

# Esthiomene: An unusual presentation of elephantiasis

Anand Pai, Umadevi V, Narayanasamy S

## To the Editors

Genital elephantiasis is an important medical problem in the tropics as it is associated with physical disability and extreme mental anguish especially in females. Genital elephantiasis is a very rare condition and due to prevalence of sexually transmitted infections (STIs) it is even rarer. Elephantiasis and chronic genital ulceration in women is called 'Esthiomene' [1].

A 40-year-old married female, mason by occupation, presented with swelling of the vulva and genital ulcer for the past five years. She complained of difficulty in micturition and difficulty in walking. She also had history of painful swellings in the inguinal region, on both sides, for the past seven years. There was no history of fever, cough with expectoration or weight loss. Her menstrual cycles were regular.

She had two healthy children. She gave a history of her husband's extramarital contacts. On genital examination two large nodular swellings of size 15×7×5 cm were present involving both the labia majora (Figure 1). Multiple nodules were present on the mons pubis. Multiple superficial and deep ulcers of varying size were present on the inner aspect of both labia majora.

Multiple, firm, non-tender, matted lymph nodes involving both the horizontal and vertical groups of inguinal lymph nodes were found bilaterally. Groove sign was present. Per rectal examination was normal. Systemic examination was normal. Mantoux test was negative. X-ray chest and ultrasound abdomen were



Figure 1: Vulval elephantiasis with chronic genital ulcerations

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normal. Hemogram, LFT and renal parameters were normal. Blood VDRL was non reactive. Serology for HIV antibodies and filariasis was found to be negative. Smear for malarial parasite and microfilaria was negative. Tissue smear for Donovan bodies was negative. Albumin/Globulin ratio was reversed (0.6). Tissue biopsy of the swelling was taken and histopathological study revealed lymphangiectasia (Figure 2). Culture of discharge in McCoy's medium showed growth of lymphogranuloma venereum strains. Polymerase chain reaction study showed features specific for Chlamydia trachomatis L1-L3 serovars which confirmed the diagnosis. The patient was started on doxycycline 100 mg twice daily for 15 days and was

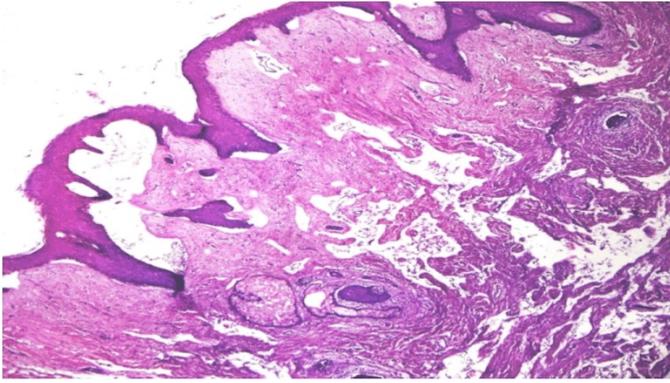


Figure 2: Histopathology of the vulval swelling showing lymphangiectatic changes (H&E, x400).

referred to plastic surgery. As no major structures were involved excision of the swelling was done. Postoperatively patient was followed up and had no further complaints.

Among sexually transmitted infections, lymphogranuloma venereum and donovanosis are the most common, others being syphilis and infection with non-LGV strains of *Chlamydia trachomatis*. *Chlamydia trachomatis* serovars L1-L3 are the causative agent of LGV and initiates the disease process primarily in the lymph channels leading to thrombolympangitis and perilympangitis [2]. Extension of inflammatory process to draining lymph nodes causes periadenitis, matting of lymph nodes, formation of abscesses, fistulae and sinuses. Healing takes place by fibrosis. A combination of chronic oedema, sclerosing fibrosis and active lymphogranulomatous infiltration in the subcutaneous tissue results in the massive enlargement of genitalia. These late complications occur in the tertiary stage of LGV and are frequent in women. The diagnosis of LGV associated elephantiasis is supported by high titre serology and identification of the organism in the pus or bubo fluid by cytology or culture [3]. *Chlamydia trachomatis* is an obligatory intracellular organism and diagnosis is usually made by tissue culture on McCoy's medium, direct immunofluorescence and polymerase chain reaction. Microimmunofluorescence test is the only serological means of distinguishing LGV strains of *Chlamydia trachomatis* from other serovars. Polymerase chain reaction amplification and sequence analysis of the omp 1 gene is also useful in identification of L1- L3 serovars [4]. History of exposure to infection, a small transitory primary lesion, followed by chronic inguinal adenitis resulting in typical suppuration and fistulation is characteristic of LGV. With the exclusion of tuberculosis, filariasis, Hodgkin's disease, gonorrhoea, syphilis, malignant disease and chancroid, the diagnosis of the condition becomes at once easy.

Treatment modalities of genital elephantiasis due to STIs require an interdisciplinary approach involving genitourinary medicine physicians, urologists, and dermatologists. The objectives of treatment are to reduce swelling, restore shape and normal sexual

function, and prevent inflammatory episodes. Medical therapy should be the first line of treatment and any surgical intervention should be undertaken only under the cover of appropriate antibiotics. In case of genital elephantiasis caused by LGV, doxycycline 100 mg twice daily should be given for prolonged periods (improvement reported with up to 13 month therapy). Surgery is the only effective option for a select group of patients in whom the disorder is disabling and persistent. Reduction procedures include labial reduction which is easily achieved by wide elliptical excision with a single suture line [5, 6]. Recurring swelling in a minority of patients will be benefited by another similar procedure. Larger labial defects may be covered with myocutaneous, floating island or fasciocutaneous flaps [7]. However, there are no studies available demonstrating the long-term efficacy of reduction procedures in STI-related genital elephantiasis. Due to the manifold aspects of the disease and of the possibility of the lesions remaining insignificant for a long time, every case of genital elephantiasis or rectal stricture needs to be subjected to a critical investigation in regard to the possibility of this specific infection. Hence we found it worthwhile to report this case.

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### Author Contributions

Anand Pai – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Umadevi V – Conception and design, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published

Narayanasamy S – Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published

### Guarantor

The corresponding author is the guarantor of submission.

### Conflict of Interest

Authors declare no conflict of interest.

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