CME

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LEARNING OBJECTIVES

- Describe Fournier's gangrene, including its comorbidities, multiple causes, and multi-organism nature
- Understand the clinical presentation of Fournier's gangrene and how it can be diagnosed
- Outline the treatment modalities, including aggressive resuscitation, utilization of broad-spectrum antibiotics, and emergent surgical therapy

Fournier's gangrene: Be alert for this medical emergency

Intense pain and tenderness in the genitalia are hallmarks of this infection. Early diagnosis, prompt antibiotic administration, and surgical debridement are essential.

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Fournier's gangrene is an aggressive and rapidly spreading infection of soft tissue, or *necrotizing fasciitis*, that involves the deep and superficial fascia of the perineum.[1] The rate of fascial necrosis in Fournier's gangrene is reported to be 2 to 3 cm/h.[1] Thrombosis of subcutaneous and cutaneous blood vessels produces gangrene, but the fascial necrosis is usually more extensive than the visible gangrene suggests.[2] Classic findings are necrosis of the superficial and deep fascial planes, fibrinoid coagulation of the nutrient arterioles, polymorphonuclear cell infiltration, and positive microorganism culture of involved tissues.

An estimated 750 cases have been reported in the literature since Fournier first described the disease in 1883.[3] Fournier's gangrene, an uncommon disease with no seasonal variation, is not indigenous to any region of the world; however, the largest number of cases originates from the African continent. The typical patient is a male in his sixth or seventh decade with comorbidities (see Table 1, page 46). Fournier's gangrene is 10 times more common in men than women.[4] Women tend to develop the disease following childbirth.[5] Regardless of gender predominance, multiple risk factors are associated with the disease[6] (see Table 2, page 46).

EPIDEMIOLOGY AND ETIOLOGY

Fournier's gangrene was originally thought to be an idiopathic gangrene of the genitalia; however, a specific etiology is found in approximately 95% of cases.[7] Anorectal abscess, genitourinary infection, and traumatic injury are the most common causes.[8] Comorbid diseases that compromise the immune system are a predisposing factor. The most frequent systemic illness associated with Fournier's gangrene is diabetes mellitus, which is seen in 10% to 60% of cases.[9] Diabetes causes defective phagocytosis, decreased cellular immunity, and microvascular disease with resultant ischemia.[10,11]

An iatrogenic or noniatrogenic injury to the perineum can initiate the development of Fournier's gangrene. Cases have been reported following hydrocele aspiration, blunt thoracic trauma, vasectomy, sparganosis (parasitic infection) of the scrotum, transrectal prostate biopsy, and penile self-injection with cocaine. Other case reports include the following etiologies: complications of varicella in a child, steroid enema use

FIGURE 1. Scrotal wall edema and skin discoloration.

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with radiation treatment for proctitis, spinal cord injury, and femoral heroin injection.[12]

The necrotizing process commonly originates with an infection in the anorectum, the urogenital tract, or the skin around the perineum.[13] Anorectal causes include an infection in the perianal glands, colonic diverticulitis, decubitus ulcers, or a colorectal injury or malignancy. Urogenital etiologies include an infection in the bulbourethral glands, a urethral injury, a lower urinary tract infection, or an iatrogenic injury secondary to stricture manipulation. Dermatologic causes include hidradenitis suppurativa, scrotal pressure ulceration, trauma, a surgical complication, or intentional trauma such as *skin-popping*—a form of injection drug abuse. Other diseases in addition to diabetes that increase the risk of developing Fournier's gangrene are systemic lupus erythematosus, Crohn's disease, and HIV infection. A less commonly reported cause is bone marrow malignancy.[14,15]

Fournier's gangrene, like most cases of necrotizing infection, has a multiorganism nature;[16] the disease is due to polymicrobial infection with a mixture of aerobic and anaerobic organisms.[17] The majority of cases are caused by normal flora of the lower GI tract,[18] most commonly *Escherichia coli*.[19] Other causative micro-organisms include *Staphylococcus*, *Streptococcus*, and *Enterobacteriaceae* species, anaerobic organisms, and fungi. The infection is rarely caused by one organism; as many as five species may be cultured.

The hallmark of Fournier's gangrene is intense pain and tenderness in the genitalia. The clinical course progresses through several phases. First, fever and lethargy develop. Next, patients experience intense genital pain and tenderness associated with edema of the overlying skin, which appears dusky, indicating subcutaneous crepitance. Soft tissue gas, a byproduct of anaerobic metabolism, is produced.[20] As genital pain and tenderness increase, obvious gangrene in a portion of the genitalia and purulent drainage materialize (see Figure 1). Systemic effects range from local tenderness to septic shock, depending on necrotic progression.

MAKING THE DIAGNOSIS

Fournier's gangrene is diagnosed primarily on clinical grounds, as diagnostic studies risk postponing treatment. In cases of rapid accessibility, uncertain diagnosis, or suspicion of retroperitoneal or intra-abdominal sources of infection, imaging studies should be considered.[21] Careful palpation of the genitalia and perineum and a digital rectal examination are important parts of the physical examination. Table 3 (page 46) lists the signs and symptoms of Fournier's gangrene. Typically, fluctuance, soft-tissue crepitance, localized tenderness, or occult wounds should alert the examiner to the possibility of Fournier's gangrene. A CBC, comprehensive metabolic panel, coagulation profile, and blood cultures should be obtained.

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FIGURE 2. CT shows small pockets of gas in the rectum (arrows).

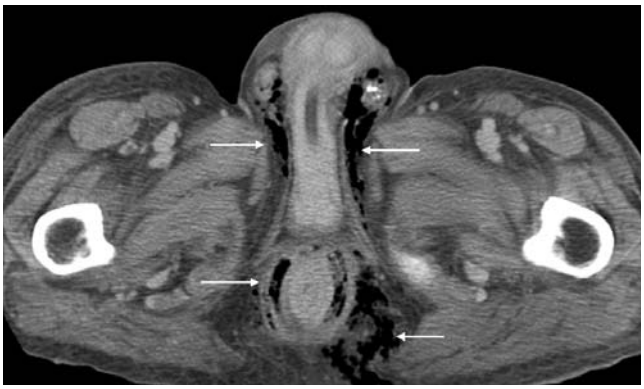

FIGURE 3. Fluid collections along the deep fascial planes (arrows) are demonstrated on CT.

KEY POINTS

- Fournier's gangrene was originally thought to be an idiopathic gangrene of the genitalia; however, a specific etiology is found in approximately 95% of cases. Anorectal abscesses, genitourinary infections, and traumatic injuries are the most common causes.
- Typically, fluctuance, soft-tissue crepitance, localized tenderness, or occult wounds in the genitalia, perineum, and anorectal area should alert the examiner to the possibility of Fournier's gangrene. A CBC, comprehensive metabolic panel, coagulation profile, and blood cultures should be obtained.
- Once the diagnosis is established, emergent surgical excision of all necrotic tissue must be performed. Given the potential fulminant nature of this necrotizing process, repeat debridement procedures are usually needed to completely eradicate the infection.

COMPETENCIES

- ●●●●● Medical knowledge
- ● Interpersonal & communication skills
- ●●● Patient care
- ● Professionalism
- ● Practice-based learning and improvement
- ● Systems-based practice

Imaging studies are more likely to detect gas within the soft tissues than is physical examination. An initial imaging study includes a plain radiograph, which may show moderate-to-large amounts of soft-tissue gas or foreign bodies. Ultrasonography also detects fluid or gas within the soft tissues[22] and is the preferred method.[23] Small pockets of soft-tissue gas are more readily detected on CT (see Figure 2, page 45). CT also can demonstrate fluid collections tracking along deep fascial planes (see Figure 3, page 45). MRI gives greater soft tissue detail than CT; however, it creates greater logistical challenges, especially in critically ill patients.

The definitive diagnosis of Fournier's gangrene is established by surgical examination under anesthesia, with an incision into the area of greatest clinical concern. If gangrenous tissue is present or purulence is drained, the diagnosis is established. Tissue samples should be sent for anaerobic and aerobic cultures, as well as histopathologic assessment. Ultimately, early identification of Fournier's gangrene is essential for a good prognosis.[24]

TABLE 1. Common predisposing comorbidities

Cirrhosis
Diabetes mellitus
High-risk behaviors (alcohol or IV drug abuse)
Immunosuppression
Malignancies
Malnutrition
Morbid obesity
Vascular disease of the pelvis

TABLE 2. Risk factors for Fournier's gangrene

Circumcision
Episiotomy
Extravasations of urine (periurethrally or through cutaneous fistula)
Hernioplasty
Hysterectomy
Local trauma or instrumentation to the perineum
Paraphimosis
Septic abortion
Urethral stricture caused by sexually transmitted diseases

TABLE 3. Signs and symptoms of Fournier's gangrene

Crepitant skin ("spongy" to the touch)
Dead and discolored (gray-black) tissue; pus weeping from injury
Fever and lethargy
Increasing genital pain and erythema or severe genital pain accompanied by tenderness and swelling of the penis and scrotum

TREATMENT

In patients who present with systemic toxicity manifesting as hypoperfusion and/or organ failure, aggressive resuscitation to return normal organ perfusion and function must take precedence.[25] Antibiotics with broad-spectrum coverage against staphylococci, streptococci, *Enterobacteriaceae* species, and anaerobes should be administered. If initial tissue stains show fungi, an antifungal should be included in the regimen. Empiric antibiotic regimens should be adjusted when the infective organisms are identified.

Once the diagnosis is established, emergent surgical excision of all necrotic tissue is required. The skin should be opened wide to expose the full extent of underlying fascial and subcutaneous tissue necrosis. Given the potential fulminant nature of this necrotizing process, repeat debridement procedures are usually needed to eradicate the infection. If perineal involvement is extensive, fecal diversion should be performed to eliminate potential contamination of the wounds; urinary diversion is accomplished via a urethral catheter. Hyperbaric oxygen (HBO) therapy has a theoretical role in treating Fournier's gangrene, but results of this therapy are mixed.[26] HBO therapy increases tissue-oxygen tension, leukocyte activation, oxygen free-radical production, capillary angiogenesis, fibroblast proliferation, and vasoconstriction and decreases anaerobe multiplication.[27] Prompt antibiotic administration and surgical debridement (with or without HBO) are the cornerstones of therapy.[28]

CONCLUSION

In the pre-antibiotic era, Fournier's gangrene was commonly fatal; even today, it poses a significant risk of morbidity and mortality.[29] Despite aggressive therapy, the mortality rate for patients with Fournier's gangrene is nearly 50% because of the aggressive nature of the infection and the presence of underlying comorbidities.[30] Delays in diagnosis or treatment increase the mortality rate. For example, a 24-hour delay in radical debridement increases the mortality rate by 11.5%; a 6-day delay is associated with a mortality rate of 76%.[31] Additional factors associated with high mortality include anorectal origin, advanced age, extensive disease, shock or sepsis at presentation, renal failure, and hepatic dysfunction.[32] Multiorgan system failure secondary to gram-negative sepsis is the most common cause of death.[33] Early clinical identification and prompt, aggressive treatment are essential for reducing mortality and morbidity in patients presenting with this disease. JAAPA

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