

# Vulval Filariasis - A Case Report

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**Abstract:** The term Elephantiasis was first described by Celsius (30 BC-50 AD). Lymphoedema is accumulation of lymph in the soft tissue due to obstruction of lymphatics which results in accumulation of protein rich interstitial fluid caused by low output failure of lymph. (1) This leads to proliferation of fibroblasts and mast cells, organization of oedema fluid and sclerosing fibrosis of subcutaneous tissue giving rise to firm, non pitting and irreversible swelling, hyperkeratosis, verrucous and condylomatous changes are feature of long standing lymph stasis collectively called as Elephantiasis. (2) Lymphoedema can be primary or secondary when accumulation of lymph results as a result of congenital and or inherited condition, it is called as primary lymph oedema and if it is caused with pathological condition, it is called as secondary lymphoedema. Commonest infective filariasis results with Wuchereria Bancrofti and Brugia Malayi. Filariasis caused by Wuchereria Bancrofti and Brugia Malayi is secondary cause of lymphedema. 90% of cases in humans is caused by Wuchereria Bancrofti. The condition may be subclinical or latent condition where the swelling is not evident despite impaired lymphatic transport to a debilitating condition with tropic skin changes such as fat deposits acanthosis and warty outgrowths, characteristic of Lymphatic Elephantiasis. (3 & 4) Vulval Elephantiasis is extremely rare and accounts with 1-2 % of total cases of genital Elephantiasis (5). Greek terminology esthioneme is used to describe elephantiasis which means to eat and carries an idea as something is gnawed, eroded or ulcerated (6). Causes of vulval lymphedema may be filarial, tubercular, chlamydial infection, postvulvectomy, postradiotherapy, after inguinal and pelvic lymph node dissection or idiopathic. (7 & 8). Non lymphatic filariasis may be caused by oncocerca volvulus, loa loa and mansonella perstans. Though rare, Genital elephantiasis when present is associated with significant physical disability and mental depression (9 & 10). It is reported as a long term complication of tubercular lymphadenitis, filariasis and sexually transmitted infection in case reports from around the world. In majority of these reports, it involved bilateral labia. (11 & 12). From Ethiopia on the other hand, there is no case report of genital elephantiasis in female. Moreover, the national lymphatic filarial management and disability prevention guidelines discloses only about cases affecting male genitalia (13). In contrast we present a neglected case of bilateral vulval elephantiasis in a 35 year old lady.

## 1. Case Presentation

The patient is a 35 year old unmarried lady from a very remote area Korchi of Gadchiroli district of Maharashtra. Patient first noticed swelling 20 years back where she first assumed to being a boil, The swelling gradually increased in size. However she neglected it for 20 years. Our Patient is unmarried due to swelling as told by her relatives. She walked with unusual gait and experienced difficulty in walking, standing upright for a long time. She chose to sit all the time with a thin towel covered over the swelling, she attained menopause two years back. She was first noticed by a very vigilant doctor in Korchi who was on house to house survey and who noticed an unusual swelling and sent her to district hospital Gadchiroli. In District hospital Gadchiroli patient was observed by a team of Physicians, Gynecologists and Surgeon. General Examination revealed patient entered the examination room with wide waddling gait. She had pallor. All her vitals were normal. There was no palpable Lymphadenopathy. Systemic Examination was also normal. On Local examination we noticed a well-defined bosselated masses, on left side measured 35 cm x 30cm and on right side measured 30 x 28 cm arising from labia majora. The swelling was pendulous and bosselated and hanging down obstructing two third of her thigh. With overlying skin being hard thickened and appearing to have extensive rugosities and hyperpigmentation with 3-4 pus discharging sinuses. Mass was non pitting and had no tenderness. Both masses obstructed the vulval cleft.

## 2. Investigation

Hemoglobin -8.2, Total leucocyte -6500 Differential leucocyte was normal with slightly elevated eosinophil count. Platelets-2.7 lakhs/cumm T3-0.87 T4-8.27 TSH-3.68,

LFT-total-1.10, Direct-0.22. Urea-3.8 Creatinine-0.98, sickling test-negative

Ultrasonography of Pelvis showed uterus 6.2x3.6x4.6, both ovaries normal in size shape and echotexture endometrial thickness was normal.

2 blood transfusions were given preoperatively to optimize her Hemoglobin. She was started on Antibiotics cefotaxime and Gentamycin and metronidazole and daily dressing was done of the pus discharging sinuses.

Catheterization was done with great difficulty and had to be done by patient in supine position with widely extended at Hip Joint and flexed at knee joint. Anesthesia could not be given in left lateral position as knee chest position could not be given and so was given with patient in sitting position. 3.2 ml of Bupivacaine of concentration 0.5% was given in L2-L3 space in First attempt.

Operative Notes: skin marking was done. Skin incision was taken by scalpel and further dissection was done with cautery. Further dissection was done by dividing and ligating larger vascular pedicles since the area is highly vascular. Division was done in between the ligated pedicles. Entire mass was removed in Toto in single setting. Left first followed by right. Vulvoplasty was done and restoration of near normal Labia was done. Operation Lasted for 4 ½ hours. No additional sedation or Mephentine was required. Excision of mass results in better quality of life with improved cosmesis.



Pre Op



Post OP

### 3. Discussion

Diagnosis of filariasis is mainly clinical and supported by histopathological examination. Neither microfilariae in blood nor adult worm in tissues can be found in blood nor can adult worm in tissue be found at all times.

Treatment of filariasis is Diethylcarbamazepine. (DEC) in three divided doses of 6 mg/kg/day for 21 days kills adult as well as microfilariae. Supportive therapy for lymph edema includes leg elevation, elastic stockings, decongestive physiotherapy and maintenance of good skin care. Surgical Treatment is used only in extreme cases in order to reduce

the weight of the affected organ and help minimize frequency of inflammation attacks and improve cosmesis and potentially reduce the risk of secondary angiosarcoma.

#### 3.1 Differential Diagnosis

Differential Diagnosis of vulvar elephantiasis include genital warts, lymphogranuloma venereum, lymphangioma circumscriptum, fibroepithelial polyp, fungal infection and carcinoma and angiomyxoma(14,15 )Vulval oedema can be a rare extraintestinal manifestation of Chrohns disease.(13)

In our patient, malignancies, post radical hysterectomy lymphadenopathy and radiation therapy are unlikely causes from history itself. Vulval elephantiasis following tubercular lymphadenitis can be confirmed by past history of Tuberculosis. In Addition, evidences of puckered scars of healed sinuses in the inguinal regions strengthens our suspicion. In Our patient though there is no treatment history and microfilariae were not detected in midnight sample, the clinical presentation 20 years back, epidemiology, the physical findings all suggest the possibility of vulval filariasis and histopathological examination confirmed our diagnosis. The other competing differential diagnosis lymphogranuloma venereum, can also cause vulval elephantiasis as a late complication due to resulting fibrosis in the inguinal area. The major risk factor for this condition, primarily seen in men who have sexual contact with HIV positive people. Even in the best setups absence of clinical presentation, low yield of cultures and low specificity of serological assessment makes it difficult to diagnose. The fact that our patient has no sexual exposure makes this possibility less likely. Other possibility of rare extraintestinal manifestation of chrohns can be ruled out as there is an absence of gastrointestinal symptoms in our patients.

### 3.2 Other case reports

- Case report by Rachana Chaudhari, Anu Maheshwari and Kshipra Nigam – 2 large masses 48x40x10 cm and another mass of 26x25x7 cm in 40 year female. The mass observed over 3-5 years.
- Case report by Satish Kumar Ranjan, Mini Sharma, Neval K. Jha – bilateral swelling of 21x40 cm on left 7x4 on right masses over 7 years.
- Case report by Wondimagengnehu, Woldeys and Dejene Asefa - 55 years old lady from Eithiopia had an unilateral mass over left labia which grew over 2 years - 45x25 cm on the left labia

In comparison to all the three case reports available till date, our case involves longest time period of 20 years and are the largest bilateral masses. Hence we conclude this case to be the rarest case of neglected vulval filariasis.

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