Peno-scrotal elephantiasis: Case report and review of the literature
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ABSTRACT
Elephantiasis results of chronic lymphedema due to a variety of obstructive diseases of the lymphatic system. It’s a rare disease outside areas where filariasis is endemic. Peno-scrotal Elephantiasis is characterized by gross enlargement of the genitalia and legs. Observation: We return a case of a 45-year-old patient referred for a giant scrotum for 10 years. The disease become disabling in that it makes sexual relations difficult, affects urination and also limited the patient mobility. He underwent a complete surgical resection of pathological tissue and peno-scrotal reconstruction, with good functional and aesthetic results. Pathology of the specimen was suggestive of either a non-specific fibrosis. We update through a review of the literature aspects of the diagnostic and therapeutic care of the peno-scrotal elephantiasis.

Introduction
Elephantiasis results of chronic lymphedema due to a variety of obstructive diseases of the lymphatic system. It is characterized by gross enlargement of the genitalia, the legs and the arms. Genital elephantiasis is a common result of filariasis and lymphogranuloma venereum. It’s a rare disease outside areas where filariasis is endemic. However, it may also follow granuloma inguinal, carcinomas, lymph node dissection or irradiation and tuberculosis, although this happens rarely [1]
The histology is essential to establish the diagnosis and refute the others like a malignant tumor which is a real obsession in such clinical presentation. From this observation, we present the clinical and pathological aspects and report our surgical strategy.

Observation
A 45-years-old patient, presented a major chronic lymphedema of the scrotum for 5 years, making both walking and the sexual activity difficult. He also complaints of psychological distress. There is no concept of stay in endemic area filarial. The patient doesn’t have any medical history such as recurrent urethritis, surgery, pelvic radiotherapy, or scrotal trauma. The mass was a reason for many referrals in health facilities and charlatans without improvement.

Physical examination revealed huge scrotum that involved the penis and limb elephantiasis with a pachyderm skin. (Figure 1) The mass was painless and so stony that the patient could seat on it. It was impossible to identify the testicles. The research of microfilaria in the blood and the Chlamydia serology were both negative; the exploration of the vascular axis of the lower limbs was normal, and the abdomino-pelvic CT scan showed a thickened scrotal wall, with no obstructive cause.

Removal of the drain was made on the 25th day. There was no recurrence with a follow up of one year. Scrotum and penis recovered their softness and had a satisfactory appearance, sexual activities resumed.

Figure 1. Pre-Operative aspect
The patient underwent surgery under spinal anesthesia. Excision of all the skin and pathological scrotal envelopes was performed: taking off all its tunics, and sparing healthy tissue scrotal at the root of purse. The affected penile skin was resected half from a ventral incision with urethra-vesical probe in place; the plastic reconstruction used penile and scrotal healthy skin. (Figure 2) The extirpated scrotal mass weighed 9.600kg. (Figure 3) Pathological analysis of the pathological examination of the surgical specimen revealed an edema of the deep dermis, associated with an inflammatory infiltration in the dermis, concluded a non-filarial idiopathic pено-scrotal lymphedema. The postoperative course was uneventful (Figure4).
Lymphedema is generally classified as primary when there is no known etiology, and as secondary when its cause is a known disease [4]. Secondary lymphedema is the most common form and usually the causes involve resection or removal of regional lymph nodes by surgery, radiotherapy, tumor invasion, direct trauma or infectious process. [5]

Regarding infection, it may be secondary to a parasitic disease, lymphatic filariasis, which is the preserve of tropical and subtropical; or a bacterial infection generally Chlamydia trachomatis but it is still potentially curable [6].

Clinically, the hypertrophy and the deformation of the external genital organ are the main manifestations of the peno-scrotal elephantiasis. Besides, the skin becomes thick, cardboard and loses its elasticity. The penis can be completely buried in the scrotum preventing intercourse and sometimes causing urinary tract symptoms [7].

The Doppler ultrasonography can eliminate an obstacle on the vascular axis of lower limbs; in case of filariasis, it shows adult worms moving in the lymphatic vessels. Bipedal lymphography or radiological isotope shows, sometimes pathognomonic aspects of lymphatic filariasis with lymph blockages storied, lymphangiectasia, and glandular hypertrophy with patchy appearance. But there isn’t always a concordance between lymphographic images and surgical exploration. [2]

Ultrasound, CT scan, MRI and found a tumor-induced compression eventual (abdominal or pelvic). When urethral stenosis is suspected it is recommended to use an urethrocytography as it is imperative to treat it the same time as elephantiasis.

In addition, the scrotal MRI is a good alternative: it is a non-invasive method, which helps to clarify the limits of surgical resection. More the latter is limited easier will be the reconstruction of the scrotum.

Inflammatory diseases of the scrotum represent the major part of the differential diagnosis.

A consensus on the treatment strategy was found in the literature: it is to realize the excision of the mass and the extraction of all the skin and subcutaneous tissue reached. Reconstruction of the skin is performed by skin grafts.

Conservative techniques aim to improve the lymphatic drainage: the lymphangioplasty by polyethylene pipes, or lymphatic anastomosis (Nielubowicz operation) between the saphenous vein and the superficial inguinal lympho-nodal group [8], nevertheless the permeability of the anastomosis is always temporary. [9]

Several plastic surgery techniques have been described for reconstruction after radical excision of the scrotum. Indeed, some advocate the use of pedicle flap removed from inguinal or pubic area [6], while others prefer to use free thin skin graft [10]. However, an alteration of the thermal control can be found and then inducing impaired spermatogenesis [3].

In addition, using the cranial-dorsal portion of the scrotum, which is often stored represent a good alternative for some authors. It seems that the functional and aesthetic results are good. [11].

Conclusion

Peno-scrotal Elephantiasis is a rare idiopathic disease usually encountered in wired endemic areas. Because of its weight and volume, it alters the quality of life of patients both physically and psychologically. Pathological examination often found a Benin cells. Filariasis often cited as etiology has been removed by the search for microfilariae that was negative. Surgical treatment consists of removing the scrotal mass with retention of both testicles. Cosmetic and functional results were good.

The authors declare no conflict of interest.

Reference


