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The EM Educator Series

The EM Educator Series: Fournier Gangrene – Simple, raging cellulitis

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Case 1:
A 35-year-old male with no past medical history presents with severe rectal pain and inability to sit still on stretcher despite pain medication. He is tachycardic and appears in severe pain.

Case 2:
A 65-year-old female with history of diabetes and obesity presents with altered mental status. She is febrile, tachycardic, and hypotensive. On examination you find redness in the perineal region that tracks up the anterior abdominal wall.

Questions for Learners:

1. What are the major risk factors for Fournier gangrene?
2. How can patients present?
3. What laboratory testing may be helpful, and what risk scores are available?
4. What is the utility of imaging, including ultrasound?
5. What are the ED management priorities?
6. What are major pitfalls with Fournier gangrene?

Suggested Resources:

- #FOAMed
  - emDOCs Fournier Gangrene
  - emDOCS Misdiagnosing and Mismanaging
  - IBCC – NSTI
  - Wik EM
- Journal Articles
  - JEM – Fournier Gangrene
  - JEM – NSTI Pearls and Pitfalls

Answers for Learners:

1. What are the major risk factors for Fournier gangrene?

Risk Factor	Frequency
Diabetes Mellitus	20-70%
Alcohol misuse disorder	25-50%
Obesity	10-60%
Immunocompromised state	30-40%
Hypertension	Up to 25%
Peripheral vascular disease	Up to 25%
HIV infection	Up to 16%
Tobacco use	Up to 15%
Chronic renal failure	Up to 15%
Hematologic disorders	Up to 14%
Congestive heart failure	Up to 11%
Chronic liver disease	Up to 11%
Malignancy	Up to 8%
Recent surgery	Up to 5%
No risk factors	Up to 10%

Table 1. Risk factors for Fournier's Gangrene.

2. How can patients present?

Fournier's gangrene (FG) is a clinical diagnosis based on the presence of fluctuance, crepitus, exquisite tenderness, and wounds of the genitalia and perineum. Although the diagnosis is straightforward in the classic presentation, failure to examine the perineal area, especially in the older or obtunded patient, can result in misdiagnosis. Furthermore, the early symptoms of FG and NSTIs are not characteristic; hence, FG is often misdiagnosed as cellulitis or abscess in 75% of cases.

The clinical presentation of FG varies widely depending on the extent of infection as well as patient comorbidities. Typically, the infection begins as a localized cellulitis adjacent to the portal of entry, commonly in the perineum or perianal region, with an insidious presentation. Early presenting features are often non-specific and common to other infectious etiologies. In one study of NSTIs, the most common initial chief complaints were swelling (80.8%), pain (79%), and erythema (70.7%). Bullae (26%), overlying skin necrosis (24%), and crepitus (20%) were less common upon initial examination, but associated with later stages of necrotizing fasciitis.

Presence of subcutaneous gas and crepitus are highly specific for clostridial infections. Fever and tachycardia are present in 40% and 61% of these patients, respectively. The affected area may also appear swollen, dusky, and/or present with a characteristic purulent "dishwater" discharge with associated feculent odor, attributable to the presence of anaerobes. The presence of hypotension and septic shock is a late and ominous sign, occurring in roughly 21% of patients with NSTIs, but associated with high specificity (93.3%). This has a strong correlation with mortality, and, along with associated multiorgan failure, is the principal cause of death in patients with NSTIs.

The presenting tenderness, erythema, and swelling may mimic less severe infections including erysipelas and cellulitis. However, a key feature of FG is pain out of proportion to physical examination and should

alert the clinician to the possibility of FG. However, this is not always present, as local anesthesia may develop secondary to local nerve ischemia or due to a preexisting neuropathy (e.g. diabetic neuropathy). Furthermore, cellulitis and erysipelas often present with well-demarcated areas of inflammation and erythema. In contrast, FG may present with areas of poorly demarcated erythema, as well as blisters and bullae during the later stages of infection. While cellulitis and erysipelas may present with symptoms of generalized infection, including malaise and fever, FG can result in severe systemic toxicity with associated multiorgan failure. Furthermore, as FG may spread rapidly along fascial planes, areas of tenderness and erythema may extend as far up as the clavicle.

The extent of necrosis is an important prognostic factor as some studies have shown that patients with a necrotic area less than 3% of total body surface area rarely die, while patients presenting with involvement of > 5% total body surface area have a worse prognosis.

3. What laboratory testing may be helpful, and what risk scores are available?

While no single laboratory test has adequate sensitivity and specificity to discern NSTIs from other soft tissue infections, The Laboratory Risk Indicator for Necrotizing Fasciitis (LRINEC) score may suggest the presence of NSTI (Table 3); however, it should not be used to exclude the diagnosis. The original data examined age, gender, serum potassium, platelet count, C-reactive protein(CRP), leukocyte count, hemoglobin, sodium, creatinine, and glucose from 89 consecutive patients with NSTIs compared to 225 control patients. Patients were stratified into low-risk (≤5), intermediate-risk (6-7), and high-risk (≥8) categories, corresponding to a probability of <50%, 50%-75%, and >75% for the development of a NSTI, respectively. A score ≥ 6 was found to have 92% positive predictive value and a 96% negative predictive value for presence of a NSTI.

Laboratory Risk Indicator for Necrotizing Fasciitis		
CRP (mg/dL)	<15	0
	≥15	4
WBC (per mm^3)	<15	0
	15-25	1
	>25	2
Hemoglobin (g/dL)	>13.5	0
	11-13.5	1
	<11	2
Sodium (mEq/L)	≥135	0
	<135	2
Creatinine (mg/dL)	≤1.6	0
	>1.6	2
Glucose (mg/dL)	≤180	0
	>180	1
Composite Score	Score <6	Low Risk
	Score 6-7	Intermediate
	≥ 8	High Risk

Table 3. The Laboratory Risk Indicator for Necrotizing Fasciitis (LRINEC) score.

Criticisms of the LRINEC scoring system include its retrospective development and poor sensitivity among ED patients. The score was not explicitly designed to exclude NF in patients with a low-risk score, and subsequent studies externally validating the score have thus far failed to replicate the high sensitivity and negative predictive value reported in the initial paper. The LRINEC score, applied in isolation to ED patients, would miss over 20% of cases of NSTIs, with an associated sensitivity between 68% and 80%. There have also been cases of NSTI with LRINEC scores of 0.56 As the LRINEC score has limited sensitivity, it should not be used as the sole determinant of clinical decision-making for the diagnosis of FG.

4. What is the utility of imaging, including ultrasound?

The diagnosis of FG is primarily clinical, and in many cases, imaging is not necessary, nor is it desirable if it leads to a delay in surgical management. However, imaging is a useful adjunct in those cases in which the presentation is atypical, or when there is concern regarding the true extent of the disease.

Conventional radiography can be used to detect the presence of soft tissue swelling as well as gas in the perineal fascial planes before crepitus is noted on exam. Subcutaneous emphysema may extend from the scrotum and perineum to the inguinal regions, anterior abdominal wall, and thighs. Evidence of gas formation is present in nearly half of all patients with FG and is highly specific (94%). However, the absence of gas formation on imaging should not exclude the diagnosis due to poor sensitivity (49%).

Point-of-care ultrasound (POCUS) allows for the evaluation of soft tissue inflammation, collections/abscesses, as well as identification of subcutaneous gas. Characteristic findings include thickened perineal tissue caused by inflammation and edema, as well as a "cobblestone" appearance throughout the subcutaneous tissue. Acoustic shadowing of subcutaneous gas secondary to bacteria may result in a "snow globe" or "dirty shadowing" appearance caused by hyperechoic foci demonstrating reverberation artifacts. While POCUS has been shown to be highly specific for NSTIs (up to 93%), it is insufficiently sensitive (approximately 88%) to exclude a diagnosis with such a high morbidity and mortality.

CT imaging plays an important role in the diagnosis of FG and to evaluate the extent of the disease. Intravenous contrast can further characterize the soft tissue and should be used if possible. Characteristic findings include asymmetric fascial thickening, fluid collections, abscess formation, fat stranding around involved structures, and evidence of subcutaneous emphysema. CT may uncover the underlying etiology of FG, such as a perineal abscess, fistula formation, or any infectious process in the intra-abdominal or retroperitoneal spaces. It assists in differentiating FG from less severe infections and allows evaluation of both the superficial and deep fascial planes.19 CT has a sensitivity of 88.5% in addition to high specificity (93.3%) for the diagnosis of NSTIs.

MRI with gadolinium contrast is an excellent imaging modality to characterize soft tissues. However, in the ED setting, MRI is of limited value due to its high cost, extended time of examination, and the fact that it requires a clinically stable patient. Although it has been shown to have a high sensitivity (100%) and specificity (86%) to diagnose NSTIs, MRI has a limited role for evaluation of FG in the ED.

Modality	Test Characteristics	Findings in Fournier's Gangrene
X-ray	Sensitivity 49% Specificity 94%	-Hyperlucencies representing subcutaneous emphysema -Soft tissue swelling
POCUS	Sensitivity 88% Specificity 93%	-Thickened, edematous soft tissue with "cobblestoning" - Reactive unilateral or bilateral hydroceles -Acoustic shadowing of subcutaneous gas resulting in a "snow globe" or "dirty shadowing" appearance
CT	Sensitivity 88.5% Specificity 93.3%	-Asymmetric fascial thickening -Underlying etiology (abscess, fistula, etc.) -Fluid collections with possible air/fluid levels -Subcutaneous emphysema -Fat stranding
MRI	Sensitivity 100% Specificity 86%	-Asymmetric fascial thickening -Partial or complete absence on post-gadolinium images of signal enhancement of the thickened fascial planes -Fluid collections with possible air/fluid levels -Subcutaneous emphysema -Fat stranding

Table 4. Imaging modalities in Fournier's gangrene.

5. What are the ED management priorities?

The cornerstones of treatment of FG include emergent surgical debridement of all necrotic tissue, broad-spectrum antibiotics, and hemodynamic resuscitation with intravenous fluids as well as vasoactive medications as needed. As the rate of fascial necrosis has been noted as high as 2–3 cm per hour, FG is considered a surgical emergency necessitating early involvement of the appropriate surgical teams, which decreases mortality. As up to 21% of patients present with symptoms of hypotension and septic shock, hemodynamic resuscitation and patient optimization prior to surgical intervention are important aspects of management.

Broad-spectrum parenteral antibiotic therapy is initiated empirically upon diagnosis of FG and then subsequently tailored based on culture results. Any initial antibiotic regimen must have a broad range of activity against commonly implicated organisms, most notably staphylococcal and streptococcal species, as well as coliforms, gram-negative bacteria, Clostridium, Bacteroides, and Pseudomonas. Empiric antibiotics must cover for MRSA, typically with linezolid or vancomycin, which is combined with a carbapenem or beta-lactam-beta-lactamase inhibitor. Clindamycin should be added, as it can suppress toxin production and modulate cytokine production, as well as decrease mortality from NSTIs. In those patients with severe penicillin hypersensitivity, clindamycin or metronidazole combined with an aminoglycoside or fluoroquinolone should be administered. Additionally, many have suggested adding penicillin for treatment of streptococci and, in particular, when Clostridium species are suspected. Doxycycline should be considered for those patients with significant risk for Vibrio vulnificus and Aeromonas hydrophila involvement, including exposure to marine exposure, and exposure to seafood. Special consideration should be given to initiating early antifungal therapy with amphotericin B or fluoroconazoles in those patients with a history of fungal infections and immunocompromised patients, as fungal sources are now an emerging cause of NSTIs.

Empiric Antibiotic Regimens for Fournier's Gangrene
Carbapenem *OR* **Beta lactam-beta lactamase inhibitor** *PLUS* **Clindamycin** *PLUS* **Vancomycin, daptomycin, or linezolid**
In patients with severe hypersensitivity to carbapenems or beta lactam-beta lactamase inhibitors, consider substituting: **Aminoglycoside** *OR* **Fluoroquinolone** *PLUS* **Metronidazole**
In patients with salt or freshwater exposure and risk for Vibrio vulnificus or Aeromonas hydrophila involvement, consider adding **Doxycycline**
In patients with significant risk for fungal involvement, consider adding **Amphotericin B or Fluoroconazoles**

Table 5. Initial empiric antibiotics for patients with suspected Fournier's gangrene.

Hemodynamic resuscitation and optimization of the patient's comorbidities play an integral role in ED management. Patients may present hypotensive or in septic shock characterized by hypoperfusion, which can result in organ dysfunction. Aggressive fluid resuscitation and hemodynamic support are often required, as evidence of end-organ dysfunction is associated with increased mortality. As many patients with FG have underlying comorbidities that may exacerbate underlying FG, most commonly diabetes mellitus, it is important to treat these underlying comorbidities. While few data exist concerning optimizing patient comorbidities in those with FG, a treatment strategy may be extrapolated from current critical care literature. Insulin therapy for glycemic control for a target blood glucose level of 140-200 mg/dl has been suggested in critically ill patients, which is reasonable for patients presenting with hyperglycemia. These patients may also present with an episode of diabetic ketoacidosis (DKA) secondary to FG and should be managed in accordance with current guidelines. Up to 50% of patients with FG have an underlying alcohol use disorder and may be suffering from concomitant alcohol

withdrawal or delirium tremens. These patients should be treated with adequate supportive care, as well as benzodiazepines administered in a symptom-triggered fashion, guided by the Clinical Institute Withdrawal Assessment of Alcohol scale, revised (CIWA-Ar).

Prompt surgical consultation is recommended for all patients in whom FG is suspected and should not be delayed by laboratory or imaging investigations. Depending on local practice patterns, as well as individual patient characteristics, this may involve physicians from the general surgery, urology, or obstetrics and gynecology services, or a combination. The most important variable affecting mortality in patients with NSTIs is time to admission and debridement. One study suggests that survival decreases from 93.2% to 75.2% with a delay in debridement from 24 to 48 hours. Similarly, in another study, the average time from admission to operation was 90 hours in non-survivors versus 25 hours in survivors, making early surgical intervention imperative.

Hyperbaric Oxygen Therapy (HBOT)is used as an adjunctive therapy for the optimization of infected tissue oxygenation and for its bactericidal and bacteriostatic effects, especially in the post-surgical period. However, the lack of randomized controlled studies limits the use of HBOT to patients unresponsive to conventional surgical and intensive care management. As such, HBOT is not routinely recommended prior to surgical debridement, and consultation with a hyperbarics specialist is typically not a consideration in the ED.

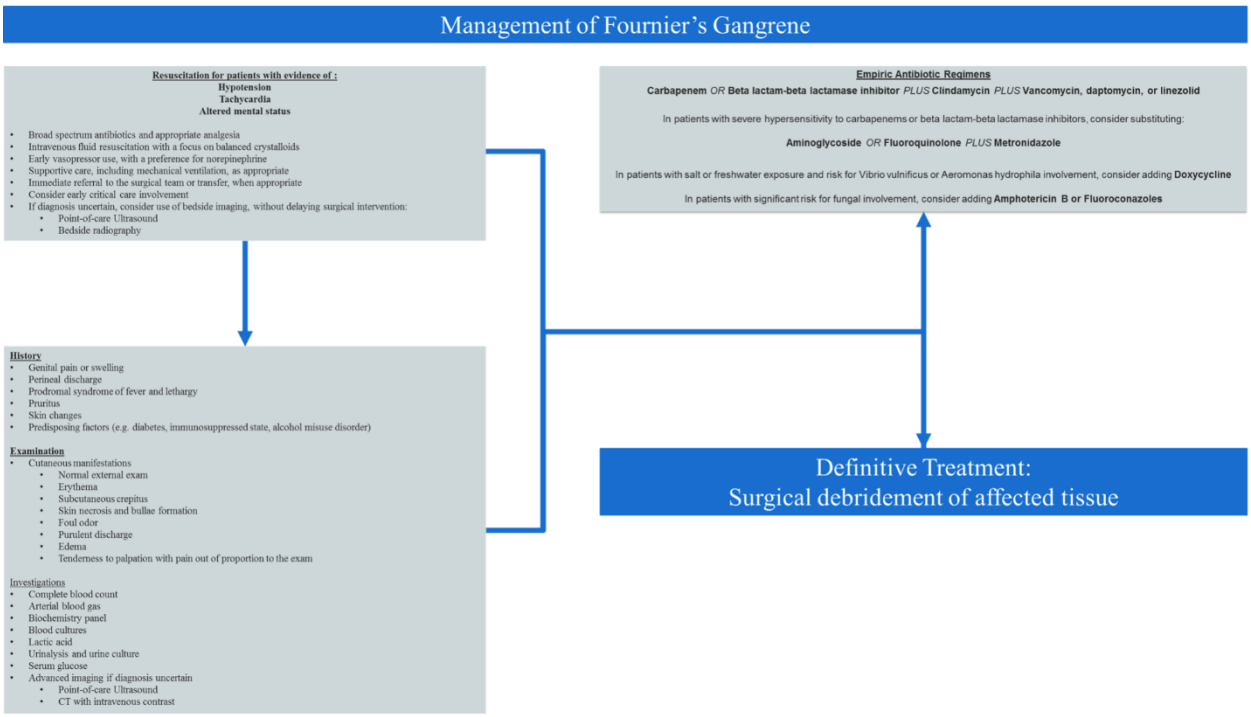

Figure 5. Management algorithm for FG.

6. What are major pitfalls with Fournier gangrene?

How is necrotizing fasciitis missed?

1. Unreliable history
2. Unreliable exam and anchoring on cellulitis
3. Failure to complete a full skin exam, especially in those unable to provide a history
4. Failure to consider if patient's pain is uncontrollable and anchoring on the "pain seeker"
5. Reliance on lab scoring and LRINEC score
6. Too much reliance on imaging for ruling out disease
7. Being told this can't be necrotizing fasciitis after surgical consultation

Where do we fail in management?

1. Forgetting this is a surgical disease
2. Failure to provide broad spectrum antibiotics including clindamycin or linezolid for toxin inhibition
3. Failure to resuscitate appropriately with fluids and vasopressors to ensure end organ perfusion
4. Believing that IVIG and hyperbaric oxygen are cornerstones of therapy