Rev Med Hosp Gen Méx. 2016;xxx(xx):xxx–xxx

REVISTA MÉDICA DEL
HOSPITAL GENERAL
DE MÉXICO
www.elsevier.es/hgmx

CLINICAL CASE

Early primary lymphoedema treated by en bloc excision: A case report

J.A. García-Espinoza[a,*], **V.B. Aguilar-Aragón**[b], **S. Vásquez-Ciriaco**[c]

[a] *Department of General Surgery, Hospital Regional de Alta Especialidad de Oaxaca, Oaxaca, Mexico*
[b] *Department of Cosmetic and Reconstructive Surgery, Hospital Regional de Alta Especialidad de Oaxaca, Oaxaca, Mexico*
[c] *Department of Surgical Oncology, Hospital Regional de Alta Especialidad de Oaxaca, Oaxaca, Mexico*

Received 6 July 2016; accepted 8 August 2016

KEYWORDS
Lymphoedema;
Filariasis;
Cancer;
ABThera system;
Graft

Abstract
Introduction: Lymphoedema is a pathological condition with a low global incidence where protein-rich fluid accumulates in the interstitium, causing inflammation and degenerative changes in the affected limbs. It is divided into primary and secondary; worldwide the most common causes are cancer or its treatment and filariasis infection. It is diagnosed through the patient's medical history and treatment depends on the severity.
Case report: Twenty-one-year-old male with the condition for 8 years, with a gradual increase in volume in the right lower limb, seen in the emergency department due to functional disability; en bloc excision plus split-thickness grafting was performed, fixed with sutures and ABThera, with 85% integration and functional improvement.
Conclusions: Lymphoedema has a high socio-economic impact. Its treatment involves several specialists and must be multidisciplinary, who then find the cause and propose management according to the stage. Combined medical and surgical therapy is the best choice for advanced stages.

* Corresponding author at: Calle Aldama sin número, paraje el Tule, San Bartolo Coyotepec, CP 71256 Oaxaca, Mexico.
Tel.: +52 951 8080501.
E-mail address: jaime_506@yahoo.com.mx (J.A. García-Espinoza).

http://dx.doi.org/10.1016/j.hgmx.2016.08.007

Please cite this article in press as: García-Espinoza JA, et al. Early primary lymphoedema treated by en bloc excision: A case report. Rev Med Hosp Gen Méx. 2016. http://dx.doi.org/10.1016/j.hgmx.2016.08.007

PALABRAS CLAVE
Linfedema;
Filariasis;
Cáncer;
Sistema Abthera;
Injerto

Linfedema primario precoz, tratado mediante escisión en bloque: reporte de caso

Resumen
Introducción: El linfedema es una condición patológica de baja incidencia mundial, que acumula líquido rico en proteínas en el intersticio, provocando inflamación y cambios degenerativos en las extremidades afectas. Se divide en primario y secundario, la causa global más frecuente es el cáncer o su tratamiento y la infección por filarias. El diagnostico se realiza a través de historia clínica y el tratamiento depende del grado de severidad.
Caso clínico: Masculino de 21 años con padecimiento de 8 años, con aumento progresivo de volumen en extremidad inferior derecha, acude a urgencias por incapacidad funcional; se realizó escisión en bloque más aplicación de injertos de espesor parcial, fijados con suturas y Abthera, con integración de 85% y mejoría funcional.
Conclusiones: El linfedema tiene alto impacto socioeconómico, su tratamiento involucra diversos especialistas, debe ser multidisciplinario, encontrando la causa y según estadio proponer manejo. La terapia médica y quirúrgica en conjunto es la mejor opción para estadios avanzados.

Introduction

Lymphoedema is a pathological condition in which protein-rich fluid accumulates due to a locoregional dysfunction of the lymphatic vessels.[1]

Estimating the precise global incidence of this condition is difficult due to unreported and poorly diagnosed cases. However, it known that cancer and its treatment is the most common cause worldwide, followed by filariasis (*Wuchereria bancrofti*),[2] common in developing countries or those with a tropical climate. It is more common in men than in women, with a 2:1 ratio, and most cases are not associated with genetic conditions.[3] Several risk factors for developing this disease, which precipitate its clinical expression, have been identified.

It is divided into two groups: primary lymphoedema and secondary lymphoedema. Primary lymphoedema is when it is caused by a malformation of the lymph nodes and vessels with no precipitating cause, which in turn is further divided by the age of onset: congenital[4] which has an onset from birth to up to two years of age, shows mutations in the VEGF gene[5] and has a frequency that varies between 77% and 94%; praecox or Meige's syndrome, with an onset at puberty or pregnancy before 35 years of age, shows mutations in the FOXC2 gene,[6] and has a frequency that varies between 6% and 12%; and tarda which has an onset after 35 years of age and an approximate frequency of 11%. All present more commonly in the lower limbs. Secondary lymphoedema results from diseases in which the natural history or their treatment deteriorates the lymph nodes and vessels. In this group the most common cause is breast cancer, sarcoma, melanoma, or malignant gynaecological tumours or their treatment (lymphadenectomy or radiotherapy)[7]; other causes include filariasis which affects 90 million people worldwide, recurrent cellulitis and cutaneous tuberculosis, rheumatoid arthritis, sarcoidosis, obesity (BMI > 30), and severe trauma.[8]

The clinical presentation has an insidious onset in cases of primary lymphoedema with no observable triggering cause, starting with soft, distal oedema with a slow progression over the course of years.[9] It is important in patients with soft, medical treatment – resistant oedema with no apparent cause to take a full medical history including age at onset and duration of disease, area involved, associated symptoms, personal history of chronic disease (diabetes, hypertension, prior history of cancer and its treatment), recent trips, history of genetic diseases, and history of infections.

The oedema progresses distally and starts to present fat and fibroblast infiltration, which cause irreversible skin hardening. Deformity occurs as well as progressive limitations to mobility in the limb which can affect daily activities. In addition, skin changes such as keratosis and papillomatosis occur and, when the degree of severity is higher, recurrent events such as cellulitis, lymphangitis, and skin breakdown start. In advanced stages, lymphoedema complications such as lymphosarcoma, Kaposi's sarcoma, and lymphoma have rarely been reported.[10]

There are different classifications of lymphoedema depending on the clinical characteristics (oedema is reversible by elevating the limbs and circumference). The International Society of Lymphoma classifies lymphoedema into four stages from 0–III according to the skin characteristics and reversibility by elevating the limb (Table 1)[11]. This system is currently the most widely used.

In most cases, a lymphoedema diagnosis is established based on the medical history and physical examination (Stemmer's sign is characteristic; it consists of the inability to form a skin fold after pinching near the first phalangeal joint). However, in some patients in whom the diagnosis is in question, it becomes necessary to use additional tests.[12] A Doppler ultrasound should be performed on the limb for diagnostic certainty to check that the deep venous system is working properly. The gold standard for diagnosis in the

Please cite this article in press as: García-Espinoza JA, et al. Early primary lymphoedema treated by en bloc excision: A case report. Rev Med Hosp Gen Méx. 2016. http://dx.doi.org/10.1016/j.hgmx.2016.08.007