



Hybrid resection of a giant thigh plexiform neurofibroma



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ARTICLE INFO

Article history:

Received 13 September 2014

Received in revised form

30 December 2014

Accepted 3 January 2015

Available online 9 January 2015

Keywords:

Neurofibromatosis

Plexiform neurofibroma

Multidisciplinary team

ABSTRACT

PRESENTATION OF CASE: Authors present a case of a 55 year old patient with Type 1 neurofibromatosis (NF1) and a very large right thigh plexiform neurofibroma. The patient had increasing difficulty with mobilization due to this enlarging bulky mass. Preoperative embolization reduced the bleeding risk at surgery allowing successful gross resection of the mass by a multidisciplinary surgical team. Limb function was restored to normal.

DISCUSSION: Massive plexiform neurofibromatosis is a rare expression of von Recklinghausen's disease or NF1. These large masses result in severe disfigurement and significant functional disability. They are extremely vascular and there is potential for malignant transformation.

CONCLUSION: These massive tumors require complex preoperative, intraoperative and postoperative management strategies with involvement from a multidisciplinary team. We discuss the challenges of surgical intervention and to discuss the current literature.

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1. Introduction

Plexiform neurofibromas are most commonly seen on the face and neck [1]. Cases of lower extremity plexiform neurofibromas are seen less frequently and in addition are extremely vascular. As a result, one single management option for plexiform neurofibroma has not been established. Surgery is often delayed due to fear of massive bleeding and significant disfigurement and debilitation occurs. The purpose of this report is to discuss a case of a large plexiform neurofibroma and review the current literature.

2. Case report

A 55-year-old woman with NF1 was referred to our institution to evaluate a very large right thigh plexiform neurofibroma measuring approximately 40 pounds (Fig. 1). The lesion was affecting the patient's ability to walk and causing her a great deal of discomfort. On examination, the patient had multiple café-au-lait patches, axillary and inguinal freckling and multiple small neurofibroma nodules varying in size from 0.5 to 3 cm scattered

over her entire body. The patient was diagnosed with neurofibromatosis at a young age. The patient's family history was significant in that both her mother and son were diagnosed with neurofibromatosis. The patient also had a remote history of an attempted resection, which was abandoned due to massive bleeding.

The patient was mobilizing independently at home, albeit with increasing difficulty due to the cumbersome mass. Imaging was ordered and the magnetic resonance imaging (MRI) revealed a mass involving the entire anterior thigh that was extremely vascular. The imaging was consistent with the diagnosis of peripheral neurofibromatosis and demonstrated enlarged and tortuous major vessels (Fig. 2A and B). A biopsy of the right thigh area confirmed a diagnosis of plexiform neurofibroma.

A multidisciplinary team of vascular and orthopedic surgeons received her care and felt a resection was feasible following preoperative embolization (Figs. 2–4). At operation the tumor was dissected off the underlying quadriceps muscle and removed as a single en-bloc mass. Following the tumor removal primary skin closure was achieved. The patient recovered in hospital for 4 days and then was discharged home. The gross specimen measured 64 cm × 35.5 cm × 20 cm. Pathological examination revealed a plexiform and diffuse neurofibroma with foci of atypia. There were areas of embolization with infarction but no malignant transformation. At follow up the wound healed well and resulted in a normal caliber and functioning limb (Fig. 5). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Abbreviations: (NF1), Type 1 neurofibromatosis; (MRI), Magnetic resonance imaging.

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<http://dx.doi.org/10.1016/j.ijscr.2015.01.015>

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Fig. 1. Large right thigh plexiform neurofibroma.

Table 1

To diagnose NF1, two of the following clinical features must be present [6].

- Six or more café-au-lait macules >5 mm in diameter in prepubertal and >15 mm in diameter in post pubertal individuals; for each lesion, the longest diameter is measured
- Two or more neurofibromas of any type of one plexiform neurofibroma
- Freckling of the axillary or inguinal regions
- Optic glioma
- Two or more Lisch nodules (iris hamartomas)
- A distinctive bony lesion such as sphenoid dysplasia or thinning of the long bone cortex with or without pseudoarthrosis
- A first-degree relative (parent, sibling, or offspring) with NF1 based upon the above criteria

and subcutaneous tissue associated with a wrinkled and pendulous appearance [4]. It is mostly considered a benign tumor condition; however malignant transformation has been reported in 2 percent of patients with NF1 (or 4.2% of those 21 years of age or older) [5].

The natural history of a plexiform neurofibroma varies, as some lesions remain asymptomatic superficial lesions, while others progress into large invasive disfiguring lesions as seen in our case [6]. Tumor progression is a serious problem for patients with a plexiform neurofibroma due to the debilitating nature of the disease to a patient's function and potential for malignant transformation. Child and adult patients with tumors that cannot be completely removed are at particular risk.

In order to investigate the neurofibroma MRI and computed tomography are used to determine the site and the expansion of the plexiform neurofibroma. Unfortunately, however, these imaging modalities are not reliable in deciphering between a plexiform neurofibroma and those that have progressed into malignant peripheral nerve sheath tumors (MPNST) [16]. MPNST usually arise from plexiform neurofibroma, they tend to metastasize widely and usually have a poor prognosis. More recently with the advent of positron emission tomography with the glucose analogue fluorine-18-fluorodeoxyglucose the diagnosis can be determined. It is a functional imaging technique which allows the visualization and quantification of glucose metabolism and reflects the increase in metabolism in malignant tumors [17]. However, the gold standard in identifying malignant transformation within a plexiform neurofibroma is histology. Unfortunately histology requires complete excision, which is not always possible. If a core biopsy is performed, the malignant change may be missed within a large heterogeneous tumor [18].

Surgical intervention is therefore an important therapeutic and diagnostic option, which has two goals: to provide histo-

3. Discussion

Neurofibromatosis appears in two different neurocutaneous autosomal dominant genetic forms. Neurofibromatosis types 1 and 2 affect nerve cell tissue development and growth [1]. NF1, also known as von Recklinghausen's disease is the most common type of Neurofibromatosis (Table 1). Plexiform neurofibromas occur in 26.7% of patients with NF1 [2]. Plexiform neurofibromas present at, or within the first 2–5 years of life. Key characteristics include hyperpigmentation, thickening of the skin and hair excess [3]. There are two types of plexiform neurofibromas, nodular and diffuse. A diffuse plexiform neurofibroma, also known as elephantiasis neurofibromatosis, is characterized by an overgrowth of epidermal

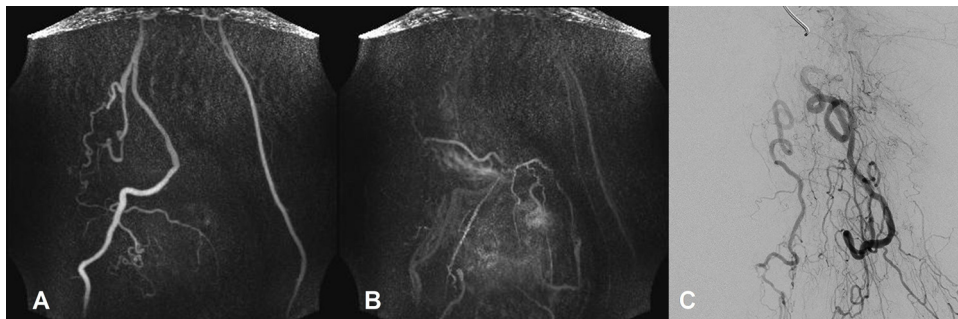


Fig. 2. MRI scan demonstrating markedly enlarged arteries going to the lesion with supply going from the profunda and tortuous superficial femoral arteries (A + B). Pre embolization peripheral angiogram demonstrating large collaterals supplying the tumor from superficial femoral artery (C).

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