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Filarial Lymphedema: A Debilitating Old World Disease in Modern Tropics - Our Experience and Review of Literature

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Introduction

Objective: Filarial lymphedema , also known as elephantiasis is a neglected chronic disease of the tropics with an estimated population of approximately 36 million population living with this condition as per WHO [1]. While the infection may be acquired in childhood, its visible manifestations may occur later in life causing severe and temporary/ permanent disability and disfigurement [2].

Objective: to present our experience with patients presenting with filarial lymphedema.

Methodology: Between September 2016 to October 2017, 11 patients presented to us with grade III / IV lymphedema involving lower limbs or penoscrotal region or both. 8 of them had already received antifilarial medication several cycles in past, 3 of them were previously operated and presented with a recurrence of the condition. In our setting patients were treated with medicine,

excisional surgery and other supportive measures appropriate as per patient's condition.

Results: In the 6 month period of follow up patients presented with better cosmetic outcome, being able to ambulate easily and perform day to day activities, while some patients with penoscrotal filariasis were able to resume sexual activity.

Conclusion: Filarial lymphedema, though associated with chronicity and recurrence, can be controlled and managed by several measures including excisional surgery. It has a high social stigma and is a debilitating condition. In lack of any definitive treatment, elimination remains the key to management of this disease worldwide.

Key words: DEC- diethylcarbamazine, ADL Aacute dermato- lymphangio- adenitis.

Introduction

Lymphatic filariasis (LF) is a vector-borne disease of the tropical and subtropical countries due to infection by filarial worms, which invade the lymphatics of humans, initiating pathological changes leading to later filarial disease manifestations. The nematode species that cause LF include mostly Wuchereria bancrofti(90%), to a lesser extent Brugia malayi (10%), and Brugia timori to a very small extent[3]. WHO launched its Global Programme to Eliminate Lymphatic Filariasis (GPELF) in 2000. In 2012, the WHO neglected tropical diseases roadmap reconfirmed the target date for achieving elimination by 2020[1]

Materials and Methods

Out of all patients presenting with lymphedema and admitted in one of the units of department of surgery, we studied those with grade III/IV lymphedema [2] of filarial etiology over a period of 14 months from September 2016 to October 2017 and a total 11 patients were evaluated and managed accordingly. Patients were evaluated clinically with a proper history and examination, followed by laboratory and radiological evaluation available (Doppler and ultrasound), and management was planned accordingly.

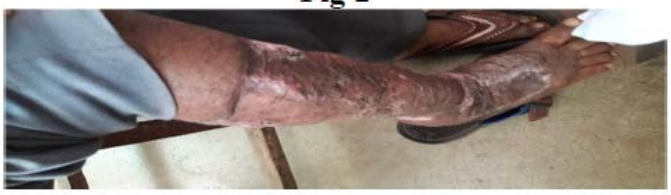
Results

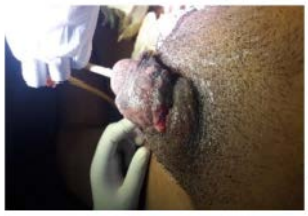
Patients presented to us with swelling and disfigurement of lower limbs or penoscrotal region or both. All the patients were males, belonging to age group 30- 50 yrs and living in rural areas. All had these complaints persisting and progressing for > 5 years. Of the total 11 patients 5 had only lower limb swelling (unilateral) of which 3 were cases with recurrence, 4 had isolated penoscrotal swelling while 2 patients had both(involvement of b/l lower limb and penoscrotal region). 9 of them gave history suggestive of recurrent episodes of adenolymphangitis. All except 3 patients had received DEC/Albendazole in past at primary care or higher centres. 3 patients had history of Charle's procedure in the past and presented with recurrence in the same limb in 5 years. None had history of any radiation, major surgery, renal, cardiac or hepatic disease. Patients with lower limb disfigurement had severe disability and it affected their work, whereas those with penoscrotal lymphedema had impaired sexual life and difficulty in ambulation and micturition. On examination all patients were afebrile and stable. No significant regional lymphadenopathy was noted. Edema was non pitting. Skin changes were present including lichenification, nodules over skin, fissurations etc. USG and Doppler showed subcutaneous edema, fibrosis and showed no signs of living parasite. Blood tests were negative for microfilariae. Evaluation was done w.r.t disease and considering surgical intervention needed.

After exclusion of any other etiology and considering the endemicity of filariasis in our country, a diagnosis of filarial lymphedema was reached. Patients were started with DEC, single dose Albendazole(400mg)[4] , compression therapy and limb elevation while they were evaluated. Excisional surgery was planned. Charle's procedure was undertaken for those with lower limb involvement. Scrotoplasty and penile skin excision with STSG was done for cases with penoscrotal manifestations. Post-operative periodwas uneventful with good uptake of graft. Patients were discharged by 7-10 days. Foot care, compression and limb elevation was advised. During the follow up period only 4 out of 11 cases turned up, 2 with limb involvement had reduced bulk, better ambulation and cosmesis while the other two with penoscrotal involvement reported better ambulation, and one of them reported.

Fig 1

Fig 2





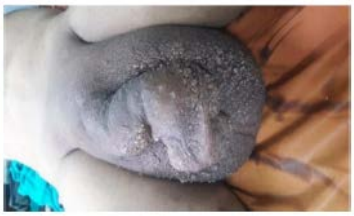


Fig 3 Fig 4

Preoperative and postoperative images of patient with penoscrotal lymphedema.

Fig 5 Fig 6

Picture showing recurrence in 5 years in patients who underwent Charle's procedure earlier. Table 1.History, clinical examination, evaluation and follow up in patients with filarial lymphedema.

Sl no.	Chief complaints		Episodes of ADL	H/o prior intake of DEC or albendaole	Past history (surgery/radiation)	Clinical examination			Investigations		Follow up
	Lower limb swelling	Penoscrotal swelling				Pitting edema	Significant Lyphadenopathy	Skin changes	Doppler (s/c edema)	Blood for mf	
1	+	–	+	+	–	–	–	+	+	–	+
2	–	+	–	+	–	–	–	+	+	–	–
3	+	+	+	+	–	–	–	+	+	–	–
4	+	+	+	+	-	–	–	+	+	–	–
5	–	+	–	+	–	–	–	+	+	–	+
6	–	+	+	–	–	–	–	+	+	–	–
7	+ (R)	–	+	+	+ (same condition)	–	–	+	+	–	+
8	–	+	+	–	–	–	–	+	+	–	–
9	+(R)	–	+	+	+ (same condition)	–	–	+	+	–	–
10	+ (R)	–	+	+	+ (same condition)	–	–	+	+	–	–
11	+	–	+	–	–	–	–	+	+	–	+

Discussion

Lymphatic filariasis, transmitted by mosquitoes is the commonest cause of lymphedema in endemic countries [2]. Approximately two third of the population affected by filariasis live in India, China and Indonesia [3]. It mostly affects the lower limbs, sometimes the arms, less commonly male genitalia, and rarely breasts and genital region in females. This disease becomes not only a physical problem for the patient but is also a major burden socially as well as economically, owing to the deformity, disability and social stigma of the disease as well as the endemicity and poor vector and disease control. [5, 6,]. Pathologically, infective larvae (L3) of filaria is deposited on skin after an infected mosquito bites a human. Many types of mosquitoes including culex, aedes and anopheles transmit this infection [1]. The deposited larvae enters the lymphatic system where they develop into adult worms. The preferred site in adult males in scrotal lymphatics

in bancroftian infections [7]. Other common locations described in women and children are larger lymph vessels and lymph nodes draining to lower and upper limbs. The adult parasites live in these sites for 6-8 years or more and are responsible for initiating the early pathology in LF. It is believed that this damage to lymph vessels is caused by the adult parasites through mediators produced by them, which cause vessel dilatation or inhibit contractility [8]. Once these vessels are damaged, there is stasis, further aggravating damage to the unidirectional valves, manifesting as soft pitting edema. Over time this progresses to accumulation of fibroblasts, adipocytes and macrophages leading to local inflammatory response. Minimal subclinical breaks and poor hygiene of skin predisposes to secondary bacterial infections (presenting as acute febrile episodes of ADL,adenolymphangitis), further aggravating the inflammatory response and fibrosis. Several such episodes eventually lead to characteristic lymphostatic elephantiasis [9] Clinically, uninterrupted it evolves from stage of asymptomatic microfilaremia to chronic disfigurement leading to permanent disability over time, with several febrile episodes in between.

Clinically it can be divided as acute and chronic manifestations and ADLA seen in any stage of the disease [10]. In an endemic area, in most patients the clinical diagnosis of filarial lymphedema can be made from the history of evolution of the disease and clinical examination of the affected limb. The usual presentation is with unilateral or sometimes bilateral but asymmetrical swelling of the limbs, which is of long duration and associated with thickening of the skin, along with history of repeated episodes of fever and pain in the affected part indicating dermato- adenolymphangitis attacks.

The routine tests like night blood examination to detect mf, Immuno-chromatographic-card test (ICT) card test for filarial antigenemia and ultrasonography for locating the adult worms are usually negative once lymphedema is established. Ultrasound and Doppler can be used to exclude other causes of lymphedema. Lymphoscintigraphy is the test of choice in patients with suspected lymphedema [9]. Contrast magnetic resonance lymphangiography appears to be one of the most promising new tests for evalution of lymphedema, it gives anatomical as well as functional status lymph vessels and lymph nodes of affected limb. Treatment strategy depends on the stage of presentation of the disease. In patients with early stage and active infection DEC is the drug of choice earlier recommended 6mg/kg over 12days, now recommended even single dose is as effective[8]. DEC has no role in chronic and advanced stages of lymphedema when neither adult worm nor micofilaria are present in the patient. It has no role in prevention or treatment of ADLA [2].

Acute episodes of dermato-lymphangioadenitis are distressing and best treated by bedrest, limb elevation, symptomatic management using paracetamol , local application of ointments and antibiotic preparations. Severe cases can be given oral antibiotics like ampicillin/amoxicillin. It is best to prevent the episodes by educating patients regarding proper lower limb care. Lymphedema initially can be taken care at home by simple measures like limb elevation at night and compression garments. With further progression or in more advanced cases, special care may be taken by therapies like complex decongestive physical therapy, compression pump therapy. These measures do not completely cure the disease but halt further progression and decrease the magnitude of lymphedema over time. Surgical intervention is needed in cases with stage II and III who have severe functional impairment, recurrent episodes of lymphangitis, and sevre pain despite optimal

medical therapy. Two main categories of operations available are: reconstructive and excisional [9].

Conclusion

A vast majority of population in endemic countries live with various manifestations of lymphatic filariasis. It affects not only the physical health of an individual but impairs a person sexually, mentally and socially. The economic burden and loos in workforce impacts the nation badly. Lymphatic filariasis (LF) is a disease not just treatable or controllable; it is a disease that can be eliminated. It carries a social stigma which is responsible for seeking healthcare only at very late stages of the disease. Apart from treatment and rehabilitation of existing cases, mass education, early seeking of medical help, vector control and strict implementation of elimination programmes will be needed to make the modern tropics free of old world disease.

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