Case - Surgical treatment of penoscrotal edema in a patient with lymphedema-distichiasis syndrome

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Cite as: Neville K, MacGilivray M, Hoare D. Case - Surgical treatment of penoscrotal edema in a patient with lymphedema-distichiasis syndrome. *Can Urol Assoc J* 2026 July 20; Epub ahead of print. http://dx.doi.org/10.5489/cuaj.9673

Published online July 20, 2026

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INTRODUCTION

KEY MESSAGES

- Scrotal lymphedema results either from primary lymphatic abnormalities, which may be genetic, or from secondary damage or obstruction of lymphatic vessels, most commonly due to infection, surgery, cancer, radiation, trauma, or chronic inflammation.
- Genital lymphedema can cause voiding dysfunction, recurrent infection, and impaired QoL, particularly in patients with primary lymphatic disorders.
- Extensive penoscrotal lymphedema refractory to conservative therapy can be effectively managed with complete excision and tailored reconstructive techniques, resulting in sustained functional improvement and reduced infectious morbidity.

Lymphedema is a disorder of impaired lymphatic drainage resulting in progressive interstitial fluid accumulation, tissue hypertrophy, fibrosis, and recurrent infection. It is classified as primary lymphedema or secondary lymphedema. Primary lymphedema is caused by agenesis or congenital malformation of the lymphatic ducts, and it can be genetic in nature. Secondary lymphedema occurs following lymphatic injury due to malignancy, surgery, radiation, trauma, or infection, or chronic inflammation.[1,2] A common cause of chronic inflammation leading to secondary lymphedema is hidradenitis suppurativa.[3] Although secondary lymphedema is more commonly encountered in urologic practice, primary forms present unique diagnostic and management challenges.

Lymphedema-Distichiasis syndrome (LDS), is a rare autosomal dominant cause of primary lymphedema, typically caused by pathogenic variants in the *FOXC2* gene.[4,5] LDS is characterized by pubertal-onset lower-limb lymphedema and distichiasis (extra row of

eyelashes), with variable involvement of the external genitalia and other systemic features.[4,6] Progressive lymphatic dysfunction can lead to recurrent cellulitis, skin breakdown, lymphorrhea, functional impairment, and significant psychosocial distress.

In men, involvement of the external genitalia can result in severe penoscrotal lymphedema, causing difficulty with voiding, hygiene, sexual function, and mobility, and is often refractory to conservative measures.[7] Surgical excision and reconstruction can provide meaningful functional improvement; however, reports describing operative management of genital lymphedema related to LDS are scarce. We present a case of severe penoscrotal lymphedema in a patient with LDS, highlighting surgical management and reconstructive considerations relevant to urologic practice.

CASE REPORT

A 66-year-old male was referred to urology with severe, progressive penoscrotal lymphedema resulting in recurrent infections, difficulty with voiding, and impaired hygiene. He had a longstanding history of primary lymphedema, with onset at puberty, initially involving the right lower extremity, followed by bilateral lower limb involvement. Over subsequent decades, the lymphedema progressed proximally and was complicated by increasingly frequent episodes of cellulitis requiring hospitalization. By his late 50s, genital involvement became prominent, with progressive enlargement of the penis and scrotum, recurrent skin breakdown, lymphorrhea, and recurrent infections despite chronic suppressive antibiotic therapy.

The patient had a family history of lymphedema affecting multiple first-degree relatives across generations. Genetic testing confirmed a *FOXC2* gene mutation, consistent with LDS. His medical history was notable for B-cell non-Hodgkin lymphoma diagnosed in 2023 and treated with R-CHOP chemotherapy, adrenal insufficiency secondary to prolonged corticosteroid exposure, hypothyroidism, dyslipidemia, gastroesophageal reflux disease, and recurrent cellulitis.

In September 2023, urology consultation was required for difficult urethral catheterization. Marked foreskin redundancy, significant penile edema, and near-obscuration of the urethral meatus necessitated cystoscopic guidance for catheter placement.

Conservative measures failed to provide meaningful symptomatic relief, and the patient continued to experience recurrent infections and functional impairment. Following multidisciplinary discussion, the patient elected to proceed with surgical resection of penoscrotal lymphedema with reconstruction in September 2025.

Surgical technique

Surgery was performed under general anesthesia with prophylactic cefazolin. The patient was positioned in dorsal lithotomy and prepped and draped in the usual sterile fashion. Initial inspection demonstrated extensive lymphedematous involvement of the penile shaft and scrotum with associated changes of papillomatosis and hyperkeratosis consistent with severe, chronic disease (Figure 1).

Care was taken to identify and preserve the glans penis, which appeared relatively spared. Circumcision was performed and sufficient glans cuff was preserved. The specimen was sent for pathological analysis. A 16-French silicone urethral catheter was placed without difficulty. The lymphedematous penile and scrotal skin was circumferentially delineated, and the outer margins were scored using electrocautery. Dissection of the diseased tissue was performed using a combination of electrocautery and LigaSure, with careful preservation of the spermatic cords and testes. Additional resections of deeper scrotal tissue were required to achieve complete excision of lymphedematous tissue. Bilateral hydroceles were drained, revealing atrophic but viable testes. The entire scrotum, including tunica vaginalis, was excised to minimize risk of recurrence and to prevent postoperative hydrocele formation. The resected tissue was sent for pathological examination (Figure 2).

Following complete resection, reconstruction was planned. Bilateral perineal advancement flaps were raised and allowed primary closure of the perineal defect using interrupted absorbable sutures. Both testes were pexied inferior to the penile base, and the remaining native skin edges were secured to the penile base.

The total defect was then measured. A split-thickness skin graft measuring approximately 10 × 18 cm was harvested from the left thigh using a dermatome. A 6 cm segment of the graft was tailored for penile shaft reconstruction and circumferentially wrapped around the shaft, secured with interrupted, quilting 5-0 polyglactin 910. Moderate suprapubic lymphatic fluid drainage was noted intraoperatively. The remaining graft was meshed at a 1:1.5 ratio and used to reconstruct the neoscrotum, fixated using 5-0 polyglactin 910 along the periphery of the wound and to quilt the graft to the underlying testes (Figure 3).

Bolster dressings were applied to both the penile and scrotal grafts using non-adherent dressings, mineral oil–soaked cotton pads, and compressive gauze wraps, secured in place with chromic sutures. A penoscrotal nerve block was administered at the conclusion of the procedure. The patient tolerated the procedure well, with no intraoperative complications.

Postoperative

The skin graft was secured with the bolster dressing for five days to avoid trauma. The catheter was maintained for the same period, and the nursing team performed daily hygiene of the genital area, avoiding manipulation by the patient. The bolster dressings were removed on the 5th postoperative day, with near-total graft integration (Figure 4). The patient was discharged on the 6th postoperative day. He was seen in follow up 3 months post-operatively with satisfactory aesthetic result and no recurrence of lymphedema (Figure 4). His erectile function remained impaired, likely secondary to previous oncologic treatments, but voiding function improved substantially with no recurrent infections or concerns regarding urinary retention.

Histopathological examination of the foreskin and excised penoscrotal tissue demonstrated changes consistent with chronic lymphedema. Sections showed subepidermal dilated thin-walled vascular channels with associated stromal edema, fibrosis, and chronic inflammation. The surface epithelium exhibited mild hyperkeratosis, acanthosis, and

papillomatosis with reactive changes. No evidence of in situ or invasive carcinoma was identified. The findings were reviewed at departmental quality assurance rounds and were felt to be concordant with the clinical diagnosis of penoscrotal lymphedema.

DISCUSSION

Advanced penoscrotal lymphedema represents a challenging reconstructive problem due to distorted anatomy, chronic inflammation, and a high risk of wound complications. In this case, longstanding primary lymphedema associated with Lymphedema-Distichiasis syndrome progressed over decades to involve the external genitalia, resulting in recurrent infection, lymphorrhea, and significant functional impairment. Surgical intervention was pursued after failure of conservative measures and recurrent hospitalizations, reflecting a common trajectory in advanced disease.

Once genital lymphedema reaches an advanced stage, excisional surgery with reconstruction remains the most effective treatment strategy. Systematic reviews and case series demonstrate that complete resection of lymphedematous tissue provides durable symptom relief, improved function, and reduced infection rates, particularly when conservative or microsurgical lymphatic procedures are no longer feasible due to extensive fibrosis.[8,9] This approach was chosen in the present case given the chronicity of disease and recurrent cellulitis.

Several operative principles illustrated here are supported by the existing literature. Preservation of the glans penis, which is often relatively spared, allows for reliable anchoring of penile reconstruction and improved cosmetic outcomes. Aggressive excision of diseased tissue, including deeper scrotal components, is emphasized to minimize recurrence, while preservation and fixation of the testes enables functional neoscrotal reconstruction.[9] Reconstruction using a combination of local advancement flaps and split-thickness skin grafts is widely described and offers adaptable coverage for large defects, with favorable graft take and long-term durability.[10] Emerging reconstructive techniques incorporating lymphatic-preserving or lymphatic-transfer flaps have shown promise in selected patients with genital lymphedema; however, these methods are technically demanding and may be less suitable in cases of extensive longstanding disease.[11]

CONCLUSIONS

Genital lymphedema, although uncommon, represents an important and often underrecognized source of urologic morbidity, with significant implications for voiding function, hygiene, infection risk, and quality of life. This case highlights a rare cause of progressive genital lymphedema and details the surgical approach which resulted in meaningful and sustained cosmetic and functional improvement.

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FIGURES AND TABLES

Figure 1. Preoperative views of penoscrotal lymphedema.

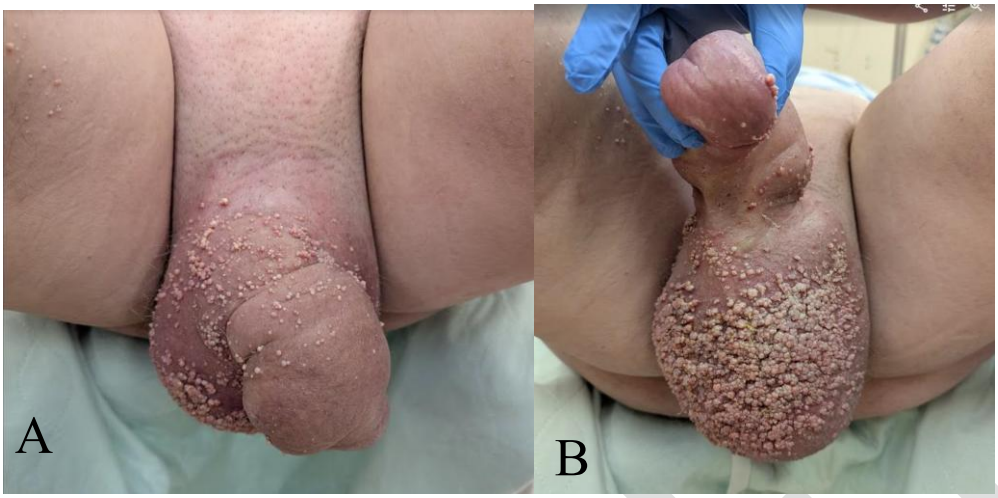


Figure 2. The resected penoscrotal lymphedema tissue.

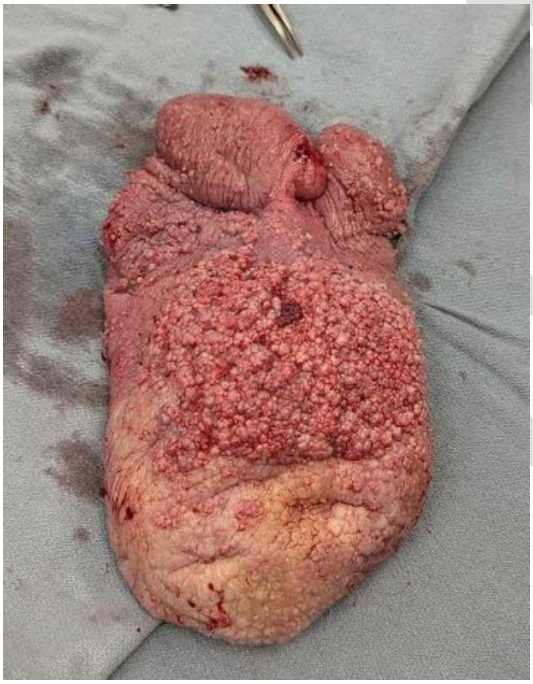


Figure 3. Intraoperative view of penile shaft and scrotal skin grafts.

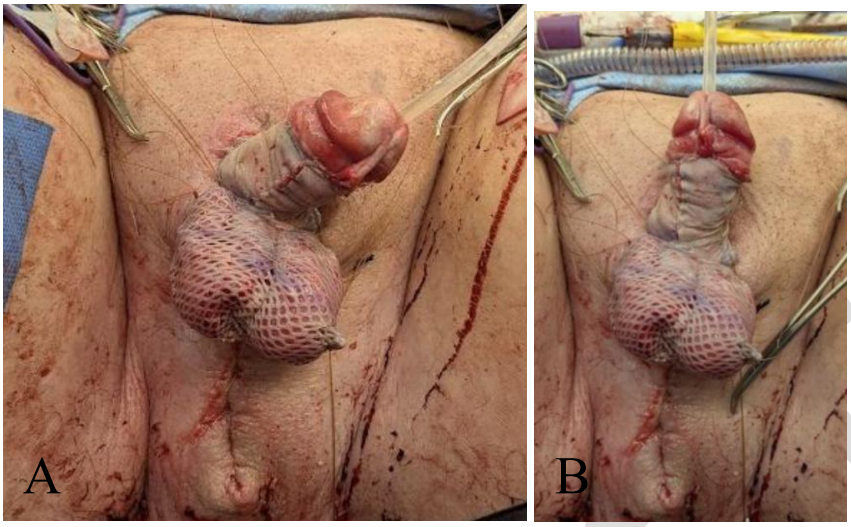
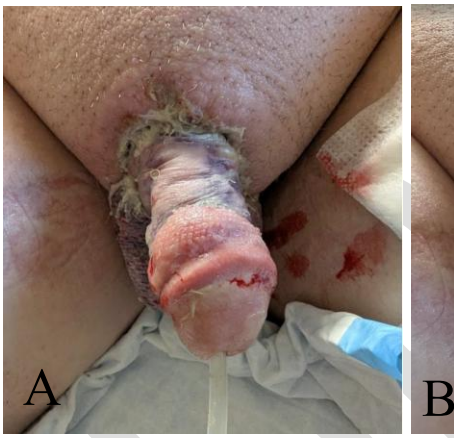
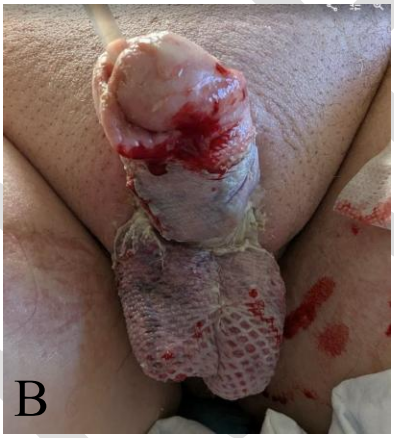


Figure 4. (A-C) Results 5 days postoperative. (D-E) Results at 3-month followup.





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