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CASE REPORT

MASSIVE PENOSCROTAL AND BILATERAL LOWER LIMB LYMPHEDEMA IN A YOUNG AFRICAN: A CASE REPORT

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ABSTRACT

Introduction: Lymphedema is a challenging disorder characterised by swelling and fibrosis of the skin and subcutaneous tissue due to obstruction of the lymphatic drainage. The penoscrotal area is sometimes affected with untoward clinical and psychosocial effect on both patients and relatives. Rarely, the external genitalia and the extremities may both be affected as seen in our index patient.

Case report: We report the case of a 23-year-old man with massive penoscrotal lymphedema and bilateral lower limb lymphedema who had surgical excision and reconstruction with the remnant normal tissue.

Conclusion: Massive penoscrotal lymphedema with bilateral lower limb lymphedema is a rare occurrence and very few has been reported in the literature especially in Sub-Saharan Africa.

Key words: Lymphedema; Filariasis; Penile; Scrotal diseases

INTRODUCTION

Lymphedema is the chronic and progressive accumulation of protein-rich lymphatic fluid in the interstitial spaces due to blockage of the lymphatic system. Worldwide, the most common cause of lymphedema is *Wuchereria bancrofti* infection which has been estimated to account for about 90 million of the cases.[1] Other causes include congenital lymphedema or other acquired aetiology like lymph node dissection, tumors, radiation therapy and lymphogranuloma venereum.[2]

Several organs of the body can be affected, notably the legs and arms but rarely the external genitalia. The involvement of the external genitalia causes a marked enlargement of its volume and an uncomfortable situation for the patient.[3] Lymphedema of the penis and scrotum are also rare occurrence that are equally associated with a lot of untoward experiences. More rarely, it may be idiopathic, often occurring in association with lymphoedema of the limbs as seen in the index case. [4]

Clinically the swelling is painless and shows pitting edema initially. Gradually the skin becomes coarse and tough as fibrosis sets in causing massive enlargement of the genital skin. The swelling is usually progressive, sometimes attaining giant sizes often larger than the patient's head, a condition often referred to as giant scrotal lymphedema. [5] The giant size scrotal lymphedema (massive lymphedema) has a grotesque appearance and

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often impairs ambulation. It makes sexual intercourse impossible, prevent voiding in the standing position and mitigate against proper hygiene of the perineal region. The condition may also lead to mal-odour and recurrent episodes of skin infection, thereby adversely affecting the patient's quality of life and self-esteem. [6]

The use of non-surgical treatment with different approaches has proven inefficient and is no longer used in giant scrotal lymphedemas. Surgical intervention is therefore the mainstay of treatment in such patients.[3] Complete excision of the affected tissue is advocated in order to prevent recurrence. The spermatic cord and the testes are however spared because they drain into the deep inguinal and abdominal lymph nodes which are usually not affected. Following surgical excision, the remnant skin is usually enough for the scrotal reconstruction. Other options that are often employed when the remnant skin is not adequate for closure include the use of skin graft and local flaps. [1]

The aim of this work is to report the rare case of a patient with massive penoscrotal and bilateral lower limb lymphedema. This report is necessary as there are very few reported cases of massive penoscrotal lymphedema with associated bilateral lymphedema in young black patients. The report also further emphasizes the need to have lymphoscintigraphy in sub-Saharan African countries in order to arrive at specific diagnosis.

CASE REPORT

A 23-year-old male patient presented with bilateral limb swelling of 12 years and 2-year history of progressive swelling of the penis and scrotum, associated with pain and skin changes with no swelling in other parts of the body (Figure 1). The scrotal swelling has since progressively worsened and significantly affected his mobility. There was no prior groin or lower abdominal surgery, no exposure to irradiation and no history of similar swelling in the family. He lived in a rural area where the major source of water supply was from a flowing river. Prior to his presentation, medical management with the use of compression stockings and albendazole had been used in another facility with no improvement. Clinical examination revealed a markedly swollen scrotum with peau d'orange changes and indurations. It measured 169cm in circumference and 53cm in length. The penis was also enlarged and almost completely buried within the scrotum.

There was a marked non-pitting edema of both lower limbs up to the proximal thigh with associated induration, dermatitis and verrucous skin changes. The inguinal nodes were enlarged but not painful (Figures1 and 2). Ultrasound scan of the scrotum revealed thickened scrotal skin with stone-paved appearance and marked vaginal hydrocoele. Full blood count results showed Packed Cell Volume of 40% with the leukocyte count within normal range. The Electrolytes, Urea and Creatinine values were also within the normal limits. Skin snip and blood samples yielded no microfilaria. Lymphoscintigraphy was not available for the assessment of the lymphatic channels.

With the patient in supine position after administration of combined spinal epidural anaesthesia, the skin preparation was made using 1% cetrimide and 5% povidone iodine solution. Sterile drapes were laid to isolate the surgical field and a size 16 urethral catheter was passed. A vertical incision was made starting from the upper border of symphysis pubis through the dorsal surface of the scrotum to expose the whole length of the buried penis. The abnormal penile skin were excised and the remaining skin flaps were used to reconstruct the penis. Two lateral incisions were then carried out on either side of the vertical incision encircling the root of the scrotum to meet at the perineum. The

lateral incisions were deepened and care was taken not to injure the structures of the spermatic cord. The cord was then followed distally towards the testes. The cord and the testes with their coverings were isolated from the surrounding structures by blunt finger dissection. The tunica vaginalis was opened and 1.5 litres of hydrocele fluid was drained out. The excess sac was excised and sutured around the testes and epididymis. The same procedure was carried out on both sides. All the abnormal skin and the edematous tissues were excised, including the median septum (Figure 3).

Haemostasis was secured and care was taken not to injure the urethra. Both testes were fixed to the scrotal wall to prevent torsion. The two remnant skin flaps were approximated in the midline to construct a new scrotum. Two active drains were left in the scrotal bed and brought out at the most dependent part. The penis was immobilized in the vertical position using bulky dressing. The scrotal wound was also covered with sterile dressings and held with crepe bandage (Figure 4).

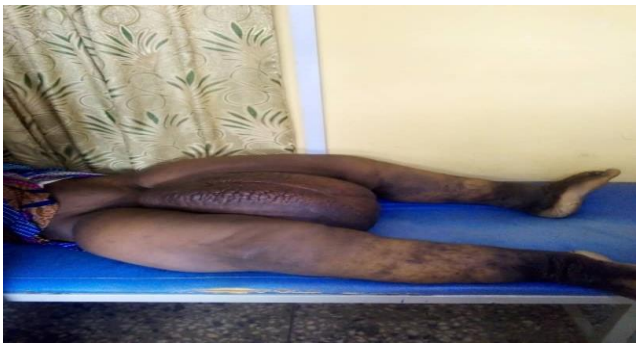
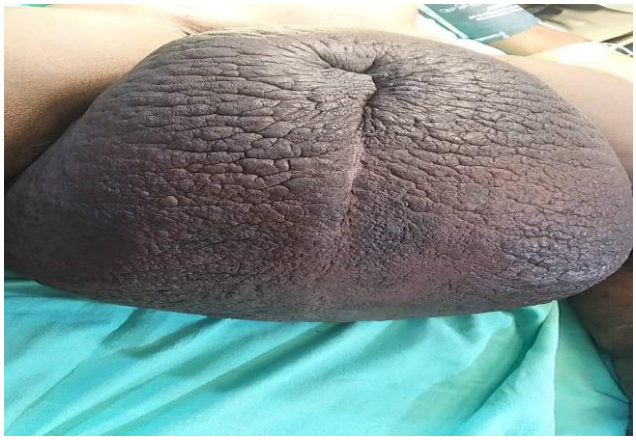


Figure1- Picture showing patient with scrotal and bilateral lower limb lymphedema

The patient was transferred to the Intensive Care Unit for post-operative monitoring on account of the prolonged surgery and he was discharged to the ward on post-operative day 2. After two weeks on admission he was discharged home and followed up in the surgical out-patient clinic (Figure 5). He was re-admitted 6 months later and had the first stage debulking surgery of both lower limb lymphedema using Homan's method. (Figure 6) He has since been discharged with good post-operative outcome and awaiting the second stage debulking surgery.



Fgure 2- A closer view of the scrotal lymphedema showing the skin changes

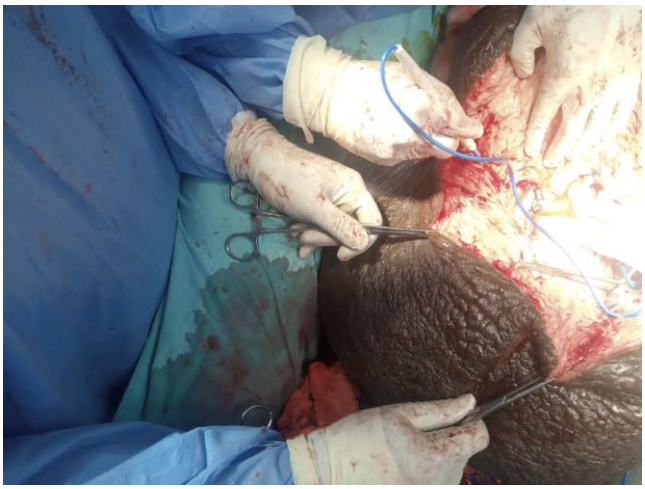


Figure 3- Picture showing intra-operative resection of the edematous scrotal tissue

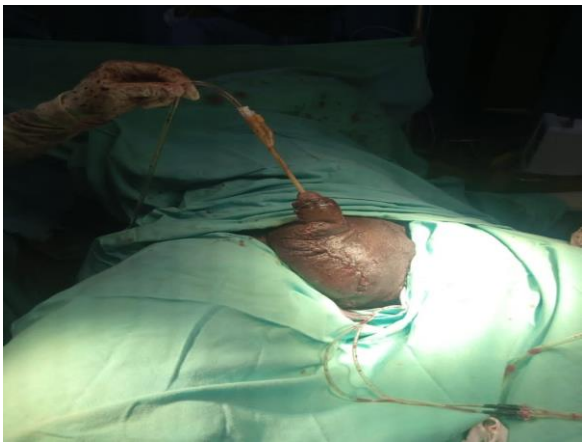


Figure 4- The picture shows the immediate post-operative outcome

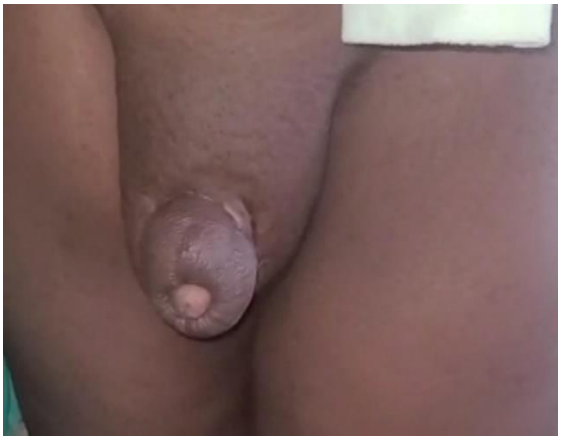


Figure 5- Appearance 3 months after surgery

Figure 6- Appearance six month after penoscrotal surgery (at the first stage debulking surgery for the lower limbs lymphedema)

DISCUSSION

From the clinical presentation of the patient we were unable to identify a specific cause of the lymphedema. Both congenital and acquired causes were ruled out but the only positive finding was that the patient lived in a rural community and used to wade through stream as a child but the skin snip and blood samples taken yielded no microfilaria. The presentation can however be best described as idiopathic primary lymphedema because there was no available facility where lymphoscintigraphy can be done for the patient. Lymphoscintigraphy is the gold standard for diagnosing lymphedema especially congenital lymphedema by establishing lymphatic aplasia, hypoplasia or hyperplasia.[2] A similar case of idiopathic primary lymphedema was reported by Fabrice *et al.*in Benin republic where he also emphasized the challenge of unavailability of lymphoscintigraphy in arriving at specific diagnosis.[7] The lymphedema in his patient was however limited to the limb without the involvement of the external genitalia.

One of the rare presentation of lymphedema like that of our patient was the case reported by Wananukul *et al* in Thailand where the patient had lymphedema of all extremities and the external genitalia.[8] The diagnosis of their patient was however confirmed with lymphoscintigraphy unlike in the index case. In both cases, the swelling started from the lower limbs before involving the external genitalia much later. In addition, there have been reports of association of chronic pyogenic skin infection with genital lymphedema. [9,10] This may also be the case in our patient who had prior progressive scrotal skin infection before the onset of the penoscrotal lymphedema.

In our patient, at the initial phase when only the limbs were affected, medical management with the use of compression stockings and albendazole had been instituted in another facility with no improvement. The patient only presented to us when the scrotum and the penis have been affected and he was already experiencing untoward associated symptoms. One of the major challenges of such conditions in Sub-Sahara Africa is the late presentation of patients to specialized centres where such cases can be effectively managed. In the literature, there are variations in the details of the surgical approaches and this may be as a result of the peculiarities of the patient's presentation. In our patient, the remnant flaps were enough to cover both the

penis and the scrotum after excision of the abnormal tissue without any need for skin grafts. In most other cases in the literature the use of skin grafts for either scrotal or penile reconstruction was predominant.[5] In addition, most of the cases reported concomitant vaginal hydrocele like in the index case while others were not associated with hydrocele.[11]

The report of a rare case of massive penoscrotal and bilateral lower limb lymphedema emphasizes the need to make lymphoscintigraphy readily available and accessible in sub-Saharan African countries for the purpose of accurate diagnosis of the condition. Such cases can be possibly prevented through community-based screening and addressing the environmental risk factors for microfilarial infection. There is also a need for long-term follow-up of patients with massive scrotal lymphedema as it obtains in the index case who is currently being followed up at our surgical outpatient clinic.

CONCLUSION

Massive penoscrotal lymphedema with bilateral lower limb lymphedema is a rare occurrence and very few has been reported in the literature especially in Sub-Saharan Africa. Recurrence after surgical excision is one of the major complications of this condition, thus the index case is currently being followed up at the surgical outpatient clinic.

REFERENCES

1. Shimony A, Tidhar D. Lymphedema: A Comprehensive Review. Ann Plast Surg. 2008; 60:223–9.
2. Szuba A, Shin WS, Strauss HW, Rockson S. The third circulation: Radionuclide evaluation of lymphedema. J Nucl Med. 2003; 44:43–5.
3. Modolin M, Mitre AI, da Silva JCF, Cintra W, Quagliano AP, Arap S et al. Surgical treatment of lymphedema of the penis and scrotum. Clinics. 2006; 61(4):289–94.
4. Salako AA, Olabanji JK, Oladele AO, Alabi GH, Adejare IE, David RA. Surgical Reconstruction of Giant Penoscrotal Lymphoedema in Sub-Saharan Africa. Urology. 2018; 112:181-185.
5. Rahman GA, Adigun IA, Yusuf IF, Aderibigbe AB, Etonyeaku AC. Giant scrotal lymphedema of unclear etiology. J Med Case Rep. 2009; 3:7295.
6. Thejeswi P, Prabhu S, Augustine AJ, Ram S. Giant scrotal lymphedema- a case report. Int J Surg Case Rep. 2012; 3:269–71.
7. Fabrice LM, Ocloo J, Wapembe E, Pierre KB, Bienvenue HM, Ericson M. Idiopathic congenital lymphedema in a 9 year old girl: a case report. Am J Surg case reports. 2023; 4(10):1098.
8. Wananukul S, Jittitaworn S. Primary congenital lymphedema involving all limbs and genatalia. J Med Assoc Thai. 2005; 88(12):1958–61.
9. Holman M, Arnold G, Walton N, Jurkiewicz J. Reconstruction of male genitalia with elephantiasis. Urology. 1977; 10:576–8.
10. Balik E, Eren T, Bulut T, Büyükuncu Y, Bugra D, Yamaner S. Surgical approach tois suppurativa in the perineal/perianal and gluteal regions. World J Surg. 2009; 33:481–5.
11. Ballo B, Dembele O, Remi A, Traore S, Ongoiba S, Kone O, et al. Discovery of scrotal elephantiasis during a free surgical treatment campaign for adult hydocoeles at Koutiala Reference Health Centre. East African Sch J Med Sci. 2023; 6(6):300–3.