Scrotal lymphoedema: staged reduction and overview of the surgical management

AA Mohamed*, SM Mahmoud, MAM Ibnouf, S Mustafa

Plastic Surgery Unit, Soba University Hospital, Khartoum, Sudan

Abstract

Background: Massive scrotal lymphedema (SL), also termed elephantiasis, can be caused by obstruction, aplasia or hypoplasia of lymphatic vessels. It is usually caused by acquired infection like in lymphogranuloma venereum or filarial infestation with Wuchereria bancrofti. Scrotal elephantiasis is extremely rare outside endemic regions in Africa and India.

Case report: A 33 year-old male presented with massive SL. Investigations illustrated a primary SL, and surgery proved to be the best management option.

Conclusion: Primary SL is more common in industrialized countries. We present a primary one where serial reduction of the scrotum was performed.

*Corresponding author: Email: abdelsamieabdalla@gmail.com

Introduction

Scrotal lymphedema (SL) is a rare clinical pathology with multiple aetiologies1. Primary lymphedema is caused by lymphatic system malformations. Filariasis is the most frequent cause all over the world 1. However, in industrialized countries, scrotal lymphedema is secondary to other pathologies or iatrogenic interventions, like in chronic infections, oncologic surgery with or without lymphadenectomy and pelvic radiotherapy 1. We report a case of neglected primary scrotal lymphedema in a Sudanese patient, with an overview of the series of surgical reductions and discuss the literature of both the disease and the surgical options for management1.

Patient and method

A 33 year-old patient from Southwest Sudan, presented with increased scrotal size and bilateral lower limb swelling since birth. He was an outcome of normal vaginal delivery. At 40 days old, the patient was first seen in a regional hospital where his parents were reassured. The swelling was progressing and eventually the penis was completely buried. At the age of seventeen, he was admitted to the same hospital because of troubling heaviness and burning micturition. He was managed by diuretics, scrotal support, and intravenous antibiotics.

Clinical examination revealed no abnormalities in the general examination. The scrotum volume was extremely enlarged and affecting the patient’s gait. There was no tenderness, hotness, or cough impulse, and the testicles were not palpable. The skin of the scrotum and both lower limbs was rough and hyperpigmented; macules were obvious (Figure 1 A).

Figure (1)
A full work up including (blood tests, tissue biopsy, ultrasound scans, Doppler scan, lympho-scintigraphy, MRI) was performed. Repeated nocturnal blood films and serology for Microfilaria were negative, tissue biopsy demonstrated no evidence of Microfilaria or lymphogranuloma venereum (LGV) and lymphoscintigraphy demonstrated hypoplasia of lymphatics of lower limbs. An ultrasound scan of the scrotum demonstrated a hugely enlarged scrotum with enlarged septal compartment with a large movable fluid collection, an abdominal ultrasound showed a huge pelvic multilocular cystic lesion of undetermined origin, and an MRI illustrated a large cystic swelling occupying the pelvic cavity, compressing and pushing the urinary bladder to the right, with a wide neck of 3.5 cm connecting to the scrotal cystic swelling and suggesting a differential diagnosis of a hydrocele ascending through a patent processus vaginalis to the pelvic cavity (Figure 2).

On assessment of the lympho-vascular status of the lower limbs and the scrotum, Doppler ultrasound scan of the lower limbs demonstrated a normal arterial flow with a slow flow and echogenic venous blood indicating aggregation of RBCs into longer units due to proximal compression. Lymphoscintigraphy with 2 mCi Tc-99 m with nano-colloid at 5 min, 10 min, 20 min and 30 minutes post injection of both lower limbs illustrated absence of tracer activity from the site of injection (toes web) up to the level of the inguinal region denoting a total lymphatic aplasia. All the above investigations suggested primary (congenital) lymphedema.

The patient was admitted and had scrotal reduction. This included excision of a crescent ellipse of the scrotum preserving the scrotal skin with buried penis and the posterior skin attached to the perineum (figure 1B).

Six months later, further reduction of the scrotum was performed where a posteriorly based scrotal flap with two lateral scrotal flaps and the anterior flap carrying the penis were thinned and preserved, all the rest of the scrotum was excised following identification of testicles and cords (Figure 1C). This approach exploited delay to avoid necrosis and preserved part of the scrotal skin to maintain thermo-regulation for spermatogenesis.

The last surgical session included degloving, reduction and thinning of penile skin before wrapping it back around (figure 1D). All the procedures were done under spinal anaesthesia.

Discussion

Although, Filariasis is a common disease in developing countries, and it is the most common cause of lymphedema worldwide, other etiologies can be found in developing countries and Africa. The most common causes of primary lymphedema are congenital defects, which affects very rarely the genital area. Infections have been reported as the origin of scrotal lymphedema, such as hidradenitis suppurativa, asymptomatic Chlamydia Trachomatis infections, or filariasis infestation. Tests for all these infective processes were negative in our patient. Furthermore, workup has ruled out scrotal lymphangioma, a benign tumour that leads to dilatation of the lymphatic vessels of the skin. Blisters and inflammatory reactions are features of the condition. In contrast, fluid and lymph collection in soft tissues were prominent features in our case. The complaints of the patient were; the heaviness intervening with his daily activity, and the social stigmata of the condition. Other complaints reported were erectile dysfunction that can be due to the elephantoid mass wrapping the penis, and/or testicular atrophy with delayed sexual characteristics. Surgical treatment is indicated in moderate or severe SL. In mild cases, conservative treatment was also reported with hygienic and compressive measures, and medical treatment also has been attempted without long-term success.
There are two main methods of surgical treatment of chronic genital lymphedema: lymphangioplasty and lymphangiectomy with reconstructive surgery. However, surgeon’s preference may play a role in the management, with either closing the gap after lymphangiectomy with split thickness skin grafts or saved scrotal flaps to cover the spermatic cords, testicles and the penile shaft. Most authors agree, that in moderate and severe cases, a complete resection of skin and subcutaneous tissue must be done, with reconstruction with skin grafts. In this patient staged reduction and delay to avoid necrosis were exploited and part of the scrotal skin was preserved to help thermoregulation.

Conclusion
SL is found to be common in its primary entity in the industrialized countries. We report a case of primary lymphedema in a developing country, with the definitive surgical option of management using reduction, delay, thinning techniques and scrotal flap reconstruction to preserve thermoregulation.

References