

A case report of surgical debulking for a huge mass of elephantiasis neuromatosa

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Abstract

Achievement of a safe outcome for an extensive mass with hypervascularity in the extremities requires a surgical team skilled in musculoskeletal oncology. We report debulking surgery for a huge mass of elephantiasis neuromatosa in the right leg of a 56-year old man using the novel Ligasure® vessel sealing system.

Introduction

Patients with plexiform neurofibroma rarely suffer from hypertrophy of the skin of the leg. Plexiform neurofibroma may become very large and deforming, and be referred to as elephantiasis neuromatosa.^{1,2} Owing to the cosmetically grotesque appearance and functional loss and pain in the affected limb, surgical treatment is usually considered. However, hypervascular structures are usually involved in this disease, and these can cause spontaneous hemorrhage and severe bleeding, especially during surgery. Removal of such huge masses seems impossible without massive bleeding. As a result, conservative treatment without surgery is usually adopted in most cases, and, even if surgery is selected, disarticulation or amputation of the affected limb is sometimes recommended to the patients.^{3,5} In the present report, we describe surgical debulking for a huge mass of elephantiasis neuromatosa in the right leg of a 56-year old man using the novel Ligasure® vessel sealing system (Valleylab, Boulder, CO, USA), and discuss how to approach an extensive mass of elephantiasis neuromatosa.

Case Report

A 56-year old man with a huge mass below both buttocks that had gradually increased in size over 35 years was referred to our hospital. At 20 years of age, the mass in the buttock was the size of a baseball. The patient had under-

gone a surgical operation ten years before his referral, and had been diagnosed with von Recklinghausen disease. This condition had been present for many years, and the tumor had continued to grow gradually. Spontaneous subcutaneous hemorrhages frequently occurred. At four years before his referral, a plastic surgeon had attempted debulking surgery at his previous hospital, but the operation was not completed because of massive bleeding, even with a small skin incision. During the previous year, he had only been able to walk short distances and remained in bed in hospital. There was no particular familial history of neurofibromatosis. On examination at his first visit, he had an extensive mass of neurofibroma over his trunk, and café-au-lait spots were scattered around the whole body. The right leg below the buttock was markedly enlarged, and he required great effort to go to the toilet and could not stand up by himself. Dilated superficial veins and multiple skin ulcerations were present on the huge mass. In view of the clinical appearance, plexiform neurofibroma was easily considered as a diagnosis. Magnetic resonance imaging (MRI) revealed an extensive mass that infiltrated the fat, skeletal muscle and adjacent bone in the lower leg. A T2-weighted sequence of MRI demonstrated dilated twisting capillaries in the high-intensity region of a superficial subcutaneous lesion, appearing as a myxomatous lesion, compatible with the histology of neurofibroma. Contrast-enhanced angiographic computed tomography (CT) demonstrated a rich vascular supply to the extensive mass, arising from both sides of the major external and internal iliac arteries. Extensive capillary pooling was also seen. After these imaging studies, biopsies were attempted at several spots to exclude the possibility of sarcomatous change. The needle biopsy specimens confirmed the histology of the neurofibroma. At that time, total tumor resection was judged to be impossible, and debulking surgery was planned piece by piece. During surgery, reduction of the mass was carried out using a novel bipolar vessel sealing device, Ligasure®. The resected surgical specimen was compatible with the histology of neurofibroma and no malignant changes were observed. Post-operatively, most parts of the operation scar gradually healed. Re-suturing was necessary at only a small part of the skin fold. At one year after the debulking surgery, he could move by himself and was extremely satisfied with the outcome.

Discussion

Neurofibromatosis is estimated to occur at rates of 1 in 2,000-3,000 births and shows an equal sex distribution. Von Recklinghausen

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disease is an autosomal dominant trait and represents a relatively common disorder characterized by neuroectodermal and mesodermal tissues. The typical characteristics include von Recklinghausen disease of the skin, café-au-lait spots and hamartoma of the iris. Plexiform neurofibroma characteristically shows diffuse irregular infiltration into the adjacent muscle and fat, and occasionally becomes huge and acquires a grotesque clinical appearance, referred to as elephantiasis neuromatosa.^{1,2} Radiological modalities for elephantiasis neuromatosa play important roles in providing anatomical data if surgery is planned. Among the radiological assessments, CT and MRI can provide many useful advantages for surgical planning.^{6,7} Elephantiasis neuromatosa may involve hypervascularity and most reported cases have exhibited massive spontaneous bleeding.^{3,7} In our case, a previous surgical attempt to debulk this lesion had resulted in massive blood loss and was not completed. Pre-operative assessment of the vascular supply to such a huge mass seemed to be inevitable, considering the importance of controlling intra-operative bleeding. The use of MRI enabled us to non-invasively assess the relationships of the lesions with the major vessels, as well as the vascular supply and angiographic features of the lesions. The contrast-enhanced angiographic CT confirmed the hypertrophic nature of the abnormal tissues, as well as the large drainage vessels and multiple tortuous collateral branches. These modalities may assist not only in the correct diagnosis but also in understanding of the anatomy of the vasculature of elephantiasis neuromatosa. Plexiform neurofibromatosis is a poorly circumscribed unencapsulated tumor, with diffusely infiltrating cutaneous lesions. Cosmetic and functional problems with the affected limb are the major concerns for the patient and doctor, regarding the indications for surgery. Plexiform neurofibroma is often hypervascular and can lead to severe bleeding during surgery.⁸ Surgeons and patients sometimes hesitate to undertake the challenging surgery required for huge plexi-

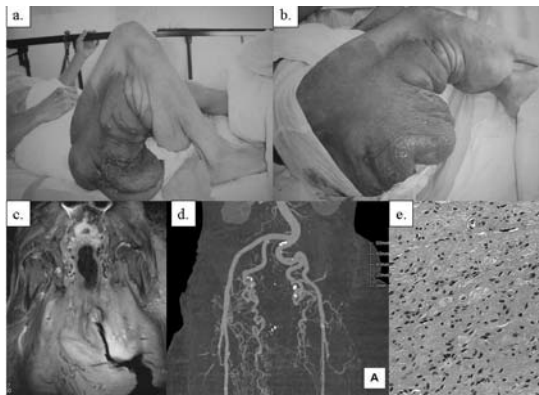


Figure 1. (a,b) The patient suffers from a huge mass of elephantiasis neuromatosa in the right leg. (c) A T2-weighted MRI sequence reveals a high-intensity myxomatous mass and hypervascularity. (d) Contrast-enhanced angiographic CT shows a rich vascular supply arising from both sides of the major external and internal iliac arteries. (e) The histopathological findings at biopsy confirm neurofibroma.

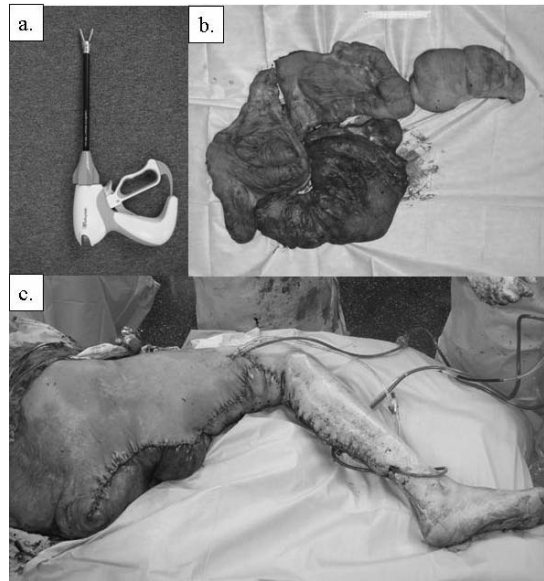


Figure 2. (a) The Ligasure® vessel sealing device. (b) Resected surgical specimens of the elephantiasis neuromatosa. (c) Gross findings of the right leg after surgery.

form neurofibromas because of the high risk. Sometimes, amputation or disarticulation, both of which are less invasive and not as technically demanding, may be recommended to the patients. For any surgical procedure, the surgeons must try to minimize the risk of intra-operative complications and to ensure that the surgery proceeds as smoothly and rapidly as possible. The Ligasure® vessel sealing system has been used in several fields of abdominal, urological and gynecological surgeries, and many clinical studies have confirmed its efficiency and safety.⁹⁻¹¹ However, little information is available concerning the clinical applications of this technology for musculoskeletal oncology of the extremities. This system is a hemostatic method that offers consistent permanent autologous sealing of vessels and tissue bundles up to 7 mm in diameter. The system is composed of an electrosurgical generator and a hand piece with a ratcheted scissor mechanism. When the sacrificed tissue is grasped and compressed by the instrument, sealing is automatically completed. After removal of the instrument, the seal is visible as a translucent

sealed zone, which can be safely divided.¹² Sealing is accomplished with minimal charring, and the delivery of thermal energy is limited to the adjacent tissues of up to 0.5-2 mm. No foreign items are left within the body, such as clipping wire and ligatures, which can act as possible sources of infection and/or allergy. The Ligasure® system seems to be safe because control of these aspects is not dependent on the experience of the operators. This system is expected to reduce the risks of intra-operative and post-operative complications compared with traditional procedures in the musculoskeletal system.

Conclusions

The Ligasure® system is a novel device for surgical debulking of extensive hypervascular masses in the musculoskeletal field, such as this case with elephantiasis neuromatosa, because of the lack of technical demands and the associated reductions in blood loss and operating time.

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