Necrotizing Fasciitis Associated with Hailey-Hailey Disease: A Rare Case

Ruo-Chen Zhang[1], Na-Na Zheng[1] and Lian-Sheng Zhong[2]

[1]*Department of Dermatology, Xuzhou Municipal Hospital Affiliated with Xuzhou Medical University, Xuzhou, China*
[2]*Department of Dermatology, Children's Hospital of Fudan University at Xiamen, Xiamen, China*

ABSTRACT

Necrotizing fasciitis (NF) is a rare and life-threatening infection of soft tissue characterised by rapid and extensive destruction of the skin, subcutaneous fat, and fascia. Early diagnosis of NF is challenging, and it can be very difficult to distinguish NF from other infectious diseases of skin and subcutaneous tissue. Imaging studies and laboratory investigations are crucial diagnostic means for NF. We diagnosed a case of NF with multiple organ dysfunction and septic shock, and this is the first case of NF associated with Hailey-Hailey disease (HHD) to our knowledge. Clinicians should be alert to signs and symptoms of NF in HHD and other skin diseases with damaged skin barrier function such as pemphigus, pemphigoid, and all kinds of ulcers, especially in diabetic and immunosuppressed patients.

Key Words: *Necrotizing fasciitis, Genodermatosis, Hailey-Hailey disease.*

How to cite this article: Zhang RC, Zheng NN, Zhong LS. Necrotizing Fasciitis Associated with Hailey-Hailey Disease: A Rare Case. *J Coll Physicians Surg Pak* 2022; **32(JCPSPCR)**:CR111-CR112.

INTRODUCTION

Necrotizing fasciitis (NF) is a rapidly progressing infection of soft tissues with extensive destruction of the skin, subcutaneous fat, and fascia. NF has high mortality, even if surgical debridement is performed promptly. It may occur from any trauma to the surface of skin, diabetes mellitus, chronic hepatitis, and immunosuppressive conditions.[1] Hailey-Hailey disease (HHD), also known as familial benign pemphigus, is a chronic genodermatosis which is characterised by multiple recurrences of vesicles, maceration and erosions in friction areas (such as the neck, the axilla, the groin, and the perineum). Herein, we describe a rare case of NF associated with HHD in a 55-year male with type 2 diabetes mellitus.

CASE REPORT

A 55-year male with a history of HHD and type 2 diabetes mellitus presented to our department complaining of an acute exacerbation of pain and swelling on the scrotum, perineum, and right thigh for 3 days. Physical examination revealed extensive tender erythema and swelling on the right thigh, scrotum and perineum, and vesicles, erosions, and fissures on both sides of the axilla and groin (Figure 1a, b). The patient also had fever at 37.8°C. The patient was hospitalised for further treatment.

Correspondence to: Dr. Lian-Sheng Zhong, Department of Dermatology, Children's Hospital of Fudan University at Xiamen, Xiamen, China
E-mail: 498716354@qq.com

Received: July 13, 2020; Revised: November 04, 2020; Accepted: November 30, 2020
DOI: https://doi.org/10.29271/jcpsp.2022.JCPSPCR.CR111

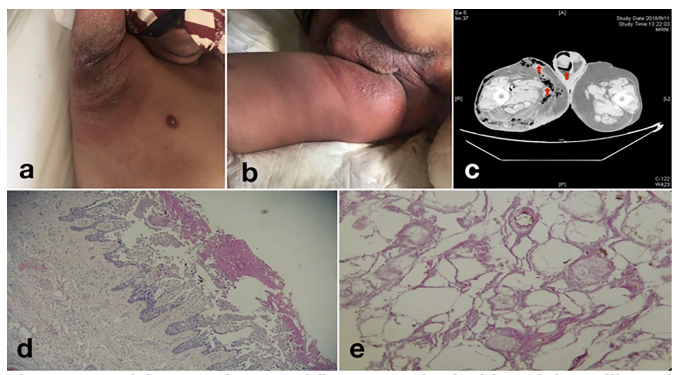

Figure 1: Vesicles, erosions, and fissures on both sides of the axilla and groin and extensive tender erythema and swelling on the right thigh, scrotum and perineum (a, b). Computed tomographic image showing fat edema and air (red arrow) in the subcutaneous fat of the right thigh, scrotum and perineum (c). Histopathology of tissues showing acantholysis throughout the epithelium giving the appearance of a dilapidated brick wall, and extensive necrosis of the dermis and subcutaneous tissues (HE, ×50 (d), ×100 (e).

The lesions of the right thigh and perineum continued to progress rapidly despite treatment with antibiotics. Computed tomography (CT) scan showed fat oedema and air in the subcutaneous fat of the right thigh, scrotum, and perineum (Figure 1c). Laboratory investigations revealed white blood cell (WBC) count of 18.3×10^9/L, neutrophil (NE) count, 15.5×10^9/L, and glucose (GLU), 13.5 mmol/L. On general examination, the patient was unconscious and with sustained fever at 38.9°C. Vitals were heart rate (HR), 124 beats/min, respiratory rate, 28 times/min, blood pressure (BP), 90/67 mmHg and pulse oxygen saturation (SpO_2), 92%. The patient was then diagnosed with NF and was treated with intravenous imipenem and vancomycin immediately. Extensive surgical debridement of the groin, leg,

and perineum was performed two hours after diagnosis. On surgical exploration, there was extensive pus, fat and fascial necrosis spreading to most of his lower limbs and perineal region. Swab culture for bacteria revealed *Escherichia coli* infection. Tissue culture for fungus was negative. Histopathology of debrided tissue from the perineum showed acantholysis throughout the epithelium giving the appearance of a dilapidated brick wall, and extensive necrosis of the dermis and subcutaneous tissues (Figure 1d, 1e). Unfortunately, postoperative septic shock and multiple organ dysfunction developed, including acute liver and renal failure. The patient was then transferred to the intensive care unit and died after two days.

DISCUSSION

NF is a destructive infection of the skin and subcutaneous tissues associated with a mortality rate of 24% to 34%.[2] Early recognition, proper antibiotic treatment and timely surgical debridement are the most important principles of managing NF. However, NF may not be easily distinguished from other soft tissue infections (*i.e.* erysipelas and cellulitis) in the early stage, but some clinical features (such as warmth, swelling, induration, and poorly defined margins) are helpful in diagnosis.[3] Laboratory investigations are also essential tools for the diagnosis of NF. The Laboratory Risk Indicator for NF (LRINEC) is assessed by six routine laboratory tests and used initially to make an early distinction between NF and other soft tissue infections, and it is also a helpful tool for early diagnosis of NF.[4] This patient's LRINEC score was 8, which strongly predicted the occurrence of NF. Moreover, it is found that patients have a high risk of NF with increasing immunosuppressive conditions (such as cancer, diabetes mellitus, HIV, and organ transplantation). Especially, diabetes mellitus is the most common cause of NF, accounting for 44.5% to 72.3%.[5] In the affected site, local trauma has usually been found to be a portal of entry for bacteria that initiate the process of infection.[6] In this case, the occurrence of NF was caused by the erosions and fissures of HHD which served as a portal of entry for the bacteria and immunosuppression brought about by uncontrolled sugar exacerbated it. To our knowledge, this is the first case of NF associated with HHD. In summary, clinicians should be alert to signs and symptoms of NF in HHD and other skin diseases with damaged skin barrier function such as pemphigus, pemphigoid, and all kinds of ulcers, especially in diabetic and immunosuppressed patients.

PATIENT'S CONSENT:

The authors certify that they had obtained all appropriate patient consent forms. In the form, the patient had given his consent for his images and other clinical information to be reported in the journal.

COMPETING INTEREST:

The authors declared no competing interest.

AUTHORS' CONTRIBUTION:

RZ: Designed and drafted the manuscript.
RZ, NZ: Contributed in the sample selection and analysed the data.
LZD: Designed the study and critically revised the manuscript.
All authors contributed to the final draft of the manuscript and approved it for submission.

REFERENCES

1. Baron TH, Morgan DE. Acute necrotizing pancreatitis. *N Engl J Med* 1999; 340(18):1412-1417. doi:10.1056/NEJM199905063401807.
2. White NR, Fowler LL. Retroperitoneal and cutaneous necrotizing fasciitis secondary to necrotizing pancreatitis. *J Emerg Med* 2014; 47(2):147-9. doi: 10.1016/j.jemermed.2014.02.009.
3. 3Kiat HJ, En Natalie YH, Fatimah L. Necrotizing fasciitis: How reliable are the cutaneous signs? *J Emerg Trauma Shock* 2017; 10(4):205-210. doi: 10.4103/JETS.JETS_42_17.(4).
4. El-Menyar A, Asim M, Mudali IN, Mekkodathil A, Latifi R, Al-Thani H. The laboratory risk indicator for necrotizing fasciitis (LRINEC) scoring: the diagnostic and potential prognostic role. *Scand J Trauma Resusc Emerg Med* 2017; 25(1):28. doi: 10.1186/s13049-017-0359-z.
5. Elliott DC, Kufera JA, Myers RA. Necrotizing soft tissue infections.Risk factors for mortality and strategies for management. A*nn Surg* 1996; 224(5):672-83. doi: 10.1097/00000658-199611000-00011.
6. Taviloglu K, Yanar H. Necrotizing fasciitis: Strategies for diagnosis and management. *World J Emerg Surg* 2007; 2:19.doi:10.1186/1749-7922-2-19.