Idiopathic scrotal, penile, and perineal elephantiasis – successful surgical treatment

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KEY WORDS

penis ◗ penile reconstruction ◗ scrotal elephantiasis ◗ scrotal reconstruction ◗ subtotal scrotectomy

ABSTRACT

Scrotal elephantiasis is both emotionally distressing and physically disabling, caused by chronic lymphedema created by obstruction, aplasia, or hypoplasia of lymphatic vessels. We present a case of a patient with massive scrotal, penile, and perineal elephantiasis of unexplained origin and discuss the diagnostic and therapeutic approach. The patient underwent four-stage repair: subtotal scrotectomy saving penis, testes, and spermatic cords followed by scrotal and penile reconstruction with adequate cosmetic and functional outcome.

INTRODUCTION

Elephantiasis is defined as deformation of the body caused by chronic lymphedema created by obstruction, aplasia, or hypoplasia of lymphatic vessels. We can also distinguish mosquito-transmitted tropical elephantiasis that is caused by filarias of *Wuchereria bancrofti*, *Brugia malayi*, or *B. timori*. Scrotal elephantiasis is extremely rare outside endemic regions in Africa and India [1, 2, 3]. In Western countries it is caused by chronic or recurrent erysipelas, herpes virus infections, chronic dermatosis, lymphogranuloma venereum, thrombophlebitis, and occasionally it has been attributed to radiotherapy, neoplasm, and lymphadenectomy [4, 5]. Scrotal elephantiasis is both emotionally distressing and physically disabling. Difficulties with hygiene, urinary incontinence, unaesthetic appearance, loss of libido, and immobility are severely debilitating symptoms.

In February 2008 a 40-year old male was admitted to the Department of Urology of the Silesian Medical University, Poland, with massive scrotal elephantiasis, perineal swelling, and regional lymphadenopathy. Medical history of this patient begins in 1996 when he was treated with antibiotics because of acute orchiepididymitis. Inguinal adenopathy and scrotal elephantiasis persisted after treatment. Between 1997 and 2007 he was diagnosed and treated in many clinical departments in Poland. The detailed immunological, dermatological, endocrinological, and imaging diagnostics was performed, but only revealed Hashimoto's thyroiditis. Laboratory tests excluded human immunodeficiency virus infection, toxoplasmosis, mononucleosis, testicular cancer, schistosomiasis, and chlamydial and filarial infection. Myeloproliferative disorders were also excluded. Histopathological examination of the inguinal node was nonspecific as well. An MRI scan of the abdomen did not reveal a cause of the condition. In 2004 the patient was admitted to the Dermatology and Internal Medicine Department with chronic nettle-rash, angioedema, and hectic fever. Procaine Penicillin therapy resulted in hectic fever remission but had no effect on skin lesions or lymph nodes. During the next few years, despite numerous medical interventions, enlargement of the scrotum was observed. After many clinical consultations the patient was hospitalized in our Department in 2008 and finally qualified for surgical genital reconstruction.

On initial physical examination the patient had a massively enlarged scrotum extending down to his knees. The huge solid

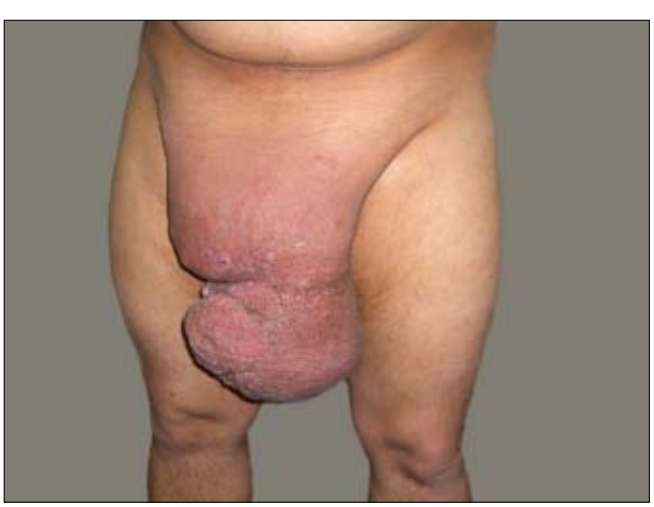

Fig. 1. The huge solid verrucous scrotal mass.

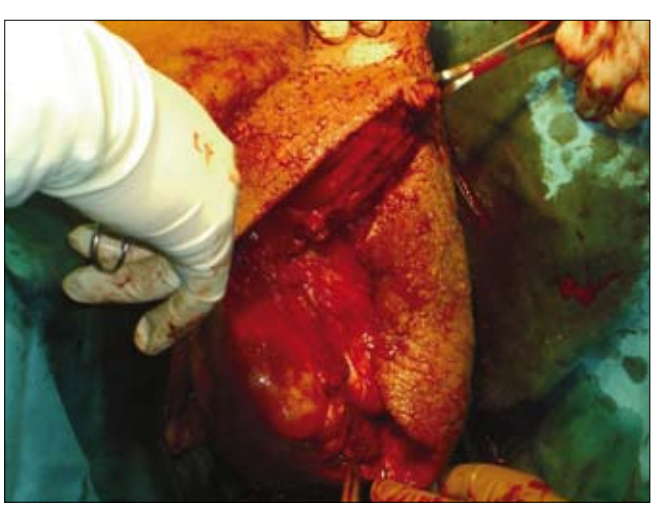

Fig. 2. Longitudinal incision of the scrotum and perineum. Subtotal scrotectomy.

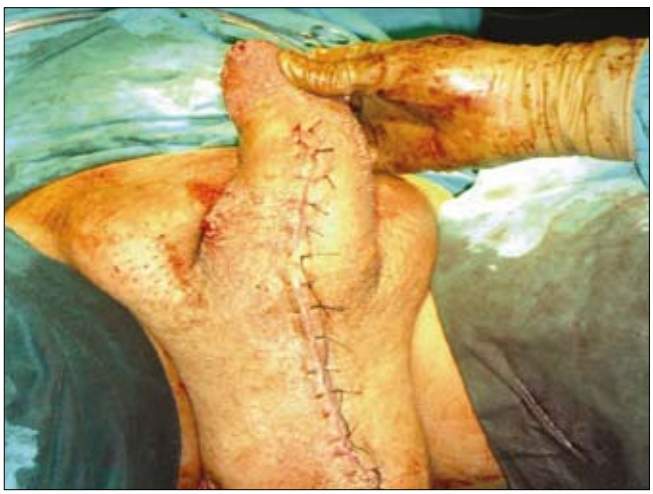

Fig. 3. View after first stage of the surgical treatment.

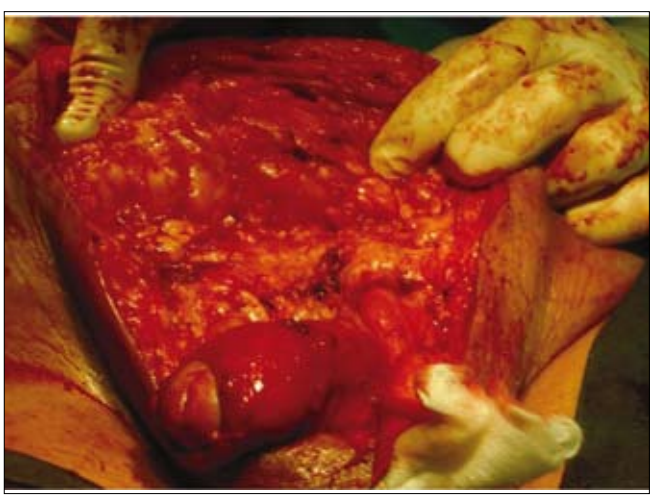

Fig. 4. Scalped penis before inoculation under the suprapubic skin.

verrucous scrotal mass, sized 70 × 50 × 30 cm, made it impossible to differentiate the anatomic structures (Fig. 1). The scrotal skin was thickened and edematous while covering the penis. There was no concomitant swelling of the lower extremities. Although the testes and cords were not palpable, they appeared properly in ultrasound and MRI scans.

We performed a multi-stage reconstruction of the scrotum and penis. During the first part, after longitudinal incision of the scrotum and perineum, subtotal scrotectomy was performed (Fig. 2). After this operation a suprapubic cystostomy was made (Fig. 3). Histopathological examination of the excised scrotal tissue showed chronic inflammation with areas of epidermal thickening and dermal fibrosis as well as focal lymphatic and capillary vessel occlusion.

In the second stage (two months later) the scalped penis (Fig. 4) was inoculated under the skin in the suprapubic area and appropriate plastic reconstruction was performed in this region (Fig. 5). We had to reduce the overabundance of subcutaneous tissue under the inoculated penis and cover it by a skin flap in the third part of the operation.

The fourth part of the surgical treatment entailed plastic reconstruction of penis with a skin graft from the suprapubic area (Fig. 6). The wounds healed by primary intention with results initially satisfactory to the patient and staff. The post-scrotoplasty follow-up revealed a further decrease in scrotal size as well as penile elongation with preservation of a normal erection.

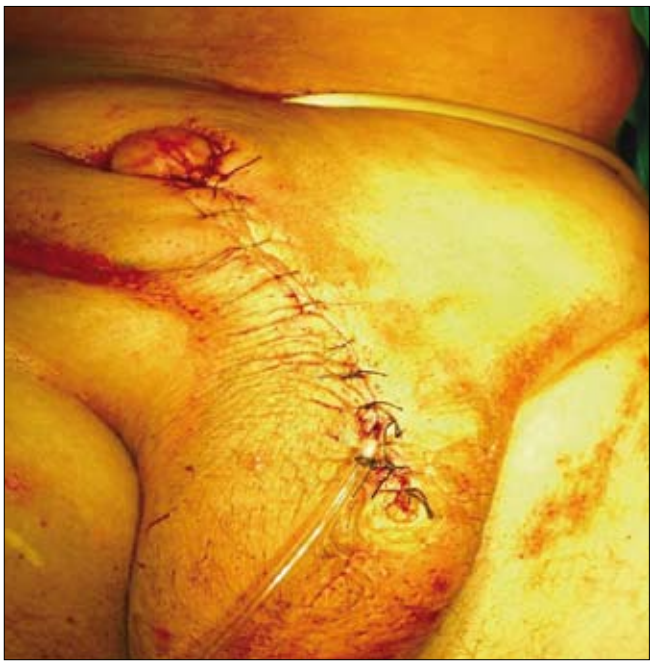

Fig. 5. Inoculated penis after second stage.

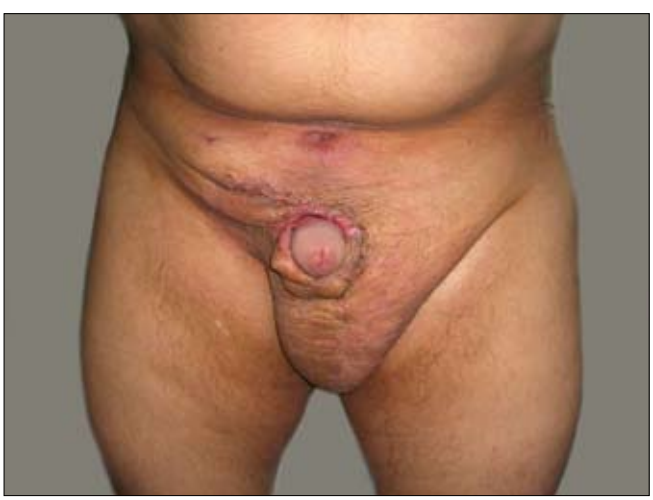

Fig. 6. View after plastic reconstruction of the penis with graft from the suprapubic area.

DISCUSSION

Penile and scrotal lymphedema causes significant functional, cosmetic, and psychological problems. It mostly develops in tropical regions and is a rare condition outside regions endemic for *Chlamydia trachomatis* or *Wuchereria bancrofti*.

Lymphedema has two types: primary and secondary. Primary lymphedema is subdivided into three categories: 1. congenital-inherited (Nonne-Milroy-Meige syndrome), 2. praecox (with early onset), and 3. tarda (with late onset) [9].

Secondary lymphedema could have four origins: 1. occlusive (secondary to neoplasm, radiation, surgical intervention, mechanical trauma, and injection of chemical agents), 2. inflammatory (parasitic, bacterial, and fungal infections), 3. phlebitic, and/or 4. angioneurotic [10].

In our region penile and scrotal lymphedema mostly occurs following an infection or as a reaction to trauma. Idiopathic lymphedema is rarely seen and is caused by a primary obstruction of lymphatic vessels of the scrotum [9].

In extensive disease, complete excision of all elephantoid tissue, preferably saving the penis, spermatic cord, and testes, is appropriate [2, 7, 8]. Lymphangiectomy of the superficial lymphatic network located above the Buck's fascia, which is derived from median raphe and prepuce lymphatics, should be performed in such cases. These lymphatics drain to superficial posterior lymphatic channels. A deeper system is located beneath the Buck's fascia and is drained into deep inguinal lymph nodes [10]. This kind of drainage leads to the success of this surgical method. It is essential to remove involved skin and subcutaneous tissue. Multi-stage reconstruction, chosen by us, guarantees better cosmetic results and allows us to see the effects of each stage after wound healing and plan future steps more adequately. Our treatment returned proper urination and sexual function, significantly improving the patient's quality of life.

CONCLUSION

Scrotal lymphedema in the Western world occurs due to variable origin. We present the patient with scrotal lymphedema of unknown origin and discuss the diagnostic and therapeutic approach. In the case presented here, four-stage extensive excision of elephantoid tissue saving penis, spermatic cord, and testes was performed with adequate cosmetic and functional results.

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