

# Angiosarcoma in Chronic Filarial Lymphedema – A Report of Two Cases

Amar Kamat<sup>1</sup>, Pramod S Chinder<sup>1</sup>, Suraj Hindiskere<sup>1</sup>, Anto Anand Gopurathingal<sup>1</sup>

## Learning Point of the Article:

Though a rare complication, angiosarcoma needs to be diagnosed correctly and treated adequately for better outcomes..

## Abstract

**Introduction:** Cutaneous angiosarcoma is an uncommon aggressive tumor known to arise in chronic lymphedema of various etiologies with post mastectomy lymphedema being the most common cause also known as Stewart–Treves syndrome. Chronic filarial lymphedema is the most uncommon etiology for development of angiosarcoma.

**Case Report:** We describe two cases of angiosarcoma developing in a setting of long-standing filarial lymphedema. The patient presented with the same clinical features of rapidly progressing nodules.

**Conclusion:** With India harboring most of the filarial load, this scenario should be kept in mind by clinicians for early diagnosis and treatment. Angiosarcoma is a rapidly progressing tumor and is fatal; hence, an idea about its diagnosis is of utmost importance.

**Keywords:** Stewart–Treves syndrome, angiosarcoma, chronic filarial lymphedema.

## Introduction

India once being an endemic country for lymphatic filariasis has many known complications. Angiosarcoma is a rare but significant complication that has received little attention in the literature. We present two examples of the same.

## Case Report 1

A 61-year-old female presented with an enlarged and edematous right leg (Fig. 1). The patient reported a history of lymphatic filariasis diagnosed 15 years previously. She complained of rapidly increasing skin nodules over her leg progressing upward for 6 months. Her prior treatment consisted of antibiotics with variable response. On examination, there was extensive skin thickening with multiple nodules and an ulcerating wound over

the anterior surface of the leg measuring approximately 6 × 3 × 3 cm. The superior most nodule was at the superior border of the patella. Magnetic resonance imaging (MRI) (Fig. 2) showed extensive skin thickening and subcutaneous edema, with normal skeletal muscle and bone. A positron emission tomography (PET) was performed, showing only local disease with no metastasis. An fine-needle aspiration cytology was suggestive of high-grade spindle cell sarcoma. Following pre-operative evaluation, it was noted that all hematological parameters were normal and serological parameters including HBsAg, HCV, and HIV I and II which were negative. The patient was planned for an above-knee amputation (Fig. 3, 4), maintaining a 2 cm margin for the same. Intraoperative frozen section was performed, showing clear margins. The post-operative histopathology confirmed the diagnosis of angiosarcoma spindle-shaped solid type expressing

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## Author's Photo Gallery



Dr. Amar Kamat



Dr. Pramod S Chinder



Dr. Suraj Hindiskere



Dr. Anto Anand Gopurathingal

<sup>1</sup>Department of Orthopaedic Oncology, HCG Hospital, Bengaluru, Karnataka, India.

### Address of Correspondence:

Dr. Amar Kamat,  
Department of Orthopaedic Oncology, HCG Hospital, Bengaluru, Karnataka, India.  
E-mail: dramar.kamat@gmail.com

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**Figure 1:** (a and b) Lymphangiosarcoma in a 61-year-old female.



**Figure 2:** Imaging suggestive of subcutaneous and skin lesion – (a) X-ray in AP and lateral views, (b) magnetic resonance imaging in coronal view, and (c) positron emission tomography scan.

CD31 and CD34. Subsequently, she received adjuvant chemotherapy regimen with doxorubicin-paclitaxel-based therapy. Prosthetic fitting was done at week 6 and the patient was made to weight bear with support for another 6 weeks. At one-year follow-up, an interval PET scan confirmed the disease-free status of the patient. The stump had healed well and consolidated. The patient is currently walking on the prosthesis without support (Fig. 5).

**Case Report 2**

A 60-year-old man presented to the hospital with multiple ulcerative nodules over his right leg for 1 year (Fig. 6). He was diagnosed with a right filarial limb 20 years previously. He was managed outside with an incision and drainage and antibiotics 6 months before the presentation. Subsequent to this procedure, he reported an increase in the size and the extent of the nodules. On examination, there was extensive skin thickening with multiple nodules and an ulcerating nodule of 10 × 5 × 3cm over the foot with palpable popliteal nodes. MRI (Fig. 7) suggested extensive skin thickening with subcutaneous edema of the right limb, with normal bone and skeletal muscles. A PET scan

demonstrated the popliteal nodes, representing a likely metastasis of the tumor. No systemic spread of the disease was observed in the PET scan. On histopathological evaluation, spindle cell sarcoma was suggested. Following the preoperative workup, an above-knee amputation was planned (Fig. 8), maintaining a 2 cm margin. Postoperative pathology confirmed angiosarcoma, expressing CD31 and CD34 and D2-40 (Fig. 9). The patient received doxorubicin-paclitaxel based adjuvant chemotherapy. At 6 months, patient had blisters on the stump and had to undergo re-fitting of the prosthesis. At 12 month follow-up (Fig. 10), the patient is disease-free as confirmed with PET scan. He is currently ambulant without any complaints.

**Discussion**

Angiosarcoma is an uncommon tumor arising in chronically edematous limbs. It has been reported to most commonly arise in females undergoing mastectomy with removal of axillary nodes (Stewart–Treves Syndrome) [1, 2]. Stewart and Treves were the first to describe a case of Angiosarcoma arising from chronic post-mastectomy lymphedema [1]. It can also occur in idiopathic, congenital, and post-operative chronic lymphedema



**Figure 3:** (a and b) Above knee amputation.



**Figure 4:** Stump postoperatively.



**Figure 5:** (a and b) One year follow-up – Patient ambulating currently on prosthetic limb.



**Figure 6:** (a and b) Lymphangiosarcoma in a 60-year-old man.

Devi and Bahuleyan [11] were the first to describe angiosarcoma in filariasis. They suggested chronic lymphatic stasis as a probable cause of tumor development. The patient had no systemic metastasis but died shortly after diagnosis due to unknown causes, before initiation of treatment. Sordillo et al. [12] described a patient with angiosarcoma in a 20-year-old filarial right lower limb. He was operated on for above knee amputation and radical

[2, 3, 4, 5, 6]. The prognosis of this tumor is very poor, with a 5-year survival rate of between 12 and 33% [7].

Lymphatic filariasis is a neglected tropical disease and is caused by parasites, known as nematodes (roundworms). They can be: *Wuchereria Bancrofti* (most common), *Brugia Malayi*, and *Brugia Timori*. Adult worms harbor in the lymphatic system for almost 6–8 years producing millions of microfilariae. These microfilariae are ingested by the mosquito (mainly *Culex* and *Anopheles*) by ingesting blood of the infected host. The microfilariae mature into infected larvae in the mosquito. When these mosquitoes bite people, the larvae are deposited into the skin and proceed to the lymphatic vessels, maintaining the cycle.

Lymphatic filariasis presents as asymptomatic, acute, or chronic stages. Asymptomatic filariasis may still be silently causing harm to the lymphatic system and kidneys. Acute filariasis may cause inflammation of the skin, lymphatic system, and nodes. Chronic lesions may present as lymphedema of limbs, elephantiasis, and/or hydrocele. Filariasis causes a massive social stigma.

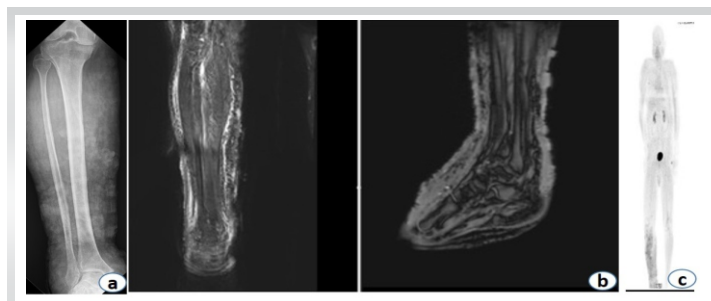
There is no clear consensus in the current literature regarding the malignant transformation of chronic filarial lymphoedema and its optimum management. El Toraei [8] did not identify a single case of angiosarcoma in a large number of cases of filarial lymphedema. Many authors claim that they have not seen this tumor arising in patients with filarial lymphedema. Hence, this tumor occurring in a filarial limb is a rare entity [9, 10].

groin dissection. No adjuvant therapy was given as the nodes were free of disease. The patient developed lung metastasis 8 years after the treatment. Muller et al. [13] demonstrated angiosarcoma in a 15-year-old filarial limb. The patient underwent a hind-quarter amputation. The patient was free from disease at 2-year follow-up. Krishnamoorthy et al. [14] described a patient with angiosarcoma in a 33-year chronic filarial lymphedema. The patient underwent an above-knee amputation, but died at 5 months duration with lung metastasis.

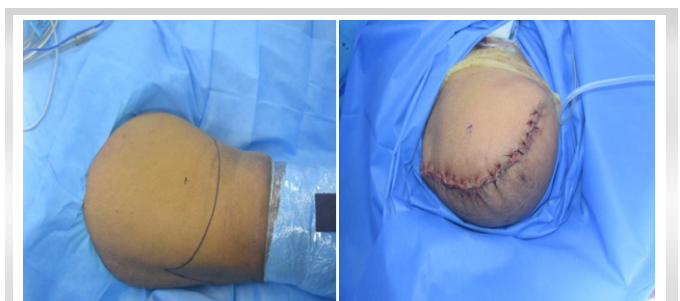
We have reported this case due to the rarity of such a phenomena and to highlight the rare complication of chronic filarial lymphedema. The physician should be made aware of this complication and how a radical and aggressive approach in a planned manner is vital to save the life of the patient. Although the number of patients with newly diagnosed filariasis has significantly decreased in the last decade due to various initiatives [15], there are still many patients with a filarial limb in the community with a potential to develop this dreaded complication.

There seems to be an established role in getting a pre-operative PET scan done, as it helps the clinical to know whether it is a metastatic presentation or not. This may guide the subsequent use of adjuvant chemotherapy. In our study, popliteal nodes were positive for one patient suggestive of metastasis and the other patient had no metastasis.

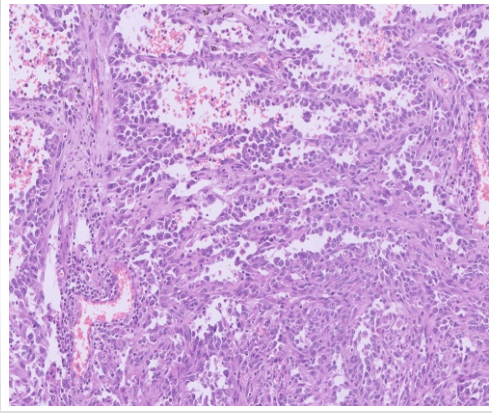
The surgical aspect for such a presentation needs attention. The



**Figure 7:** Imaging – (a) X-ray AP and lateral view, (b) magnetic resonance imaging coronal and sagittal view, and (c) PET scan.



**Figure 8:** (a and b) Above knee amputation same as above.



**Figure 9:** Ovoid to spindle-shaped cells in vague sheets and nests in scanty stroma. Brisk mitosis with no necrosis.



**Figure 10:** One-year follow-up of the stump.

**Conclusion**

Angiosarcoma setting in a scenario of chronic filarial lymphedema is a rare but significant complication with only a few cases reported world-wide. With India having harbored most endemic areas for lymphatic filariasis, close follow-up of these patients along with awareness of this phenomenon is crucial in early diagnosis and treatment. The study of adjuvant chemotherapy shall be done to judge the long-term survival in such cases.

mb should be wrapped preoperatively to prevent soiling of the surroundings and also of the operation theater. Limb elevation and steroids are started for the patient to decrease the lymphedema as much as possible as it minimizes wound complications. Antibiotics should be started a day before the surgery.

**Chart 1:** The Yellow Ribbon (TYR) protocol for management of leg ulcers before surgery

Day 1-3: Start steroids and maintain limb elevation



Day 4: Start antibiotics and wrap the limb



Day 5: Surgery

**Clinical Message**

This case report strives to achieve early diagnosis and treatment in a potentially curable case.

**Declaration of patient consent:** The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given the consent for his/ her images and other clinical information to be reported in the journal. The patient understands that his/ her names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**Conflict of interest:** Nil **Source of support:** None

**References**

1. Stewart FW, Treves N. Lymphangiosarcoma in post-mastectomy lymphedema. *Cancer* 1948;1:64-81.
2. Hajdu SI. *Pathology of Soft Tissue Tumors*. Philadelphia: Lea and Febiger; 1979.
3. Mackenzie DH. Lymphangiosarcoma arising in chronic congenital and idiopathic lymphedema. *J Clin Parhol* 1971;24:524-9.
4. Memck TA, Erlandson RA, Hajdu SI. Lymphangiosarcoma of a congenitally lymphedematous arm. *Arch Pathol* 1971;21:365-71.
5. Burnett WO, Hardy JO, Hendrix JH. Lymphangiosarcoma following postmastectomy lymphedema. *Ann Surg* 1969;169:960-8.
6. Calnan J, Cowdell RH. Lymphangio-endothelioma of the anterior abdominal wall: Report of a case. *Br J Surg* 1959;46:375-9.
7. Fletcher CD. *Diagnostic Histopathology of Tumors*. 3rd ed., vol. 1. London, United Kingdom: Elsevier Churchill



Livingstone; 2007. p. 63-7.

8. El-Toraei I. Filial lymphedema. *Angiology* 1966;17:644-60.

9. Enzinger FM, Weiss SW. *Soft Tissue Tumors*. St. Louis: CVMosby; 1983.

10. Scott RB, Nydick I, Conway H. Lymphangiosarcoma arising in lymphedema. *Am J Med* 1960;28:1008-12.

11. Devi L, Bahuleyan CK. Lymphangiosarcoma of the lower extremity associated with chronic lymphedema of filarial origin. *Indian J Cancer* 1977;14:176-8.

12. Sordillo EM, Sordillo PP, Hajdu SI, Good RA. Lymphangiosarcoma after filarial infection. *J Dermatol Surg Oncol* 1981;7:235-9.

13. Muller R, Hadju SI, Brennan MF. Lymphangiosarcoma associated with chronic filarial lymphedema. *Cancer* 1987;59:179-83.

14. Krishnamoorthy N, Viswanathan S, Rekhi B, Jambhekar NA. Lymphangiosarcoma arising after 33 years within a background of chronic filariasis: A case report with review of literature. *J Cutan Pathol* 2012;39:52-5.

15. Das PK, Ramaiah KD, Augustin DJ, Kumar A. Towards elimination of lymphatic filariasis in India. *Trends Parasitol* 2001;17:457-60.

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