

A Case of a Huge Unilateral Lower Limb Swelling of an Adult in South India

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Abstract: ***Background:** Our objective is to present a clinical case of elephantiasis in an adult living in South India which progressed over 4 years. The objectives of this case report are to discuss the clinical presentation, and appropriate management of this malformation. **Materials and Methods:** A clinical case of a patient with a huge unilateral lower limb swelling is presented. The patients presentation, management and the follow up procedures are discussed. **CONCLUSION:** Lymphatic obstruction causes an increase in the protein content of the extravascular tissue, with subsequent retention of water and swelling of the soft tissue. The increase in the extravascular protein stimulates proliferation of fibroblasts, organization of the fluid, and the development of a non-pitting swelling of the affected extremity.¹*

Keywords: elephantiasis

1. Introduction

Lymphedema is caused by a compromised lymphatic system that impedes and diminishes lymphatic return. In primary lymphedema, the failure is caused by congenital hypoplasia or aplasia of the peripheral lymphatics or by valvular incompetence. In secondary lymphedema, the lymphatic drainage is altered by an acquired blockade of the lymph nodes due to infection, malignancy, Radiotherapy or by disruption of the local lymphatic caused by one of the following etiologies: recurrent attacks of lymphangitis, malignancy, obesity and surgery.^{2,3,4}

Lymphatic filariasis affects nearly 120 million people in the tropical and subtropical regions of the world and 66% of the people at risk of the disease live in WHO's Southeast Asia region—including India. The filarial nematodes (*Wuchereria bancrofti* and *Brugia malayi*) that cause these diseases are transmitted by blood-feeding insects and produce chronic and long-term infection through suppression of host immunity. Disease pathogenesis is linked to host inflammation invoked by the death of the parasite, causing hydrocoele, lymphoedema, and elephantiasis in lymphatic filariasis.⁵

2. Case Report

A 40 year old male presented with chronic and massive left lower extremity swelling—clinically graded as Stage III Lymphedema (Figure 1,2) Massive disfiguring lymphedema of his lower leg caused pain, difficulty ambulating, and the inability to lift his leg.

O/E- Gross swelling extending from foot upto groin. Apparent skin thickening of left LL. Skin hangs in folds. Trophic skin changes such as acanthosis nigricans and warty overgrowths present. Scrotum and external genitalia appear normal.

Palpation- Pitting is absent. Fibrosis of skin of left lower limb. Hyperkeratosis+



Doppler USG was done of left LL which reported excess dermal subcutaneous fluid accumulation and normal arterial and venous vasculature with no change in the muscle mass.

Nocturnal peripheral smear for *Microfilaria* was Negative.

Since patient did not have filariasis, the full dose of diethylcarbamazine (DEC) (6 mg/kg/day divided tid PO) was given at the beginning on day 1 and consecutively for 14 days.

Surgical management was not considered for this patient in view of co-morbidities and relative risk for operation.

3. Discussion

Lymphatic filariasis and onchocerciasis are parasitic helminth diseases that constitute a serious public health issue in tropical regions.

Initial damage to lymphatic vessels may remain subclinical for years. Manifestations of chronic lymphatic filariasis occur mostly in adults 30 yr of age or older and result from anatomic and functional obstruction to lymph flow. This obstruction results in lymphedema of the legs, arms, breasts, and/or genitalia. Male genital involvement, such as hydrocoele, is very common in *W. bancrofti* infection, but uncommon in *Brugia* species infection. Chronic lymph stasis predisposes affected extremities to bacterial superinfections,

sclerosis, and verrucous skin changes, resulting in elephantiasis, which may involve 1 or more limbs, the breasts, or genitalia. They cause long-term chronic infection as reported in this case with a few infected individuals developing overt disease⁶.

Drugs used to treat and control filariasis include diethylcarbamazine, ivermectin, and albendazole. Given as a single dose of 6 mg/kg, diethylcarbamazine is effective for reduction of acute and chronic cases of microfilaraemia for at least 1 year, and is the basis of mass drug distribution by the global programme to eliminate lymphatic filariasis in areas without co-endemic onchocerciasis^{7,8,9}. A regimen of 6 mg/kg diethylcarbamazine for 12 consecutive days; where the dose of DEC may increased gradually to avoid treatment-associated complications such as pruritus, fever, generalized body pain, hypotension, and even death in persons with high microfilarial levels,, is better than is the single dose at leading to absence of blood microfilariae, and can be given to individuals if supervised by a doctor.

Lymphoedema is chronic and progressive and affects a significant proportion of the population. The goal of lymphedema therapy is to restore function, reduce physical and psychologic suffering, and prevent the development of infection by initiating therapy as early as possible before extensive, irreversible fibrosclerotic changes occur in the interstitium.^{10,11,12}

Complex decongestive therapy (CDT) is a comprehensive lymphedema reduction program that combines elevation, "remedial" exercise, manual lymphatic drainage massage, and compression wraps, and represents the current international standard of care and first-line therapy for stage II and III lymphedema.^{11,12}. The program involves two phases, each with four discrete components .

Phase I CDT includes four components: manual lymph drainage, an exercise regimen, multilayered low-stretch wrapping, and skin care. It is the current "gold standard" of lymphedema management^{13,14}. Although phase I CDT can achieve abrupt and dramatic reductions in limb volume, long-term success requires ongoing maintenance with a phase II home-based program. In the absence of consistent adherence, a patient's lymphedema volume will inevitably re-accumulate.

Elastic wrapping is the principal compression technique used in phase I CDT. Wrapping techniques are complex and favour the use of low-stretch, in lieu of the more traditional high-stretch, bandages as it maintains unyielding resistance that increases the pressure created by the muscle pump with movement also. During phase I CDT, low-stretch wraps are worn 24 hours a day (except when the patient is receiving a massage or bathing). In the maintenance phase, a compression garment is worn during the day and the wraps continue to be worn at night.

Graded compression garments are designed to provide the maximum compression in the distal portion of the limb, with progressively less pressure as the garment extends proximally. Lower limbs with chronic lymphedema

generally require 30 to 40 mm Hg of compression at the ankle to control the swelling.

Exercise, although often undervalued by patients and practitioners alike is integral to lymphedema management. It involves repeated contraction and relaxation of muscles within a lymphedematous body part. Muscles are contracted and relaxed in specific sequences, depending on the location of the lymphatic congestion in an attempt to mobilize lymph from the distal to proximal areas of the affected body part. When performed within compression bandages, it supposedly creates an internal pumping mechanism that promotes lymph absorption and transport.

Surgical options available can broadly be divided into physiological and reductive types. While the former aims to restore lymphatic flow and includes flap transposition, nodal transfers and bypass procedures, the latter attempts to treat the consequences of sustained lymphatic stasis by removing the fibrofatty tissue that has been pathologically created by excision or liposuction.

Delayed diagnosis and treatment of non-cancer-related lymphedema result in increased severity and impaired mobility, making this a unique, complex condition requiring modifications to CDT in the outpatient setting.

Late-onset malignancies are a potentially devastating, but fortunately rare, complication of long-standing lymphedema. In most series, they develop in no more than 1% of patients with lymphedema.

We were able to show that an integrative concept including surgery is a good additional option for the treatment of severe cases of lymphedema in appropriate candidates. Furthermore, an adequate perioperative conservative setting helps to minimize possible complications.

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