JMS 2021; 4(1):126-129
UDC: 617.586-005.98-089.168.1-06

SURGICAL TREATMENT OF PRIMARY LYMPHEDEMA COMPLICATIONS – A CASE REPORT

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Abstract

We present a case of a 28-year-old woman with primary lymphedema of the right leg, with tumor formations on her right foot, causing functional disabilities. Tumor formations were removed and histologically confirmed as dermatofibromas. Five years later, the patient was readmitted due to foot problems, keratosis of the heel, as well as increased edema of the dorsum of the foot and lower leg.

Liposuction and excision of lymphoedematous tissue from the dorsum of the foot, release of the constrictive band at the level of the anterior surface of the ankle with "Z" plasty, as well as liposuction of lymphoedematous tissue of the lower leg and thigh were performed. The postoperative course was without complications, with a light seroma at the dorsum of the foot.

During the 4-year follow-up period, the patient showed a noticeable improvement, with a slow increase in the volume of the lower leg and thigh, which did not reach the preoperative dimensions. Unfortunately, lymphedematous tissue increased again on the dorsum of the foot as prior to surgery.

Keywords: lymphedema, tumor, surgery

Introduction

Lymphedema is a progressive swelling of the intercellular tissue of a certain part of the body, most often one or more extremities, trunk, head, genitals, where lymph accumulates in the intercellular space, because it cannot drain through lymphatic or venous circulation.

Lymphedema is a severe, chronic, progressive, long-lasting and incurable disease. Swelling, feeling of fullness and heaviness reduce the range of motion and tightness in the joint, cause a slight discomfort with the possibility of pain or tingling sensation, repeated episodes of infection because lymphedema is a chronic inflammatory process (skin warmer and slightly redder).

In addition, there are episodes of cellulitis or lymphangitis cases accompanied with thickening of the skin, hardening of the extremities, leakage of lymph (lymphorrhea) and extremely large swelling – elephantiasis [1, 2].

A case report

A 28-year-old woman came to the Clinic for Plastic and Reconstructive Surgery due to pronounced tumorous changes in her right foot which made performing normal functions quite strenuous. From the anamnesis we realized that the lymphedema of the right leg was diagnosed from birth and the patient had two uncomplicated pregnancies. Seven years before her first admission to the hospital, tumor formations had started to grow on her right foot, which had been treated only conservatively, using elastic stockings and bandages (Figure 1).

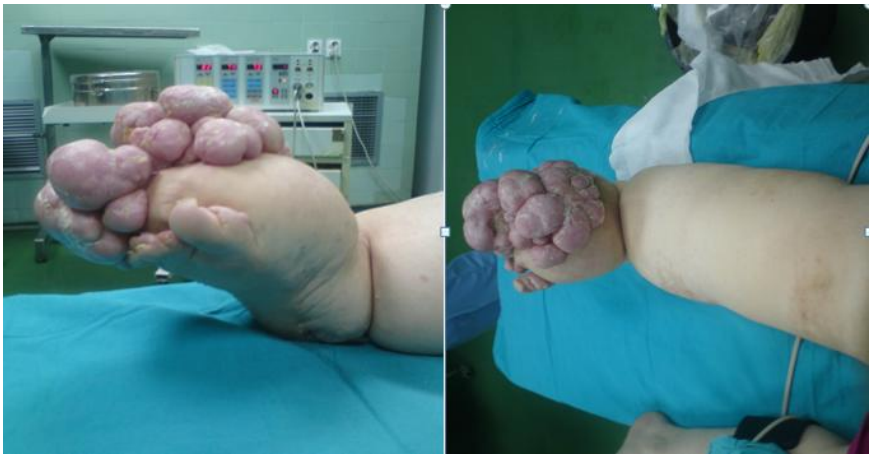

Figure 1.

After admission, lymphoscintigraphy was performed, and interruption of the lymphatic drainage in the upper third of the femoral region was observed. A standard preoperative preparation was performed; the patient underwent surgery and the tumors were removed (histologically confirmed as dermatofibromas).

Most of the lymphoedematous tissue of the dorsum of the right foot was removed during the surgery. The postoperative period was without complications.

The patient was advised to apply manual drainage at home and to wear elastic socks. Five years after the first surgery, the patient was readmitted due to foot problems, keratosis of the heel, as well as increased edema of the dorsum of the foot and lower leg.

The dimensions of the lower and the upper leg were measured preoperatively in two places. After the treatment options were explained to the patient, a second surgery was performed.

Liposuction and excision of lymphoedematous tissue from the dorsum of the foot, release of the constrictive band at the level of the anterior surface of the ankle with "Z" plasty, as well as liposuction of lymphoedematous tissue of the lower leg and thigh were performed (Figure 2).

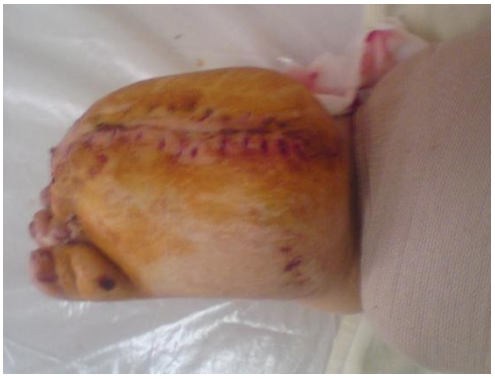

Figure 2.

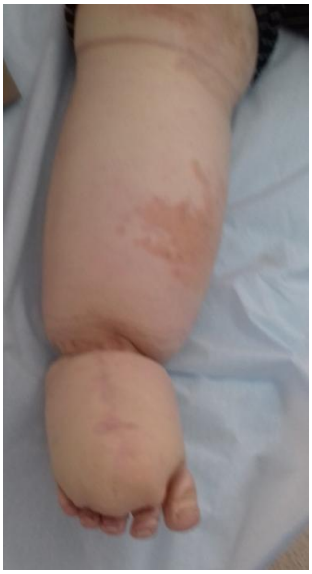

Figure 3.

The postoperative course was without complications, with a slight seroma at the dorsum of the foot. During the 4-year follow-up period, the patient showed a noticeable improvement, with a slow increase in the volume of the lower leg and thigh, which did not reach the preoperative dimensions. Unfortunately, lymphedematous tissue increased again on the dorsum of the foot as prior to the surgery (Figure 3). The patient continued with conservative treatment, wearing elastic stockings, satisfied with the result of the surgical treatment.

Discussion

Lymphedema is classified as primary or secondary according to the underlying etiology. Primary lymphedema represents developmental lymphatic vascular deficiency which can be either congenital or hereditary. A number of mutations of genes involved in lymphatic development are associated with primary lymphedema (i.e., GJC2, FOXC2, CCBE1, VGFR-3, PTPN14, GATA2, and SOX18).

Although developmental anomalies are present at birth, lymphedema may develop at some time later in life. Congenital lymphedema is clinically evident at birth or within the first two years of life. Many lymphatic malformations have been defined as different diseases, such as Milroy and Meige disease. If primary lymphedema presents at birth, but exists before the age of 35 years, it is named lymphedema praecox.

This form is common and it frequently arises during puberty or pregnancy. Lymphedema tarda is relatively rare and develops after the age of 35. Lymphangiomas are uncommon, congenital, benign, often cystic malformations of the lymphatics and may be associated with other vascular malformations [1, 2, 3, 4].

Secondary lymphedema can be acquired and may arise as a result of cancer surgery, radiation therapy, chronic venous insufficiency, lipedema, trauma, infection, immobility, or underlying systemic diseases. The most common cause of lymphedema worldwide is filariasis.

We used the standard diagnostic test of measuring the circumference of the extremities every 8-12 cm and comparing them with the healthy one; a difference of over 2 cm in circumference in terms of increase was an indication for lymphedema. We also measured the dimensions of the lower and the upper leg preoperatively and postoperatively.

Surgical treatment

Physiological reconstruction of lymphatics and resection operations do not yield good results and usually result in complications (long lymphorrhea, wound infection, skin dehiscence, skin necrosis, thrombosis, scarring, and scarring contractures to amputation).

They are divided into: resection, lymphosuction, reconstructive (lymph-venous, lympho-venous, lympho-lympho anastomoses) and drainage operations or a combination of the before mentioned. The multiplicity of operations only speaks of their lack of success.

The goals of surgical treatment are to improve the function, reduce the weight of the extremities, patient's ailments, prevent complications of lymphedema (cellulitis, lymphangitis, lymphorrhea, chyloderma, lymphangiosarcoma) and a better aesthetic appearance of the extremities. Today, about 100 types of surgical procedures are applicable, and none of them gives the expected results [5-9].

Resection involves excision of lymphedematous tissue, the most commonly used type, but is burdened with frequent and significant complications.

Lymphosuction is a reduction operation for lymphedema and is always accompanied with postoperative external compression.

Drainage bridges the 'lymph block' and stagnation or removes lymph from the affected limb without direct interventions on the affected lymph vessels.

Reconstructive surgery represents reconstruction of lymph flow by a direct intervention on lymphatic vessels, current with the development of microsurgery.

Combined procedures involve the use of multiple methods. Central lymphatic block, i.e. hylos reflux is also an unresolved surgical problem [5-9].

We performed liposuction and excision of lymphoedematous tissue from the dorsum of the foot, release of the constrictive band at the level of the anterior surface of the ankle with "Z" plasty, as well as liposuction of lymphoedematous tissue of the lower leg and thigh.

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