



Leukocytoclastic vasculitis leading to penile necrosis and ischemia of the bladder and urethra

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ABSTRACT

We report a case of leukocytoclastic vasculitis-induced penile necrosis in a 69-year-old male with no previous urologic history. After suffering an ischemic event to the distal shaft of the penis as well as an ischemic event involving the right side of the bladder, he underwent suprapubic tube placement for urinary diversion. Despite attempts at anticoagulation for penile salvage, he lost the distal aspect of the glans penis to auto-necrosis. Ultimately, the underlying disease was determined to be systemic ANCA-associated vasculitis, and the patient was treated with rituximab and prednisone as well as penile wound debridement.

1. Introduction

Leukocytoclastic vasculitis (LCV) is a common form of small vessel vasculitis with histopathology demonstrating neutrophil-predominant inflammation, fibrinoid necrosis, and leukocytoclasia.¹ LCV can be caused by infections, paraneoplastic conditions, or systemic diseases.¹ The leading clinical presentation of LCV is palpable purpura, but frank necrosis, including penile necrosis, has been reported.¹ Here we report a complex case of LCV-associated penile necrosis associated with segmental bladder ischemia.

2. Case report

A 70-year-old male (Patient "M") having history of hypertension, hyperlipidemia, segmental colitis associated with diverticula (SCAD), and unprovoked extensive bilateral lower extremity DVT and PE six months prior, with a recent diagnosis of leukoclastic vasculitis (LCV), presented to the emergency department with two days of penile pain, swelling, and discoloration. On anticoagulation for the past six months, this was held two days prior to presentation for upcoming endoscopy. He had been diagnosed with LCV from a skin biopsy after developing spontaneous large bilateral flank hematomas, thought to be warfarin-induced, and thus had been transitioned to apixaban.

Physical exam was notable for ischemic, cold glans tissue with demarcation at the distal shaft commensurate with his circumcision

scar. The shaft was edematous, but the corpora were palpably soft (Fig. 1). A penile blood gas showed pH 6.88 and pO₂ < 30. Despite the ischemic blood gas, history and physical examination were not consistent with priapism; thus, no further drainage was performed.

Labs demonstrated a leukocytosis to 19.8, hemoglobin 9.4, and acute kidney injury (AKI) with creatinine 2.38. Urinalysis demonstrated >180 RBCs. CT of the abdomen and pelvis revealed bilateral corpora cavernosum were expanded and hypodense, with inflammatory change along the corpora into the preperitoneal space. CT cystogram was performed due to history of gross hematuria and stranding around the bladder which showed no evidence of bladder perforation.

Hematology and Rheumatology initiated heparin high intensity protocol with further workup over concern for thrombotic event because of held anticoagulation. He was started on high dose steroidal therapy due to concern for a vasculitic process. Vascular imaging was considered but unfortunately, due to his AKI, he could not receive intravenous contrast. A foley catheter was placed for initial urinary drainage.

Mild improvement in skin discoloration after initiation of the above therapy was noted (Fig. 2). Due to significant discomfort from the foley catheter, options for alternative urinary diversion were discussed with the patient, who opted for suprapubic tube (SPT) placement. Cystourethroscopy during SPT placement revealed evidence of vascular congestion/necrotic change of the bulbar urethra and devitalization of the right lateral bladder wall, causing concern for bladder infarction.

The diagnosis was ANCA-associated leukocytoclastic vasculitis

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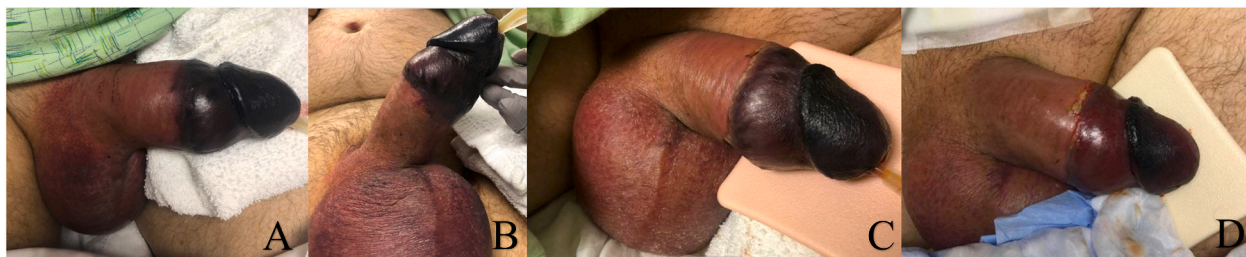


Fig. 1. a and b. Penis at time of presentation to ER. c and d. Improvement in ischemia on HD4 after initiation of heparin and steroids.



Fig. 2. Autoamputation of the penile glans.

which was treated with rituximab and prednisone and transition back to apixaban. On hospital day sixteen, he was discharged.

Followed closely by urology as an outpatient, approximately two months later M's glans was noted to have autoamputated (Fig. 3). His penile wound received local care and clobetasol cream to allow healing

via secondary intention. Cystoscopy three months post-injury was notable for significant debris but showed healthy and well-perfused bladder and urethral mucosa. Urodynamics revealed small bladder capacity at 163 ccs and hypoactive detrusor muscle with max pdet of 33 cm H2O. When his SPT was removed, he was voiding on his own 18 months post-injury with improving frequency and urge incontinence. He continued pelvic floor physical therapy and anticholinergic medication.

3. Discussion

This case highlights the importance of care coordination between specialties for complex presentations. M's medical history contained features concerning for numerous reported causes of penile necrosis. For example, isolated gangrene of the penis has been reported in patients with ESRD as a localized manifestation of vascular calcification.² Based on M's proteinuria and acute kidney injury, calciphylaxis in the setting of ESRD warranted consideration; however, the workup was negative. Hematology investigated a potential underlying hypercoagulable or paraneoplastic process. Thrombus was not noted on imaging; tissue biopsies, chest and abdominal imaging, and colonoscopy results revealed no concerns for malignancy.

The edematous shaft made ischemic priapism an immediate concern, requiring urology consultation. Anticoagulant use is a known risk factor for ischemic priapism, and penile blood gas analysis revealed oxygen deprivation.³ However, the corpora appeared soft on physical examination, so priapism treatment was not pursued.

The possibility of Fournier's gangrene also required immediate attention; treatment delay is associated with up to 90% lethality.⁴ In Fournier's gangrene, bacteremia of the urogenital tract, anorectal area, or genital skin initiates a cytokine cascade, leading to endothelial damage and subsequent activation of the coagulation cascade with inhibition of fibrinolysis and disseminated microthrombosis of vessels



Fig. 3. Admission CT abdomen and pelvis.

feeding the fascia. With inflammation, these factors produce ischemic necrosis of the fascia. Local manifestations begin with ulceration of the glans, prepuce, skin of the penis, or scrotum, progressing to tissue necrosis within hours, sometimes leading to sepsis, multi-organ failure, and death. Despite the penile ischemia and necrosis, our patient was alert and oriented, demonstrating no signs of altered mental status, making Fournier's highly unlikely.⁴

Per Rheumatology, due to M's atypical p-ANCA of 1:320, positive PR3 of 1.7, and biopsy proven LCV, ANCA-associated vasculitis (AAV) was a potential cause of his presentation. Prior case studies described LCV-associated penile necrosis with underlying AAV, as well as several other LCV-associated systemic conditions, including Crohn's Disease (CrD), Henoch-Schönlein purpura (HSP), cryoglobulinemia, polyarteritis nodosa (PAN), antiphospholipid syndrome (APLS), and Behcet's Disease (BD).^{2,5}

Considering M's biopsy-proven LCV, recurrent clots, positive atypical p-ANCA and PR3, and the involvement of multiple vascular territory infarcts, ANCA-associated LCV was concluded as the most likely cause of M's presentation. The high level of collaboration between our medical and surgical teams optimized the care M received at our institution.

4. Conclusion

Although penile necrosis is rare due to the rich circulation network of the perineum and lower abdomen, a wide range of causes have been reported, including ischemic priapism and Fournier's gangrene in

addition to numerous paraneoplastic and systemic conditions related to LCV. In patients suffering from penile necrosis, surgical management must be combined with multidisciplinary evaluation to determine the best course of treatment.

Consent

Written consent was obtained from the patient for use of medical health information and pictures.

Declaration of competing interest

The authors of this article have no conflicts of interest to disclose.

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