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Surgical treatment of a large plexiform neurofibroma of the lower extremity

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Plexiform neurofibromas are benign nerve tumors occurring in approximately 30% of patients with neurofibromatosis type 1. They develop as neural proliferations of single or multiple nerve fascicles, and are typically highly vascular in nature. In this case report, we describe a 28-year-old male with a paternal family history of neurofibromatosis type 1 and a large plexiform neurofibroma of his left lower extremity present. Following consultation and shared decision-making, the patient underwent surgical debulking primarily to reduce pain, to improve shoe gear fit, and to improve ambulation.

Keywords: plexiform neurofibromas, neurofibromatosis, surgery

A 28-year-old male with a past medical history of neurofibromatosis type 1 was seen for evaluation and management of a painful mass on his lateral left leg (Figure 1). He was no longer able to wear normal shoes, which in turn affected his activities of daily living. His surgical history included a prior debulking procedure of his left medial leg and foot at the age of 3. His social history included active tobacco use of a ½ pack of cigarettes a day, and had not graduated from high school. The patient related an extensive paternal family history of neurofibromatosis type 1, affecting multiple family members. He reported a paternal family member passing away from a peritoneal malignancy caused by plexiform transformation into a malignant peripheral nerve sheath tumor.

Preoperative surgical planning included a coordinated effort between podiatric and plastic surgeries. Surgical and conservative options were discussed in detail with the patient. The elected plan for the surgery was to debulk the lateral leg mass, with the goal of reducing the associated pain and to allow the patient to fit into a shoe. Risks and benefits were discussed in detail with the patient, and the patient was educated regarding the likelihood and speed of mass regrowth.

Results

The patient underwent surgical debulking as an outpatient. Preoperatively, blood was typed and crossed in anticipation of blood loss secondary to the highly vascular nature of plexiform neurofibromas. A thigh tourniquet was used, and the patient was placed into a lateral decubitus position. A large semi-elliptical incision was utilized, oriented in line with the mass. The mass was identified and carefully dissected, with electrocautery used as necessary. The mass was noted to readily extend through tissue planes, and was not sharply defined. Local neurovascular structures were carefully avoided during dissection of the mass.

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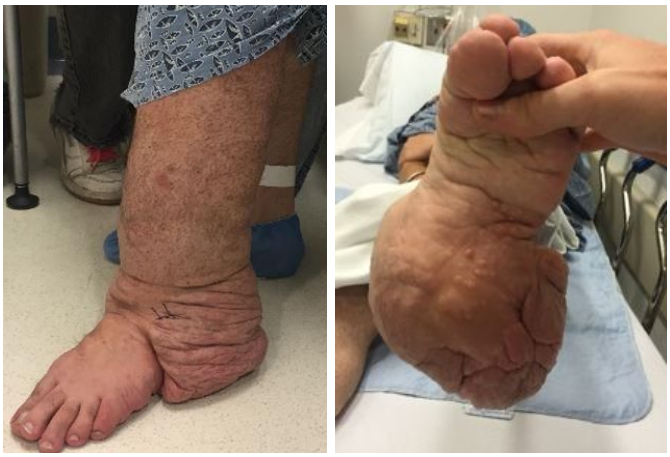

Figure 1 Preoperative weightbearing and nonweightbearing clinical appearance of the left lower extremity, depicting the large mass.

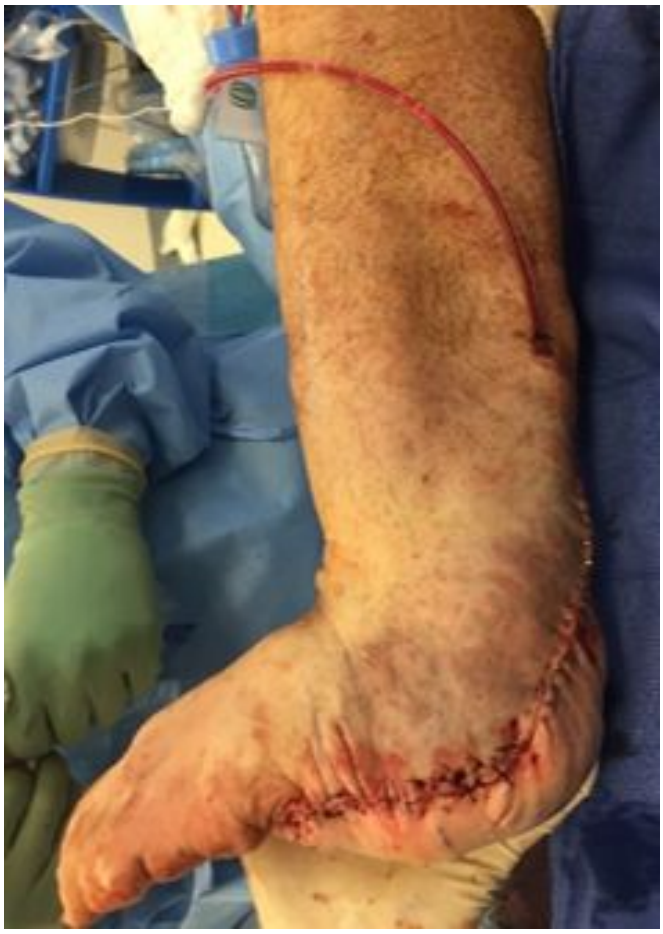

Figure 2 Immediate postoperative image of the left lower extremity following surgical debulking, with the surgical drain visible.

With direct and unobstructed exposure obtained, the large mass was debulked with representative samples sent to pathology.

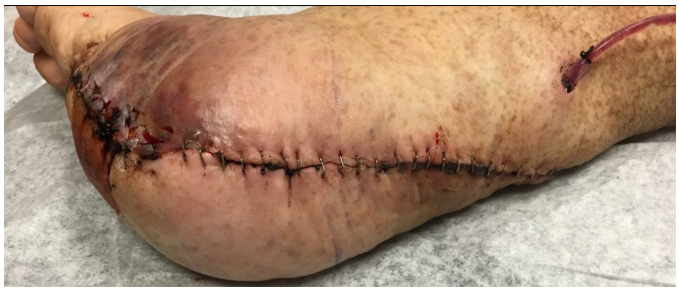

Figure 3 Postoperative image of the left lower extremity at the first postoperative visit, with surgical drain visible.

The mass was noted to extend into the peroneal tendons, lateral ankle ligaments, and the fat pad of the heel; these anatomic structures were carefully preserved during the debulking process.

Following debulking of the mass, the tourniquet was released. Electrocautery was again employed to assist in obtaining hemostasis. Epinephrine soaked gauze was also employed as a hemostatic agent to promote vasoconstriction, further reducing blood loss during dissection. The surgical site was closed in layers, with Floseal hemostatic matrix (Baxter International, Deerfield, Illinois) applied during closure. A passive, closed, surgical drain was inserted prior to skin closure (Figures 2 and 3).

Discussion

Neurofibromatosis type I (NF-1), formerly known as Recklinghausen's or von Recklinghausen disease, is a subtype of neurofibromatosis accounting for 90% of cases [1]. NF-1 is an inherited, autosomal dominant, single-gene disorder of chromosome 17: this non-sense mutation takes place on the NF-1 gene, with a prevalence of 1/3000 births and an equal distribution between males and females [2]. NF-1 usually presents in childhood, and manifestations include café au lait spots, neurofibromas, skeletal dysplasia, and neuropathy secondary to space-occupying neurofibromas [3,4].

Plexiform neurofibromas occur in approximately 30% of the patients with neurofibromatosis type I [5]. Malignant transformation occurs in about 2-16% of cases and is diagnosed with histopathologic biopsy [4,6]. Treatment planning requires consideration of the patient's goals of treatment, the extent of the deformity, and the presence of malignant transformation.

It is of vital importance to plan preoperatively in order to anticipate the atypical surgical dissection. Surgical time may be longer than anticipated, as anatomic layers will be obscured and violated by the invasive and vascular nature of these masses. Preoperatively, blood should be typed and crossed with the anticipation of significant levels of blood loss. Careful, layered closure should be performed, with the incorporation of hemostatic agents. A closed surgical drain should be considered as well.

Due to the invasive and diffuse invagination of the mass, multiple tissue planes were carefully dissected with the anticipation of overall "debulking" rather that complete marginal resection of the soft tissue mass.

Though rarely encountered, management of large plexiform neurofibromas should include a shared-decision making process and a realistic depiction of the surgical outcome. Operative management should be deferential to the highly vascular and invasive nature of these soft tissue tumors.

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