and using cutting and electrocoagulation devices such as LigaSure™ (Valleylab, Tyco International Healthcare, Boulder, CO)[3].

Recently, the use of Selumetinib, a selective inhibitor of type 1 and type 2 MAP kinases, has been described. When administered orally, Selumetinib seemingly reduces progression and avoids early MFPN recurrence[12]. However, the surgical approach is still required when it comes to reducing large lesions.

Hivelin et al. described the esthetic subunit approach in MFPN, which allows ptotic deformity to be treated effectively[11]. Facial esthetic subunits are topographic divisions that allow asymmetries and facial disharmonies to be analyzed. The face is divided into segments (Fig. 4) with similar skin shapes and textures inside, establishing bordering lines known as facial shadows where scars can be more easily hidden[13].

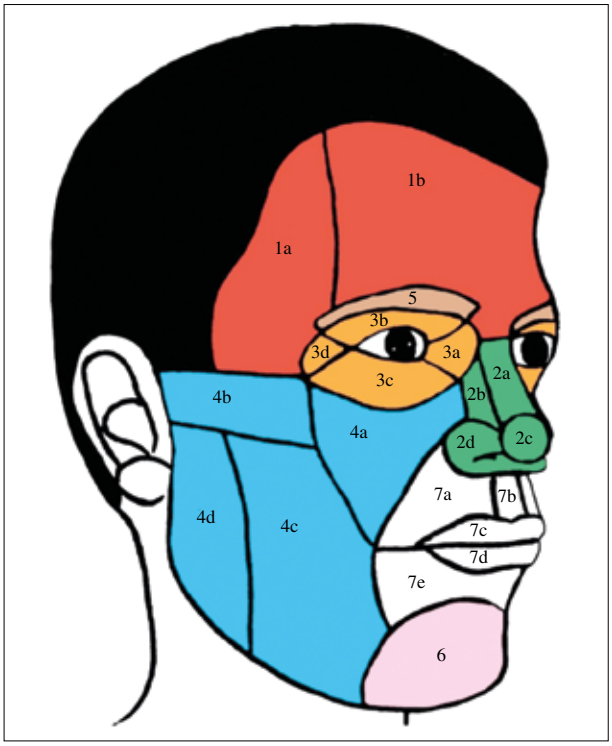

Figure 4. Facial esthetic subunits: red, frontal units: temple (1a), central frontal region (1b), and eyebrow (5); green, nasal region: nasal dorsum (2a), lateral wall (2b), tip (2c), and wing (2d); orange, periocular region (3a), upper eyelid (3b), lower eyelid (3c), and external canthus (3d); blue, nasal region: infraorbital (4a), zygomatic (4b), buccal (4c), and parotid (4d); pink, chin (6); white, perioral region: lip side (7a), philtrum or Cupid's bow (7b), upper vermilion (7c) and lower vermilion (7d), and lower lip (7e).

The postoperative use of Steri-Strip™ or Micropore™ (3M, St. Paul, MN) paper tape has been demonstrated to reduce the risk of scar narrowing, as it diminishes tension at the injury borders and minimizes tension forces[14].

In large facial tumors, multi-stage surgery and separate analysis of the various elements involved allows for a safe surgical approach[6]. In addition, the risk of re-intervention is higher in large >4.5 cm MFPNs or in MFPNs involving the periorbital tissue, especially when canthopexy is required[15].

CONCLUSIONS

Neurofibromatosis is a complex condition which requires cross-disciplinary management. Today, Selumetinib is considered the treatment of choice, since it prevents

progression and avoids early recurrence. Surgery is indicated in refractory cases – those causing great deformity or posing a risk to life. It is typically indicated right after completing growth, but it can be performed at an earlier age in case of malignization suspicion or great functional and/or esthetic alteration. Surgical planning should be carried out through CT-scan and MRI, while considering the possibility of bleeding. The esthetic subunit approach with multi-stage surgery provides with satisfactory results. Massive and potentially mutilating resections should be avoided.

REFERENCES

1. Jett K, Friedman JM. Clinical and genetic aspects of neurofibromatosis 1. Genet Med. 2010; 12(1): 1-11.
2. Hivelin M, Wolkenstein P, Lepage C, Valeyrie-Allanore L, Meningaud JP, Lantieri L. Facial aesthetic unit remodeling procedure for neurofibromatosis type 1 hemifacial hypertrophy: Report on 33 consecutive adult patients. Plast Reconstr Surg. 2010; 125: 1197-207.
3. Konno, E, Kishi, K: Use of LigaSureTM vessel sealing system in neurofibroma excision to control postoperative bleeding. J Plast Reconstr Aesthet Surg. 2012; 65: 814-7.
4. DeBella K, Szudek J, Friedman JM. Use of the national institutes of health criteria for diagnosis of neurofibromatosis 1 in children. Pediatrics. 2000; 105: 608-14.
5. Hirbe AC, Gutmann DH. Neurofibromatosis type 1: a multidisciplinary approach to care. Lancet Neurol. 2014; 13: 834-43.
6. Bogdasarian RN, Fientisch AM, Langer PDand Datiashvili RO. Massive Facial Neurofibromas: Review of the Literature, New Classification System and Case Presentations. Ann Surg Perioper Care. 2016; 1(2): 1007.
7. Lee V, Ragge NK, Collin JR. Orbitotemporal neurofibromatosis. Clinical features and surgical management. Ophthalmology. 2004; 111: 382-8.
8. Avery RA, Katowitz JA, Fisher MJ, Heidary G, Dombi E, Packer RJ, et al. Orbital/periorbital plexiform Neurofibromas in children with Neurofibromatosis type 1: multidisciplinary recommendations for care. Ophthalmology. 2017; 124: 123-32.
9. Hua C, Zehou O, Ducassou S, Minard-Colin V, Hamel-Teillac D, Wolkenstein P, et al. Sirolimus Improves Pain in NF1 Patients With Severe Plexiform Neurofibromas. Pediatrics. 2014; 133(6): e1792-7.
10. Wolkenstein P, Durand-Zaleski I, Moreno JC, Zeller J, Hemery F, Revuz J. Cost evaluation of the medical management of neurofibromatosis 1: a prospective study on 201 patients. Br J Dermatol. 2000; 142: 1166-70.
11. Vélez R, Barrera-Ochoa S, Barastegui D, Pérez-Lafuente M, Romagosa C, Pérez M. Multidisciplinary management of a giant plexiform neurofibroma by double sequential preoperative embolization and surgical resection. Case Rep Neurol Med. 2013; 2013: 987623.
12. Dombi E, Baldwin A, Marcus LJ, et al. Activity of Selumetinib in Neurofibromatosis Type 1–Related Plexiform Neurofibromas. N Engl J Med. 2016; 375(26): 2550-60.
13. Fattahi TT. An overview of facial aesthetic units. J Oral Maxillofac Surg. 2003; 61(10): 1207-11.
14. Atkinson JA, McKenna KT, Barnett AG, McGrath DJ, Rudd M. A randomized, controlled trial to determine the efficacy of paper tape in preventing hypertrophic scar formation in surgical incisions that traverse Langer's skin tension lines. Plast Reconstr. Surg. 2005; 116: 1648-56.
15. Pessis R, Lantieri L, Britto JA, Leguerinel C, Wolkenstein P, Hivelin M. Surgical care burden in orbito-temporal neurofibromatosis: Multiple procedures and surgical care duration analysis in 47 consecutive adult patients. J Craniomaxillofac Surg. 2015; 43: 1684-93.