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Review Article

Fournier's gangrene - Case report and a brief review

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Abstract Fournier's gangrene is a serious condition with very high mortality. It is usually associated with various co-morbidities. Early diagnosis with multispecialty approach and aggressive management is the key to successful outcome. We successfully managed Fournier's gangrene in a 32-year-old diabetic male. This case is reported and the condition is reviewed in the light of recent literature.

Key words
Fournier's gangrene (FG), emergency, infection, mortality, co-morbidity, debridement.

Introduction

Fournier's gangrene (FG) is a rare but potentially life threatening condition. This condition is named after Professor Jean-Alfred Fournier, the French venereologist who in 1883, used the term "fulminant gangrene" of the penis and scrotum for a sudden onset, rapidly progressing idiopathic scrotal gangrene in young men.[1] This condition is seen usually in debilitated patients with co-morbidities and systemic disorders such as diabetes mellitus, alcoholism, immunosuppression and perianal infections and in spite of recent advances in medical sciences; mortality rates can be as high as 12-45%.[2] Early diagnosis and aggressive management are the keys to decrease the morbidity and mortality.[3]

Case Report

A 32-year-old diabetic male reported with fever, diffuse body aches and increasing scrotal pain of one day duration. The severity of scrotal pain was 8-9/10 on 0-10 numeric pain rating scale. The pain had no relieving factors and would get aggravated with movements. There was no other past medical or surgical history of significance and diabetes was being managed with oral hypoglycemic drugs. There was no history of trauma, genital instrumentation or recent travel. On examination, the patient was conscious, well-oriented, ill-looking, in discomfort, febrile (Temp. 39°C) but hemodynamically stable. Local examination of perineum revealed patch of malodorous parchment like eschar over scrotum (**Figure 1A**) with edema and tenderness. There were no significant findings on general physical examination. Blood reports revealed leucocytosis with shift to left, normal RBC mass and platelet count, normal coagulation profile and normal arterial blood gas analysis. Serum chemistry showed hyperglycemia (random blood sugar-13.8 mmol/l). Urine analysis revealed traces of sugar. No flora was grown from blood or urine culture. Blood sugar levels were optimized by insulin and the patient was operated upon under general anesthesia. Wide debridement of scrotum (**Figure 1B**) followed by dedicated wound care

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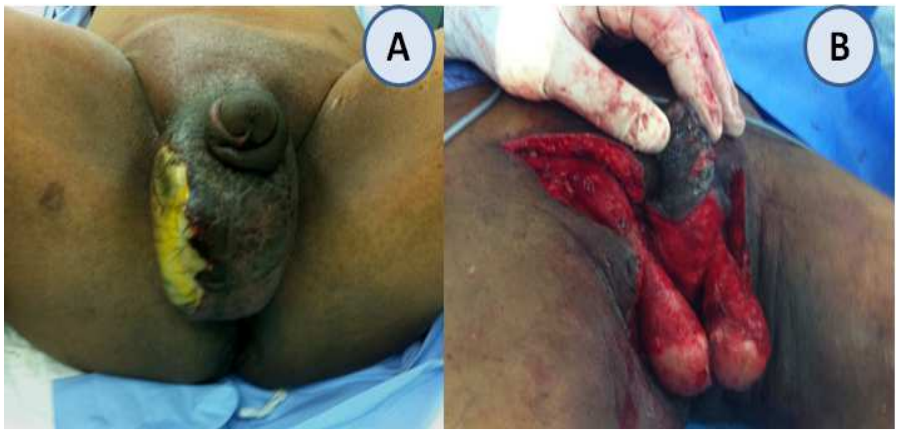

Figure 1 Fournier's gangrene with necrosed scrotal skin (A) and after wide debridement (B).

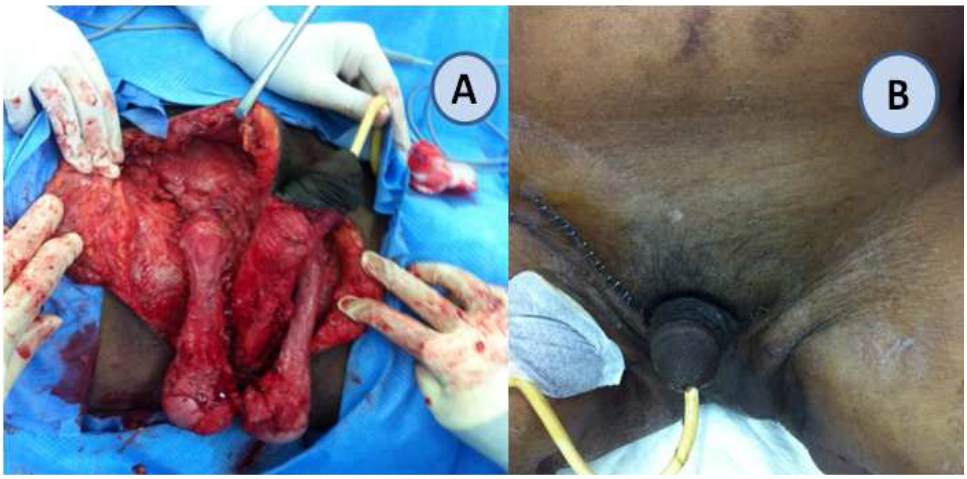

Figure 2 Reconstruction with local flaps (A) and 1 week postreconstruction (B).

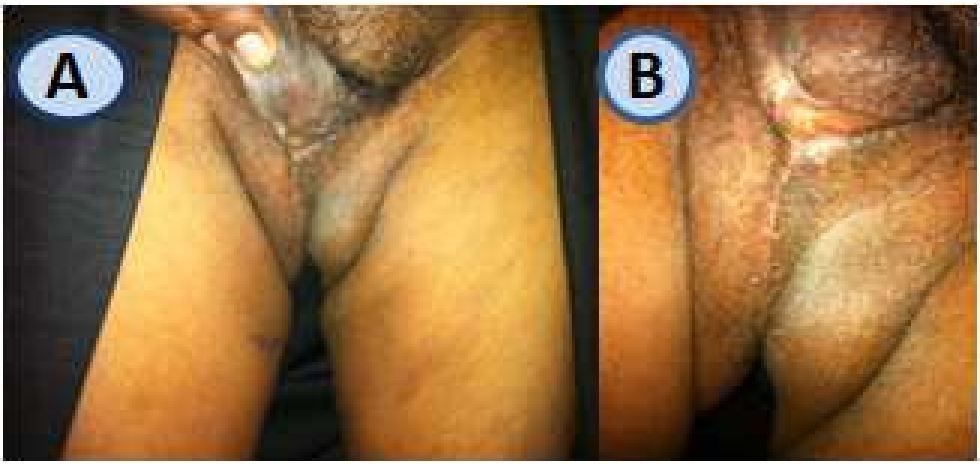

Figure 3 Perineal scar at three months follow-up, panoramic view (A), close-up view (B).

with regular dressings was done. Histopathological analysis of debrided tissues showed necrotic changes in the fascia with acute inflammatory cellular infiltration and fibrinoid thrombosis of the nutrient arterioles, thereby confirming the diagnosis of FG. After one week, the perineal raw areas were reconstructed by mobilizing local flaps (**Figure 2 A-B**). There

were no postoperative complications and patient was discharged home after inpatient management of two weeks. The patient was attached to the services of diabetologist besides being followed up in our clinic at two weekly intervals for three months. At three months, patient had healed with acceptable perineal scar (**Figure 3A, B**) and was discharged from our services.

Discussion

Historical background

Fournier's gangrene (FG) is a type of life threatening necrotizing infection or gangrene usually affecting the perineum. This condition is named after Jean-Alfred Fournier, a French dermatologist who specialized in the study of venereal diseases. He delivered a clinical lecture in 1883 and presented a case of perineal gangrene in an otherwise healthy young man.

Synonyms

In literature, many terms have been used synonymously to describe this clinical condition which include idiopathic gangrene of the scrotum, periurethral phelgmon, streptococcal scrotal gangrene, phagedena, and synergistic necrotizing cellulitis.

Epidemiology

FG is uncommon, though the exact incidence is unknown. A retrospective case review by Eka revealed 1726 cases documented in the literature from 1950-1999, with an average of 97 cases per year reported from 1989-1998.[4] The disease typically occurs from fourth to seventh decade of life. However, uncommonly cases are reported in pediatric age group.[5-7] The male-to-female ratio is approximately 10:1[2] and this lower incidence in females is attributed to better drainage of the perineal region through vaginal secretions. Men who have sex with men (MSM) are at higher risk, especially for infections caused by community-associated methicillin-resistant *Staphylococcus aureus* (MRSA).

Etiology

FG was originally described as idiopathic gangrene of the genitalia, but identifiable cause can be found by diligent search in 75-95% of cases. The disease process commonly originates from a nidus of infection in the anorectum (30-50%), the urogenital tract (20-40%), or the skin of the genitalia (20%). Anorectal causes of FG include perianal, perirectal, and ischiorectal abscesses; anal fissures; and colonic perforations. Urogenital tract causes include infection in the bulbourethral glands, urethral injury, iatrogenic injury secondary to urethral stricture manipulation, epididymitis, orchitis, and lower urinary tract infection. Dermatologic causes include hidradenitis suppurativa and scrotal ulcers. Cases are reported in literature where FG occurred as a complication of genital piercing, masturbation, prosthetic penile implants, circumcision, steroid enemas, strangulated inguinal hernia and penile self-injections.[8] Comorbid systemic disorders that depress cellular immunity frequently act as a predisposing factor.[3,9] Diabetes mellitus is reported to be present in 20%-70% of patients[3,9,10] and chronic alcoholism in 25%-50% patients. Other such factors include malignancy,[11] vascular diseases of pelvis, cirrhosis, systemic lupus erythematosus, morbid obesity, HIV infection[12,13] malnutrition, filariasis,[14] long-term steroid intake, hot humid climate and extremes of age. In neonates, the predisposing factors mentioned in literature include prematurity, a diaper rash, and varicella infection.[4-7] Patients with spinal cord injury are at higher risk for FG secondary to neurogenic bladder, neurogenic bowel, genital flora and

impaired sensation.[15,16] Czymek *et al.*[17] found the female gender to be an independent risk factor for mortality in patients with FG due to association to anatomical predisposition to higher incidence of inflammation of the retroperitoneal space and abdominal cavity. FG can however occur without any associated comorbidity.[18,19]

Microbiology

Cultures from the wounds in FG show polymicrobial infection by aerobes and anaerobes, including coliforms, *Klebsiella*, streptococci, staphylococci, clostridia, bacteroids, and corynebacteria.[20] *Escherichia coli* is the predominant aerobe, and bacteroides is the predominant anaerobe, with an average of 4 isolates for each case.[20] Most of these microbes are normal commensals in the perineum and genitalia, but because of the impaired host cellular immunity, they become virulent and act synergistically to invade tissue and spread at alarming pace along the tissue planes.[21] The synergistic microbial activity results in production of various exotoxins and enzymes like collagenase, heparinase, hyaluronidase, streptokinase, and streptodornase, which lead to tissue destruction and spread of infection. The platelet aggregation and complement fixation induced by the aerobes and the heparinase and collagenase produced by the anaerobes lead to microvascular thrombosis of the small subcutaneous vessels and gangrene of the overlying skin. Blood cultures are however positive in only 1/3rd of cases in spite of extensive infection.[20]

Clinical course

FG most commonly has rapid onset and fulminant course though uncommonly the disease can have insidious onset and slow progression. The hallmark of Fournier gangrene is intense pain and tenderness in the genitalia with pronounced systemic signs which are out of proportion to the extent of perineal signs and symptoms. The usual progression of clinical course occurs through following stages:

i. Prodromal symptoms of fever and lethargy, lasting for 2-7 days.
ii. Intense genital pain and tenderness with edema of the overlying skin; pruritus may occasionally be present.
iii. Increasing genital pain and tenderness with progressive edema and erythema of the overlying skin.
iv. Dusky appearance of the overlying skin; subcutaneous crepitus may be felt due to the action of gas forming microbes.
v. Necrotic patches start appearing over the overlying skin. If the patient is not managed aggressively at this stage, the condition worsens and sepsis with multiple organ failure follows.

Spread of infection

The infection spreads along the facial planes (**Figure 4**) and is usually limited by the attachment of the Colles' fascia in the perineum. Infection usually spreads to involve the scrotum and penis but can spread up the anterior abdominal wall and even reach up to the clavicle. The testes are usually spared due to the intraabdominal origin of testicular vessels and their involvement indicates retroperitoneal source or spread of infection. The course taken by the spreading infection may provide some insight into the possible origin of infection. Urogenital infections spread posteriorly along the Bucks and Dartos fascia to involve the Colles' fascia, but do not reach the anal margin due to the attachment of the Colles' fascia to the perineal body. Anorectal infections in contrast usually start from perianal area.[20]

Diagnosis

The diagnosis of FG is often made by clinical assessment. Imaging modalities may be required to evaluate the true extent of the disease or to arrive at the diagnosis, if the patient reports with atypical presentation. Plain radiographs may show air bubbles within the tissues. Ultrasonography[22] can show scrotal wall edema with gas. Computed tomography (CT) and magnetic resonance imaging are useful in selected cases to diagnose or rule out retroperitoneal or intra-abdominal disease process.[23,24] CT findings include asymmetric fascial thickening, subcutaneous emphysema, fluid collections, and abscess formation. Imaging can provide the evidence of gas within tissue planes even in the absence of clinical crepitus.

Histologic findings

Histology of the debrided tissue may reveal the following pathognomonic findings of Fournier gangrene:

i. Necrosis of the superficial and deep fascial planes.
ii. Fibrinoid thrombosis of the nutrient vessels supplying the superficial and deep fascia.
iii. Polymorphonuclear cell infiltration
iv. Microbes identified within the involved tissues.

The skin itself often has minimal inflammation until late in the disease and the extensive inflammatory process proceeds deep to intact skin.

Differential diagnosis

The differential diagnosis of FG include cellulitis, scrotal abscess, strangulated hernia, gonococcal balanitis, pyoderma gangrenosum, polyarteritis nodosa, ecthyma gangrenosum, allergic vasculitis, vascular occlusion syndromes, Wegener granulomatosis, necrolytic migratory erythema, testicular trauma, herpes simplex and warfarin necrosis.[20] The importance of proper diagnosis and differentiating from conditions like vasculitis or pyoderma gangrenosum is very high as the optimal treatment of these diseases (high doses of corticosteroids) is opposite to that of FG.

Fournier's gangrene severity index (FGSI)

To prognosticate the illness and predict the mortality, Laor *et al*.[25] has developed an objective and simple scoring system (Fournier's gangrene severity index- FGSI), using nine common vital sign and laboratory parameters including temperature, heart rate, respiratory rate, serum sodium, potassium, creatinine, bicarbonate levels, hematocrit and leukocyte count. The degree of derivation from normal is graded from 0 to 4 and the individual values are summed to obtain the FGSI score. With a score of over 9, a 46% probability of death has been found while a score of less than 9 is associated with 96% probability of survival.[26] Multiple studies have confirmed the clinical application of FGSI scale.[27] However, a recent study by Salami et al failed to confirm the application of FGSI scale and found only hematocrit and serum sodium levels to be of prognostic value.[28] Janane also could not find correlation between FGSI score the patient's survival but rather found metabolic aberrations and extent of disease on presentation as the reliable predictors of outcome.[29] In another recent study by Altarac *et al*. severe sepsis on admission and hypotension below 90 mmHg were found to be significant predictors for higher mortality.[30]

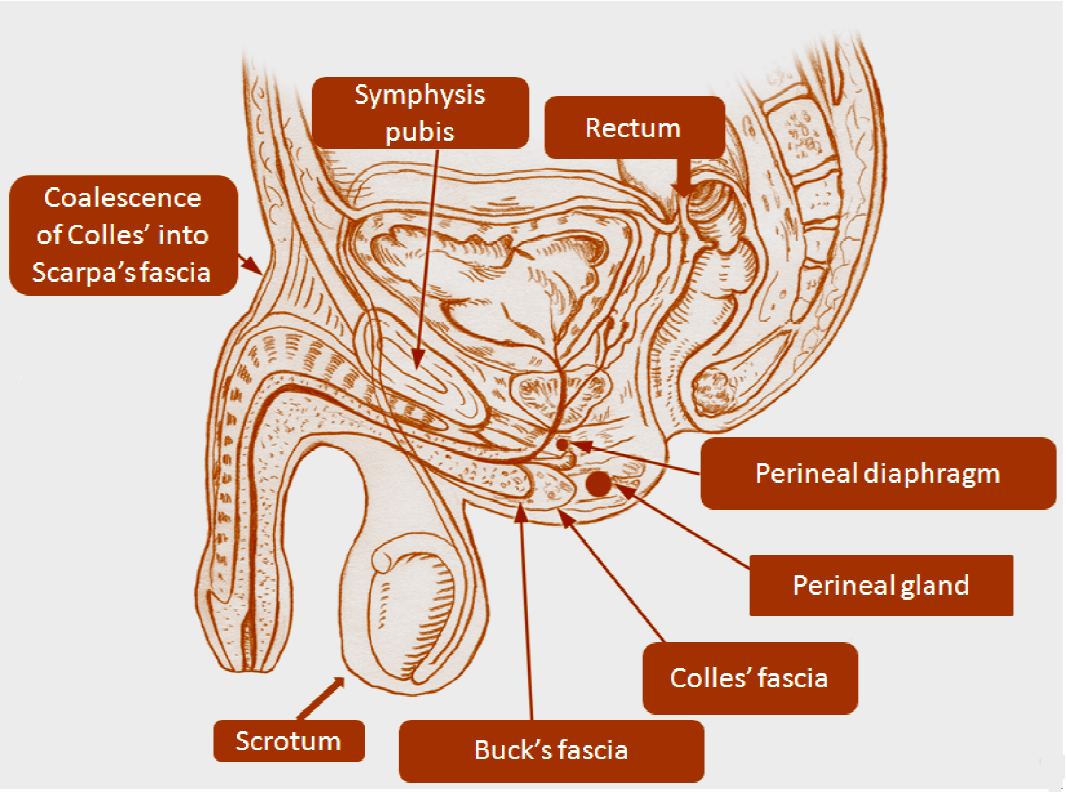

Figure 4 Anatomy of pelvis with description of perineal fascia.

Management

FG warrants aggressive multidisciplinary management including earliest surgical debridement, hemodynamic stabilization and broad spectrum antibiotics.[9,10,20] The full extent of the disease may become apparent only on operating table and is usually greater than the areas of skin involvement as we saw in our reported case. All nonviable tissue must be excised completely and the multiple sessions of debridement are generally required. Delay in debridement has significant negative impact upon survival.[3] Urinary or fecal diversion and even orchidectomy may be required in selected cases depending upon the foci of origin and extent of the disease. The raw area created as a result of debridement needs dedicated wound care and once fit reconstruction with skin grafts or flaps.[3] In recent years, vacuum-assisted closure (VAC) system dressings have significantly improved the post-debridement wound care, minimizing the skin defects and speeding up the healing process.[31-33] In some series, hyperbaric oxygen therapy has also been found to be effective in wound management.[21,29,34,35] It is believed to increase oxygen pressure in tissues thereby decreasing the number of anaerobic bacteria and reducing toxemia. Moreover, hyperbaric oxygen has been found to restore the normal phagocytic function of neutrophils, reduce inflammation, increase the proliferation of fibroblasts and enable angiogenesis.[21] Honey has also been reported to be cost-effective for wound management in FG.[36,37] Honey has been found to be capable of controlling the multiplication of bacteria due to its low pH value, high osmolarity and presence of elements like enzymes, hydrogen peroxide, flavonoids, and phenolic acid.

In patients who report with features of systemic toxicity like hypoperfusion or organ failure, aggressive resuscitation to restore normal organ

perfusion and function and continuous monitoring in intensive care units saves lives. Broad-spectrum antibiotics are to be initiated as soon as possible and aim is to provide effective coverage for gram-positive, gram-negative, aerobic, and anaerobic bacteria. Antifungal therapy (amphotericin B) is indicated if initial tissue stain with potassium hydroxide shows evidence of fungal growth.[37,38]

Prognosis

Due to better understanding of the etiopathogenesis of the disease, the availability of broader spectrum antimicrobials, improved intensive care facilities and the trend towards early and aggressive surgical intervention, outcome of FG has improved and mortality had decreased from more than 80% to around 12-45%.[20,25,38,39] Long term complications among survivors of FG are common and include chronic pain, sexual dysfunction, temporary stool incontinence and extensive scarring.[20] Since the disease continues to be a life-threatening emergency and has high associated health care costs in spite of low incidence, it is suggested in recent literature that that primary and secondary prevention measures should be implemented.[40]

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