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GIANT NEUROFIBROMA OF THE LABIA MAJORA, EXCISION WITH A MODELLING TECHNIQUE

Giulia Argentino, Simone La Padula, Oana Hermeziu, Rosita Pensato, Jean Paul Meningaud, Barbara Hersant

Department of Plastic, Reconstructive and Maxillo facial Surgery, Henri Mondor Hospital, University Paris XII, Créteil, France

Summary

Background. Neurofibromatosis type 1 (NF1) is an autosomal dominantly inherited neurocutaneous disease caused by a mutation in the neurofibromin gene on chromosome 17q11.22. NF1 is a multisystem disease, and patients with this condition are at an increased risk of developing both benign and malignant tumors of the central and peripheral nervous systems. Although rare, NF can affect the genital tract, with the vulva being the most common site and involvement of the vagina, cervix, and ovaries being reported less frequently. Genital involvement often causes pain and psychological discomfort for patients, and surgical removal is the mainstay of treatment due to the large innervation of the area.

Material and methods. This work describes the case of a 41-year-old patient with a known diagnosis of NF1 who presented with a large painful mass in the left labium majus. The mass was surgically treated with a modeling excision technique.

Results. At the six-month follow-up visit, the patient reported being very satisfied with the results, and no complications were observed.

Conclusions. Conservative modeling surgical excision appears to be the most appropriate technique for managing giant genital NF. This technique minimizes the risk of bleeding, restores correct anatomy and sexual function, and has a positive impact on the patient's psychological well-being.

Key words: neurofibromatosis type 1, genital neurofibromatosis, modelling excision, fibrin glue, labia majora neurofibroma, plexiform neurofibroma

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Correspondence
Simone La Padula
Department of Plastic, Reconstructive and Maxillo facial Surgery, Henri Mondor Hospital, University Paris XII, Créteil, France
E-mail: drsimonelapadula@gmail.com

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INTRODUCTION

Neurofibromatosis is a group of rare genetic diseases classified as neurocutaneous syndromes. These conditions primarily affect the skin and nervous system. Among the various forms of neurofibromatosis, NF1 is the most common, with a prevalence of approximately 1 in 3,000 cases. The NF1 gene is located on chromosome 17's long arm, and it regulates the production of a protein called neurofibromin. The disease manifests when this protein malfunctions [1-32].

Typically, the cutaneous symptoms of NF1 consist of neurofibromas and café au lait spots. Genitourinary involvement is rare, and vulvar involvement

is exceedingly rare. Neurofibromas typically appear in small sizes, and complete surgical excision is the preferred treatment when possible. We present the case of a 41-year-old woman who had a giant neurofibroma of the labia majora, which was treated surgically.

MATERIALS AND METHODS

We present the case of a 41-year-old woman with familial NF1, inherited paternally, who presented with perineal masses with genital involvement that had been slowly growing for several years and causing pain. The patient, who had an active sex life, sought evaluation at our clinic for therapeutic options due to discomfort and dyspareunia during sexual intercourse. The patient's medical history revealed a cerebellar glioma, related as an NF manifestation, and a previous history of thrombocytopenia. Physical examination revealed an enlargement of the external genitalia, particularly the labia majora, with a hyperpigmented mass of soft-elastic consistency that was painful on palpation. The largest lesion, a plexiform neurofibroma, was located on the right labium majus and had previously been operated on several times but had relapsed (Fig. 1). A follow-up CT scan of the abdomen and pelvis showed an increase in the size of the right labium majus and a left latero-rectal mass with invasion of the left buttock, suggestive of a neurofibroma measuring 55 x 35 mm and associated with hypertrophy of the labia majora. In recent months, the patient had noticed rapid growth of the left lesion, causing anatomical deformation of the vulvar region. The case was evaluated by a multidisciplinary team. Due to the painful symptoms and rapid growth causing significant relational problems for the patient, we decided to perform palliative surgical removal of the left mass using a modeling technique. Before proceeding, the patient was informed of the increased risk of hematoma and bleeding due to her history of thrombocytopenia and the inability to perform a radical excision of the lesion.

OPERATIVE TECHNIQUE

Surgery was performed under general anesthesia with the patient in a gynecological position. The lesion was located on the labia majora, and a cutaneous lozenge was marked along its entire length. To minimize the risk of bleeding during the surgery, the lesion was infiltrated with a solution containing 2 mg of adrenaline and 2 g of tranexamic acid, which was diluted in 1 L of physiological solution (Fig. 2). The incision was made with a cold blade following the design, and the dissection of the planes was carried out with an electric scalpel and bipolar forceps, with careful haemostasis to prevent intraoperative bleeding. These lesions are difficult to dissect because they lack cleavage planes, and the excision should not include the lateral and deep margins. The dissection was performed in the subcutaneous tissue until satisfactory anatomical appearance was achieved with anatomical restoration of the labium majus.

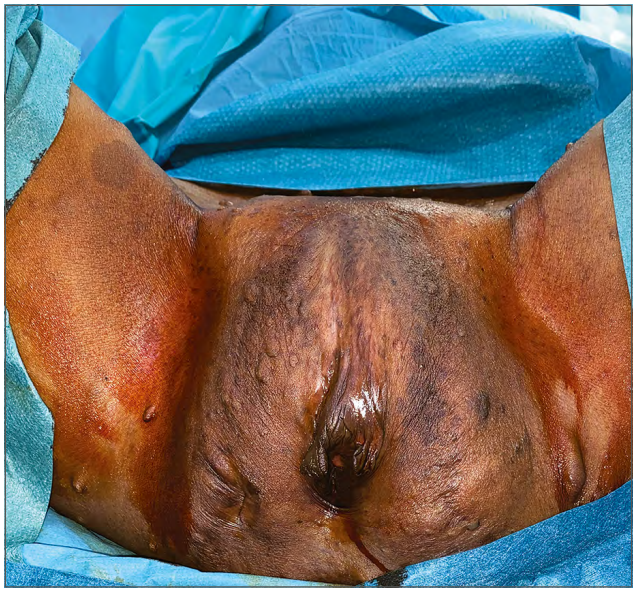

Figure 1. Clinical presentation of giant neurofibroma of the labia majora.

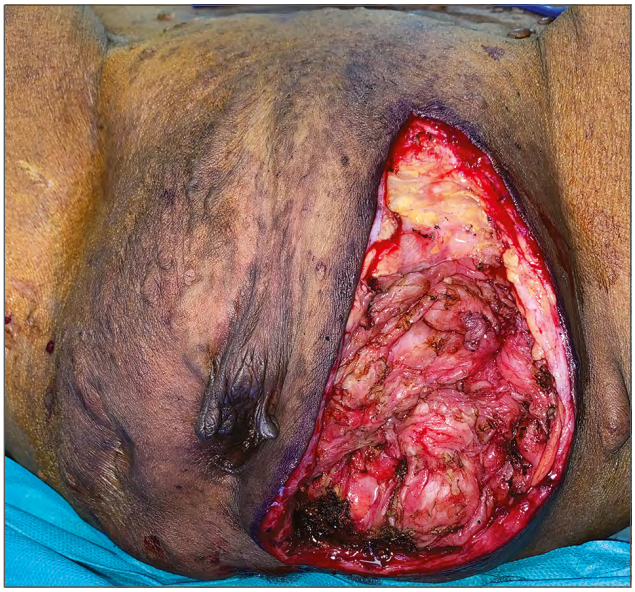

Figure 2. Note the large defect following surgical modelling excision of the lesion.

This excision technique is referred to as the "modelling excision technique" and differs from a simple ellipsoid excision. Its primary objective is to remove a maximal amount of plexiform neurofibroma, and it is used not only in the genital area but also in other locations such as the head and neck. In certain cases, achieving complete removal of the neurofibroma in these areas may be challenging or even impossible [8].

The surgical specimen obtained at the end measured 10 x 6.5 cm. Sections of the mass showed a lesion composed of a mixture of spindle cells and nerve fibers arranged in a haphazard pattern. No atypical or malignant cells were identified. The margins of the specimen were well-defined and appeared to be free of tumor involvement.

The defect was closed by primary intention, using deep anchoring points in Vycril 0, and direct skin suture for simple points in Vycril 2/0.

To minimize the risk of bleeding, we employed autologous platelet-rich plasma (A-PRP) glue.

Preparation of A-PRP glue

The peripheral blood was collected using Regen-Kit-Surgery tubes (RegenLab, Le Mont-sur-Lausanne, Switzerland) to prepare 10 mL of autologous platelet-rich plasma (A-PRP) glue. The RegenKit-Surgery kit included 2 RegenKit-blood cell therapy tubes for PRP preparation and 1 Regen autologous thrombin serum tube for autologous thrombin isolation. After centrifugation at 1,500 g for 5 minutes, PRP was obtained from 8 mL of blood using the Regen-blood cell therapy tube, resulting in 4 to 5 mL of A-PRP with a platelet recovery rate exceeding 80% and a concentration factor of 1.6 (corresponding to a platelet concentration of 400 billion platelets per millimeter). A second centrifugation at 1,500 g for 5 minutes was solely performed for the Regen autologous thrombin serum tube to extract the autologous thrombin serum. The PRP was subsequently mixed and activated with autologous thrombin using the Regen spray applicator in a 9:3 ratio (PRP: thrombin) to form PRP glue. The PRP glue was sprayed at the subcutaneous tissue and applied under the suture [21].

The use of fibrin glue proved to be helpful in controlling post-operative haemostasias [21] (Figs. 3-5). At the six-month follow-up, the patient reported satisfaction with both the aesthetic and functional outcomes, which led to a reduction in pain. No complications were observed, suggesting that the use of fibrin glue may be useful in controlling post-operative hemorrhage, which is the most frequent complication in cases of plexiform neurofibromas.

DISCUSSION

Peripheral or classical neurofibromatosis (NF1) is an autosomal dominant inherited condition that is relatively common, with an estimated frequency of 1:3000 [2]. It was first described by Von Recklinghausen in 1882 as a genetic ectodermal anomaly with systemic and progressive involvement, primarily affecting the skin, nervous system, bones, eyes, and other organs [3]. The

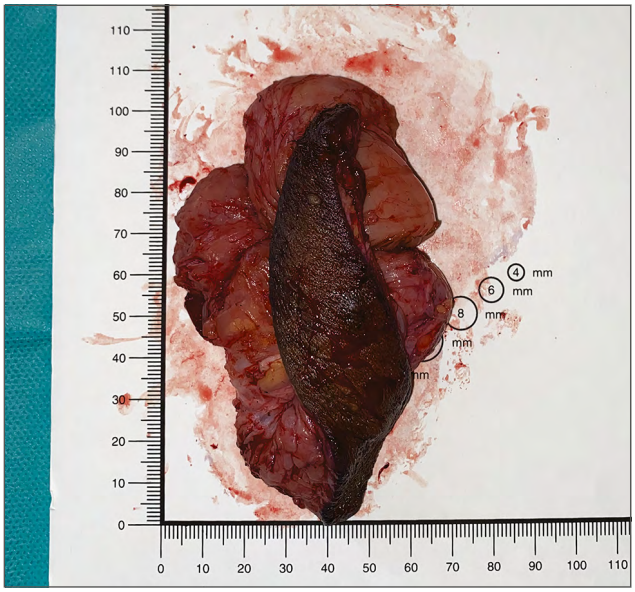

Figure 3. The surgical piece obtained at the end measured 10 x 6.5 cm.

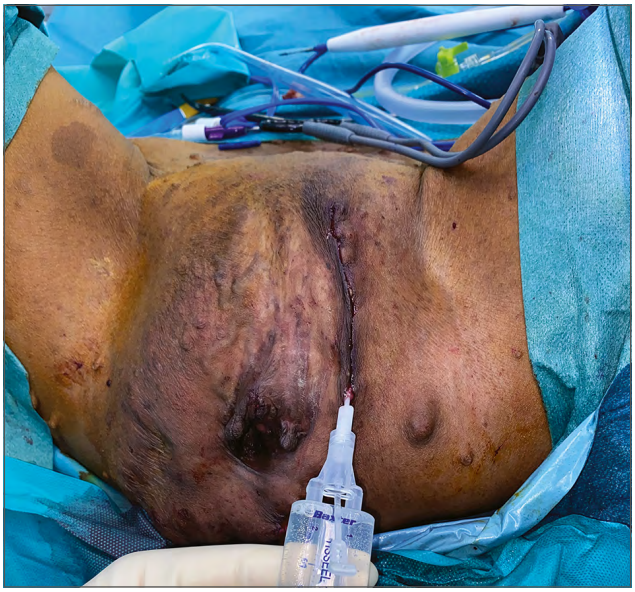

Figure 4. Fibrin glue was injected before proceeding with the skin suture to reduce the risk of post-operative bleeding.

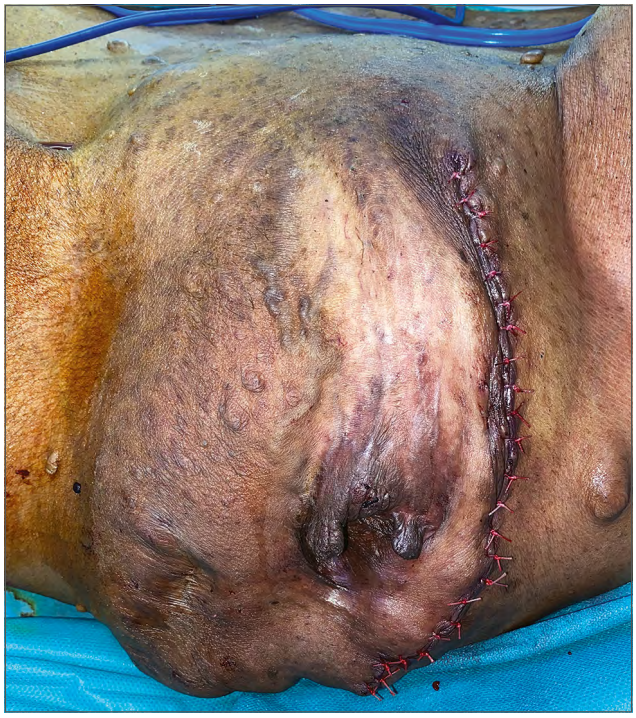

Figure 5. Immediate result at the end of the procedure shows satisfactory removal of the giant NF.

condition is characterized by several types of mutations in the NF1 gene located on the long arm of chromosome 17q11.22, which encodes a tumor suppressor protein called neurofibromin. Gene mutations lead to uncontrolled cell proliferation resulting in the formation of neurofibromas, which are benign tumors of the peripheral nerve sheath and are considered pathognomonic of type I neurofibromatosis [4-29]. There are four main types of neurofibromas: cutaneous (> 95%), subcutaneous, nodular, and plexiform [5]. Plexiform neurofibroma (PN) is the main type of neurofibroma when the localization of NF1 affects the genitourinary tract, and it can involve any organ, although genital involvement is extremely rare. The vulva is the most frequent genital site [6]. Usually, these tumors are small and slow-growing, but other examples of giant neurofibromas in the genital tract have been reported in the literature. In a case series by Iavazzo et al. [30], six patients with giant neurofibromas of the vulva underwent surgical excision. The tumors ranged in size from 6 to 20 cm in diameter, and all patients had a favorable outcome with no recurrence of the tumor during follow-up. In another case report by Lin et al. [31], a patient with a giant neurofibroma of the labia majora underwent surgical excision using a modified technique that involved preserving the neurovascular bundle of the clitoris. The patient had a successful outcome with no complications or recurrence of the tumor. In a case report by Trastour et al. [32], a patient with a giant neurofibroma of the vagina underwent surgical excision using a vaginal approach. The patient had a successful outcome with no complications or recurrence of the tumor. These data suggest that surgical excision is the treatment of choice for giant neurofibromas in the genital tract. However, the surgery can be challenging due to the delicate nature of the genital organs and the potential for damage to adjacent structures. Therefore, a careful surgical approach is necessary to minimize the risk of complications. In cases where the tumor is located in the vulva, excision can be performed using a standard surgical approach. In cases where the tumor is located deeper in the genital tract, a vaginal approach may be necessary. In some cases, a modified surgical technique may be necessary to preserve the neurovascular bundle of the clitoris and minimize the risk of sexual dysfunction. Patients with NF1 are at risk of malignant transformation, as they have an increased risk of developing malignant peripheral nerve sheath tumors (MPNST), and transformation into neurofibrosarcoma is also possible [7]. The rapid growth of any tumor is usually associated with malignant changes. Therefore, as in our case, it is important to intervene surgically to exclude malignancy and reduce the mass effect that causes severe pain and discomfort to the patient due to the important innervation of the genital area.

CONCLUSIONS

The cornerstone of neurofibromatosis management is providing longitudinal and multidisciplinary patient-centered care. Specific monitoring of clinical manifestations by age is crucial for early recognition and symptomatic treatment of lesions and complications, with the aim of improving the patient's quality of life. Giant neurofibromas are difficult to fully resect, and they are known for their high relapse capacity and increased risk of malignant transformation. Furthermore, their localization in the genital tract can cause severe pain and physical and psychological discomfort in patients. In our opinion, conservative modeling surgical excision is the most appropriate technique, as it minimizes the risk of bleeding, restores correct anatomy and sexual function, and has a positive impact on the patient's psychological well-being.

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Conflict of interest statement

The authors declare no conflict of interest.

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Author contributions

The authors contributed equally to the work.

Ethical consideration

Not applicable.

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