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Plexiform Neurofibroma: A Case Report

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Abstract **Case Report**

Plexiform neurofibroma is a characteristic lesion of Von's disease Recklinghausen. Currently, surgery is the only effective treatment. This is difficult form due to its hemorrhagic nature and the pervasive appearance of the lesions.
Keywords: Plexiform neurofibroma, neurofibromatosis type 1.

INTRODUCTION

The gluteal region is a frequent site for the development of various generally benign swellings. Among them, plexiform neurofibroma is a benign nerve tumor, most often developed in the cephalic region due to its rich innervation, occurring early in childhood and adolescence and considered pathognomonic of type 1 neurofibromatosis (NF1) [1]. Its evolution is slow and the extension is done locally and to neighboring regions resulting in monstrous deformities [2]. The treatment is surgical, with no certainty as to the relevance of its indication at one or another stage of development. The aim of this study was to highlight the clinical, evolutionary and therapeutic aspects of this condition through a study of a case of plexiform neurofibroma of the gluteal region.

OBSERVATION

This is a 17-year-old student, followed for type 1 neurofibromatosis without any other notable medico-surgical history who was referred to our plastic and aesthetic surgery department and burned for better management of a straight buttock mass. The onset of the disease was more than 3 years old, marked by the appearance of a mass in the right buttock gradually increasing in size and developing in a context of apyrexia and maintenance of general condition.

Table-1: Diagnostic criteria for NF1 - Conference of consensus on neurofibromatosis (NIH-Bethesda, 1988)

Six or more Café au lait spots (≥5mm in children, ≥15mm in adults)
Two or more neurofibromas of different type or a plexiform neurofibroma
Axillary or inguinal lentigines
Glioma of the nervus opticus
One or more Lisch's nodules
Bone disorders

Physical examination showed a firm, painless lump in the right buttock, slightly adherent to the deep plane of the right buttock. The patient was recognized as a carrier of type 1 neurofibromatosis according to NIH diagnostic criteria [2] (Table 1), hence the suspicion of plexiform neurofibroma as the most probable diagnosis.

The patient underwent an MRI of the soft parts of the pelvis demonstrating a multicentric diffuse deep myofibromatosis producing a mass of the right buttock measuring 12 cm long axis (image 1).

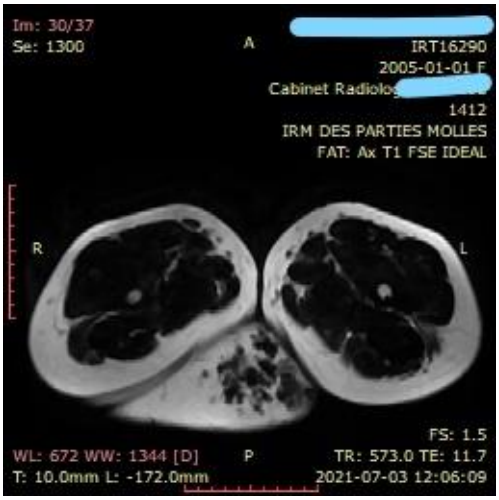

Image-1: MRI section showing a plexiform neurofibroma

The preoperative assessment was unremarkable. The aim of the surgical treatment is to reduce the size of the tumor, to improve the aesthetic aspect and the psychosocial aspect. Surgical treatment consisted of partial excision of the mass (figures 2, 3, 4). The anatomopathological examination concluded the histopathological appearance of plexiform neurofibroma in the context of type 1 neurofibromatosis. The postoperative course was simple and with satisfactory healing tempered by a localized unsightly appearance.

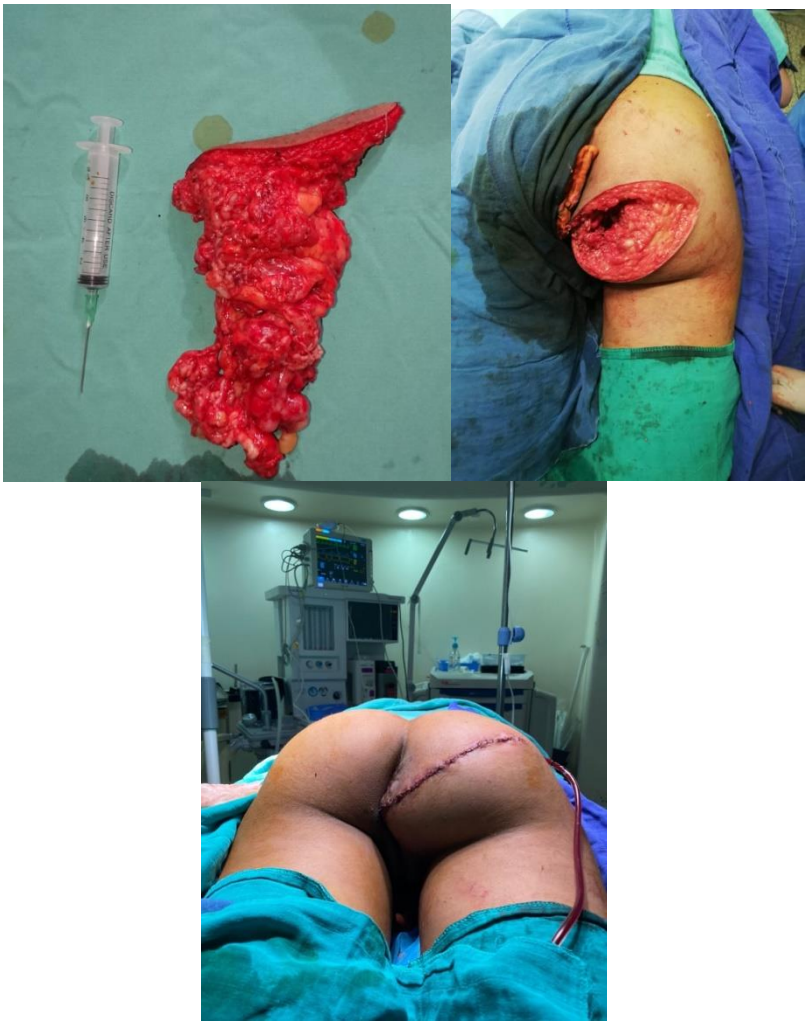

Fig-2, 3, 5: The course of the surgical procedure

DISCUSSION

Plexiform neurofibroma is a benign tumor of the perineurium of the peripheral nerves. It consists of a proliferation of Schwann cells arranged in myxoid tissue of varying abundance comprising numerous fibroblasts and collagen fibers with a so-called "grated carrot" appearance [3]. Plexiform neurofibroma is strongly associated with neurofibromatosis [1], in particular with Von Recklinghausen's disease (NF1) where it occurs in 24 to 32% of affected patients [4, 5]. Plexiform neurofibroma belongs to the four types of neurofibromas found in NF1 according to the classification of the 1988 consensus conference of the National Institute of Health Development [6]:

- Discrete cutaneous neurofibromas of the epidermis or dermis;
- Discrete subcutaneous neurofibromas that are deeper;
- Deep nodular neurofibromas;
- Diffuse plexiform neurofibromas.

Formerly called "plexiform neuroma" or "royal tumor", plexiform neurofibroma differs from other types of neurofibroma by the importance of its Schwannian component [3]. The diagnosis of plexiform neurofibroma is essentially pathological, in particular outside the context suggestive of type I neurofibromatosis. The clinical aspect is not specific, it may suggest a tumor or a vascular malformation, a skin or conjunctive tumor, benign or malignant [7]. The opposite skin is classically pathological (hypertrophy, hypertrichosis, brown pigmentation, angiomatous appearance), with two main characteristics: hyperextensibility and lack of elasticity [8]. This skin or subcutaneous tumor, sometimes painful, varies in size, ranging from a few centimeters to an entire body segment. Plexiform neurofibroma can be present at birth, but often appears in children between the ages of two and five [1, 6, 8, 9]. In the limbs, it generally follows a nerve axis organizing itself into a cord or cluster. Craniofacial localization occurs in three to seven percent of cases when it is part of type I neurofibromatosis [10, 11]. In addition to these locations, plexiform neurofibroma can develop at the expense of all peripheral nerves [12, 13].

The imagery is fairly unspecific. Ultrasound can still rule out a tumor or a rapidly circulating vascular malformation. MRI has the classic appearance of peripheral nerve tumors (low to moderately intense T1 signal, T2 hypersignal often with a heterogeneous appearance). Areas of hypersignal correspond to areas of myxoid tissue or cystic degeneration. The hyposignal nodular areas correspond to the collagenous and fibrous tissue and can be enhanced with gadolinium.

The literature does not abound with descriptions of cases of plexiform neurofibromas with prose difficulties in surgical management. In 2010, Levy Bencheton *et al*. [14] described a case of plexiform neurofibroma isolated in an 85-year-old man. They insisted on the difficulties of surgical management due to the invasion of the subcutaneous tissues and the frequency of recurrences.

Neurofibromas can degenerate in 3 to 15% of cases into neurofibrosarcoma. This incidence is 9000 times higher than in the general population [4, 5, 15, 16]. The risk of degeneration alone justifies systematic resection, but it is most often performed in front of a painful tumor and / or the cause of aesthetic damage. Given their often large size, hemorrhagic and poorly limited nature, excision is most often delicate and most of the time cannot be "carcinological". These incomplete resections do not appear to be the source of an increased risk of degeneration into neurofibrosarcoma [17]. In the context of type 1 neurofibromatosis, annual clinical monitoring is recommended [16]. The risk of malignant degeneration of a possible tumor residue should be considered in the face of any rapid increase in the size of the lesion, the appearance or increase of pain. These signs should require a radiological assessment (magnetic resonance imaging) to be carried out without delay and a rapid resumption of surgery should be considered.

The clinical diagnosis of neurofibromatosis is made, according to the 1988 consensus conference (Table 1), by the presence of at least two of the seven cardinal criteria of type 1 neurofibromatosis. The presence of a plexiform neurofibroma does not allow not, on its own, to make the diagnosis of neurofibromatosis type 1 as shown by Lin *et al*. [15].

Symptoms vary according to age and clinical diagnosis can be difficult, especially in children, if they have no family history of type 1 neurofibromatosis. However, at the age of eight years, type 1 neurofibromatosis is considered to have almost complete penetrance (Table 2). Most patients with type 1 neurofibromatosis are therefore diagnosed before the age of ten.

Long-term postoperative monitoring of patients is difficult. The results are generally disappointing and unstable over time, which often leads to discontinuation of treatment and monitoring.

CONCLUSION

Cutaneous neurofibromatosis is a condition that causes significant aesthetic damage. Plexiform neurofibroma surgery plexiform is not stereotypical and depends on several factors related to the injury, the patient and the experience of surgeon. Complete excision is not always possible. The surgery must be conservative so as not to replace the plexiform neurofibroma by another aesthetic and functional damage more important. Prolonged monitoring is necessary for detect progressive complications related to the disease.

Declaration of interests

The authors declare that they have no conflicts of interest in relation to this article.

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