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Trauma and reconstruction

Penoscrotal elephantiasis: A severe form of genital lymphedema

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A B S T R A C T

Penoscrotal lymphedema is a manifestation of disrupted lymphatic drainage, causing a significant increase in scrotal volume. impacting both aesthetics and quality of life.

Elephantiasis, classified as stage III by the International Society of Lymphology, represents the advanced stage of scrotal lymphedema, often linked to parasitic diseases.

The diagnosis is clinical, and the treatment involves mass excision. The lymphatic reconstruction is an innovative therapeutic approach that improves post-operative results and better quality of life.

In this case report, we present the case of massive genital elephantiasis managed surgically, and we will focus on surgical techniques and reconstructive approaches.

1. Introduction

Elephantiasis is the most advanced stage of lymphedema and it include large deformities. It is considered as clinical condition where we observe a retention of macromolecules in the interstitial space.

Genital elephantiasis is considered as severe scrotal swelling with the hypertrophy of soft tissue, generally chronic. It significantly affects the patient's quality of life due to cellulitis and fibrosis in soft tissue and the oedematous condition, causing a sexological and psychological impact.

The underlying cause could be primary cause or secondary to a parasitic disease (such as filariasis)[1] or due to intrinsic or extrinsic lymphatic obstruction. The diagnosis is often obvious, based on clinical history and clinical examination.

The management of this pathology depends on the stage of lymphedema, starting with conservative treatment relying on physical and drugs therapy in early stages. At the stage of elephantiasis, the attitude is surgical consisting on resection of affected tissue and lymphatic vessel reconstruction.

Through this case report, we delve into the treatment of genital elephantiasis, placing a particular focus on surgical interventions and reconstructive approaches.

2. Case presentation

A 39-year-old patient with no medical or surgical history except a severe obesity (BMI at 35.2 kg/m^2).

Symptoms began to manifest two years ago, marked by the gradual increase in scrotal volume in the absence of fever and there were no preceding instances of trauma, neoplasms, or travel outside the country. According to the patient, the mass became bothersome over the past year, strongly prompting the medical consultation.

Clinical examination revealed a significantly enlarged scrotum, sparing the penis. However, the testicles were not palpable, and the scrotal tissue was hard and sclerotic, with no signs of inflammation noted (Fig. 1).

The patient then underwent surgical treatment due to the advanced stage of the disease. The surgical approach involved excising all swollen tissues weighting 2 kg (Fig. 2) while identifying and preserving the penis and both testicles, with the possibility of approximating the edges without the need for a skin graft.

The patient showed a good clinical evolution and was maintained in our facility for a week with the application of negative pressure therapy.

The histopathological study revealed a whitish fibrous appearance with a soft consistency upon cutting the skin flap. Under microscopy, the sections showed an oedematous, fibrous connective tissue, containing blood vessels with thickened walls, and certain areas displayed a dense

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Fig. 1. Image showing the aspect of penoscrotal elephantiasis
A: Frontal view
B: Lateral view.

Fig. 2. Image showing the macroscopical aspects of the penoscrotal elephantiasis post excision.

lymphoplasmacytic inflammatory infiltrate (Fig. 3). Malignant tissue or Microfilariae and/or adult worms were not found in the tissue analysis.

The follow-up was maintained to one year, there was no signs of local or distant relapse and characterized by the satisfaction of patient on cosmetic outcome.

3. Discussion

Genital elephantiasis is known as chronic and serious scrotal swelling associated with an hypertrophy of soft tissue. It is considered as a rare condition specially when the patient history reveals no significant risk factors as in our patient beside the obesity. The true incidence of this condition is poorly understood, however, Clinckaert et al. have demonstrated that the risk of occurrence of elephantiasis is important in case of surgery with pelvic lymph node dissection or pelvic radiation for prostate cancer.[1]

The onset of such condition comes from an uncontrolled and untreated lymphedema, leading to the hypertrophy of soft tissues, followed by dermal hyperkeratinization, ultimately progressing to elephantiasis. Such mass poses a significant risk of infections due to mechanical stress and hygiene constraints. Additionally, it significantly impairs the patient's quality of life, affecting even basic aspects such as clothing choices, leading to diminished self-esteem and compromised sexual function.[2]

The elephantiasis or lymphedema stage III can be primary where it is associated to an hypoplastic lymphatic system or secondary to sclerosis or obstruction of lymphatic system, or an infection by Wuchereria Bancrofti which is the most common cause of lymphedema worldwide.[1]

According to the International Society of Lymphology (ISL),

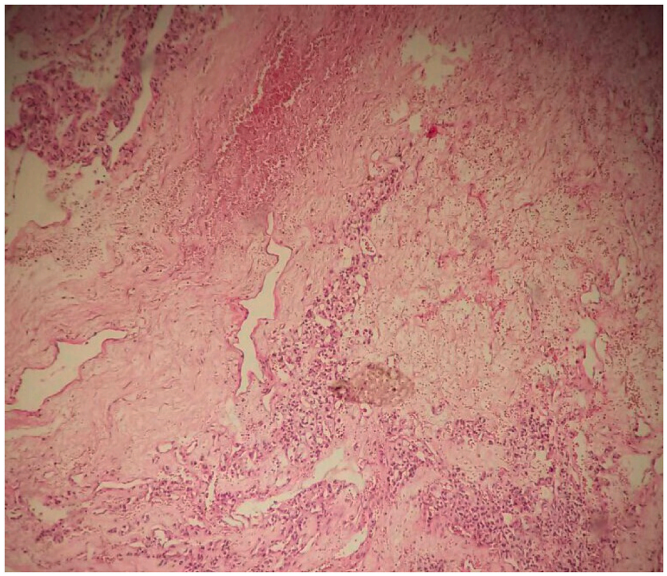

Fig. 3. Histological Image showing edematous fibrous tissue associated to a dense lymphoplasmacytic inflammatory infiltrate.

lymphedema can be classified into four stages reflecting also its progression. Minimal alterations begin to appear with no observable swelling, corresponding to stage 0 or Ia. Subsequently, the appearance of pitting with reversible swelling characterizes stage I. As the swelling

becomes irreversible with evident pitting and the onset of fat and fibrosis deposition, this corresponds to stage II. Finally, the most advanced stage, stage III, corresponds to elephantiasis, involving extensive deformations with thickened skin and increased deposition of fat and fibrosis.[3]

On the other hand, it is crucial to determine the severity of the disease, especially since there is no universally accepted reporting nomenclature assessing it, which will determine the type of lymphatic surgery. There is a classification proposed by Capuano et al.[4] that focuses on hydrocele characteristics based on laterality, size, and the degree of penile burial without specifying the degree of lymphedema. As for the latter, there is a genital lymphedema score [Table 1] based on subjective signs. This score provides a thorough evaluation of genital lymphedema, with results showing a strong correlation with indocyanine green lymphography.[2]

For decades, surgical treatment relied solely on excising lymphedematous tissue. Nowadays, there is a shift towards incorporating lymphatic reconstruction to prevent recurrence and enhance functional outcomes. Yamamoto et al.[5] described a technique of radical resection and reconstruction (RRR) in a single operative session, achieving satisfactory results and improved lymphatic drainage. This technique involves three steps:

- Radical resection of elephantiasis tissue.
- Soft tissue reconstruction using chimeric SCIP flaps.
- Lymphatic reconstruction via the LIFT concept.

The reconstruction according to Yamamoto is based first on identifying lymphatic pathways in both the donor and recipient sites using lymphography. Subcutaneously, 0.2 ml of a lymphatic tracer (Diagnogreen 0.25 %) is injected, and circumferential fluorescent images of lymphatic flow are captured using a portable near-infrared camera system, thereby obtaining a lymphatic map. Then, the skin flap, serving as a graft, is transferred to position the lymphatic vessels contained within the flap to bridge the lymphatic gap at the recipient site, bringing the stumps of the lymphatic vessels as close together as possible. The suturing is performed using Vicryl 3.0.[5]

Through this technique, postoperative complications are less common such as infection, seroma, and recurrence of elephantiasis. Moreover, it enhances the patient's quality of life by improving the genital lymphedema score, leading to the restoration of genital form and function.[5]

4. Conclusion

Lymphedema is classfied into 3 stages with elephantiasis as the most advanced. It is considered as a challenging situation with issues like wound healing and relapsing lymphedema.

Various treatment exists in literature relying on the excision of lymphedematous tissue that might pose for clinicians. However, the emergence of new therapeutics approaches based on lymphatic reconstruction offers better postoperative results and enhance quality of life.

Declarations

Our institution does not require ethical approval for reporting individual cases or case series.

Table 1
Genital lymphedema score (GLS).

Subjective symptoms	No	Yes
Sensation of heaviness	0	1
Sensation of tension	0	1
Swelling	0	1
Urinary troubles	0	2
Cutaneous lymphatic cyst	0	2
Genital lymphorrhea	0	2

Total = GLS Range 0–9.

Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

Availability of data and material

Not applicable.

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Contribution

All the authors have contributed to the production of this book by studying the medical records, and ensuring the interventions, the care discussions as well as the reading of the book after its writing.

CRediT authorship contribution statement

Jihad Lakssir: Writing – original draft, Methodology, Investigation, Data curation. **Youssef Kadouri:** Writing – review & editing, Validation, Conceptualization. **Omar Bellouki:** Writing – review & editing, Methodology. **Ahmed Ibrahimi:** Supervision, Methodology. **Hachem EL-Sayegh:** Supervision, Methodology. **Yassine Nouini:** Supervision, Methodology.

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