A rare case report of vulvar necrotizing fasciitis in a healthy patient

Takshaka Patel, Rajul Kothari, Antonio Gangemi*

*Department of Surgery – Division of General, Minimally Invasive & Robotic Surgery, University of Illinois at Chicago, 840 S. Wood Street, Suite 435E, Chicago, IL 60612, USA

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A B S T R A C T

INTRODUCTION: Necrotizing fasciitis (NF), a necrotizing infection of the soft tissue, is a medical emergency usually occurring in the lower extremities and abdominal regions and often difficult to diagnose promptly.

PRESENTATION OF CASE: This case report looks at one atypical presentation of NF with the unusual location of the vulva and no known associated comorbidities or risk factors.

DISCUSSION: Diagnosing this patient was particularly difficult due to the inconsistent clinical, laboratory and imaging findings. The CT scans and WBC count were indicative of NF, but the LRINEC score was not high enough to make the diagnosis of NF. As a result, we relied on the hemodynamic instability and clinical findings of the physical exam to be strong indicators of NF, and act on that indication.

CONCLUSION: Acting quickly on the hemodynamic findings and suspicion as opposed to waiting for a confirmed diagnosis resulted in a good prognosis since immediate surgical debridement is imperative to surviving this acute condition. Despite major advancements in the imaging modalities and the introduction of a laboratory score, our case suggests that the diagnosis still heavily relies on clinical findings, such as hemodynamic instability. Furthermore, our case suggests that NF should be included in the differential regardless of atypical location and lack of common clinical associations.

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Introduction

Necrotizing fasciitis (NF) is a relatively rare and potentially fatal condition that involves acute infection of soft tissue [1]. The most effective method of treating someone with this condition is to perform immediate surgical debridement [1,2]. Higher mortality rates in patients with NF are linked to greater delays in providing the needed surgical attention [2–4]. Specifically, delay in the surgical treatment beyond 24 h of presenting to the hospital is linked to increased mortality rates [5]. Since the clinical presentation of NF, which includes pain, fever, and edema [1,6], is similar to the clinical presentations of other disorders, such as cellulitis and other forms of gangrene [7], further testing is often imperative to accurately diagnose this condition. However, current diagnostic tests are not absolutely reliable [7–9] and a great amount of physician discretion is often required in making the correct decision. A valuable and validated diagnostic tool currently available is the Laboratory Risk Indicator for Necrotizing Fasciitis (LRINEC) score which has a positive predictive value of 92.0% but has not achieved wide-spread use, mostly due to the fact that the parameter with the highest weight, C-reactive protein (CRP), cannot be promptly obtained [10]. Therefore, a rapid clinical diagnosis based off the knowledge of the risk factors, comorbidities and clinical presentation most commonly associated with this condition is still essential to establish appropriate and life-saving surgical treatment. A well known diagnostic criterion is the location of NF, which most commonly occurs in the lower extremities, upper extremities, and abdomen [11,12]. Additionally, useful diagnostic criteria are the presence of risk factors, such as diabetes, prior history of trauma, or use of IV drugs, and the presence of co-morbidities most commonly associated with this condition [1,7]. Also, the acute (usually hours) onset of NF, characterized by extremely rapid spread once the infecting microbiome gains access to the fascial layer after the initial injury, represents a useful adjunct to the diagnosis [12,13].

Although a few cases of NF originating in the vulva have been identified, it is an extremely rare presentation and most often linked with diabetes or post-partum status [14–16]. Herein, we describe an exceptionally unusual case of NF with vulvar location and no association with known risk factors or co-morbidities. This case occurred at an academic institution and the case report has been presented in accordance with the SCARE criteria [17].

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and spreading to the abdominal wall. The gynecological and general surgery teams worked together to perform first aggressive debridement. During the second look exploration surgery, the gynecology team performed further debridement of the vulvar area. When the abdominal area was examined, a few additional pockets of necrosis in the abdominal wall were discovered and debrided. The following three surgeries composed of additional debridement of the genital and abdominal regions and definitive closure of the wound in both areas. The patient’s stay in the surgical intensive care unit was five days long. Microbiology samples determined that the offending agents were Clostridium clostridiforme and Bacteroides. The Jackson-Pratt drain placed at the time of the first surgical exploration was removed during follow-up visits, which indicated that the patient healed well from the surgery with no complications and proper bowel function. The cosmetic outcome was also favorable to the patient.

Discussion

The diagnosis of NF was very challenging in our patient for several reasons that will be discussed ahead. Suspicion of NF was supported by the elevated WBC and pelvic CT findings. However, neither of those findings are conclusive towards the diagnosis of NF. The elevated WBC may be due to a variety of pathologies, including the much more common cellulitis. Additionally, the presence of gas and fat stranding reported with the CT scan (Figs. 1 and 2) may suggest NF but is not a conclusive indication of NF due to the limited reliability of CT scans in the diagnosis of this condition [7,9,18]. Furthermore, there is a significant number of NF cases where these radiologic findings may not be present at all [7,9,18].

The LRINEC score is, to our knowledge, the only laboratory tool validated on scientific ground with accurate and positive predictive value. Although there is some debate regarding the usefulness of LRINEC in early recognition of NF, multiple studies have validated the ability of the LRINEC in detecting NF and differentiating it from other soft tissue infections that may clinically present in a similar fashion [10,19,20]. Some studies have found that LRINEC, although highly specific, is not adequately sensitive, therefore a low LRINEC score should not be used as reasoning to eliminate the diagnosis of NF [21]. The LRINEC scoring system has not reached widespread use yet mostly due to the fact that the parameter associated with the highest weight (4 points) is not readily available in an acute setting (CRP is usually assessed in an elective setting with radio-immunologic assay and requires 24–48 h for the result) [10]. In our case this was the main reason why we could not fully rely on the LRINEC Score to make the call for emergent surgical exploration.

The demographics of our patient contributed to much extent to the difficult diagnosis, as NF is more commonly found in elderly males, IV drug users, immunocompromised or diabetic individuals [1,7,22,23]. Additionally, among the few reported cases of NF originating in the vulva, the affected patients are typically diabetic or in a post-partum state [14–16]. Our patient met none of these requirements and was an otherwise healthy female.

The location of the infection contributed perhaps the most to the difficulty of the diagnosis, as NF is most often seen in the extremities and abdomen [11,12,22]. Locations apart from the extremities and abdomen are generally considered to be less likely involved with NF and more likely due to a different soft tissue infection, such as cellulitis [7].

These diagnostic difficulties explain why the patient was initially admitted to the medical ward and a surgical consultation was requested several hours after being admitted to the hospital. The patient was in fact initially misdiagnosed with cellulitis. A further delay in the correct diagnosis and most importantly in the appropriate surgical treatment could have likely had fatal con-

**Presentation of case**

A 56-year-old African American female with no relevant past medical or surgical history presented to the Emergency Department with a swelling over her left labia developing for six days that had opened and begun to drain two days prior. Upon admission to the medical ward, her temperature was 36.8 °Celsius, the blood pressure was 100/60 mmHg despite IVF resuscitation, the pulse was 96–100 beats per minute, the respiratory rate was 20 breaths per minute, and the arterial oxygen saturation was 98%. Of note, her white blood cell count (WBC) was 17.4 x 10⁹ per liter. The patient was started empirically on vancomycin and piperacillin/tazobactam, a surgery consult was requested, and a pelvic CT scan was obtained which showed a cutaneous ulceration over the left labia and air in the subcutaneous fat of the left groin and left lower abdominal wall (Figs. 1 and 2). Inflammatory fat stranding was also present along the left lateral abdominal wall (Fig. 2). The LRINEC score was 3 based on the available data (CRP not available), and had the potential to go up to a 7 if CRP data was available and the levels were greater than 150 mg/l. Although none of the diagnostic measures indicated NF over other soft tissue infection, the findings of the CT scan combined with the apparent sepsis upon presentation and an LRINEC score of 3–7 raised a suspicion of NF.

The decision to perform immediate explorative surgery was made. Since the vulva was affected, the gynecology team was also involved with this case. Explorative surgery of the pelvic and abdominal regions performed by a surgery attending physician and obstetrics and gynecology attending physician confirmed the diagnosis of necrotizing soft tissue infection originating at the left vulva

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**Fig. 1.** CT scan showing cutaneous ulceration overlying the left labia with air in the subcutaneous fat.

**Fig. 2.** CT scan showing inflammatory fat stranding extending superiorly along the anterior margin of the abdominal rectus muscles.
sequences. The tachycardia and hypotension, despite intravenous fluid resuscitation and broad spectrum antibiotic treatment having been implemented on our patient, raised the highest suspicion for NF as this condition progresses quickly to septic shock if left untreated.

The two learning points of our case report are:

1. Hemodynamic instability is the strongest and most valid indicator for emergent surgical exploration when there is a clinical suspicion of NF.
2. Unusual location, lack of associated co-morbidities and/or risks factors for NF, negative history of initiating trauma, or an low LRINEC score cannot be used to eliminate the diagnosis of NF.

Conclusion

Despite major advancements in the imaging modalities and the introduction of a laboratory score, the diagnosis of NF still heavily relies on clinical findings, most importantly hemodynamic instability, as our case suggests. Furthermore, our case suggests that NF should be included in the differential of soft tissue infections regardless of atypical location and lack of common clinical associations.

Surgical intervention was performed in the case we are herein reporting as soon as suspicion of NF was raised. Our approach is in line with the evidence from currently available literature indicating prompt surgical debridement as the most important and independent prognostic factor [2–5].

Conflicts of interest

The Authors declare no conflict of interest pertinent to this case report.

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Ethical approval

No Institutional Review Board is required for publication of a case report at our institution

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request

Authors contributions

Study Concept and design: A. Gangemi.