

## Research Article

# Contrasting Lymphatic Filariasis with Kaposi Sarcoma in a Known HIV Disease Patient: A Clinical and Histopathological Presentation and Implication to Care

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## Abstract:

Lymphatic filariasis is a systemic disease arising from infestation with filarial worms. The species that commonly infest the lymphatic vessels include *Wuchereria bancrofti* and *Brugia malayi*. The larval stages of the worms are transmitted by biting mosquitoes or flies. *Wuchereria bancrofti* is transmitted by night biting anopheline mosquitoes. The infection is common in tropical Africa. The adult worms infiltrate the lymphatics and produce large numbers of microfilaria into the circulation. The species *B. malayi* is inoculated by *Mansonia* or *Anopheles* mosquitoes and is known to cause less severe disease. Infection with *W. bancrofti* may present diversely from subclinical infection to overt manifestation such as elephantiasis. Acute infection may present with febrile illness, pain or tenderness in the area of the inflamed lymphatics. The chronic phase is usually characterized by presence of lymphadenopathy in lower limbs, retroperitoneal tissues, lymphedema, hydrocele and elephantiasis. The clinical presentation of lymphatic filariasis can mimic that of Kaposi Sarcoma, a malignancy common in advanced HIV disease and may present a diagnostic dilemma especially in the absence of histopathologic findings. We report a case of a 73 year old known HIV disease patient presenting with chronic indurating leg swelling. Histopathology report revealed papillomatus and hyperkeratotic skin lesions with no evidence of malignancy and no presence of microfilaria on a blood smear.

**Keywords:** Lymphatic filariasis, Kaposi sarcoma, HIV, clinical, histopathology

## Introduction

Lymphatic filariasis (Lf) is a tropical disease caused by infection with filarial nematodes especially of the species *Wuchereria Bancrofti* and *Brugia Malayi*. The slender thread-like filarial worms have an affinity towards the skin and subcutaneous tissue or lymphatic system. Humans are the definitive host for the infections while mosquito acts as an intermediate host [1, 2]. The transmission of the larva form of the parasite leading to disease is through the bite of a mosquito. Mosquitoes are infected with microfilariae by ingesting blood when biting an infected host. Microfilariae mature into infective larvae within the mosquito. When infected mosquitoes bite people, mature parasite larvae are deposited on the skin from where they can enter the body. The larvae then migrate to the lymphatic vessels where they develop into adult worms, thus continuing a cycle of transmission. When lymphatic filariasis develops into chronic conditions it leads to lymphoedema (tissue swelling) or elephantiasis (skin/tissue thickening) of limbs and hydrocele (scrotal swelling) [3]. *Wuchereria bancrofti* filariasis characteristically presents with microfilaremia and paradoxical eosinophilia in the acute phase. The chronic phase is usually characterized by presence of lymphadenopathy in lower limbs, retroperitoneal tissues, lymphedema, hydrocele and elephantiasis [2, 4]. The disease resulting from infestation with the filarial worms to affect the lymphatic system and the

skin is what is referred to as Elephantiasis. Lesions associated with Lymphatic filariasis may not easily be distinguished from those of Kaposi sarcoma (KS) and should thus justify the need for biopsy and histopathology for definitive diagnosis. Kaposi Sarcoma is an angioproliferative tumor associated with human herpes virus type 8 virus (HHV-8) [5, 6] with a predilection to the skin, lymph nodes and lymphatic vessels and the viscera.

In other cases KS may present in a rare manifestation known as Elephantiasis nostras verrucosa (ENV). Elephantiasis nostras verrucosa is a progressive cutaneous hypertrophy resulting from chronic non-filarial lymphedema secondary to obstruction of the lymphatic system [7]. It usually presents as non-pitting edema superimposed by hyperkeratotic papules, nodules, and verrucous cobblestone-like plaques [8, 9]. This case report seeks to highlight a probable case of lymphatic filariasis in contrast with Kaposi sarcoma and the implications to care based on the clinical and histopathological presentation.

## Clinical presentation

We present a case of a 73 year old female patient referred from a level 2 hospital to a tertiary hospital for further evaluation and management of suspected Kaposi sarcoma in a known HIV disease patient on anti-retroviral therapy. Patient reported having been generally unwell for about 6 years prior presentation. In the current presentation she complained of

bilateral leg swelling, painful ankle and knee joints and burning sensation in the feet for a period of 2 months. She is a known hypertensive on Atenelol and also on Tenofovir, Lamivudine and Dolutegravir for HIV disease. She reported no history of Diabetes, Cardiac disease or Tuberculosis. Her CD4+ count was 330 cells/ul and viral load of target not detected status. She is a divorced peasant farmer based in a rural area of the country. She reports no history of any form of substance abuse. She was a receipt of anti-filarial medications in the form of Diethylcarbamazine and Albendazole administered in a mass vaccination exercise by the Zambian Ministry of Health to combat filariasis. This was about 3 years prior presentation to hospital.

On examination, she was stable, afebrile, not pale or jaundiced and not in any obvious distress. She had gross bilateral lower limb swelling with hyperpigmentation and induration. The swellings were mildly pitting and non-tender. The right mid-calf diameter was 44.5cm and the left was 43.5cm. There were no varicosities or calf tenderness. Findings in the other systems were largely unremarkable. Images in figures 1 and 2 further depict the lower limb findings.

### Images depicting a case of chronic lymphedema



Figure 1.



Figure 2.

The blood pressure was 167/98, pulse rate 51beats/minute and temperature of 36.7 degrees celcius. A provisional diagnosis of Chronic Lymphedema secondary to Lymphatic filariasis with a differential diagnosis of Kaposi's sarcoma was made. It was also noted that the patient presented with uncontrolled hypertension.

Results of the investigations ordered revealed a microcytic anemia with hemoglobin of 10.4g/dl and an MCV of 76.1 fl (79.1-98.9). The ESR was 27 mm/hr. (0-29) and biochemistry suggested relatively normal liver and kidney function. The blood smear for microfilaria was negative. The histopathology findings on the skin section suggested moderate papillomatosis and hyperkeratosis with slight lymphocytic infiltration in the dermis. There were no signs of malignancy on the biopsy tissue.

### Discussion

Clinical features of Lymphatic filariasis presenting as Elephantiasis can mimic those of Kaposi's sarcoma and present a challenge with diagnosis more so in patients with HIV disease where Kaposi's sarcoma is a common malignancy. The distinction in this case may only be clearly drawn on histopathological findings. However, a high index of suspicion of one over the other can be raised through a carefully taken

history and physical assessment of the patient. Lymphatic filariasis (LF) is a disease arising from infestation with a filarial worm. In Sub Saharan Africa, LF is caused by the filarial nematode *Wuchereria bancrofti*, transmitted by mosquitoes of the species of *Culex quinquefasciatus*, *Anopheles gambiae* s.l. and *Anopheles funestus* [10, 11]. The disease is also widely endemic in Zambia with an overall prevalence rate estimated at 7.4% in 2011 from more than 10, 000 sampled individuals across 108 sites in all regions of the country [12]. Patients in the endemic areas can be asymptomatic for many years. Microfilaremia is usually detected in blood or skin specimens, but its absence does not rule out the presence of filariasis [2].

Our patient presented with symptomatology quite consistent with filariasis and more so coming from a region (Southern Province), where the prevalence was reportedly highest in the 2009 to 2011 mapping [12]. The negative blood smear in our patient could be attributed to her prior history of receiving the anti-filarial medications in the mass vaccination campaign.

The possibility of HIV/AIDS associated chronic lymphedema was also considered based on the HIV status and clinical presentation. However, patient reported a relatively stable clinical, immunological and virological course while on anti-retroviral therapy. The biopsy and histopathology report did not also favour classic Kaposi's sarcoma. In this case findings would suggest in early stage; thin walled, dilated vascular spaces in the epidermis with interstitial inflammatory cells and extravasated red cells. Later lesions would suggest spindle shaped stromal cells with irregular slit like spaces filled with red blood cells.

A plausible differential for consideration is Elephantiasis Nostras Verrucosa (ENV). This is described as a progressive progressive cutaneous hypertrophy characterized by non-pitting edema with superimposed hyperkeratotic papules, nodules, and verrucous cobblestone-like plaques [13]. In our patient there were no obvious nodules or verrucous cobblestone-like plaques. Histopathology of ENV suggests pseudoepitheliomatous hyperplasia. Other histological features observed during the early stages of the disease include dilated lymphatic channels, loss of sweat glands, and dermal papillae [14]. Similarly, findings in our patient suggested moderate papillomatosis, hyperkeratosis and slight lymphocytic infiltration in the dermis but were devoid of dilated lymphatic channels.

Based on the clinical presentation, the referring hospital's plan was to institute treatment for Kaposi's sarcoma. However, on re-evaluation, the history, physical findings and histopathological report could not justify the indication for chemotherapy. Patient was advised to remain fully compliant to ART and anti-hypertensives for optimal viral suppression and BP control respectively. Additional prescriptions received were Albendazole, Haemup and Brustan. On subsequent clinical reviews patient remained stable and with no evolution or worsening of symptoms.

### Conclusion

The clinical presentation of Lymphatic filariasis and that of Kaposi's sarcoma can be conflicting. Although a carefully taken history and physical examination may help in establishing a probable diagnosis, a biopsy for histopathology determination must be indicated to avoid misdiagnosis and misapplication of treatment. Apart Kaposi's sarcoma from other potential causes of chronic non-filarial lymphedema such as tumors, trauma, radiotherapy and chronic venous stasis [7] should also be considered on the differential diagnoses.

### Consent:

Formal consent was obtained from the patient

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**Conflict of Interest:** None

### Author Contributions:

The concept and design of the case report were devised by Christopher Nyirenda. All authors contributed towards the content, review, and ultimate write-up of the manuscript.

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