CASE REPORT

Surgical management of chronic genital lymphoedema

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SUMMARY

A 44-year-old man with a spinal cord injury was referred to a specialist urology service with a 7-year history of significant genital swelling. His condition had eluded diagnosis and was refractory to all previous treatments. The considerable swelling both impacted his quality of life and prevented the patient from adequately managing his neurogenic bladder. He was diagnosed with chronic idiopathic genital lymphoedema and underwent total scrotoectomy, wide penile skin excision and split skin graft to the penile shaft. The patient made an excellent recovery. We present this unusual case with preoperative, intraoperative and postoperative images.

BACKGROUND

Genital lymphoedema is the abnormal retention of lymphatic fluid in the subcutaneous genital tissues. It is caused by lymphatic obstruction. In <20% of cases, the disease is idiopathic, due to the development of abnormal lymph networks.¹ However, a secondary cause is usually identified, such as surgical or other trauma, tumour infiltration, parasitic infection, radiotherapy or sexually transmitted infections. Less common aetiologies include sarcoidosis, vasculitis, Crohn's disease and peritoneal dialysis. Genital lymphoedema can affect men or women. Henceforth, this paper will discuss only male genital lymphoedema. This can involve the scrotum, penis or both. The lower limbs may also be affected. The genitals drain to the bilateral inguinal nodal beds, so both must be altered to create lymphoedema.

If left untreated, recurrent cellulitis leads to connective tissue collagenous hyperplasia and fibrosis and loss of elastic fibres. These irreversible microscopic changes cause permanent swelling, with debilitating loss of function and appearance. Management goals in genital lymphoedema are to obtain satisfactory voiding, sexual and cosmetic outcomes. Treatment consists of correcting the precipitating cause and trial of conservative mechanical and antibiotic therapies. Failing these, surgery may be required.

In this case of idiopathic genital lymphoedema, the patient went untreated for several years due to failure to recognise and refer the condition. At first urological review, the patient had advanced disease with skin changes that made daily activities, bladder management and sexual life difficult. Due to the delay in presentation, surgical management was unavoidable. We present this case to increase recognition of genital lymphoedema.

CASE PRESENTATION

A 44-year-old man with a complete T7 level spinal cord injury sustained 23 years prior, presented with a 7-year history of progressive scrotal swelling. This was complicating his ability to transfer from bed to chair, made bladder management difficult and prevented him from driving and precluded him from sex as his penis was buried in the swelling. The patient reported that following a case of bacterial epididymitis that was related to a urinary catheter, that his scrotum began to enlarge progressively. He denied any surgery, venereal infections, foreign travel or other known causes of genital lymphoedema.

After his injury, the patient's initially performed intermittent self-catheterisation (ISC). However, the progressive genital lymphoedema eventually made this management of his neuropathic bladder impossible. The patient then resorted to attaching a condom catheter to the gradually disappearing penile shaft. Ultimately, this too failed, and an indwelling urethral catheter had to be changed via flexible cystoscopy.

On examination, the patient had a swollen penis and scrotum. The genital skin was hyperpigmented, with giant rugae. The penis was entirely buried and entirely obscured by the scrotal tissue (figure 1). The scrotal had diameter 30 cm. On palpation, the swelling was non-tender, non-pitting and non-reducible. The skin was extremely thickened and did not transilluminate. The testicles were undetectable, and the glans of the penis was only palpable by introducing a gloved finger into the fold of skin from which urine emerged.

INVESTIGATIONS

Full blood count, serum electrolytes and liver function tests were normal. A culture of genital skin swabs and catheter urine specimens detected only mixed growth. On scrotal ultrasound, both testes were of standard size and echogenicity. There were no hydroceles. The scrotal skin thickness was 2–3 cm.

Since the patient had no history of foreign travel to areas where *Chlamydia trachomatis* or *Wuchereria bancrofti* are endemic, serological testing was not performed. As the patient was currently not sexually active and never had a history of risky sexual encounters, testing for venereal disease was also not performed.

DIFFERENTIAL DIAGNOSIS

By exclusion, the working diagnosis was of idiopathic genital lymphoedema, exacerbated by the

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Figure 1 Preoperatively; the scrotum is massively enlarged and the penis not visible. The adjacent surgical forceps are 10 cm in length.

patient's sedentary lifestyle due to his spinal cord injury. Parasitic and venereal infective causes, as well as malignancy and rarer causes, were thought unlikely.

TREATMENT

Given the significant impact and irreversibility of this patient's lymphoedema, it was clear that the patient would not benefit from conservative management. He underwent testes-sparing total scrotoectomy. Dissection was meticulous to preserve the testes, spermatic cord, urethra and penile shaft. [Figure 2](#) shows the degloved penis with catheter in situ. The spermatic cord is visible bilaterally, and the testes are wrapped in a warm saline soaked pack for protection during the operation. The excised tissue is shown in [figure 3](#).

Bilateral orchidopexy was performed to prevent later testicular torsion, and his scrotum was reconstructed with perineal skin spared from the disease process ([figure 3](#)). A split skin graft was harvested from his left anterior thigh, quilted onto the penile shaft and a compressive dressing applied ([figure 4](#)).

Dressings were removed after 10 days. A urinary catheter was left in place for 3 weeks. Erections did not need to be inhibited with cyproterone acetate as the patient required medical assistance for sexual intercourse.

Specimen histology revealed skin tissue with extensive papillomatosis. There was also dense lymphoplasmocytic infiltration and dense fibrocollagenous tissue with dilated lymphatic channels of the dermis. All findings were consistent with chronic lymphoedema. There was no evidence of parasites, tumour, vasculitis or granuloma.

Figure 2 Intraoperatively; total scrotoectomy and wide excision of penile skin has been performed. The spermatic cords are visible, with the testis retracted superiorly under warm saline soaked gauze.

OUTCOME AND FOLLOW-UP

Two weeks postoperatively, the patient enjoyed a dramatic improvement in his quality of life. He now experienced no difficulty transferring or driving, had resumed performing ISC and was planning on resuming an active sex life. [Figure 5](#) shows the surgical site just prior to the urethral catheter was removed, 3 weeks post-operatively.

Figure 3 Intraoperatively; the excised penoscrotal specimen.

Figure 4 Intraoperatively; after penile split skin grafting was performed, compressive dressings are applied. The defect in the perineum is easily closed to give an excellent cosmetic result.

DISCUSSION

Genital lymphoedema is a rare condition, with the literature currently limited to case reports and small case series.¹⁻³ The most common aetiologies in the Western and developing world are elective inguinal node dissection and infection, respectively.^{1,4} The condition can be extremely debilitating and left untreated will require surgery. The critical step in the management of genital lymphoedema is early diagnosis and intervention before genital skin changes become irreversible. The initial management of the lymphoedema is conservative with compression and elevation of the scrotum and administration of antibiotics to prevent cellulitis.¹

Extensive attempts must be made to elucidate a secondary and potentially reversible cause. After careful history and examination, patients should undergo standard blood tests, then the selective use of venereal and parasitic infection screening and imaging. The latter may include scrotal ultrasound or CT of the abdomen and pelvis, especially if malignancy is suspected. Further tests would be directed by associated symptoms. Sexually transmitted infections should be treated with antibiotics, parasitic infections with antifilarial medications and inflammatory diseases with steroids.¹ Should these measures fail, more aggressive therapy is warranted, to prevent or treat the subsequent pendulous, heavy, cumbersome sequelae.

There are two surgical options described in the literature for the treatment of chronic genital lymphoedema; lymphangioplasty and lymphangiectomy with reconstructive surgery.^{5,6} Lymphangioplasty, the establishment of new lymphatic drainage patterns from the tissue, is technically demanding and results

Figure 5 Three weeks postoperatively; the split skin graft remains viable and the penis accessible. The inconvenient scrotal bulk is gone.

have been disappointing. It has been abandoned as a reasonable treatment for chronic genital lymphoedema.^{1,7}

Lymphangiectomy with reconstructive surgery is the current favoured treatment. This involves resection of the affected tissue and closure either primarily or with skin grafts. Due to their independent lymphatic drainage, the testes, spermatic cord and penis are unaffected by lymphoedema. Genital lymphoedema recurrence requiring reoperation is much less common with total rather than partial scrotoectomy.¹ Since total scrotoectomy and reconstruction yields excellent cosmetic results, it should be

Patient's perspective

'This operation has changed my life. It has given me back my independence. I will be able to drive again, manage my bladder better and have a sex life. It has not been easy, but was totally worth it'.

Learning points

- ▶ The common causes of genital lymphoedema are a tumour, surgical trauma, infection or idiopathic.
- ▶ Early intervention prevents irreversible disease progression.
- ▶ Conservative management is compression, elevation and antibiotics.
- ▶ If this fails, surgery is required, with total scrotoectomy and skin grafting.

considered the standard of care. Managing penoscrotal lymphoedema, surgeons may elect to defer penile lymphangiectomy and perform only total scrotoectomy, as in some cases the penile lymphoedema will subsequently resolve.¹ However, if the penile skin is chronically avulsed off the penile shaft by the weight of the affected scrotum, as in our case, full excision and skin grafting are required.

Contributors DBH and JC conceived the idea of the case report. DBH and JC collected the intraoperative clinical images. DBH wrote the manuscript with support from NK. NK and JC refined the final script. JC supervised the project. All authors discussed the paper and agreed to be accountable for all aspects of the work.

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