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Case report and literature review

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Surgical management of massive penoscrotal lymphedema: Case report and literature review

INTRODUCTION: *Giant lymphedema of the penis and scrotum is a rare and peculiar condition that carries many challanges for both the patient and the physician. The etiology of the disease is often difficult to trace, especially in geographical areas where filariasis is not endemic.*
CASE PRESENTATION: *We reported an advanced case of massive penoscrotal lymphedema presented to our plastic surgery department. Extensive physical examination and imaging investigations allowed for a wide surgical excision of the tumor with the preservation of the penile body and testicles. The reconstruction was performed with adjacent healthy skin flaps and a split-thickness skin graft for the penis. The outcome was a success and the follow-up revealed no recurrences.*
DISCUSSION: *Although the onset of the disease was poorly established, we regarded it as a secondary lymhpedema resulting from chronic local infection. The particularity of this case was the exclusive involvement of the superficial structures, as seen in similar cases from the literature. The clinical presentation explained by the distinctive lymphatic drainage offered a guided and rather secure surgical approach. The reconstruction techniques are varied and should not pose recurrence risks as long as functional tissue is used.*
CONCLUSION: *Massive penoscrotal lymphedema cases display unique evolution and features leading to great impairment. In most instances, surgery is the treatment of choice as the pathological changes are irreversible. Our technique was innovative and comprised similarities and differences compared to other research, nevertheless, the results were a success.*

KEY WORDS: Penoscrotal Lymphedema, Surgical Excision, Reconstruction Techniques

Introduction

Lymphedema is a pathological accumulation of lymph in the interstitial space due to a disruption of the lympatic drainage system. This condition exhibits localized soft-tissue swelling that can lead to serious physical and emotional impariment. Lymphedema can be primary, as a result of congenital malformations, or secondary. Among acquired etiologies, most common are filariasis, cancer treatment complications, recurrent infections and trauma [1-3]. Giant lymphedema of the penis and scrotum is a rare disease in many geographic areas, raising genuine difficulties regarding the diagnosis and treatment.

Case presentation

A 50-year-old male patient with massive penoscrotal lymphedema was admited to the Plastic Surgery Department

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at Emergency County Hospital (Fig. 1). According to the anamnesis and his medical records, it all started at the age of 18 with the slowly progressive enlargement of the external genitalia. Although the cause could not be established, early biopsies revealed lymphedema associated with lymphangioma, which was later identified as lymphangioma circumscriptum. The patient did mention the occurrence of repeated local infections during the onset of the disease, but thorough investigations were not performed.

Furthermore, he did not report relevant comorbidities or surgical interventions. The gradual growth of the organs led to extensive proportions, depriving the patient of a normal lifestyle. In the process, frequent infections arose, worsening the symptoms, while for short periods of time, remissions were obtained with the help of corticosteroid treatment. According to his statement, even with the penis being severely modified, he could still urinate and achieve erection. The impressive size of the scrotum interfered with the sitting position, locomotion and personal hygiene.

On clinical examination, the scortum measured 39x32x21 cm. The penis, 22 cm in length and 26 cm in circumference, was curved and displaced leftward. The glans and the external urethral orifice were buried deep in the thickened penile mass. The skin was covered with papules, ulcers and foul purulent discharge. The adjoining lower abdominal area presented similar changes. The testicles could not be palpated because of the dense consistency of the scortum and the hardened epithelial surface. During the physical examination, the patient experienced no pain and enlarged inguinal lymph nodes could not be identified.

The patient did not suffer from other illnesses and was under no chronic treatments. The laboratory values were in the normal range, except for C-reactive protein which was elevated (70 mg/L). There was no data to suggest an immunocompromised status. The abdominal and pelvic MRI showed pronounced inguinoscortal cellulitis and mild testicular hypotrophy. Aside from these features, the genitalia appeared unaffected. This observation was in accordance with the previously conducted investigations.

The therapeutic options in advanced lymphedema are limited when the alteration of the skin and subcutaneous tissue is irreversible. In these cases, ablative surgery is the best choice for the removal of the lesions [3-5]. This method challenges the ability of the surgeon to precisely separate the tumor from the healthy surroundings without endangering the reproductive organs [3]. Therefore, an urologist also joined the medical team.

Intravenous broad-spectrum antibiotics were given prophylcatically. The patient was positioned supine on the operating table with the thighs slightly flexed and abducted and the knees flexed at 90 degrees. Two incisions were made on the anterior and posterior aspects of the penis to expose the glans. The urethra was secured with

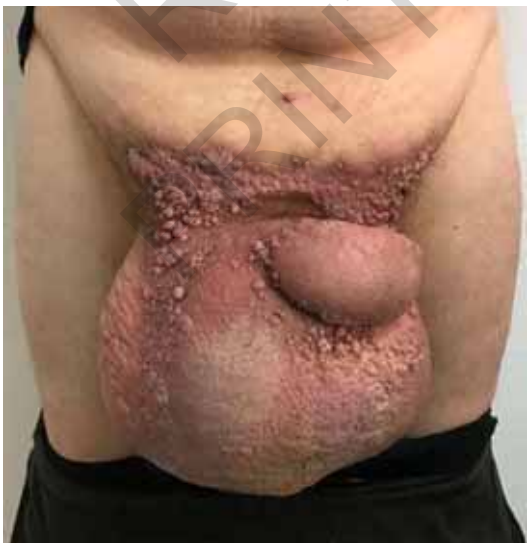

Fig. 1: Preoperative aspect showing giant penoscrotal lymphedema.

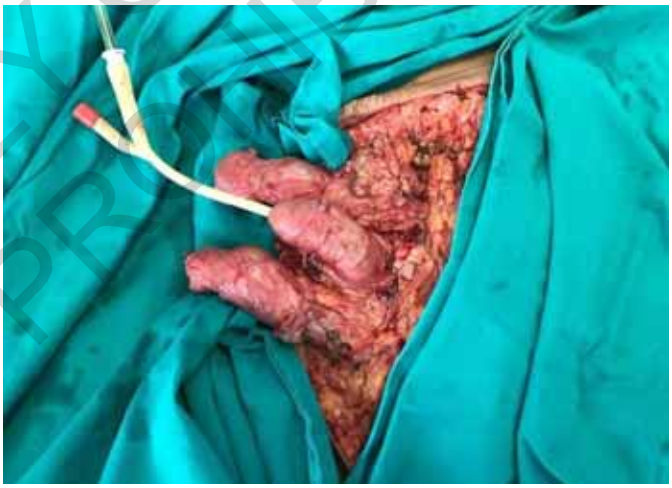

Fig. 2: Intraoperative image after resction of affected penile and scrotal skin.

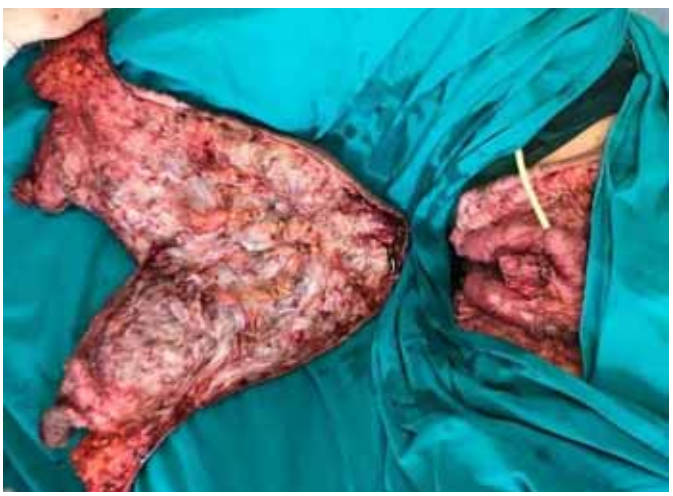

Fig. 3: The degloved penis and scrotum with the resected lesion.

Fig. 4: Immediate postoperative period showing sutures of the flaps and skin graft.

a Foley catheter which was used as a guide throughout the operation [6]. Meticulous dissection of the infiltrated tissues was carried out down to the Buck's fascia in order to delineate the penis circumferentially and the excess was resected. The dorsal incision was extended cranially to remove the modified skin from the lower abdomen. The Scarpa's fascia was preserved. The ventral incision was continued downwards on the middle line of the scrotum. The testicles and the spermatic cords were identified through bilateral progressive dissection. After the complete and safe isolation of these organs was achieved, the incisions were prolonged laterally and towards the perineum to outline the entire lymphedematous mass (Fig. 2).

All the resected specimens weighed 4.8 kg (Fig. 3). The remaining lateral flaps of healthy scrotal skin were used to reconstruct the scrotum. Bilateral orchidopexy was completed to avoid testicular torsion [3].The suprapubic deffect was closed with abdominal advancement flaps. The body of the penis was covered by a split-thickness skin graft harvested from the anterior left thigh. Two drainage tubes were placed in the new scrotal sack and two under the abdominal flaps (Fig. 4).

The operative time was 240 minutes, blood loss 300 ml, there were no intraoperative incidents and the recovery of the patient was optimal.

The hygiene of the genital area was performed daily, with careful handling of the tissues. The paraffin dressing of the skin graft and the urinary catether were maintained for 5 days [3].

Consequently, total graft integration was achieved. The scrotum was initially supported with a cotton bandage roll placed underneath and then with a suspensory jockstrap in an effort to avoid postprocedural swelling. During the first days after surgery, the flaps displayed a slight venous congestion. Thereafter, the overall evolution progressed favorably.

Fig. 5: Late postoperative period - 3 months – showing good skin coverage and no retraction scars.

The abdominal and scortal drains were removed on the 5th and 13th postoperative day respectively. The CRP value continued to decline till the end of the hostpitalization period. The patient was discharged on the 14th day in good general condition, with healed stitches and a significant improvement in locomotion. The follow-ups at 3 and 5 months revealed no recurrences (Fig. 5). The patient was satisfied with the result, both esthetically and funcationally.

Discussion

This is a case of acquired lymphangioma with giant penoscrotal lymphedema. In the literature, and especially in our country, this type of lymphatic condition is a rare entity.

The disease evolved over a period of three decades, in the absence of an obvious cause. Hence, an argument could be made regarding the etiology. From a clinical standpoint, the tumor resembled a masive localised lymphedema, still not all the characteristics were present [8,9]. Surgery, cancer treatments or parasitic infestation could be easily rulled out as a possible source, based on the medical history of the patient. The chronic local infections at the time of debut, although inadequately investigated, could have determined the initial symptoms and lymphatic obstruction.

Hidradenitis supurativa is mentioned in numerous studies as the starting point for lymphatic drainage disorders in the genital area [10-13]. Lymphangioma circumscriptum, identified together with lymphedema on the histopathological examinations, is a benign tumor that affects the superficial lymphatic vessels of the skin[17]. These dermal and subcutaneous channels become dilated, they accumulate lymph and promote the appearance of various cutaneous lesions [17]. Generally, lymphan-

giomas are congenital malformations and are diagnosed in children less than 2 years of age [14]. Nonetheless, lymphangiectasias, the acquired type of lymphagioma [15,16], can be the consequence of recurrent infections and lymphatic stasis. Chronic lymphedema can further cause a disruption in the lymphatic drainage process [21].
Therefore, lymphedema and lymphangioma not only coexist, they also sustain each other [15]. The persistent infections contribute to this vicious circle, as the probable etiology and yet a repercussion of the deficient lymphatic system [18,19]. These arguments could explain the relentless growth of the tumor that led to the given clinical presentaion.
Without the adequate treatment, the continuous swelling of the genitals turned into lymphosclerosis. While the subcutaneous tissue developed cellulitis and fibrosis, the skin became hyperkeratotic, covered with warts and infected ulcers.
The peculiarity of this disease is the irreversible character of the lesions and the confinement of the changes to a restricted region, limited in both surface and depth. The pattern of defined spreading is due to the ways in which the lymph is drained from this specific segment of the body. The lymphatic drainage of the penis and scrotum is completed by the means of two separate systems [2,4,7].
The superficial chain of inguinal nodes drains the skin and the subcutaneous tissue, whereas the deep pelvic nodes drain the testis and the body of the penis through a network located deep to the Buck`s fascia [2,4,7]. This particular involvement of the structures, in which the deep organs remain unaffected, is described in many articles and offers favorable treatment options [1-6].
Ablative surgery is the recommended choice in advanced cases [3-5]. Clinical therapies and functional anastomosis are inefficient when there is major stasis and fibrosis present [3-5]. The incisions at the transition to unaltered skin and the dissection with respect to the deep fascia helped guide a clean resection of the tumor.
The preservation of the deep lymphatic network and the coverage using healthy flaps prevented subsequent drainage issues and lowered the chances of recurrence. There are different reconstruction techniques, especially concerning the penis, however, the long-term results are similar [1-6,21,22]. Although we did not apply the skin graft in an oblique fashion [6] or used the zigzag suture (Z-plasty) [3], contracture scars did not occur.

Conclusion

Massive penoscrotal lymphedema is an unusual condition that causes great functional impairment, depriving the patient of a normal lifestyle. The etiology is frequently hard to identify and the treatment options must be carefully evaluated. The lymphatic tumor surgery and the reconstruction techniques are diverse. In our case, we based the therapeutic decision not only on the clinical presentation and medical history of the patient, but also on the review of the literature. The cosmetic and functional outcomes, without any recurrences, contributed to an improvement in the quality of life and to the social and professional reinseriton.

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