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Vacuum-assisted closure treatment of leg skin necrosis after angiographic embolization of a giant plexiform neurofibroma

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SUMMARY: **Vacuum-assisted closure treatment of leg skin necrosis after angiographic embolization of a giant plexiform neurofibroma.**

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Type 1 neurofibromatosis is a relatively common inherited disease of the nervous system, with a frequency of almost 1 in 3000. It is associated with neurofibromas of various sites. Our case report is about the surgical management of a giant neurofibroma of the right gluteal fold in a 46-year-old male with NF1. The patient presented with increasing edema and accelerated growth of the mass; he underwent percutaneous embolization of lesion vessels that induced necrosis of the neurofibroma. The patient was taken to the operating room, where surgical resection of the bulk of the lesion was undertaken. The postoperative course was complicated by delayed wound closure managed with antibiotics and vacuum-assisted wound closure.

Giant neurofibromas similar to this tumor require complex preoperative, intraoperative and postoperative management strategies. Surgical debulk is best managed with preoperative percutaneous embolization that help to avoid surgical bleeding. Postoperative delayed wound closure was managed with the application of negative pressure in a closed environment that triggers granulation and tissue formation.

RIASSUNTO: **VAC nel trattamento di estesa necrosi dei tessuti molli dell'arto inferiore post-embolizzazione di neurofibroma plessiforme.**

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La NF1 è una malattia del sistema nervoso relativamente comune (1:3000), associata alla presenza di neurofibromi in diverse sedi anatomiche e delle più disparate dimensioni. Essi si possono presentare con diversi quadri clinici, dovuti in maggior parte a compressione di strutture anatomiche adiacenti e a sanguinamento. Di seguito presentiamo il caso clinico di un uomo di 46 anni con malattia di von Recklingausen che si presenta alla nostra osservazione per la presenza di un neurofibroma gigante che si estende dal gluteo alla coscia destra, compromettendo la funzionalità e l'estetica dell'arto destro. Proponiamo un trattamento integrato con embolizzazione transfemorale preoperatoria, debulking chirurgico e medicazione postoperatoria con presidio VAC. La scelta dell'impiego preoperatorio dell'embolizzazione percutanea dei numerosi vasi neoformati trova indicazione nel limitare il sanguinamento intraoperatorio, che normalmente rende tecnicamente difficile l'asportazione chirurgica. L'utilizzo della terapia a pressione negativa trova riscontro in ferite in cui la perdita di sostanza è di notevole entità. In queste lesioni non è possibile effettuare una sintesi cutanea per prima intenzione e quindi, utilizzando la VAC, è possibile innescare la granulazione e la rigenerazione tissutale.

KEY WORDS: Vacuum-Assisted Closure - Neurofibromatosis.
VAC - Neurofibromatosi.

Introduction

Type 1 neurofibromatosis is a relatively common inherited disease of the nervous system, with a frequency of almost 1 in 3000. It is associated with neurofibromas of various sites. Our case report is about the surgical management of a giant neurofibroma of the right gluteal fold in a 46-year-old male with NF1.

Case report

We report a case of a 46-year old patient with sporadic type 1 neurofibromatosis diagnosed by the presence of *café-au-lait* spots, cutaneous and subcutaneous neurofibromas, axillary and inguinal freckles.

The patient was admitted to our Department because of a large ulcerated plexiform neurofibroma extending from the right gluteal fold to the leg, overhanging the posterior aspect of his thigh and causing severe functional impotence.

The patient underwent surgical biopsy of the neurofibroma in the right gluteus. Histological finding showed classic neurofibroma.

Angio-CT scan revealed a large soft tissue mass arising from the subcutaneous tissues of the lower back and extending to the lower part of the leg. The procedure demonstrated significant arterial supply to the lesion from some hypertrophied branches of right hypogastric and femoral arteries.

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To potentially reduce the perioperative blood loss considering the vascularity and size of the lesion, the patient underwent percutaneous embolization of some lesion vessels that induced massive necrosis of the neurofibroma (Fig. 1).

At angio-CT scan performed two weeks after radiological treatment several fluid collections were identified with satisfactory therapeutic occlusion of the lesion's vessels.

Surgical resection of the bulk of the lesion was undertaken. It was not possible to widely excise the lesion as it was infiltrating almost circumferentially the leg. Postoperatively the patient had central areas of skin break down and the closure of the surgical wound was achieved with V.A.C. (Vacuum Assisted Closure) device (Figs. 2 and 3). The dressings were changed every 48/72 hours and on discharge from the hospital the patient wound was managed for several weeks using a portable V.A.C. machine. The foam dressing was cut to the same dimensions of the wound and than placed into its bed. To overcome the skin defects the V.A.C. drape was cut into strips.

The patient mobilised independently three months after surgery with improved mobility and cosmetic results. Skin closure was complete without concomitant loss of tissue volume.

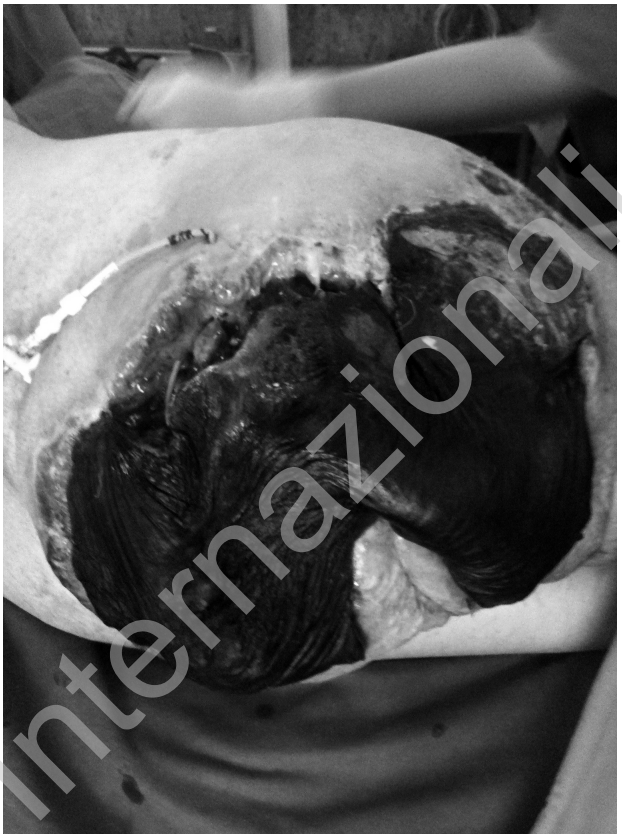

Fig. 1 - The necrosis after embolization treatment.

Discussion

NF1, known as von Reclklinghausen disease, is one of the most common inheritable disorders with an autosomal dominant transmission, an incidence of 1:3,000, and a prevalence of 1:4-5,000 (1,2). The clinical expression is extremely variable, including neoplastic and non-neoplastic disorders, mainly involving tissues of neuroectodermal or mesenchymal origin in different districts

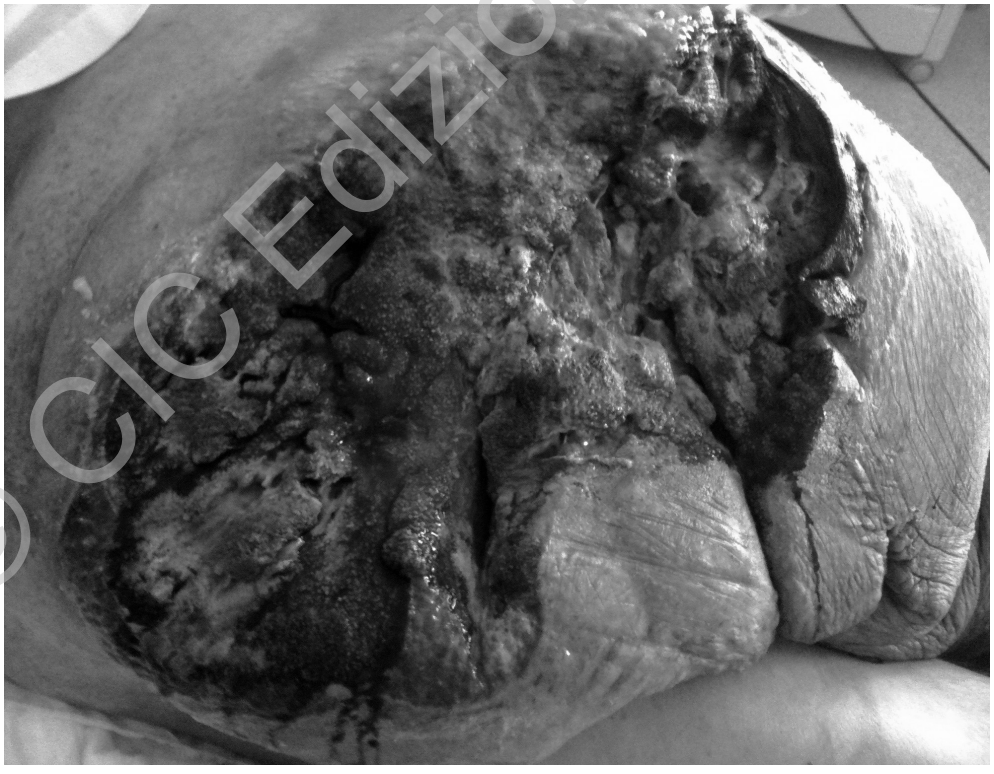

Fig. 2 - The lesion after wide excision of necrotic tissue.

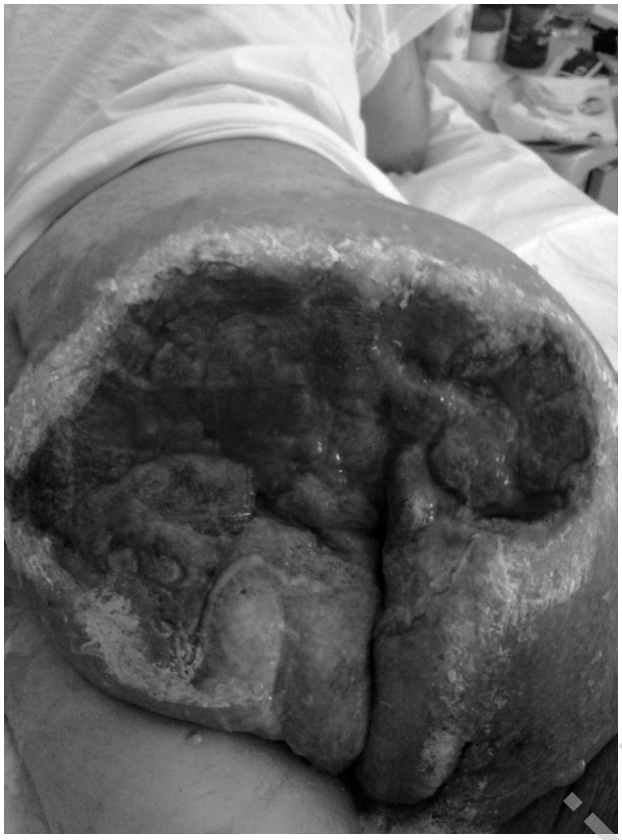

Fig. 3 - One week after VAC therapy: the granulating tissue is growing up rapidly.

such as skin, central nervous system and eye.

Plexiform neurofibromas are most often congenital tumors associated with NF 1 that can cause accelerated bone and soft tissue growth (3).

Plexiform neurofibroma, found in approximately 5% of NF 1 (4), occurs at childhood and enlarges over the years, occasionally to an enormous size.

Although this tumors are usually benign, there is a 2%-5% chance of malignant transformation in the setting of NF 1 (5).

Surgical excision of giant plexiform neurofibroma is a well accepted mode of management but there is a number of surgeons who feel it is controversial and recommend observation because of the vascular vulnerability of this type of tumor (3,6,7). In fact, neurofibromas show an increased risk of rupture of their friable vasculature secondary to arterial dysplasia or vascular invasion by the tumor (8,9).

Some Authors reported severe bleeding that needed multiple blood transfusions. Several techniques have been suggested to avoid intraoperative haemorrhage. Among these vessel embolization ensures better treatment for life-threatening bleeding.

In the case of our patient we could not perform a primary closure because of tumor size and therefore our strategy was a second-intention treatment with vacuum-assisted wound closure that demonstrated to improve wound healing successfully.

V.A.C. therapy is used in management of diffuse wounds. The principle of negative pressure relies on foam sponges being used to fill the wound void. (10). The exact mechanism by which the V.A.C. system accelerates healing is still unknown (11-13). Proposed mechanism shows that the application of negative pressure in a closed environment triggers and increases rate of granulation tissue formation (14). In addition V.A.C. increases local blood flow, decreases wound colonisation and interstitial edema (13).

Conclusion

We hope this case serves to confirm the benefit of preoperative percutaneous embolization of giant neurofibromas and the advantage of postoperative use of negative pressure healing in managing of this diffuse wound arising from giant neurofibroma located in lower extremity.

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