

## Case Report

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# Lower limb giant plexiform neurofibroma: preoperative evaluation and surgical planning

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## ABSTRACT

Few cases of lower limb giant plexiform neurofibroma have been reported in literature and there are no available guidelines for defining its preoperative evaluation and surgical approach. Lower limb giant plexiform neurofibroma is a rare condition that requires clinical and radiological preoperative evaluation, mainly with MRI. Surgery is the only treatment with no preoperative trans-arterial embolization or vessel ligation. Early excision of superficial lesions may prevent further progression.

**Keywords:** Lower limb gigantism, Plexiform neurofibromas, MRI

## INTRODUCTION

Lower limb gigantism is a rare condition, which can be due to the presence of a giant plexiform neurofibroma.<sup>1,2</sup> The complete excision of a giant neurofibroma is a great surgical challenge. Preoperative planning with MRI, CT and arteriography may enable the surgeons to define the extent of the mass and develop an operative planning. The aim of the study is to discuss the principles of treatment by carrying out a review of literature and making a case report of this uncommon condition.

## CASE REPORT

A 69 year old woman (weight 47 kg, height 156 cm) was affected by a left thigh plexiform neurofibroma, which has grown in size over the last five years leading to consequent difficulty in standing and walking. She did not suffer from any other symptoms or disorders pertaining to other systems as shown in Figure 1.

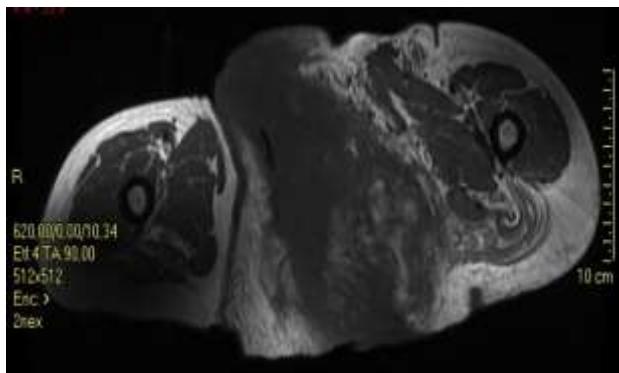


**Figure 1: Giant plexiform neurofibroma of the left lower limb (a: preoperative; b: postoperative).**

The patient reported that the mass had appeared following her second pregnancy with concomitant presence of multiple cafè au lait patches. A biopsy was performed when the mass appeared which confirmed that it was a neurofibroma. A neurologic evaluation diagnosed

type 1 neurofibromatosis (NF1). In the following years the patient underwent clinical and MRI follow-ups.

There was no family history of NF1. On carrying out a physical exam, the patient showed weakness and a loss of sensation for touch in the left lower limb. The mass extended across the medial aspect of the left thigh from the popliteal fossa to the gluteal fold and had a fluctuant to firm consistency. Imaging showed a displacement of the thigh medial muscles and lower abdomen visceral organs, with no functional symptoms related to abdomen implications as shown in Figure 2.



**Figure 2: Preoperative MRI evaluation (thigh level).**

The patient underwent partial excision of the mass under general anesthesia. During this procedure a 7 kg mass was excised, which corresponded to most of the thigh extension of the neurofibroma, and local advancement flaps were used for reconstruction as shown in Figure 3. During the operation the patient received four units of packed-red blood cells. No intraoperative complications were observed. Following surgery the patient regained normal ambulation after five months of physiotherapy rehabilitation.



**Figure 3: Partial excision of neurofibroma.**

## DISCUSSION

Plexiform neurofibroma is a rare benign tumor, which affects 30-40% of NF1 individuals. These benign tumors are caused by a proliferation of cells in the nerve sheath

extending across the length of a nerve and involving multiple nerve fascicles.<sup>3-6</sup> Plexiform neurofibromas are not generally well circumscribed or encapsulated, the overlying skin is thick and friable with nontensile strength; in plexiform neurofibromas there is an abundance of dilated and friable blood vessels inside the tumor tissues.<sup>7</sup> Plexiform neurofibromas can grow at any time of life; no risk factors for growth have been identified, although trauma has been suggested as a possible triggering factor. It is unknown if surgery can cause excessive growth in postsurgical residual tumors.<sup>1</sup>

Giant plexiform neurofibroma is a poorly defined term used to describe a neurofibroma that has grown to a significant but undefined size.

We reviewed the literature for giant plexiform neurofibroma and few articles which reported its treatment planning were found.<sup>2,8-10</sup>

Ross described a giant plexiform neurofibroma as a  $\geq 20$  kg mass which is probably related to the patient's weight since the mass represented 14% of her entire weight and following its removal the patient's BMI decreased from 19 to 16. For this reason we cannot identify a "giant" lesion by only considering its own weight, as we believe it can be better understood by a tumor excision/Patient BMI ratio.<sup>2</sup>

Ross also reported on possible postoperative complications such as lymphedema, cellulitis and delayed wound healing. The author underlined that lymphatic drainage can be altered as a consequence of the excision and could be managed with surgical drains and compression dressings.

Anatomical preoperative evaluation is mandatory. Neurofibromas develop from nerve sheaths and for this reason their surgical removal is attributed to possible nerve injuries. Lower limb neurofibromas often involve popliteal sciatic nerves, which is responsible for foot flexion-extension and partial lower limb sensitivity.

Rekha reported on a lower limb giant plexiform neurofibroma by stating that this type of situation is uncommon and should be distinguished from arteriovenous malformations, hemangiomas and elephantiasis.<sup>8</sup>

Preoperative evaluation by means of radiological imaging is of paramount importance both for diagnosis and subsequent surgical treatment.

MRI is a valuable tool for analyzing and monitoring the growth of plexiform neurofibromas, which can follow three different patterns: superficial (25%), with asymmetric diffuse extension, displacing (26%), with multinodular smoothly defined borders compressing adjacent structures, primary, along the main nerves, or

invasive (49%), if there is a dissolution of the original structure of the tissue affected.<sup>11</sup>

By carrying out angiographic studies it is possible to identify the type of neurofibromas that are likely to bleed more extensively, as an increased risk of rupture of their friable vasculature secondary to arterial dysplasia or vascular invasion by the tumor can be observed.<sup>12,13</sup>

The main treatment of plexiform neurofibromas is surgery. As suggested by other authors, pre-operative embolization can reduce the intraoperative bleeding yet it can also cause extensive tissue damage, delay wound healing and require VAC Therapy; therefore preoperative imaging plays an important role in evaluating each specific case. Preoperative trans-arterial embolization is not a gold standard approach, especially in plexiform neurofibroma due to the subsequent tissue necrosis.<sup>9,14-16</sup>

Pre-operative ligation of feeding vessels is regarded as a stimulus for increasing vascularity by opening up the choke vessels.<sup>17</sup>

Hypotensive anesthesia is ideal for operations of this kind because severe hemorrhage is frequent in neurofibroma resection.

When analyzing the case of an NF1 patient with an 11 kg massive lower limb plexiform neurofibroma, Power reported on the right time for performing surgical excision. The author suggested that early excision of this type of tumor was preferable since it limits disfigurement and morbidity associated with large lesions and may prevent their progression.<sup>9</sup>

Complete resection is often difficult due to the extensive growth of the tumor and its invasion of surrounding tissues. Regrowth after surgery is common: 20% of cases after total resection, 45% of cases after partial resection.<sup>18</sup> Total excision is a worthy aim. Excision must be carried out in stages in most cases of extensive growth. Final shaping is occasionally carried out with local flaps and skin grafts.

## CONCLUSION

Lower limb giant plexiform neurofibroma is a rare condition that deserves both clinical and radiological preoperative evaluation, mainly with MRI. Treatment is directly surgical with no preoperative trans-arterial embolization or vessel ligation. Proper surgical planning should be supported by MRI as it indicates the vasculature of the plexiform neurofibroma. Early excision of superficial lesions may prevent progression.

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