Bilateral Lower Limb and Abdominal Elephantiasis Due to Erysipelas

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To the Editor: Elephantiasis results from chronic lymphedema and is characterized by gross enlargement of the arms, legs, or genitalia. It occurs due to various obstructive diseases of the lymphatic system. The most common form of lymphedema is secondary lymphedema involving resection or ablation of the regional lymph nodes by surgery, radiation, tumor invasion, direct trauma, or an infection.[1,2] We present an unusual case of bilateral lower limb and abdominal elephantiasis due to extensive lymph node destruction by erysipelas.

A 31-year-old man presented with progressively increasing bilateral lower limb and abdominal swelling, with changes of skin over 1 year. He also described a history of repeated outbreaks of erysipelas in the right leg 5 years prior to this presentation, for which he had multiple hospital admissions and received treatment with intravenous antibiotics, whose names he could not recollect.

Physical examination revealed that he was obese, weighing about 244 kg, with a body mass index of 75.3 kg/m^2. The patient had giant-sized bilateral lower limbs and abdominal swelling, with a chronic disseminated dermatosis of the skin, characterized by edema, hyperpigmentation, hyperkeratosis, and elephantiasis nostras verrucosa (ENV) [Figure 1a and 1b]. His blood lipid parameters and serum cortisol were normal, without microfilaria.

The patient underwent vascular ultrasound examination of lower limbs, which showed swollen lymph nodes in bilateral inguinal region [Figure 1c and 1d]. A histopathological examination of the specimen showed lymph node changes in the left inguinal region. The features were suggestive of nonspecific inflammation of lymph node, cortical atrophy, lymphatic sinus dilation, and interstitial vascular proliferation with dilation [Figure 1e and 1f]. However, there was no clear evidence of erysipelas, malignancy, filariasis, or donovanosis in the specimens.

The term "elephantiasis" describes an elephant-like appearance or overt enlargement of the legs, arms, or vulva.[3] Lymphedema manifests as soft-pitting edema in the affected tissues that results in a local inflammatory response, which finally leads to nonpitting edema. The affected tissues sustain further injury as a result of the local inflammatory response and recurrent infections. The common mechanism is an underlying lymphatic obstruction leading to impaired lymphatic drainage with abnormal accumulation of interstitial fluid and subsequent development of lymphedema. This eventually results in excessive subcutaneous fibrosis and scarring, with associated severe skin changes characteristic of lymphostatic elephantiasis.[2]

ENV is a rare clinical condition associated with chronic nonfilarial lymphedema caused by bacterial or noninfectious lymphatic obstruction. Mossy papules, plaques, and cobblestone-like nodules are clinical features of ENV.[4] The patient's history and characteristic skin changes are typically sufficient to diagnose ENV. Our patient with bilateral lower limb elephantiasis and abdominal ENV is a very rare case. The absence of erysipelas histology from the lymph node ruled out direct infiltration. Moreover, the patient had a past history of repeated erysipelas outbreaks. The etiology in our case was extensive destruction of the inguinal lymph nodes and their channels due to past erysipelas outbreaks, leading to a blockage of lymphatic drainage, resulting in lower limb elephantiasis and abdominal ENV.

Many factors, including surgery, radiation, or an infection, may lead to chronic lymphatic obstruction and stasis. Filariasis, caused by infestation of the lymph nodes by the parasite *Wuchereria bancrofti*, is the most common and global cause of secondary lymphedema.[2] The precise role of some pathogens in lymphatic obstruction is uncertain.

Treatment options of elephantiasis include use of elastic bandages, pneumatic stockings, mechanical massage, oral

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Access this article online

Quick Response Code:

Website: www.cmj.org

DOI: 10.4103/0366-6999.228244

Received: 22-12-2017 **Edited by:** Li-Shao Guo
How to cite this article: Yang YP, Huang WX, Zhong WX, Fu YM, He PA, Zhao G, Feng QM. Bilateral Lower Limb and Abdominal Elephantiasis Due to Erysipelas. Chin Med J 2018;131:873-4.

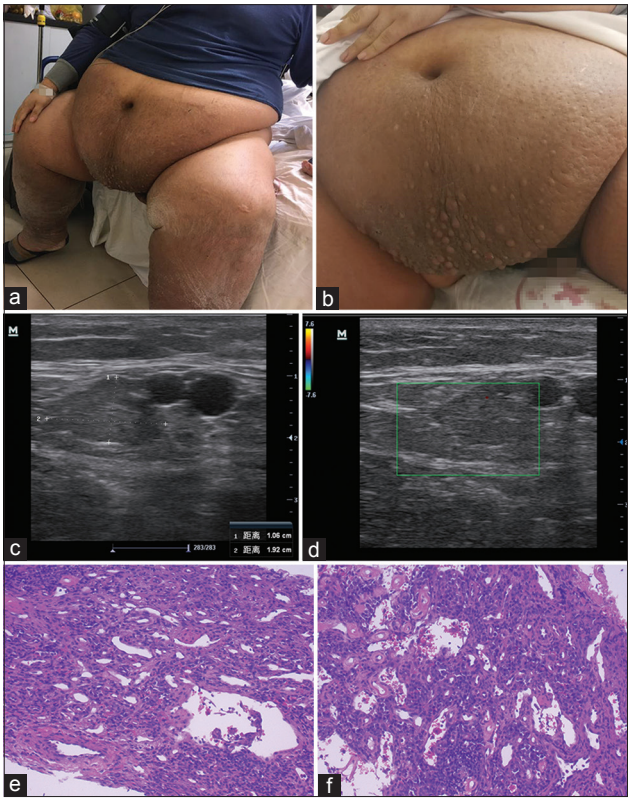

Figure 1: Chronic lymphedema in lower extremities and abdomen (a), and hyperpigmentation, hyperkeratosis, and a verrucous aspect involving the abdomen (b). Vascular ultrasound examination of the lower limb shows the sizes (1.06 cm × 1.92 cm) (c) and no significant blood flow (d) in the swollen lymph node in the left groin. Histopathological examination of the specimen of swollen lymph node in the left groin. The features were suggestive of nonspecific inflammation of lymph node cortical atrophy, lymphatic sinus dilation, and interstitial vascular proliferation with dilation (e and f) (H and E staining, original magnification ×200).

retinoids, and surgery. Reconstructive surgery is often considered to be the only treatment for serious penoscrotal elephantiasis.[5] The goal of the therapy is to re-establish function and reduce physical disability.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

This work was supported by a grant from the Foundation of Shanghai Jiao Tong University Affiliated Sixth People's Hospital (No. ynlc201709).

Conflicts of interest

There are no conflicts of interest.

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