

## Approach to Elephantiasis Nostra of Unclear Etiology: A Case Report with a Brief Review

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### Abstract

Lower extremity lymphedema is an important clinical condition causing morbidity and is frequently encountered by the phlebologists. Elephantiasis Nostra is the status characterized with the extraordinary massive swelling of one or both legs with subsequent thickening and fibrosis of the overlying skin. It is an exaggerated manifestation of a longstanding chronic lymphedema. Etiologically, secondary lymphedema is caused by an external effect such as a chronic lymphangitis, removal of the lymph nodes, trauma, mechanical obstruction, radiotherapy, venous insufficiency, obesity, heart failure, bacterial or helminthic infection (Lymphatic Filariasis), in contrary to the primary lymphedema in which an inherent malfunction of lymphatic channels exists. A morbidly obese female patient with a bilateral Elephantiasis Nostra and our effort on setting the etiological definition and the treatment approach is presented.

**Key words:** Lymphoedema; obesity; leg ulcers

### Introduction

Elephantiasis is a progressive enlargement of an extremity or body part accompanied by a chronic inflammatory fibrosis of dermal and subdermal tissues caused by chronic lymphatic or venous stasis<sup>1</sup>. It is an end symptom or a manifestation of a spectrum of different etiologies rather than a disease itself<sup>2</sup>. Elephantiasis usually affects limbs and scrotum resulting that part of the body are enormously bulked<sup>3</sup>. It is characterized with massive swelling together with thickening and fibrosis of the skin and underlying tissues due to the chronic nature of the underlying lymphedema<sup>4</sup>. Elephantiasis can be classified into five main title according to the underlying etiology; Elephantiasis Nostra is the condition caused by long lasting chronic lymphedema or lymphangitis (Non-Filarial Elephantiasis), Elephantiasis Tropica (or Lymphatic Filariasis) is the condition caused by the tropical parasites particularly *Wuchereria bancrofti*, Podoconiosis is an immune condition affecting the lymphatic vessels, Genital

Elephantiasis is the end result of lymphogranuloma venereum, and lastly the Proteus Syndrome which is an genetic disorder widely called as "Elephant Man"<sup>4</sup>. Although Elephantiasis Nostra and Elephantiasis Tropica both present similar clinical manifestations and both are mainly characterized by impaired lymphatic drainage, the former is caused by infections or trauma on the basis of chronic lymphedema while the latter is caused by helminthic infections<sup>5</sup>.

### Case Report

A 46-year-old woman with morbid obesity (Weight:130 kg, Height: 156 cm, Body Mass Index-BMI: 53,43) admitted to our clinic complaining of bilateral leg swelling which is worse in the left side. She was also complaining of weight gain in addition with a decrease in effort capacity. As a medical history, she claimed that the swelling began when she was 24-year-old, immediately after a left ankle and tibial fracture

occurred 22 years ago. Her symptoms started gradually in a month and progressively worsen from that time on. She also stated that she has progressively gained weight after that immobilization process due to this fracture, otherwise she was in normal weight. After 17 years from the initial onset of the enlargement on the left lower extremity, namely 5 years ago, the right leg also started to swell as the other. She had an open heart surgery for the secundum type atrial septal defect 17 years ago with no postoperative complication. She had three eventless pregnancies ending up with normal deliveries via spontaneous vaginal way without a history of puerperal sepsis. She had no travel history of tropical regions and no family history of familial lymphedema.

The physical examination of the significantly enlarged and malodorous lower extremities revealed massive hypertrophic changes of chronic non-pitting edema with hyperkeratosis, hyperpigmentation, crusts, purpuras which are extended over the inguinal region up to the abdominal adipose tissue bilaterally (Fig 1). The signs of the left leg were much worse than the right. As an acute manifestation, generalized erythema and warmth of both legs and weeping superficial ulcer on the posterior aspect over the left ankle were observed. Measurements of both legs at the levels of ankle, mid-calf and mid-thigh were; 52, 63 and 87 cm in the left and 46, 54 and 80 cm in right leg respectively.

Her temperature on admission was 36,8 C. Blood pressure was 120/70 mmHg and heart rate was 80/min. Patient was suffering dyspnea with a mild decrease in oxygen saturation of 92%. Laboratory investigations revealed a slightly elevated C-reactive protein level of 0.83 mg/dL (normal, 0 to 0.5) and a low iron level of 23 ug/dL (normal, 60 to 180). Complete blood count (CBC) revealed slightly decreased hemoglobin level of 10,8 g/dl (normal, 11 to 16.2), hematocrit level of 32.9% (normal, 36 to 47.9) with a mean corpuscular volume (MCV) of 68.6 fL (normal, 80 to 100) indicating microcytic iron deficiency anemia. CBC revealed no increase in leucocyte (WBC) level suggesting acute bacterial infection. Biochemical and serologic test including lipid profile together with thyroid, liver and renal functions were in normal range. A peripheral blood smear and test for specific microfilaria antigen were negative.

Direct X-ray showed bilateral partial interstitial pulmonary edema and slight cardiomegaly. Lower extremity arterial Duplex Ultrasound (DUS) investigation revealed bilateral triphasic currents in all arterial segments down to the dorsalis pedis artery. Lower extremity venous DUS images revealed bilateral grade 2 (reflux of 1.0-1.5 sec) greater saphenous vein insufficiency with augmentation and no venous thrombosis regarding the deep venous system. Lower

extremity Magnetic Resonance Imaging (MRI) revealed massive lymphedema and structural deformity of the tissues with honeycomb view which is a characteristic appearance for elephantiasis (Fig 2). (6) No tumor or space occupying lesion were inspected. Echocardiographic evaluation revealed Left Ventricular Ejection Fraction (LVEF) of 50% with grade 3-4 mitral valve insufficiency.

According to these clinical, radiological and laboratory findings, she was diagnosed as chronic lymphedema which progressed to the state of Elephantiasis Nostra. Third-generation cephalosporin at 3 gr per day empirically and furosemide 80 mg per day were started intravenously to control pulmonary and peripheral edema and to prevent probable superimposing bacterial infections. Low molecular weight heparin 6000 IU per day was started subcutaneously to prevent venous thromboembolism due to immobilization. Acetylsalicylic Acid (ASA) 100 mg and Diosmin-Hesperidin 1000 mg per day were started orally for an extra antiaggregation and to augment the venous tonus. Antimicrobial and scatrizing ointments (Ethylenglicol-monophenyl-ether and Dexapanthenol) were applied topically following the daily dressing with iodine solution to heal the weeping sore. Elevation of both legs above the heart level and pneumatic compression therapy were both continued until discharge.

At the end of the second week, the therapy resulted in partial resolution of the lymphedema and decrease in diameters of the limbs. The open sore was totally cured and exfoliations of the skin were showed slight progression. However, due to the chronic nature of the disease arising from the impaired lymphatic drainage, no great curative achievements apart from the palliative relief of the patients discomfort, would be expected. The patient was recommended for the elevation of the limbs and application of an elastic bandage as a home care and discharged with a prescription including ASA, venotonics, diuretics, iron replacement and topical medication. We also encouraged her to loose weight and increase ambulation while scheduling for a one-month follow-up.

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

**Figure 1.** Elephantiasis Nostra: Anterior and posterior views of the patient.



**Figure 2.** MRI of lower limbs showing the fibrotic and thickened skin and edematous subcutaneous tissue with a honeycomb appearance (T2 images of thigh and calf on Transverse plane).

### Discussion

Elephantiasis which is called 'Herculesian Disease' by the ancient Greeks has always been of interest among the population as well as the physicians<sup>2</sup>. Until the middle ages, it was commonly confused with leprosy as they represent some identical symptoms considering dermal hypertrophy in certain phases<sup>2</sup>. In certain African tribes, the victims of this disease were sainted just because it was thought to be the favor of gods to have massively enlarged scrotum or leg<sup>2</sup>. In 1934, Aldo

Castellani, an Italian microbiologist and physician made an etiologic description and he used the term 'Elephantiasis Nostra' to distinguish the non-filarial etiology from others<sup>4</sup>. The Italian word 'Nostra' means 'ours' and enables to differentiate Elephantiasis Nostra from Elephantiasis Tropica (Filariasis) which is caused by helminthic *Wuchereria* worms that invade and occlude the lymphatics<sup>1</sup>.

An edematous leg may be due to regional or systemic causes. Systemic causes constitute the congestive cardiomyopathy, hypoalbuminemia, renal insufficiency and protein-losing nephropathy. Regional causes include primary and secondary lymphedema, lipedema, chronic venous disease, deep venous thrombosis, cellulitis and postoperative complication following intraabdominal surgery<sup>6</sup>.

Primary lymphedema is caused by a congenital abnormality or dysfunction of the lymphatic system and is further classified according to the age of onset. The congenital lymphedema detected at birth is known as *Milroy Disease* and is rare<sup>7</sup>. The congenital lymphedema detected between the ages of 1 and 35 is known as *Lymphedema Praecox*, whereas the third type *Lymphedema Tarda* is seen after the age of 35<sup>8</sup>. Lymphedema Praecox is the mostly encountered variety and is particularly common in females<sup>9</sup>. Symptoms may be triggered by a minor trauma<sup>10</sup>.

Secondary lymphedema is caused by an acquired reduction in lymph flow. Trauma, recurrent infections, metastatic disease and radiotherapy are the responsible causes<sup>11</sup>. The most common cause of lymphedema in the developed world is mainly the interventions for the treatment of the malignancies such as radiotherapy and radical lymph node dissections<sup>5</sup>. However, lymphedema is much more common in the developing world, due to a parasitic nematode infection named *Wuchereria Bancrofti* (also known as filariasis), making it the most common cause of lymphedema over the world<sup>6</sup>.

In the light of all these information, etiological discrimination of this reported case may be debated. So what was the real etiology of this chronic lymphedema? Was it primary or secondary lymphedema?

Regarding a patient with an onset of lymphedema at 23 years of age, without any significant history except from a history of a negligible fracture, the etiology can easily be thought as primary lymphedema, namely Praecox lymphedema. Praecox lymphedema (Meige's Disease) usually affects young women and is typically unilateral<sup>5</sup>. It should be also remembered that the initial symptoms of this presented case arise after a minor trauma and are unilateral. But she also had no familial history regarding lymphedema. However, absence of a familial history do not exclude the possibility of primary

lymphedema because it can also occur sporadically as well as familial inheritance<sup>6</sup>.

On the other hand, her etiology may also be due to a secondary reason. The possible reason may be a recurrent infection like onychomycosis or recurrent cellulitis due to an incompletely treated bacterial infection that the patient is unable to remember. These infections may be underestimated during their onset, but they impair the lymphatics and further progress along the extremity soon after, ultimately resulting in a sign as this patient has. So, all this process of vicious circle may result in a such catastrophic manifestation called "Elephantiasis Nostra", an exaggerated manifestation of the chronic secondary lymphedema which has been accompanied and complicated by fungal or bacterial infections<sup>5</sup>.

Another scenario can be attributed to the bone fracture occurred just previous to the onset of the symptoms. Considering the patient's declaration, 22 years ago, the initial treatment of the fracture was made by a non-qualified bonesetter instead of a medical specialist. Accordingly, lymphatic vessels might be impaired and an infection might further be superimposed and progressed to the entire lymphatic system of the fractured side following this mistreatment. In addition, progressively gained weight in the following period might also aggravated the clinic. This might be the simplest reason of all.

Venous stasis dermatitis, congenital lymphedema, lipedema, lipodermatosclerosis, pretibial myxedema, filariasis should all be considered as the differential diagnosis of the Elephantiasis Nostra<sup>1,12,13</sup>. In the case of venous stasis dermatitis, due to the chronic venous hypertension caused by the reflux of the deep, superficial or perforator venous system, some skin changes including pitting edema, pruritic brown patches may also occur. Thus, chronic venous insufficiency can be difficult to differentiate from the initial phase of lymphedema because both have the same pitting edema while the typical skin changes of the late-stage lymphedema are not yet existed<sup>5</sup>. DUS imaging can be used to distinguish the two, since chronic venous insufficiency may actually cause secondary lymphedema<sup>8</sup>.

Considering all types of lymphedema progressed to Elephantiasis, detailed history and physical examination, laboratory tests including biochemistry, CBC, blood smears and cultures for parasites, DUS imaging (to distinguish lymphedema from any other venous pathology), Computerized Tomography and MRI (to evaluate the dermal and subdermal trophic changes), lymphoscintigraphy, echocardiography (to exclude cardiac failure) should be performed to find out the exact etiological reason<sup>8,11,14,15</sup>. In the

presented patient, lymphoscintigraphy was done in a different center, reported as lymphatic interruption in both inguinal lymphatics.

Whatever the etiology is, treatment should aim to reduce the size of the affected region, healing the ulcers, improving skin condition and mobilization of the patient. Along with the medication, the surgical option should be kept reserved. Basically two types of surgical approach (debulking and restorative) are available<sup>16</sup>. However, the impaired regenerative capacity of the tissues due to chronic changes reduce the success of the surgery. In addition to all these physical considerations, the emotional status of the patient should not be ignored. Consultation for a psychotherapy should be arranged if needed.

As a conclusion, although most of the articles regarding the Elephantiasis manifest in the literature of dermatology, clinical presentation mostly addresses the phlebologist. In this comprehensive review which exemplified with a tangible case report, it was aimed to provide an overall perspective to this subject from a vascular surgeons point of view.

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