

Vulvar Filarial Elephantiasis in A Tanzanian Woman; Rare Presentation of Lymphatic Filariasis: A Case Report and A Review of Literature

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ABSTRACT:

Lymphatic filariasis is frequently caused by *Wuchereria bancrofti* a widely distributed filarial worm throughout tropical regions of Asia and Africa. This worm is particularly prevalent in wet and humid areas. The common clinical manifestations of Bancroftian filariasis are acute adenolymphangitis, hydrocele, lymphedema and elephantiasis. Of these, elephantiasis appears to be most common in the legs. Pathology in the arms, scrotum and penis are also common. On the other hand elephantiasis of the vulva and the female breast is extremely rare and its occurrence deserves a mention in medical literature. Presented here is a 21 years old female who presented with progressive unilateral vulvar swelling over a period of two years. Microfilariae were found in peripheral blood film. The diagnosis of vulvar filarial elephantiasis was reached. The patient underwent reconstructive surgery and planned to be initiated on Diethylcarbamazine Citrate (DEC) which is a drug of choice in cases of filarial infestations.

Keywords: Elephantiasis, Filariasis, Vulvar.

Elephantiasis is a relatively rare disorder resulting from blockage of lymphatic channels of the affected part of the body. The disorder is usually characterized by gross enlargement of the part of the body that has been involved¹. The commonest sites of involvement are the lower limbs and the scrotum in the male and occasionally in other sites such as the trunk, breasts, upper limbs and has been found to be exceedingly rare in the female external genitalia according to one study done in Tanzania². Genital elephantiasis in males is an important medical problem in the tropics. It can be the result of both infectious and non-infectious diseases. The majority of cases are due to filariasis. Other causes include bacterial sexually transmitted infections (STI's), especially lymphogranulomavenereum (LGV) and donovanosis³; tuberculosis¹, haematological malignancies, dermatological diseases and in some cases from unknown causes⁴. If untreated, lymphatic filariasis is a major cause of debilitating and disfiguring chronic

disease manifestation especially if it happens in the form of lymphedema, elephantiasis and hydroceles⁵.

Filariasis is endemic in both tropics and subtropics. *Wuchereria bancrofti*, the commonest cause of filarial infections, is estimated to infect approximately 120 million people in the tropics one third of whom in Africa⁶. Despite these high figures, filarial elephantiasis of the vulva is extremely rare. It is estimated to occur in only approximately 1-2% of all cases of filarial elephantiasis encountered in clinical practice¹.

Here we present a case report of vulvar filarial elephantiasis in a 21 years old female who was referred to our hospital with a 2 year long history of progressive swelling in the right labia majora. The diagnosis was confirmed by finding microfilariae in peripheral blood film.

Case Report

A 21 years old Para 1 woman was referred to our Hospital from a District Hospital about 130km away with a longstanding history of a painless progressive right sided vulvar swelling. The patient gave a history that the swelling began about two years prior to

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admission as a small painless swelling on the right labia majora and had been progressively increasing in size over time. Initially it was not causing any noticeable problem but as time went on, she started to experience heaviness in the vulva associated with difficult coitus. With time passing she further had difficulty walking due to the size and location of the swelling. Prior and during the course of the illness there had been no history of fever or generalized body malaise. She denied to have had travelled to known filarial endemic areas in the country though she, her husband and close relatives are mobile pastoralists. They had moved from western Tanzania over ten years ago and currently residing in Central Tanzania as pastoralists. As well, no history of trauma was reported prior to the current illness. Her menses were reported to be normal and of regular pattern despite the vulvar swelling.

On general examination she was found to be healthy looking, afebrile and no lymphadenopathy was appreciated in all lymph node regions. Local examination revealed a huge soft-firm mass on the right labia majora (See Figure (1) for clarity). The consistency of the swelling differed at different points of the mass. The contralateral labia looked normal in size and consistence. Inguinal lymph nodes were not palpable.



Figure (1): Picture of The vulvar mass

There was no lower or upper limb swelling. Our initial impression was filarial vulvar elephantiasis pending exclusion of a fibroma and kaposi's sarcoma.

Mid-night peripheral blood smear for filarial worms was positive for *Wuchereria bancrofti*. Fine needle aspiration of the mass was inconclusive and a decision to proceed with wide excision was reached (Basically cosmetic reconstructive surgery). The patient was counseled and consented for the procedure. The procedure was done successfully under spinal anesthesia and a wide excision was done to remove the mass and hemostasis was achieved. (See figure (2) for clarity). The excised mass was sent for histological diagnosis.



Figure (2): Vulvar appearance immediately post operatively

Histology Report:

No filarial worms were seen on histology and the pathologist reported chronic inflammation with fibrosis, no evidence of neoplasia. Post operatively the wound had healed uneventfully and we made arrangement to secure DEC for treatment of filarial worm as these drugs are not readily available in our hospital pharmacy because filariasis are not endemic in the central region of Tanzania. The patient will start medication in the subsequent follow-up clinics when the drugs become available.

DISCUSSION:

Genital elephantiasis is defined as incongruous enlargement of the genitals secondary to lymphatic obstruction by a number of causes infective and non-infective.

Globally the most common cause of elephantiasis is Filariasis. Filariasis can present as asymptomatic/subclinical microfilaraemia, acute disease characterized by lymphadenitis or lymphangitis, or chronically as elephantiasis¹.

The vector for transmission of filarial parasitic worm is a mosquito (Anopheles, Culex, Aedes and Mansoni species) and a couple of filarial worms are known to infect human beings after a long life cycle in the vector; they include *Wuchereria bancrofti*, *Brugiamalayi* and *Brugiatimori* though the commonest filarial infection is that due to *Wuchereria bancrofti* accounting for well over 90% of all cases⁶.

Men are more susceptible to the chronic sequelae of elephantiasis as compared to women. In descending order, elephantiasis usually manifests as hydrocele, followed by elephantiasis of the entire lower limb, the scrotum, the entire arm, the vulva, and the breast².

Cases of lymphatic filariasis of the external genitalia are generally rare⁷. Likewise, Vulvar elephantiasis is extremely rare as compared to hydrocele in males, known to contribute approximately 1-2% of all cases of filarial elephantiasis¹.

In the process of finding the diagnosis, exclusion of other possible causes of vulvar elephantiasis is particularly important because the presentation may resemble other known pathological conditions. Other possible causes of vulvar elephantiasis can be non-filarial infectious or non-infectious at all and they include; lymphogranuloma venereum, donovanosis, tuberculosis and malignancies⁷. Other secondary causes include post-radical hysterectomy, lymphadenectomy and radiation therapy. Fibro epithelial polyp, fungal infection and granuloma inguinale all make important differentials for this condition¹. The presented case represents one of the uncommon manifestation of filarial elephantiasis of the vulva and warrants publication to expand our medical understand of these rare occurrences.

The adult filarial worms live in human lymphatic system causing obstruction of

lymphatic drainage and hence swelling of the involved body part and are believed to be the cause of lymphatic filariasis though it is not clear as to whether the resulting swelling is due to the worm itself or the result of the immune response towards the parasite⁸⁻¹⁰. Absence of microfilariae in the peripheral blood in symptomatic patients is a common finding in clinical practice^{11,12} thus it is generally concluded that microfilaremia in the peripheral blood does not exclude filarial disease nor its presence denote the diagnosis. In the present case however we were able to demonstrate microfilaremia in the peripheral blood film which gave us a high degree of certainty of the presence of the disease process.

As well, very rarely adult worms are recovered from excised tissue samples which make the laboratory diagnosis even more challenging in some cases and therefore most of the time the diagnosis of vulvar filariasis is made on clinical bases. Surprisingly microfilaria occurs frequently in hydrocele fluids and may occasionally be seen in urine or other body fluids.

The mosquito vectors transmit larvae that develop into adult worms in the human body. While in the human host, the adult worms usually release microfilaria into the blood stream usually in large numbers in the early part of night. Based on this, detection of microfilaria in a night blood sample through a variety of techniques becomes a convenient tool for diagnosis as it was with our case. However, as pointed out earlier, demonstrating microfilaria in the peripheral blood has been shown to be increasingly difficult in the late stage of the disease.

Ultrasound has been shown to be useful in detection of bancroftian live adult worms or their characteristic movements particularly in lymphatic vessels of the scrotal area in men ("filarial dance")¹³⁻¹⁶. But they are more dispersed and more difficult to detect within the lymphatics system in infected females,¹⁷ which possibly explains why negative findings were encountered in our case.

Regardless of the cause, the process towards development of elephantiasis/lymphedema

remains the same. This involves permanent obstruction of lymphatic channels lymphatic stasis stimulation and activation of fibroblasts destruction of lymphnodes lymphedema and elephantiasis¹⁸. Globally filariasis remains the most common cause of secondary lymphedema¹⁹.

Management of vulvar filarial elephantiasis usually requires a multidisciplinary approach as surgical, parasitological and medical specialties may be called into place to manage patients depending on the stage of the disease. Treatment goals include swelling reduction, prevention of inflammation and restoration of the normal shape and sexual function.

Medically, diethylcarbamazinecitrate (DEC) is given in three divided doses to kill adult worms and microfilaria. Ideally a course of DEC is recommended before any surgical procedure though in this case surgical procedure was done prior to initiation of the drug because DEC is not readily available in this non-endemic region of the country. The death of the worms following DEC however provokes both local and systemic reactions which do not usually necessitate withdrawal of treatment. Medical therapy using DEC alone does not produce regression of elephantiasis⁷. Another drug, Ivermectin is only effective against microfilaria and has been used in the control programs.

Surgically, skin and subcutaneous tissue of the involved part has to be excised and the defect being closed with normal skin from an adjacent area. There is no data on the benefit of surgery for the case of vulvar elephantiasis though with massive swelling like our case reconstruction surgery may be of benefit. In males on the other hand, surgery for scrotal elephantiasis has been shown to produce satisfactory results than that of the legs²⁰. Surgery for hydrocele has been found to bring about improvements in physical and social wellbeing²¹.

This case report is important because genital elephantiasis is rare, and vulvar elephantiasis is even rarer. On top of that demonstration of microfilaria in peripheral blood to confirm filariasis as the cause makes it more interesting because most of the previous case

reports have used serological tests to suspect vulvar filarial elephantiasis.

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Ethical Issues:

Informed consent was obtained from the patient to take photographs and publish her case. The permission to publish patient information was obtained from Hospital Management Team. A copy of the consent form is available for the editor of this journal to view.

Conflict of interest:

The authors declare no conflict of interest

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