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CASE REPORT

Giant plexiform neurofiberoma

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***Abstract*:** We report a rare case of giant plexiform neurofibroma in a patient affected by type-1 neurofibromatosis and we describe the correct surgical management of such lesions in order to avoid intra- and post-op blood loss related complications.

***Keywords*:** Plexiform neurofibroma; giant; surgery; blood loss; arterial embolization

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Introduction

Neurofibroma is considered as one of the characteristic features of neurofibromatosis type 1 (NF1). Plexiform neurofibroma is a congenital tumor which continuously grows, interferes with the normal function of the affected areas[1], and is characterized by enlargement along the branches of the parent nerve and rich vascularization. Complications related to this kind of lesion include malignant transformation (rare) and massive hemorrhages[1-5]. We describe a rare case of giant neurofibroma discussing its management.

Case

A 65-year-old female, affected by type-1 neurofibromatosis, presented to our attention showing diffuse neurofibromas as well as a discrete and voluminous pedunculated mass originating from the right scapular area, which had appeared 40 years before and had slowly enlarged (**Figure 1**). Such growth measured 60 cm in length, while the diameter ranged from 15 to 25.3 cm. This lesion had become a substantial burden for the patient, impeding everyday activities, and compensatory scoliosis was evident. A computed tomography-scan was performed prior to surgery, showing a superficial subcutaneous and cutaneous mass (**Figure 2**). Pre-operatory embolization was

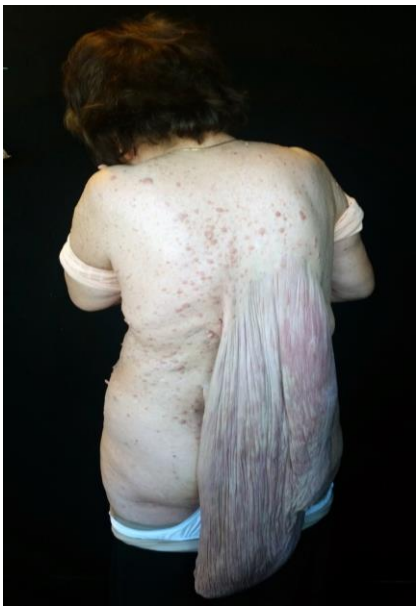

Figure 1. Pre-op view. Massive pedunculated mass.

performed in order to reduce the blood inflow and make the planned resection safer. Local flaps were used to cover the surgical defect. The excised mass, which histologically was proved a plexiform neurofibroma, weighted 12 kg, amounting to 24% of the patient's weight (50 kg). At 12-months follow-up the functional and cosmetic surgical outcome was satisfactory and no signs of recurrence were noted (**Figure 3**).

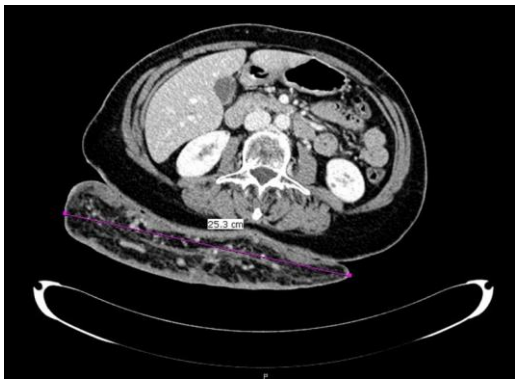

Figure 2. Computed tomography-scan at the level of max diameter

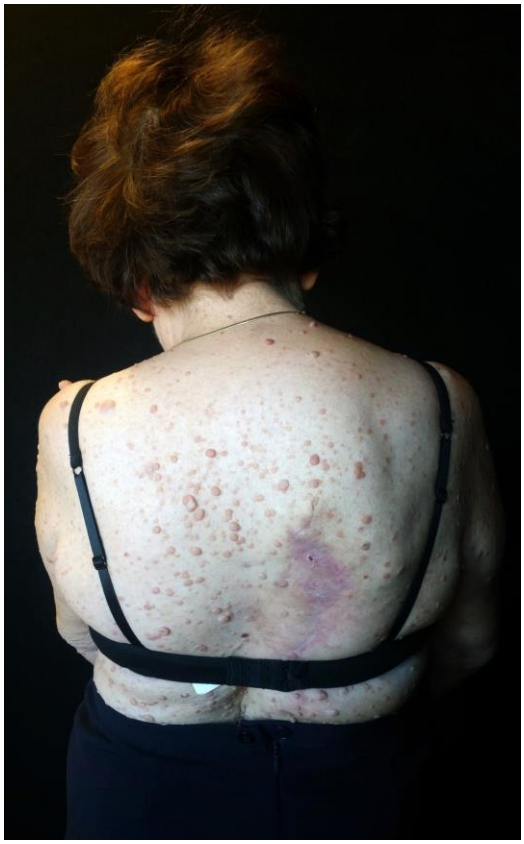

Figure 3. Post-op view. Satisfactory functional and cosmetic outcomes.

Discussion

Neurofibroma is a hallmark of type-1 neurofibromatosis. The rare plexiform variant is characterized by continuous growth. The use of the term "giant" is still controversial. Recently, Vélez *et al*. suggested to limit this nomenclature for lesions weighing 20% or more of the patient's weight[2].

Surgical intervention responds to various needs: to provide a histological diagnosis and to restore function and cosmesis. Peri-operative hemorrhage may be challenging in these patients, and various cases have been described with severe and potentially life-threatening blood-loss[1–5]. As a matter of fact, the lesion intrinsic propensity to bleed must always be considered and prevented with arterial embolization, by means of different techniques[3-5], as well as pre-operative stabilization of general conditions by means of sustaining the circulatory volume which is expected to be lost during surgery.

Conclusion

Giant neurofibroma is a rare disfiguring manifestation of NF1, and it is correctly identified when its weight amounts to at least 20% of total body weight. The treatment of this type of lesion is associated to potentially dangerous complications related to blood-loss. Comprehensive and careful pre-operative management is therefore necessary to allow safe surgery.

Conflict of interest

The authors declare no potential conflict of interest with respect to the research, authorship, and/or publication of this article.

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