

Scrotal Elephantiasis: A First Case Report from Bahrain

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Background: Scrotal elephantiasis is a disease that is rarely seen outside the tropical sectors of Africa, Asia, Central and South America. Unlike those endemic regions, where the disease is caused mostly by a filariasis; in developed countries, it is usually secondary to other non-infectious conditions or rarely hereditary.

A seventy-eight year old patient presented to the urology clinic with a painful gross scrotal swelling for more than 6 years. The disease proved to be inflammatory and was assumed to be due to filariasis. Subtotal scrotoectomy was undertaken followed by scrotal reconstruction with satisfactory cosmetic and functional outcome.

To our knowledge, this is the first case of inflammatory probably infectious scrotal elephantiasis to be reported from Bahrain.

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Elephantiasis is a term used in medicine to denote an extremely ancient phenomenon that has been described many decades in the old Persian and Indian literatures; it simply means an abnormal massive accumulation of lymphatic fluid in subcutaneous tissue leading to a gross enlargement of the affected body structure(s), usually an arm, leg, or genitals, to a degree that looks elephantoid in size, texture, and colour¹.

Scrotal elephantiasis (SE), also known as massive scrotal lymphedema, presents with a massive scrotal swelling, and frequently associated with urinary symptoms, such as, dysuria, impotence and erectile dysfunction. SE due to filariasis and Lymphogranuloma venereum infection is a quite endemic disease in the tropical regions of Africa, southern and south-eastern Asia, the Pacific islands, and the tropical and subtropical regions of South America and the Caribbean with more than 120 million infected people². SE is extremely rare outside those endemic regions and most cases, which have been reported from non-endemic countries have proved to be secondary to other underlying conditions such as obstruction of the lymphatics by neoplasm, post-radiation fibrosis, surgical intervention and mechanical traumas. Hypoproteinemia, dermatological diseases such as hidradenitis suppurativa and lichen sclerosus et atrophicus are examples of other medical disorders which have been reported. SE due to infection by parasites, bacterial or fungi are much less frequently described³⁻⁶.

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Primary elephantiasis is rarely encountered; the patient usually has a congenital lymphatics vascular aplasia or malformation. Such diseases are usually characterized by the development of oedema at certain age with characteristic clinical presentation⁶⁻⁸. Irrespective of the diseases background, almost all patients show an unaesthetic appearance and suffer severe psycho-physiological disturbances reflecting badly on poor personal hygiene, urinary incontinence, loss of libido and immobility.

This paper aims to present the first case (to the best of the authors' knowledge) of inflammatory probably infectious scrotal elephantiasis diagnosed in Bahrain.

THE CASE

On 23rd June 2007, seventy-eight year old patient presented to the urology clinic with a painful, gross scrotal swelling, dysuria, impotence and erectile dysfunction for more than six years. He was heavy smoker, 20 cigarettes a day for many years. He was a known case of chronic obstructive pulmonary disease and had a single attack of deep vein thrombosis 15 years ago. No other significant finding, but he gave history of frequent voyages to India. On physical examination, gross enlargement of the genitalia was found along with variable thickening of the overlying skin, mainly affecting the scrotum. There was an associated mild pitting edema of both lower limbs and a palpable (2x3 cm) left inguinal lymph node. The working diagnosis at that time was: A painful scrotal swelling, filarial elephantiasis to be ruled out. Laboratory investigation revealed normal CBC, normal biochemistry including renal function and serum PSA level. Mid-stream urine was unremarkable and serology for Chlamydia trachomatis and other sexually transmitted disease were negative. No peripheral blood film examination was requested for filarial larvae. Ultrasound of the bladder, kidneys and prostate were normal. Scrotal ultrasound was requested but was not performed.

Based on the history, the diagnosis was assumed filariasis. The patient was started on Doxycycline 100mg BD and Co-trimoxazole 960 mg OD, along with Distalgesic. On 26th June 2007, the patient was discharged in less distress; and has been requested to continue his medications. On 8th July 2007, excision of the scrotal, pubic and penile skin (subtotal scrotoectomy) was performed under spinal anesthesia, followed by subsequent reconstruction of the scrotum by a rotational flap and primary closure of the wound. Preservation of the penis and both testes was achieved. The excised pubic-scrotal tissues were three pieces of thick skin and attached connective tissue, the largest measures up to 14.0 x 8.0 x 3.5 cm. The dermis was diffusely fibrosed 2.5 cm in thickness. Microscopically, the disease was reported as 'Inflammatory (Non-infectious) scrotal elephantiasis'; based on the presence of non-specific chronic inflammation and the associated marked scrotal dermal fibrosis, mild to moderate lymphoplasmacytic inflammatory cell infiltrate mainly around dilated, thin walled lymphatic channels, and the absence of any specific infection or microangiopathy.

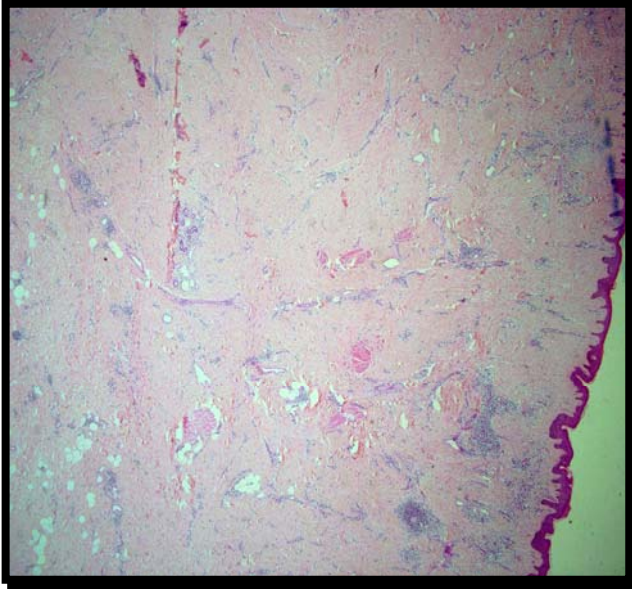


Figure 1: Scrotal Skin Showing Marked Diffuse Dermal Fibrosis and Non-specific Chronic Mononuclear Inflammatory Cell Infiltrates, Mainly around Dilated Lymphatic and Blood Capillary Channels

Post operatively, the patient was stable and the surgical wound was healing. The patient was discharged after four days; however, two weeks later; he was re-admitted for having wound infection with secondary sinuses. Wound debridement was performed under general anesthesia. Besides daily dressing, he was given three days courses of IV Flagyl and Cefuroxime antibiotics. The patient was discharged in a satisfactory condition and advised to continue oral antibiotics (Cipro-floxacin and Doxycycline). Further follow up visits showed complete healing and no functional disability.

DISCUSSION

Lymphedema can be either primary or secondary. Primary (Hereditary) lymphedema usually is not associated with other malformations, affects the lower limbs and is inherited autosomal dominant disease⁶.

Primary (Hereditary) lymphedemas are classified into: congenital lymphedema type I - (Milroy disease): presents at birth with legs swelling, lymphedema type II (Meige disease): having an onset at childhood or around puberty, and lymphedema type III - (Lymphedema Tarda) with late onset⁶. The diagnosis of hereditary lymphedema can be made, based on a positive family history of similar conditions and confirmed by lymphoscintigraphy test⁹.

On the other hand, secondary lymphedema is a group of diverse entities with variable clinical presentations. Secondary lymphedema can be classified into four subtypes, namely: Obstructive lymphedema, secondary to a neoplasm (including primary from prostate and hematological malignancies), post-radiation, iatrogenic, mechanical trauma or injection of a chemical agent; inflammatory lymphedema complicating parasitic infection, such as Filariasis, bacterial infection (e.g. tuberculosis, repeated streptococcal infection and sexually transmitted diseases, such as, Chlamydia and Donovanosis); Phlebitis associated lymphedema seen in some dermatological diseases like Hidradenitis suppurativa, and lastly Angioneurotic lymphedema^{7,10}.

Scrotal elephantiasis due to Filariasis affects about 20-50% of males living in the endemic tropical regions of Africa, Asia and America¹¹. It is estimated that over 120 million have acquired the disease and over 40 million are seriously debilitated. This disease still puts more than a billion people in more than 80 countries at risk^{2,12}. India harbors about one-third of the people infected with the disease and contains numerous breeding sites for the mosquitoes that transmit it⁶, it can be strongly argued that our patient might have contracted the disease during one of his frequent visits to the region, the parasitic filarial worms *Wuchereria bancrofti* or *Brugia malayi* can live up to 6 years in patient's blood circulation.

All patients from non-endemic areas with scrotal elephantiasis, unless it proves otherwise, must be considered potentially filariasis induced. Preoperative investigations should include a routine thick peripheral blood film microscopic examination for the filarial larva. Negative result, does not rule out filariasis, it is easily overlooked. In this case, it was assumed that the patient had filarial elephantiasis based on the history of frequent visits to India.

Filariasis is easily overlooked because it has a "nocturnal periodicity" i.e. it can be detected in the blood few hours around midnight. The "card test" can detect circulating parasitic antigens, it is a simple, very specific, and needs only few blood droplets to perform, unfortunately, it is not available. Renal, liver and cardiac functions tests are important to be established and monitored during treatment for a possible derangement.

Establishing the diagnosis of secondary inflammatory non-infectious scrotal elephantiasis is rather by exclusion; CBC, Urine routine and microscopy, ESR and blood biochemistry are usually normal. Serologic or PCR studies should prove negative for most major infectious agents mainly filariasis and Chlamydia. Serum tumor markers such PSA must be requested too. Abdominal, pelvic and Scrotal Ultrasonography reveal normal prostate, testes and lower urinary tract without evidence of outlet obstruction.

Microscopic examination of the excised affected tissue shows a rather non-specific change reflecting the effect of chronic tissue damage, mainly dermal fibrosis, and variable degrees of chronic inflammatory cells around dilated lymphatic channels.

Regardless, of the etiology of scrotal elephantiasis, the full blown disease manifests following a marked decrease in the lymphatic outflow with subsequent accumulation of interstitial oedema, hypertrophy of the involved connective tissue and the influx of chronic inflammatory cells^{3,13}. In earlier stages of the disease, usually, there is no permanent damage to the affected skin, lymphatics or subcutaneous tissue; however, persistent, long standing, chronic scrotal lymphedema eventually leads; to thrombolymphangitis, perilymphangitis and lymphadenitis leading to an irreversible tissue damage and fibrosis associated with the risk of necrotizing fasciitis^{3,14,15}. Our patient has presented in late stage where the condition is not reversible with medical therapy alone and needs to be reduced surgically. Surgical intervention is indicated in late presentation and usually aims: 1) to excise all elephantoid tissue, 2) to spare the penis, spermatic cord and testes, 3) to reconstruct the area by simple scrotoplasty usually by using skin flaps from the posterior part of the scrotum whenever possible. Medial thigh flaps can be used in the absence of adjacent scrotal tissue; however, it was not needed in our case.

In medical therapy, Doxycycline is given to treat a possible filariasis whether *Wuchereria bancrofti*, *Brugia malayi*, *Brugia timori*, or *Loa loa*. However this antibiotic is effective in

the early stages of infection and needs to be taken for 8-week to achieve complete elimination of the infection. Doxycycline has some toxic side effects and needs to be monitored^{16,17}.

There are several surgical techniques for reversing the chronic scrotal lymphedema; however, two main methods are popular, namely: Lymphangioplasty (Physiologic method) to drain the excess lymphatic from the involved region to newly established lymphatic channels and Lymphangiectomy with reconstructive surgery. The second technique was performed on our patient; it is relatively quicker and more effective especially in case of recurrent lymphedema, provided the underlying cause has been eliminated. It was claimed that this method is unsuccessful in cases of chronic fibrosis or lymphedema caused by radiation because of the lack of appropriate lymphatic channels. It is essential to remove involved skin and subcutaneous tissue completely (reduction scrotoplasty) to prevent lymphedema recurrence followed by reconstructive surgery of penis and scrotum. The posterior scrotal skin usually uninvolved and can be used as a source of skin flap for reconstruction of scrotum¹³.

CONCLUSION

Scrotal elephantiasis is a common descriptive condition, which is caused by several diverse underlying diseases, mostly secondary to infection by Filariasis. This disease is caused by a group of tropical thread-like parasites and it is quite rare disease outside those endemic regions; however, due to the modern ease of travel from one part of the world to the other, the disease can be acquired and imported to a non-endemic area, such as Bahrain.

To the best of the author's knowledge, this is the first case of scrotal elephantiasis to be reported from Bahrain. The management of this case was based on clinical suspicion, for which combined medical and surgical approaches were undertaken to control the disease progress. The former was achieved by an empirical 8-week course of Doxycycline to eradicate the potential filarial infection and the latter by scrotoplasty and scrotoplasty to regain satisfactory functional and cosmetic recovery. The patient responded well and has no further complaint or recurrence of the disease on follow up.

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