

## Giant Vulvar Elephantiasis of Filarial Origin: A Rare Case Report

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**Abstract:** Genital lymphedema is rare outside the endemic filariasis regions. Elephantiasis is the manifestation of chronic lymphedema. Involvement of limbs, trunk and male genitalia is common but elephantiasis of vulvar origin is rare even in endemic areas. we report a case of giant vulvar elephantiasis in a young female. A 21 year old unmarried female presented with swelling of bilateral labia since 7 years with dragging pain and discomfort. Patient was managed by excision of filarial tumor with vulvoplasty and discharged uneventfully.

**Keywords:** Elephantiasis, Lymphedema, Vulval, Wuchereria bancrofti

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### I. Introduction

Genital elephantiasis is caused by a variety of infective and non infective causes leading to blockage of lymphatics. Vulvar elephantiasis is rare and it accounts of only 1-2% of total elephantiasis (1). Greek terminology Esthioneme used to describe elephantiasis which means to eat and carries an idea something gnawed, eroded or ulcerated (2). Etiology of elephantiasis may be filarial, tubercular, chlamydial infection, post vulvectomy, post radiotherapy, after inguinal and pelvic lymph node dissection or idiopathic (3,4). Commonest infective Filariasis results from infection with Wuchereria bancrofti and Brugia malayi. Wuchereria bancrofti accounts for 90% of the case of human lymphatic filariasis. Nonlymphatic filariasis is caused by onchocera volvulus, loa loa or mansonella perstans. Lymphedema is due to accumulation of protein rich interstitial fluids, leads to proliferation of fibroblasts and mast cell, edema fluid organised progressively and fibrosis of the subcutaneous tissue giving rise to irreversible, firm and non pitting swelling. Hyperkeratosis, rugosities, verrucous and condylomatous changes are features of long-standing lymph stasis and are collectively termed 'elephantiasis'(5)

### II. Case Report

A 21 year old unmarried girl presented in department of general surgery of Rajendra institute of medical sciences, Ranchi, India with mild grade fever, dragging pain and swelling in genital area with difficulty in walking and working normally since 7 years. Patient was also in significant psychological distress.

There was no history of trauma, previous surgeries, radiotherapy, or chyluria. No history of swelling since childhood or lymphedema in family. All general and systemic examinations were within normal limit.

On laboratories examinations Hb-10.5gm/dl, leukocyte count 10,400 cells/mm<sup>3</sup>, neutrophilia of 72%, lymphocyte 17%, eosinophil 08%, ESR 50mm in first hour was found. Patient was hepatitis B positive (HBsAg) with titer <6 IU/ml.

On local examination bilateral vulval swelling was found [fig. 1]. Size of swellings were 21x14 cm on left side and 7x4 cm on right side with illdefined margin, irregular bosselated surface, firm in consistency, freely mobile. There was no ulceration or lymphadenopathy.

Cinical diagnosis of vulvar filariasis was made and total excision of tumor mass done under spinal anesthesia [fig. 2]. Two large irregular mass of weighs about 6 kg and 2 kg was excised [fig. 3].

Histopathological reports showed proliferation of stratified squamous epithelium showing acanthosis and increased junctional activity with interlacing band of fibroblasts and collagen and perivascular mixed inflammatory infiltrate [fig. 4]. ZN and PAS staining was negative. Filarial antigen assay was positive. Pelvic ultrasonography was unremarkable.

### **III. Discussion**

The term elephantiasis was first described by Celsius (30BC-50AD) (6). Lymphatic filariasis of external genitalia was first described in 1673. Vulval lymphedema can be a rare extraintestinal manifestation of Crohn's disease (7,8). Differential diagnosis includes genital warts, lymphogranuloma, Lymphangioma circumscriptum, and fibro epithelial polyp, fungal infection, and carcinoma and angiomyxoma (9,10).

Diagnosis of elephantiasis is mainly clinical and supported by histopathological examination. Neither microfilarae in blood nor adult worm in tissue can Found all the times (not sensitive).

Treatment of filarial lymphedema is Diethylcarbamazine (DEC) in three divided doses of 6 mg/kg/day for 21 days kills adults and microfilarae. Leg elevation , elastic stocking , decongestive physiotherapy and maintenance of good skin care is also recommended. Surgical treatments includes excision of skin and subcutaneous tissue of involved part and primary closure of defect with normal adjacent skin. Partial thickness graft or rotation flap can be used if raw area is large.

Surgical treatment is used only in extreme cases in order to reduce the weight of the affected organ, to help minimize the frequency of inflammatory attacks, to improve cosmesis, and to potentially reduce the risk of secondary angiosarcoma.(11,12).

In our case patient came with large vulvar swelling with difficulty in walk and work normally, after excision of mass we improved her life style and also provide good cosmosis which decreases her anxiety about future marital status. Rare presentation of our case evoks us to report the case.

### **IV. Figure**



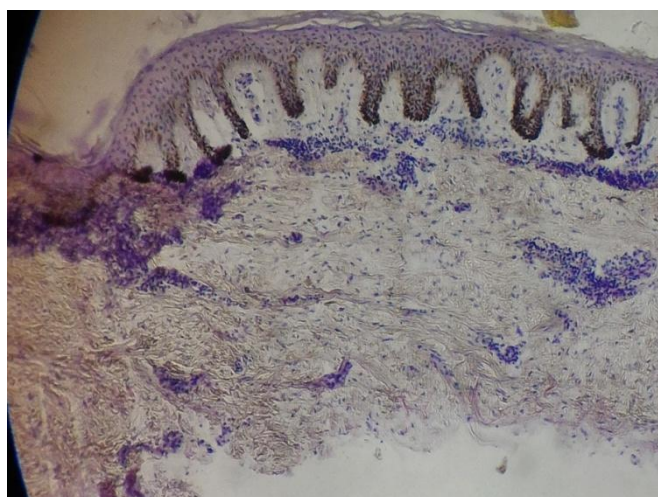
**Figure 1 . Bilateral vulvar elephantiasis**



**Figure 2. –Surgical excision of filarial tumor**



**Figure 3- Specimen obtained after excision**



**Figure 4. – Histopathological report showing acanthosis and increased junctional activity with Interlacing band of fibroblasts and collagen and perivascular mixed inflammatory infiltrate.**

## V. Conclusion

Although genital elephantiasis is rare in developed countries, it is still a challenge for tropics and subtropics. Genital elephantiasis significantly affect physical, social as well as mental status of the patient. So early detection and treatment of lymphedema is only option to prevent it.

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