

Case Report

Preoperative Transarterial Embolization in the Treatment of Plexiform Neurofibroma: A Case Report and Literature Review

Reinaldo Tovo Filho^{1*}, Francisco César Carnevale², Tatiane Zago Curi¹, Fernando Melhado Tovo¹, Silmara da Costa Pereira Cestari¹, Anna Paula Vaz de Oliveira Bomtempo¹, Grazielle Cristina Palâncio Morais¹, Anaisa Raddo Venancio de Souza¹, Reinaldo Falavigna Tovo³, Luciana Carmem Zattar², Antônio Sérgio Zafred Marcelino², Giovanni Guido Cerri², Isabella Zurita Dehó¹, Maria Eduarda Ribeiro Pinto Wanderley¹ and Nathalia Araujo Lisboa¹

¹Department of Dermatology, Institute of Education and Research, Hospital Sírio Libanês, São Paulo Brazil

²Interventional Radiology Service of the Hospital Sírio Libanês, São Paulo (SP), Brazil

³Academic of Medicine Faculty at Pontifices Catholic University from Sorocaba, Sorocaba (SP), Brazil

Abstract

Neurofibromatosis type 1 (NF1) is an autosomal dominant multisystem genodermatosis resulting from a mutation on chromosome 17q11.2, characterized by diverse clinical expression with involvement of the skin, nervous system, bones, eyes and other systems. Neurofibromas are tumors located along nerve bundles; when they become large, they are called plexiform neurofibromas. Complications of neurofibromas are rare, but include malignant transformation and potentially life-threatening hemorrhages. A comprehensive perioperative management, including arterial embolization of feeding vessels, is required in order to perform a safe surgical procedure and to reduce potential surgical complications. We report a case of a large plexiform neurofibroma on the right

*Corresponding author: Reinaldo Tovo Filho, Department of Dermatology, Institute of Education and Research, Hospital Sírio Libanês, São Paulo, Brazil, Tel: +55 11987441111; E-mail: tovoclinic@uol.com.br

Citation: Filho RT, Carnevale FC, Curi TZ, Tovo FM, Cestari SCP, et al. (2020) Preoperative Transarterial Embolization in the Treatment of Plexiform Neurofibroma: A Case Report and Literature Review. J Clin Dermatol Ther 6: 063.

Received: January 23, 2021; Accepted: February 03, 2021; Published: February 10, 2021

Copyright: © 2021 Filho RT, et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

flank, treated in a two-stage approach: first, embolization with polyvinyl alcohol particles of the inferior epigastric, lumbar and superior gluteal arteries on the right and, second, 24 hours after embolization, exeresis of the lesion, which weighed 2.5 kg. Two years after surgery, the patient was asymptomatic, without tumor recurrence and with an important improvement in self-esteem and quality of life.

Keywords: Embolization; Neurofibromatosis; Plexiform neurofibromas; Surgery

Introduction

Neurofibromatosis type 1 (NF1), also known as Von Recklinghausen's disease is an 40 autosomal dominant multisystem genodermatosis resulting from a mutation on chromosome 17q11.2, which encodes neurofibromin [1]. It is one of the most common autosomal dominant disorders, with an estimated incidence of 1 in 2500 to 3300 live 43 births [2].

It is characterized by diverse clinical expression with involvement of the skin, nervous system, bones, eyes and other systems. The main cutaneous manifestations are Café-au-Lait spots, axillary freckles and neurofibromas [1]. Neurofibromas occurs in 26.7% of the cases, they are defined as tumors located along nerve bundles; when they become large, 48 they are called plexiform neurofibromas [1,3]. They often lack function and aesthetic value, thereby negatively impacting quality of life. Cases of lower extremity are less frequent than the ones located on the face and neck, and are extremely vascular [3].

Complications of neurofibromas are rare, but include malignant transformation in 2% of the cases and potentially life-threatening hemorrhages [2,3].

There is no specific treatment for NF1, and clinical manifestations are treated individually. Surgery is the preferred treatment for plexiform neurofibromas, but the significant vascularization of the tumor and the consequent possibility of large intraoperative hemorrhages make the procedure difficult [3]. Thus, preoperative embolization becomes useful to reduce potential surgical complications.

Case Report

We report the case of a 34-year-old female patient with NF1 with a large plexiform neurofibroma (39.5 x 20 x 15.7 cm) on the right flank, impairing appearance and mobility (Figure 1). High-frequency ultrasound (24 MHz) with Doppler flowmetry demonstrated a hyperechoic solid mass with irregular borders with multiple 64 hypoechogenic nodules and permeating large-caliber vascular pedicles. Due to the intense vascularization, angiotomography of the abdomen and pelvis was performed to characterize the arterial supply, and vascularization with multiple intercostal, lumbar, iliac and gluteal branches was observed. According to the clinical-radiological findings, a two-stage approach was planned: first, embolization with polyvinyl alcohol particles of the inferior epigastric, lumbar and

superior gluteal arteries on the right (Figure 2), second, 24 hours after embolization, exeresis of the lesion, which weighed 2.5 kg (Figure 3). Arteriography performed before (A) and after (B) the embolization, demonstrating devascularization of the embolized region with preservation of the other non-target arterial branches. Anatomopathological examination and immunohistochemistry confirmed a non-malignant neurogenic tumour with degenerative atypia, confirming the diagnosis of neurofibroma. Intraoperative blood loss was moderate, but the haemoglobin drop from 14.2 g/dL to 8 g/dL on the day after the procedure, requiring transfusion of two packed red blood cell units. The patient was managed by a multidisciplinary team that included individuals from the Dermatology, Plastic Surgery and Interventional Radiology departments. Two years after surgery, the patient was asymptomatic, without tumor recurrence and with an important improvement in self-esteem and quality of life (Figures 3&4).

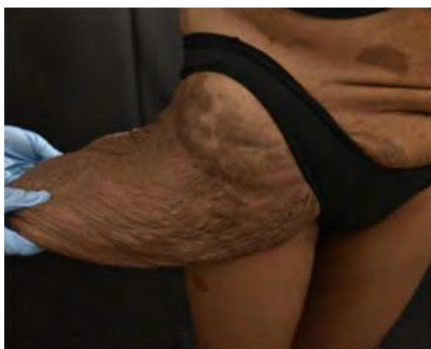


Figure 1: Large plexiform neurofibroma (39.5 x 51 x 20 x 15.7 cm) on the right flank.

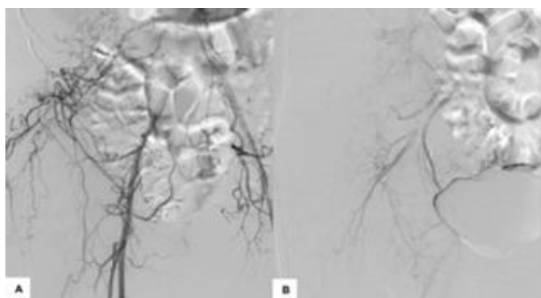


Figure 2: Intra operative: embolization with polyvinyl alcohol particles of the inferior epigastric, lumbar and superior gluteal arteries on the right.



Figure 3: Large plexiform neurofibroma (39.5 x 51 x 20 x 15.7 cm), which weighed 2.5kg.

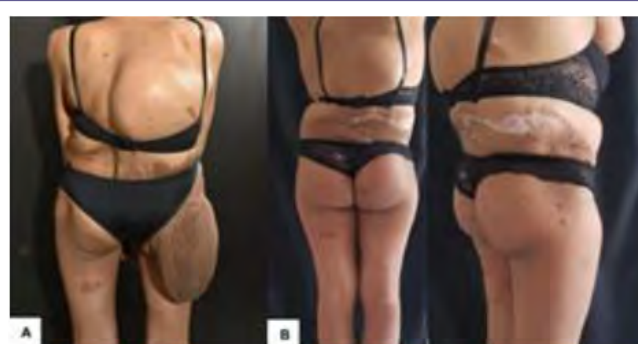


Figure 4: Plexiform neurofibroma. Patient before (A) and after two years (B) the treatment with surgery and embolization.

Discussion

Plexiform neurofibromas have variable clinical presentations, ranging from asymptomatic to invasive and disfiguring superficial lesions. In some cases, transformation to malignant tumors is observed [1]. Surgical intervention is important for histopathological diagnosis as well as for therapeutics, restoring appearance and function [3]. The intense vascularization and characteristic capillary fragility of the disease predisposes patients to a high risk of potentially fatal intraoperative haemorrhages. Therefore, preoperative embolization can be performed in an attempt to devascularize the area, reduce intraoperative bleeding and, consequently, achieve rapid haemostasis [4]. In the literature reports there are 9 cases of preoperative embolization followed by surgery in patients aged 15 to 55 years, 5 males and 4 females with plexiform neurofibromas, mainly located in the trunk. To minimize complications, it is important to properly select the material that will be used for embolization based on safety, efficacy, polymerization properties and ease of use [4]. Different embolization techniques were used by the authors [5-10], lyophilized dura mater strips, fascia lata strips, coil embolization, gelatine sponge, microsponges and an ethylene-vinyl alcohol copolymer. In the present case, polyvinyl alcohol was chosen as the embolizable particle, also used by Cavallaro, et al. and Vélez, et al. which was found to be safe and effective [5,6]. There is no consensus on the ideal interval between embolization and surgical resection. Littlewood et al. embolized a plexiform neurofibroma 3 weeks prior to surgery [7], with significant blood loss and the need for transfusion of 11 units of packed red blood cells. In our case, we performed selective embolization followed by surgery after 24 hours. This approach is important for reduction of flow in the vascular pedicles at the level of dissection and lowering the risk of adjacent tissue necrosis. Resection of plexiform neurofibromas is a difficult procedure due to the rich vascular proliferation, infiltration of deep planes and reconstruction of large areas. Therefore, adequate preoperative planning using new technologies combined with a multidisciplinary approach is important for achieving satisfactory results through functional restoration and improved appearance.

Conclusion

We describe our experience with a two-stage approach in the treatment of a large plexiform neurofibroma, using current technologies; preoperative embolization, ultracision scapel and high frequency ultrasound. The adequate preoperative management and a multidisciplinary collaboration are important in reducing

life threatening complications during the surgical exeresis of the tumor and in ensuring a safe surgical procedure. This patient was successfully treated, without any recurrence after two years of follow-up, and with an important improvement in self-esteem and quality of life.

References

1. Antônio JR, Goloni-Bertollo EM, Trídico LA (2013) Neurofibromatose: Histórico cronológico e aspectos atuais. *An Bras Dermatol* 88: 333-347.
2. Chen L, Guo C, Song X, Yan C, Hu X (2018) Long-Term Results for a One-Stage Surgery Technique for Patients With Craniofacial Plexiform Neurofibroma. *J Craniofac Surg* 29: 746-750.
3. Kolker S, Wunder JS, Roche-Nagle G (2015) Hybrid resection of a giant thigh plexiform neurofibroma. *Int J Surg Case Rep* 8: 1-4.
4. Tomei KL, Gupta V, Prestigiacomo CJ, Gandhi CD (2013) Spontaneous Hemorrhage of a Facial Neurofibroma: Endovascular Embolization Before Surgical Intervention. *J Craniofac Surg* 24: 514-517.
5. Cavallaro G, Pedullà G, Crocetti D, Giustini S, Calvieri S, et al. (2012) Vacuum-assisted closure treatment of leg skin necrosis after angiographic embolization of a giant plexiform neurofibroma. *G Chir* 33: 239-242.
6. Vélez R, Barrera-Ochoa S, Barastegui D, Pérez-Lafuente M, Romagosa C, et al. (2013) Multidisciplinary Management of a Giant Plexiform Neurofibroma by Double Sequential Preoperative Embolization and Surgical Resection. *Case Reports in Neurological Medicine* 2013: 987623.
7. Littlewood AHM, Stilwell JH (1983) The vascular features of plexiform neurofibroma with some observations on the importance of pre-operative angiography and the value of pre-operative intra arterial embolization. *Br J Plast Surg* 36: 501-506.
8. Jones RG, Kiatisevi P, Morris DC, Munket PL, Clarkson PW, et al. (2010) Intravascular embolisation and surgical resection of a giant neurofibroma with intratumoural haemorrhage. *The British Journal of Radiology* 83: 225-229.
9. Tanaka J, Kuramochi A, Nishi N, Yuasa M, Heshiki A (2005) Preoperative Transarterial Embolization Enhances the Surgical Management of Diffuse Plexiform Neurofibroma: A Case Report. *Cardiovasc Intervent Radiol* 28: 686-688.
10. D'Arpa S, Tuttolomondo A, Melloni C, Cordova A, Moschella F (2014) Management of large dorsal diffuse plexiform neurofibroma. *Eur J Plast Surg* 37: 349-352.



- Advances In Industrial Biotechnology | ISSN: 2639-5665
- Advances In Microbiology Research | ISSN: 2689-694X
- Archives Of Surgery And Surgical Education | ISSN: 2689-3126
- Archives Of Urology
- Archives Of Zoological Studies | ISSN: 2640-7779
- Current Trends Medical And Biological Engineering
- International Journal Of Case Reports And Therapeutic Studies | ISSN: 2689-310X
- Journal Of Addiction & Addictive Disorders | ISSN: 2578-7276
- Journal Of Agronomy & Agricultural Science | ISSN: 2689-8292
- Journal Of AIDS Clinical Research & STDs | ISSN: 2572-7370
- Journal Of Alcoholism Drug Abuse & Substance Dependence | ISSN: 2572-9594
- Journal Of Allergy Disorders & Therapy | ISSN: 2470-749X
- Journal Of Alternative Complementary & Integrative Medicine | ISSN: 2470-7562
- Journal Of Alzheimers & Neurodegenerative Diseases | ISSN: 2572-9608
- Journal Of Anesthesia & Clinical Care | ISSN: 2378-8879
- Journal Of Angiology & Vascular Surgery | ISSN: 2572-7397
- Journal Of Animal Research & Veterinary Science | ISSN: 2639-3751
- Journal Of Aquaculture & Fisheries | ISSN: 2576-5523
- Journal Of Atmospheric & Earth Sciences | ISSN: 2689-8780
- Journal Of Biotech Research & Biochemistry
- Journal Of Brain & Neuroscience Research
- Journal Of Cancer Biology & Treatment | ISSN: 2470-7546
- Journal Of Cardiology Study & Research | ISSN: 2640-768X
- Journal Of Cell Biology & Cell Metabolism | ISSN: 2381-1943
- Journal Of Clinical Dermatology & Therapy | ISSN: 2378-8771
- Journal Of Clinical Immunology & Immunotherapy | ISSN: 2378-8844
- Journal Of Clinical Studies & Medical Case Reports | ISSN: 2378-8801
- Journal Of Community Medicine & Public Health Care | ISSN: 2381-1978
- Journal Of Cytology & Tissue Biology | ISSN: 2378-9107
- Journal Of Dairy Research & Technology | ISSN: 2688-9315
- Journal Of Dentistry Oral Health & Cosmesis | ISSN: 2473-6783
- Journal Of Diabetes & Metabolic Disorders | ISSN: 2381-201X
- Journal Of Emergency Medicine Trauma & Surgical Care | ISSN: 2378-8798
- Journal Of Environmental Science Current Research | ISSN: 2643-5020
- Journal Of Food Science & Nutrition | ISSN: 2470-1076
- Journal Of Forensic Legal & Investigative Sciences | ISSN: 2473-733X
- Journal Of Gastroenterology & Hepatology Research | ISSN: 2574-2566
- Journal Of Genetics & Genomic Sciences | ISSN: 2574-2485
- Journal Of Gerontology & Geriatric Medicine | ISSN: 2381-8662
- Journal Of Hematology Blood Transfusion & Disorders | ISSN: 2572-2999
- Journal Of Hospice & Palliative Medical Care
- Journal Of Human Endocrinology | ISSN: 2572-9640
- Journal Of Infectious & Non Infectious Diseases | ISSN: 2381-8654
- Journal Of Internal Medicine & Primary Healthcare | ISSN: 2574-2493
- Journal Of Light & Laser Current Trends
- Journal Of Medicine Study & Research | ISSN: 2639-5657
- Journal Of Modern Chemical Sciences
- Journal Of Nanotechnology Nanomedicine & Nanobiotechnology | ISSN: 2381-2044
- Journal Of Neonatology & Clinical Pediatrics | ISSN: 2378-878X
- Journal Of Nephrology & Renal Therapy | ISSN: 2473-7313
- Journal Of Non Invasive Vascular Investigation | ISSN: 2572-7400
- Journal Of Nuclear Medicine Radiology & Radiation Therapy | ISSN: 2572-7419
- Journal Of Obesity & Weight Loss | ISSN: 2473-7372
- Journal Of Ophthalmology & Clinical Research | ISSN: 2378-8887
- Journal Of Orthopedic Research & Physiotherapy | ISSN: 2381-2052
- Journal Of Otolaryngology Head & Neck Surgery | ISSN: 2573-010X
- Journal Of Pathology Clinical & Medical Research
- Journal Of Pharmacology Pharmaceutics & Pharmacovigilance | ISSN: 2639-5649
- Journal Of Physical Medicine Rehabilitation & Disabilities | ISSN: 2381-8670
- Journal Of Plant Science Current Research | ISSN: 2639-3743
- Journal Of Practical & Professional Nursing | ISSN: 2639-5681
- Journal Of Protein Research & Bioinformatics
- Journal Of Psychiatry Depression & Anxiety | ISSN: 2573-0150
- Journal Of Pulmonary Medicine & Respiratory Research | ISSN: 2573-0177
- Journal Of Reproductive Medicine Gynaecology & Obstetrics | ISSN: 2574-2574
- Journal Of Stem Cells Research Development & Therapy | ISSN: 2381-2060
- Journal Of Surgery Current Trends & Innovations | ISSN: 2578-7284
- Journal Of Toxicology Current Research | ISSN: 2639-3735
- Journal Of Translational Science And Research
- Journal Of Vaccines Research & Vaccination | ISSN: 2573-0193
- Journal Of Virology & Antivirals
- Sports Medicine And Injury Care Journal | ISSN: 2689-8829
- Trends In Anatomy & Physiology | ISSN: 2640-7752

Submit Your Manuscript: <https://www.heraldopenaccess.us/submit-manuscript>