CASE REPORT

Penile necrosis secondary to purpura fulminans: a case report and review of literature

David B. Hogarth1,*, Paul M. Cheon2,†, Javeed Kassam3, Alexander E. Seal4, and Alexander G. Kavanagh1

1Department of Urologic Sciences, University of British Columbia, Vancouver, Canada V5Z 1M9, 2Faculty of Medicine, University of Toronto, Toronto, Canada M5S 1A8, 3Department of Neuroscience, University of Southern California, Los Angeles 90007, USA, and 4Division of Plastic Surgery, Department of Surgery, University of British Columbia, Vancouver, Canada V5Z 1M9

*Correspondence address. Tel: +1-604-875-4301; Fax: +1-604-875-4637; E-mail: dhogarth@alumni.ubc.ca

INTRODUCTION

Purpura fulminans is a hematological condition which involves both disseminated intravascular coagulation (DIC) and skin necrosis [1]. There have been many reports of limb amputations as a result of purpura fulminans, but no such report for the involvement of the penis has been made [2-4]. We highlight a case of a 60-year-old male patient who presented with purpura fulminans secondary to idiopathic protein S deficiency, and subsequently underwent a partial glansectomy for necrosis of the penis.

CASE REPORT

A 60-year-old male presented to the emergency department with pre-syncope and general malaise, following a 6-day history of an enlarging erythematous lesion over his right hip. Past medical history was significant for a provoked posterior tibial deep vein thrombosis (DVT) 16 months prior treated with 6 months of warfarin, C3-C6 spinal fusion due to C4 ASIA D central spinal cord injury 22 months prior with significant neurologic recovery, and circumcision as a child.

DIC was diagnosed on the basis of clinical findings suggestive of coagulopathy plus abnormal coagulation parameters including a fibrinogen level <0.7 g/L and an INR in excess of 10. The patient’s hemoglobin was 64 g/L from a previously normal baseline, with associated thrombocytopenia. Multiple transfusions of packed red blood cells, platelets, fresh frozen plasma and cryoprecipitate over several days were required to stabilize the patient. On post-admission Day 2 the patient’s right hip lesion had become entirely necrotic and secondary necrotic lesions had...
developed on his glans penis, distal penile shaft, left hemi-scrotum, bilateral medial thighs and suprapubic area (Fig. 1). In consultation with the plastic surgery team, this was highly suggestive of purpura fulminans. Further workup revealed an acquired protein S deficiency with no other cause of DIC identified. There were no findings suggestive of Fournier’s gangrene.

The patient was incidentally found to have right basilic and bilateral common femoral DVT’s on post-admission Days 8 and 12, respectively. Subsequently an IVC filter was inserted.

At this time, transfer to Vancouver General Hospital tertiary referral facility for coordinated multi-specialty surgical care was completed. Surgical debridement was initially delayed given his multiple DVT’s. Once stable, partial glansectomy with debridement to Buck’s fascia and sparing of the corporal bodies was completed (Fig. 2). Two subsequent surgical debridement’s were completed in conjunction with plastic surgery, resulting in debridement to and including the dorsal neurovascular bundle but sparing the spermatic cord.

Reconstruction was performed by the plastic surgery team using a $2.5 \times 12$ cm$^2$ full thickness skin graft, which was applied circumferentially to the distal penile shaft (Fig. 3). A single graft was used with the seam positioned on the ventral aspect of the penis to aesthetically recreate the midline penile raphe.

Excellent cosmetic and functional outcomes were obtained at 8 weeks of follow-up. The patient’s urinary and erectile function remained unchanged at follow-up.

![Figure 1: Prior to surgical debridement.](image1)

![Figure 2: Following surgical debridement.](image2)

![Figure 3: Following full thickness skin grafting (dorsal aspect).](image3)

![Figure 4: Following full thickness skin grafting (ventral aspect).](image4)
DISCUSSION

Purpura fulminans is a hematological condition involving both DIC and necrosis of the skin [1]. It is characterized by widespread activation of the coagulation cascade, resulting in small-vessel thrombosis and the subsequent development of ischemia [2]. This manifests as erythematous macular lesions that develop central necrosis, becoming painful, blue-black and raised due to hemorrhage into the necrotic dermis [3]. These lesions can progress to necrosis of full-thickness skin or soft tissue [3].

The literature describes three forms of purpura fulminans based on causative mechanism: acute infectious, seen in association with the systemic inflammatory response syndrome; neonatal, which results from hereditary deficiency of protein C or S; and idiopathic, which usually follows a febrile illness and is thought to be mediated largely by protein S deficiency [4]. Each form is in turn associated with a particular clinical picture [4].

Much of the evidence on surgical management of purpura fulminans comes from studies of children with the acute infectious form of the disease, most often related to meningococcalemia. Initial interventions in such patients are largely medical, aimed at resolving the inciting sepsis via antibiotic therapy [1]. However, early surgical consultation is important given the importance of early treatment [1].

A retrospective review of five patients who underwent partial penectomy due to penile gangrene and review of seven diabetic patients with penile gangrene showed that insufficient debridement may progress to liquefaction, wound infection, extensive tissue loss and sepsis [5]. Partial penectomy negatively impacts patients’ self-image and sex life and may induce anxiety and depression in few [6]. However, along with treatment of comorbid conditions, it may assist in prevention of wound liquefaction, preservation of penile length, and improvement of quality of life [9].

REFERENCES

1. Mazzone L, Schiestl C. Management of septic skin necroses. Eur J Pediatr Surg 2013;23:349–58.
2. Lowery K, Shirley R, Shelley OP, Kaniorou-Larai M, Philp B, Dziewulski P. Purpura fulminans skin loss: surgical management protocols at a regional burns centre. J Plast Reconstr Aesthet Surg 2008;61:1520–3.
3. Morris ME, Majoub JG, Walker SK, Gardner GP, Jones RG. Meningococcal sepsis and purpura fulminans: the surgical perspective. Postgrad Med J 2013;89:340–5.
4. Roughton MC, Agarwal S, Gottlieb LJ. Surgical management of acute infectious purpura fulminans. J Burn Care Res 2011;32:231–6.
5. Chalmers E, Cooper P, Forman K, Grimley C, Khair K, Minford A, et al. Purpura fulminans: recognition, diagnosis and management. Arch Dis Child 2011;96:1066–71.
6. Edlich R, Cross CL, Dahlstrom JJ, Long WBIII. Modern concepts of the diagnosis and treatment of purpura fulminans. J Environ Pathol Toxicol Oncol 2008;27:191–6.
7. Sung DK, Jung SH, Young-Joo K. Necrosis of the penis with multiple vessel atherosclerosis. World J Mens Health 2014;32:66–8.
8. Guvel S, Yaycioglu O, Kilinc F, Kayasalcuk F, Ozkardes H. Penile necrosis in end-stage renal disease. J Androl 2004;25:25–9.
9. Chiang IN, Chang SJ, Kuo YC, Liu SP, Yu HJ, Hsieh JT. Management of ischemic penile gangrene: prompt partial penectomy and other treatment options. J Sex Med 2008;5:2725–33.
10. Ficarra V, Mofferdin A, D’Amico A, Zanon G, Schiavone D, Malossini G, et al. Comparison of the quality of life of patients treated by surgery or radiotherapy in epidermoid cancer of the penis. Prog Urol 1999;9:715–20.