

CASE REPORT

Massive lymphedema as elephantiasis in the background of an alcoholic chronic liver disease

Linfedema grave tipo elefantiasis secundario a insuficiencia hepática crónica alcohólica

M^a Elena Arnáiz-García^{1*}; Ana María Arnáiz-García²; Juan Francisco Nistal Herrera³; Aida García-Martín⁴; David Alonso-Peña⁵; Javier Arnáiz García⁶.

¹Cardiac Surgery Department. University Hospital of Salamanca. Salamanca. Spain.

²Internal Medicine Department-Infectious Diseases Unit. Marqués de Valdecilla University Hospital. Santander. Spain

³Cardiovascular Surgery Department. Marqués de Valdecilla University Hospital. Santander. Spain

⁴Faculty of Podiatry. University of Madrid. Spain

⁵Plastic Surgery Department. Hospital Universitario Rio Hortega. Valladolid. ⁶Radiology Department. Aspetar-Orthopaedic and Sports Medicine Hospital. Al Buwairda St. Doha. Qatar.

*Correspondencia: M^aElena Arnáiz-García. Cardiovascular Surgery Department. University Hospital of Salamanca.Spain.Paseo San Vicente 58-182.Salamanca.Spain. Phone: 0034923291263.Fax:0034923291263.Email:elarnaiz@hotmail.com.

Abstract

The cluster of lymph fluid subcutaneously in a specific body region or limb is called lymphedema. Two types of lymphedema exist: primary or hereditary, and secondary or acquired lymphedema. Herein we describe the singular case of a massive lymphedema of lower extremities called elephantiasis. After a complete anamnesis and differential diagnosis, an alcoholic and chronic liver disease as cirrhosis was considered the main etiology of this extreme case of lymphedema. Although lymphedema is one of the symptoms of chronic liver disease, the association between cirrhosis and elephantiasis has not been previously described as described in this clinical case. Through this case report, we discuss and develop all those etiologies of lymphedema, as well as diagnosis and treatment management.

Key Words: Elephantiasis; lymphedema; cirrhosis; liver insufficiency.

Resumen

El cúmulo de líquido linfático a nivel subcutáneo en una determinada región corporal y/o extremidad se denomina linfedema. Existen dos tipos fundamentales de linfedema: primario o hereditario, y secundario o adquirido. A continuación describimos el caso de un linfedema extremo y masivo localizado a nivel de ambas extremidades inferiores denominado elefantiasis. Tras una adecuada anamnesis y diagnóstico diferencial, la presencia de una enfermedad hepática crónica de causa alcohólica tipo cirrosis se determinó como agente causal del mismo. A pesar de que el linfedema es uno de los síntomas de la enfermedad hepática crónica, no se ha descrito con anterioridad la asociación entre cirrosis y elefantiasis como se describe en este caso clínico. A través de este caso clínico, discutimos y desarrollamos todas aquellas posibles etiologías del linfedema, así como su manejo diagnóstico y tratamiento.

Palabras Clave: Elefantiasis; cirrosis; linfedema; insuficiencia hepática.

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Introduction

Lymphedema is the accumulation of subcutaneous lymph fluid in a particular body region or limb. There are two basic types of lymphedema: primary or hereditary lymphedema, and secondary or acquired lymphedema that should be considered in the absence of related familiar history. We describe the case of a massive lymphedema of both lower extremities type elephantiasis secondary to alcoholic chronic liver disease. Throughout this clinical case, we develop in detail the diagnostic process carried out and its treatment.

Case Report

A 60 year-old male patient was admitted to hospital presenting a twelve years long evolution of extreme edema located at both lower extremities (Figure 1).



Figure 1. Bilateral elephantiasis with cutaneous lesions as hyperpigmentation and hyperkeratosis associated.

He had a previous history of hypertension, tobacco consumption and chronic alcohol abuse. The patient worked as a sailor and lived in Africa and Asia several years before. Physical examination revealed ascites and hepatomegaly. Differential diagnosis was established in order to determine the origin of edema (Figure 2). First suspicion was lymphatic filariasis because of his previous epidemiologic history. Serologic tests to detect *Wuchereria bancrofti*, *Onchocerca*, and *Brugia Malayi*, were carried out, and all resulted negative. Serum aminotransferases, gamma-glutamyl transpeptidase, and bilirubin levels were elevated.



Figure 2. Lateral view of right leg showing the massive lymphedema.

Prolonged prothrombin time, hypoalbuminemia, leucopenia and thrombocytopenia were also detected. Normal levels of eosinophils were confirmed in blood tests achieved.

Because of previous history of patient working as a sailor in filaria endemic countries and taking into account that filariasis is the most common cause of secondary lymphedema, we consider lymphatic filariasis as first diagnosis option. However, blood test and serology resulted negatives and therefore revealed signs of liver insufficiency. Previous history of alcohol abuse supported the clinical suspicion of secondary lymphedema to cirrhosis. Liver biopsy confirmed the diagnosis, and cirrhosis was confirmed as cause of elephantiasis.

Alcohol abstinence, fluid restriction, and intensive diuretic therapy were established as treatment.

Main causes of lymphedema are classified in hereditary and acquired etiologies (1). Primary cases are related with lymphatic dysplasias or malformations genetically encoded. Three main categories of primary lymphedema exist often involving both lower extremities. The congenital hereditary lymphedema or Milroy disease (with autosomal dominant transmission), is presented at birth and often associated to other hereditary disorders as Noonan or Turner syndromes. Meige disease is a lymphedema praecox with onset around puberty, and the lymphedema tarda is a similar disease presented in people older than 35 years.

Acquired causes are investigated in the absence of family history. Multiple causes are considered: secondary lymphatic obstruction related with tumours (lymphoma or prostate carcinoma) or iatrogenic cancer treatments such as lymph node dissection and radiation therapy; inflammatory joint diseases (psoriatic arthritis or rheumatoid arthritis); deep vein thrombosis and trauma (2).

However, lymphatic filariasis with parasitic lymphatic obstruction is the most common cause of secondary lymphedema. It is caused by infections of nematodes as *Wuchereria bancrofti*, *Brugia timori* or *Brugia malayi*, which are endemic parasitic in tropical areas of Africa and Asia. Laboratory findings sometimes are nonspecific. Eosinophilia, positive serology test and detection of microfilariae on blood are characteristics of filariasis. Onset of secondary lymphedema is insidious, usually associated with cutaneous lesions as hyperpigmentation and hyperkeratosis. In advanced stages it is often referred as elephantiasis (3-4).

Alcoholic cirrhosis is the consequence of chronic liver disease secondary to alcoholism. It is characterized by replacement of liver tissue by fibrosis and nodules leading to loss of liver function. Physical examination of patient with cirrhosis reveals a number of physical findings as: ascites, gynecomastia, hepatomegaly and portal hypertension signs. Bilateral lower extremity edema is not frequent but also can be present in a small number of patients. Laboratory testing results show aminotransferases and gamma-glutamyl transpeptidase elevation, high levels of bilirubin, hypoalbuminemia, and hematologic abnormalities as thrombocytopenia, leucopenia and prolonged prothrombin time.

Patients with alcoholic cirrhosis commonly have complications at moment of diagnosis. Noninvasive tests for cirrhosis diagnosis have been proposed but final confirmation of alcoholic cirrhosis is achieved by liver biopsy. Management includes alcohol abstinence, nutritional supplementation, sodium and fluid restriction. Intensive diuretic therapy includes spironolactone, furosemide and aldosterone antagonists (5).

Diuretics and hypokalemia avoidance are main principles of treatment. After few weeks of treatment, an improvement of systemic edemas and symptoms can be observed.

Edema is one of symptoms of chronic liver disease. However, in previous medical literature, association between elephantiasis and chronic liver disease has not been reported previously.

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