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Plexiform Neurofibroma: Case Report

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Abstract **Case Report**

Plexiform neurofibroma (PNF) is a benign nerve tumor, most often developing in the cephalic region. It is usually considered pathognomonic for neurofibromatosis type 1 (NF1) or Von Recklinghausen disease. We report the case of a patient treated in our training for plexiform neurofibroma on NF1. This is a 28-year-old patient, without any particular ATCD, who consulted for a left scalpo-fronto-palpebral subcutaneous swelling that had been present for 12 years, causing major aesthetic discomfort. Furthermore, it presented numerous (> 6) café au lait spots greater than 15mm in diameter. The patient underwent excision of the left scalpo-fronto-palpebral tumor under general anesthesia with direct closure. The pathological examination confirmed the diagnosis of plexiform neurofibroma. The postoperative course was simple and the result satisfactory. Surgical excision remains the only treatment for plexiform neurofibomas in Von Recklinghausen disease.

Keywords: Plexiform neurofibroma – Neurofibromatosis – Von Recklinghausen disease.

INTRODUCTION

Von Recklinghausen disease or neurofibromatosis type 1 (NF1) is an autosomal dominant phacomatosis that affects approximately 1 in 4000 individuals [1]. There are several clinical expressions but the pathognomonic sign is the presence of benign tumors developed at the expense of the perineurium of the peripheral nerves: neurofibromas. We report a case of scalpo-fronto-orbital plexiform neurofibroma in a patient treated in our department.

OBSERVATION

This is a 28-year-old patient, without any particular ATCD, who presents with a left scalpo-fronto-palpebral subcutaneous swelling, soft, depressible, painless (fig 1). There was no underlying skeletal deformity. This tumor has been gradually evolving over the past 15 years. It is responsible for major aesthetic discomfort for the patient, and also for functional discomfort (left ptosis). He had no other similar lesions.

Furthermore, he presented numerous (more than 6) café-au-lait spots greater than 15mm in diameter, scattered over the entire body surface, predominantly in the chest area. An ophthalmological examination revealed the presence of Lisch nodules. Given all these elements, the diagnosis of Neurofibromatosis type 1 was made.

The patient underwent excision of the left scalpo-fronto-palpebral tumor under general anesthesia with direct suture (figs 2 and 3). Pathological examination confirmed the diagnosis of plexiform neurofibroma. The postoperative course was simple. The result was satisfactory. The patient is regularly followed in our training, there is a stabilization of the result and no sign of a new tumor.

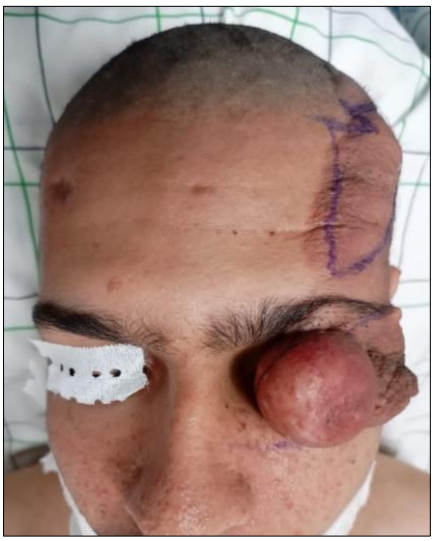

Fig. 1: Scalpo-fronto-orbital plexiform neuroma (front view)

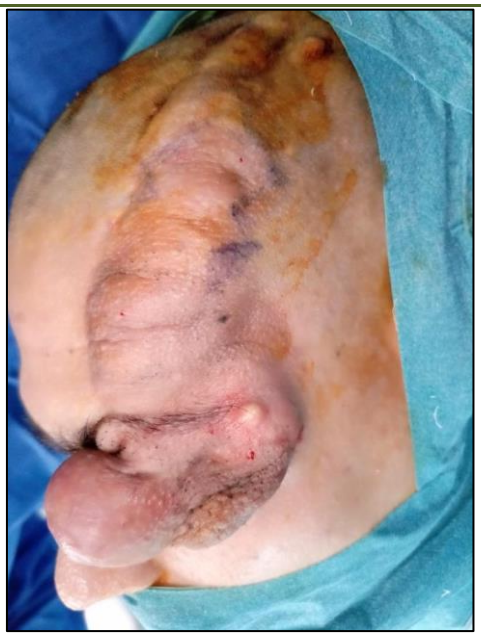

Fig. 2: Scalpo-fronto-orbital plexiform neuroma (lateral view)

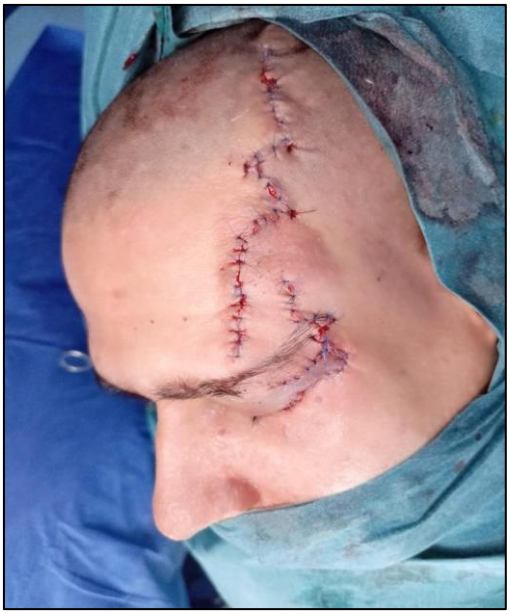

Fig. 3: Result on table after excision (lateral view)

DISCUSSION

Neurofibroma is a benign tumor resulting from a proliferation of Schwann cells and fibroblasts along peripheral nerve threads. Plexiform neurofibroma is considered pathognomonic for neurofibromatosis type 1 by some [2], and not by others [3, 4]. It occurs in 3-7% of cases at the level of the cephalic extremity [5].

Surgery remains, to date, the only effective treatment. The question that arises is the timing of surgery. Some opt for early surgery [6-8], with the aim of slowing the progression of the tumor and avoiding complications (meningoencephalocele in the orbital form). Others, however, opt for surgery in the post-pubertal period to wait for the lesions to stabilize [9]. And still others opt for later surgery in order to reduce the number of iterative interventions [10, 11].

Neurofibroma surgery is a difficult surgery, generally due to the extent of the lesions and the major bleeding risk. Extensive lesions which generally affect several aesthetic units of the face sometimes justify the need to multiply the procedures in order to obtain a satisfactory result.

If left unoperated, the risk of malignant transformation of a plexiform neurofibroma (most often into a sarcoma) is estimated at 2 to 5% of lesions in the context of NF1 [12]. Hence also the interest in surgery.

CONCLUSION

Plexiform neurofibroma is a typical NF1 lesion. Its diagnosis is clinical and easy, confirmed by pathological examination. It is responsible for aesthetic and sometimes functional discomfort, justifying its treatment. Its treatment, to date, remains surgery. This surgery generally requires several interventions, taking certain precautions to guarantee a satisfactory result and the safety of the patient.

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