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SCROTAL ELEPHANTIASIS: A RARE PATHOLOGY (ABOUT AN OPERATED CASE)

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ABSTRACT

Scrotal elephantiasis is a condition characterized by a considerable increase in scrotal volume which can reach a very large size; common in filarial endemic areas. Outside of this epidemiological context, sporadic cases of different etiologies are reported in the literature. It is a pathology that generally occurs in men in their fourth decade. is very disabling for the patient from a functional, sexual and aesthetic point of view. The symptoms are characterized by an edematous infiltrate of the skin and subcutaneous tissue, cardboard in appearance and purplish in color. The scrotum, penis and perineal region are gradually affected. Scrotal elephantiasis is a pathology of primary, secondary or very often idiopathic etiology. We report the case of a 56-year-old patient with scrotal elephantiasis measuring 65 cm in circumference, in whom we were able to perform a resection with preservation of the noble structures and a reconstruction of the scrotal bursa by suture detachment of the healthy skin of the either side of the elephantiasis and the reconstruction of the loss of skin substance of the penis by skin graft of thin skin. We take stock, through our experience and several literature reviews, of the diagnostic aspects and therapeutic management of penoscrotal elephantiasis.

KEYWORDS: Lymphedema, elephantiasis, reconstruction.

INTRODUCTION

Progressive and chronic tissue swelling due to poor lymphatic drainage causes lymphedema. The latter is classified as primary (due to abnormal lymphatic development) or secondary/acquired (due to damage to the lymph nodes or vessels).[1] Affected tissues initially swell due to accumulation of subcutaneous lymph, and the resulting inflammation then stimulates adipose deposition and fibrosis, causing further enlargement over time.[1,2]

Primary lymphedema is rare; it affects 1.2 per 100,000 people under 20.[3] 90% of scrotal lymphedemas are secondary, but remain rare in developed countries. Its most common cause is a parasitic infection with Wuchereria bancrofti, Brugia malayi or Brugia timori.[4] Other causes of secondary lymphedema in developed countries include radiation therapy, surgical removal of lymphatic tissue, penetrating trauma, and bacterial infections. Complications of lymphedema include infection, functional disability, and chronic skin changes.

Genital lymphedema is often associated with localization in the lower limbs and occurs when swelling due to inadequate lymphatic drainage affects the male or female genitals. Genital lymphedema is rare. Men and women are equally affected.[9]

We report a case of scrotal elephantiasis treated in the plastic and reconstructive surgery department of the IBN ROCHD University Hospital in CASABLANCA in Morocco with a review of the literature.

PATIENT AND OBSERVATION

This is the patient MF, aged 56, married and father of 2 children, originally from and resident in Fkih Bensaleh, professional driver, of low socio-economic level. Operated in 2016 for anorectal abscess. Five months later, he presented with scrotal lymphedema (figure 1). The onset of his symptoms dates back 8 years with the installation of scrotal edema gradually increasing in volume until the disappearance of the proximal half of the penis surrounded by the infiltrated skin. Furthermore, the patient does not present any urinary symptoms but has a handicap from a sexual and aesthetic point of view with psychological repercussions such as anxiety. All evolving in a context of apyrexia and preservation of general condition (figure 1). The patient's general examination did not show any particularity (BP = 12/6

cmhg, HR = 73 bpm). Local examination revealed scrotal elephantiasis measuring 65 cm in circumference.

Rectal examination did not reveal any prostatic hypertrophy. Examination of the lymph node areas did not show any particularity. The rest of the clinical examination, particularly cardiac and pleuropulmonary, was unremarkable. A pelvic MRI was requested as part of the locoregional exploration assessment which showed diffuse edematous infiltration and thickening of the scrotal membranes without detectable tissue mass. Both testicles are atrophic and pushed outward with an elongated ureter.

A biological assessment showed Hb = 13 g/dl; GB = 5400 and Pq =.430000 Urea = 0.3g/l; creatinine = 7 mg/l; fasting blood sugar = 0.9 g/l and a normal hemostasis report.

A serology test for filariasis and chlamydia in the blood came back negative.

The patient was scheduled for a single-block scrotal excision procedure under spinal anesthesia with release of the two testicles covered directly by the remaining skin and the penis which was covered by a thin skin graft taken from the internal face of the left thigh (Figure 2, 3). The surgical specimen weighed 2.6 kg and was sent to the pathologist. The post-operative aftermath was simple. The removal of the wires was done on postoperative day 10 with the graft holding 85%. The anatomopathological result showed that it was an acute and chronic inflammatory change without signs of malignancy or specificity. The long-term outcome was favorable with good healing (Figure 4)

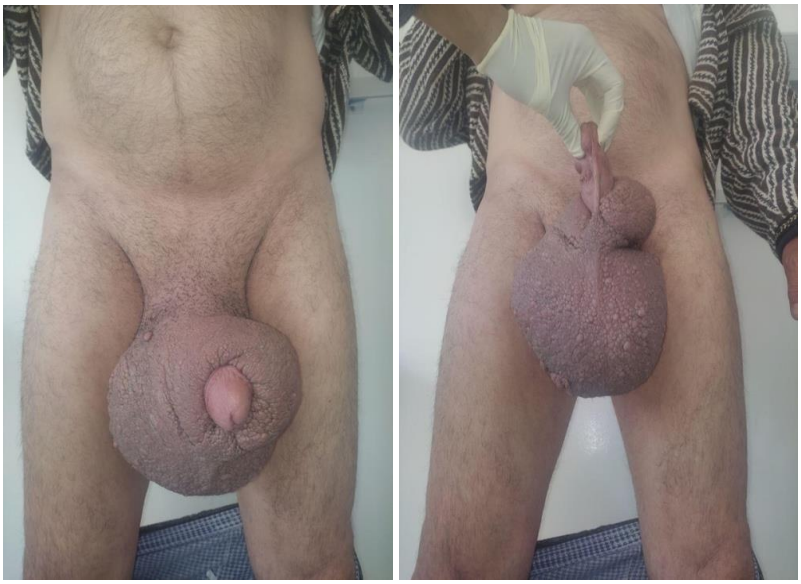

Figure 1: Front View and Preoperative Profile of The Scrotal Bursa.

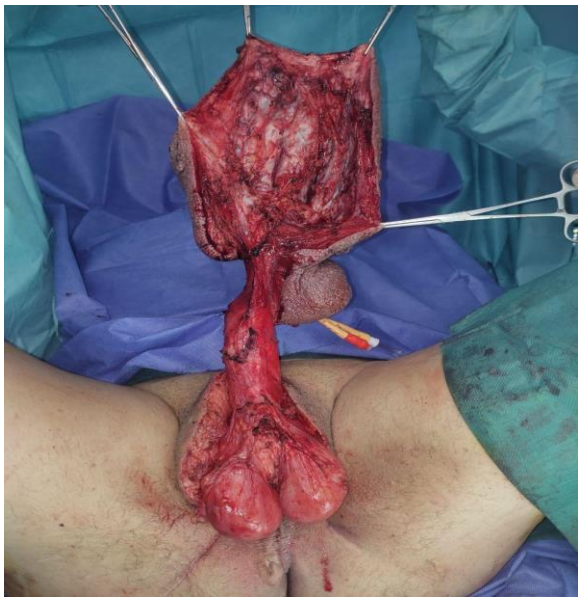

Figure 2: Dissection and Resection of Penoscrotal Skin Tissue with Release of The Testicles.

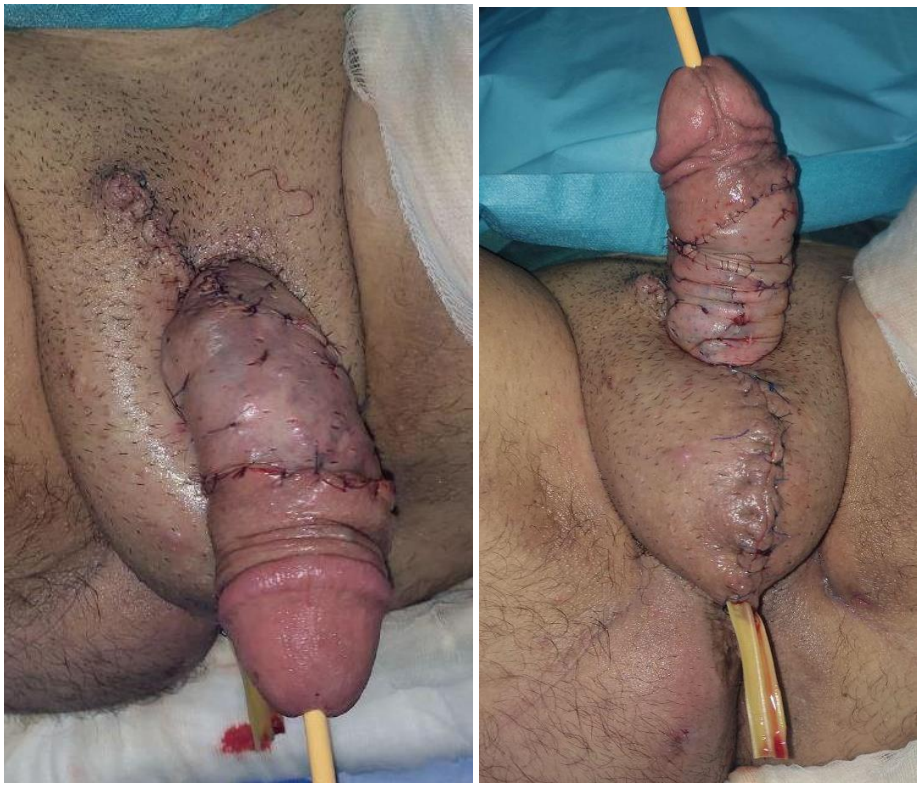

Figure 3: Immediate Post OP After Suture of The Bursae and Skin Graft of The Penis.

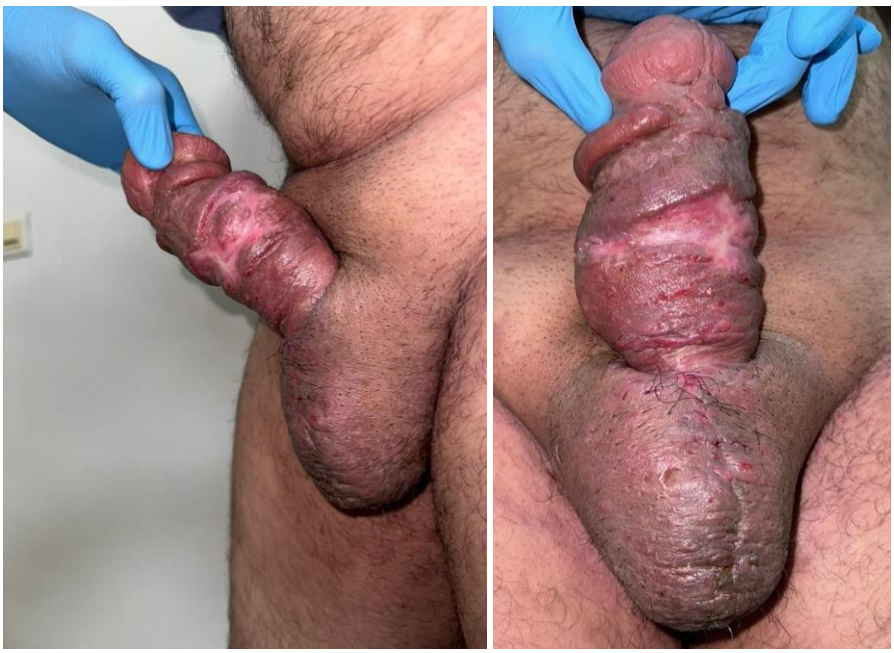

Figure 4: Photo a D30 post-operative with good healing and recovery of function and patient comfort.

DISCUSSION

Scrotal lymphedema is characterized by the presence of a protein-rich fluid effusion at the scrotal and penile level. It often affects the male sex. The lower limbs can be affected in certain cases.[1] The etiology can be primary (idiopathic) due to irreversible congenital dysplasia of the scrotal lymphatic system.[2,3] The composition of the soils has been incriminated, especially volcanic soils rich in aluminum silicate which would be absorbed transdermally, through contact of the feet with the clayey soil. It can be secondary (acquired) to a parasitic infection (filariasis) in filarial endemic countries, inducing ductal obstruction. To intrinsic or extrinsic lymphatic obstruction by: abdominal or pelvic cancer surgery, radiotherapy, chronic inflammatory disease or after chronic venous stasis.[4,5]

Kaposi's sarcoma as well as certain chronic venereal infections have been described as causes of certain cases of elephantiasis.[6] Continuous ambulatory peritoneal

dialysis or previous trauma can also be the cause of scrotal lymphedema.

The cause of primary genital lymphedema is currently unknown, but is believed to be due to a failure in the development of the regional lymphatic system which results in stasis and accumulation of lymph. Lymphedema may extend to the genitals in hereditary lymphedema. These include Milroy's disease, where lower limb lymphedema is present at birth, and there is also a family history of lymphedema[11]; Meige's disease, in which patients develop limb lymphedema lower than adolescence; and lymphedema-distichiasis, where lymphedema is associated with an extra row of eyelashes. Nonsense mutations in vascular endothelial growth factor receptor 3 (VEGFR3) have been associated with primary lymphedema.[12] VEGFR3 expression is restricted to the lymphatic endothelium after early angiogenesis[13] and it has been demonstrated that it induces cell proliferation, migration and survival, via activation of the ERK, AKT and JNK pathways.[14] Lymphedema with distichiasis is caused by mutations in the splice site of the C2 protein, a transcription factor involved in lymphatic development.[15]

Scrotal and vaginal swelling can also be caused by lymphatic malformations and obstruction or dysfunction of the central conducting lymphatic channels. Both of these disorders have been linked to activating somatic mutations in the PIK3CA gene, which encodes the catalytic subunit of the enzyme phosphatidylinositol 3-kinase, and is involved in the regulation of the AKT pathway. Although not fully understood, post-zygotic PIK3CA mutations are thought to result in lymphatic tissue proliferation in affected tissues.[16]

Clinically, penoscrotal involvement is the most common with a bulky scrotum that can reach significant dimensions leading to the burying of the penis as was the case in our patient. The impact of the latter is significant on the micturition function inducing urinary infections, urethral stricture sometimes going as far as pyelo-calyceal dilation; as well as on the patient's sex life.[7] The time between the onset of symptoms and the first consultation is most often long until the bursa reaches a significant volume as is the case in our patient.[8]

History and physical examination are the primary mode of diagnosis, followed by imaging in limited cases. A detailed history should document when lymphedema first developed and also exclude causes of secondary lymphedema such as infection, trauma, previous radiation therapy, or previous surgical procedures. A family history should also be sought, as genital lymphedema can be part of hereditary diseases such as Noonan syndrome, lymphedema-distichiasis, or Milroy disease.[12,15] Lymphoscintigraphy when done for certain patients with concomitant manifestations of the lower limbs shows a delay in transmission and/or cutaneous reflux of the radiolabeled colloid, indicating lymphatic dysfunction with a sensitivity of 92% and a specificity of 100%.[17] Ultrasound is less specific and CT scan exposes the reproductive organs to radiation and should therefore be avoided. MRI can help distinguish lymphedema from other vascular abnormalities, such as venous or lymphatic malformations, and may also be helpful in surgical planning. Patients with a central conduction abnormality may benefit from lymphangiography to localize the lymphatic blockage or abnormality. Biopsy should only be used if malignancy or other conditions, such as noninfectious granulomatous disease, are suspected. Other biological tests can be used to look for chlamydia or microfilariae in the blood[9] and which were negative in our patient.

Therapeutically: The goal of treatment is to ensure function and take care of aesthetic damage. Conservative treatment consisting of lymphatic diversion to the venous network or dilation of lymphatic vessels has been abandoned.

Treatment begins with patient education on the management of genital lymphedema. Infection prevention is of great importance. Daily washing of the genitals and application of a moisturizing agent should be emphasized to prevent skin damage. The patient and their family should learn to recognize the signs or symptoms of cellulitis, as this complication can be life-threatening. Treatment with oral or intravenous antibiotics should be initiated quickly once cellulitis is diagnosed. Low-dose prophylactic antibiotics should be considered in patients who have had three or more episodes of cellulitis within a year. Patients with lymphatic malformations are also at increased risk of infection and should practice good hygiene. Patients with lymphatic malformations may develop intralesional hemorrhages, which are often painful and difficult to manage. The bleeding is often self-limiting and is treated with compression medications and nonsteroidal anti-inflammatory drugs. Lymphatic leaks from skin lymph vesicles or genitals of patients with central conduction abnormalities should be kept dry. Patients can apply absorbent dressings to the affected areas. Cutaneous lymphatic vesicles, particularly on or near the genitals, act as a portal of bacterial entry and can be treated aggressively by CO2 laser ablation of lesions[18] and/or macrocystic lymphatic duct sclerosis.[19] Often with CO2 laser treatments, multiple sessions are necessary to create enough scar tissue to slow or stop lymphatic leakage. Sclerosis near the meatus or urethra risks causing stenosis and should only be attempted by experienced clinicians. Sclerotherapy within the scrotum may also risk damage to the testicles.

Initiation of compression therapy of elastic shorts, pneumatic devices is the basis of non-operative treatment in patients with genital lymphedema.[19] Due to the shape and location of genital lymphedema, the pressure dressing can be difficult to apply and may require ingenuity on the part of the patient and family.

Some patients apply a wedge under the pelvis to elevate the genitals while sleeping to improve lymphatic return. Our institutional experience has shown that when tolerated, compression therapy is useful in maintaining and sometimes reducing genital size. Post-operative penis wrapping is particularly effective in reducing the risk of recurrent swelling.

Surgical treatment of genital lymphedema is indicated in patients with significant disfigurement or morbidity for whom nonoperative management has failed. Indications for surgery include excess mass, functional impairment, and/or disfigurement. Surgical intervention for female genital lymphedema is rare. It is best to wait until the child has reached puberty and is fully developed before attempting to reduce excess tissue. For extreme cases of genital lymphedema, early conservative reduction can be performed, pending possible additional intervention when the child is older. Patients and their families must be informed of their expectations. Although disfigurement can be greatly improved, the genitals will likely never appear completely normal. It should be emphasized that surgery will not alter reproductive function or abilities if performed correctly. In our plastic and reconstructive surgery department in Casablanca, we use an excisional procedure for penile and scrotal lymphedema. The goal of the procedures is to excise the lymphedematous tissue along with the removal of redundant overlying skin. Alternative surgical approaches have been described previously with different incisions without removing redundant skin.[20–21]

Scrotal reduction of lymphedema or lymphatic malformations occurs as follows: An incision is madeeither transversely at the level of the most dependent part of the scrotum, orvertically encompassing the median raphe. The testicles and spermatic cord are identified and protected. A full-thickness portion of scrotal tissue, encompassing skin, adipose tissue, and dartos fascia, is excised. In cases of hydroceles, the tunica vaginalis is also excised. The excess scrotal septum is resected and the testicles are fixed with non-absorbable sutures to the reduced septum. Scrotoplasty is thus performed to facilitate vertical closure to create a new median raphe. For the patient with extreme scrotal hypertrophy, additional redundant tissue can be resected from the lateral aspect of each hemiscrotum and closed using a VY advancement method. Closure is accomplished with multiple layers of absorbable suture in the tunica vaginalis, dartos, subcutaneous tissue, and skin. Postoperatively, bed rest with scrotal elevation for one week (allowing limited ambulation for showers, toileting activities, and meals) and patients are encouraged to wear compression underwear thereafter. shows patients after scrotal reduction. The outcome is most often satisfactory but there is always a risk of recurrence due to persistent or recurrent lymphatic obstruction.

CONCLUSION

Scrotal elephantiasis is a pathology that remains rare outside filarial endemic areas. Its etiologies remain different, hence the importance of a good history, clinical examination and well-conducted paraclinical exploration. The treatment remains surgical with very satisfactory results. Prevention remains a means to reduce clinical worsening.

Conflicts of interest

The authors declare no conflict of interest.

Author contributions

The authors contributed to the care of the patient and the completion of this work. All authors contributed to the conduct of this work. They also declare having read and approved the final version of the manuscript.

Abbreviation

VEGFR3: Vascular endothelial growth factor receptor-3
JNK: Jun N-terminal Kinase
ERK: Extra-cellular regulated kinase
AKT: “protein kinase B”

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