Necrotizing fasciitis following episiotomy in a woman with Hailey-Hailey disease: A case report

Heba Abu Saleem a, Osama Al-Natour b, Ismaiel Abu Mahfouz c,*, Mohd Abu Assamen b, Said Al-Natour b

a Department of Obstetrics and Gynaecology, Specialty Hospital, Amman, Jordan
b Department of Surgery, Specialty Hospital, Amman, Jordan
c Department of Obstetrics and Gynaecology, Faculty of Medicine, Al Balqa Applied University, Al Salt, Jordan

ARTICLE INFO

Keywords:
Infected episiotomy
Necrotizing fasciitis
Hailey-Hailey disease

ABSTRACT

A healthy 25-year-old primiparous woman had an uncomplicated pregnancy and spontaneous vaginal delivery with mediolateral episiotomy. Twenty-four hours postpartum, she developed increasing perineal pain and swelling. Initial examination showed a localized erythema and tissue oedema at the episiotomy site. The woman was admitted to hospital for management of the infected hematoma at the site of the episiotomy. Thereafter, she was started on intravenous antibiotics, and exploration under anaesthesia was planned. The woman’s medical condition deteriorated rapidly, and necrotizing fasciitis (NF) was strongly suspected. Therefore, aggressive medical and surgical management was undertaken, including broader-spectrum antibiotics and multiple surgical debridement. A biopsy of the debrided tissue showed acantholysis and dyskeratosis, which are features of Hailey-Hailey disease of the skin (familial benign chronic pemphigus), a rare condition. The woman eventually had a V-Y advancement fascial flap and made a complete recovery. In this case report, the details of the development of NF in a woman who was found to have Hailey-Hailey disease are discussed.

1. Introduction

Necrotizing fasciitis (NF) is a rare life-threatening soft-tissue infection caused by a wide variety of pathogens and may spread quickly, leading to sepsis, shock, multi-organ failure and death [1]. The incidence has been estimated to be 0.4 per 100,000 and the mortality rate as high as 35% [2]. Stephenson et al. [3] reviewed the clinical details of 29 non-pregnant women with NF of the vulva and found that a delay in diagnosis of more than 48 h was associated with a mortality rate of 73%. Prompt recognition, accurate diagnosis, immediate administration of broad-spectrum antibiotics, and surgical intervention are associated with a favorable outcome.

Patients with risk factors for NF and a history of soft-tissue trauma or surgical procedure typically present with excruciating pain with or without skin changes. This is usually followed by rapid deterioration of general health. Therefore, close observation in these clinical scenarios should include monitoring of vital signs, fluid resuscitation, blood investigations, wound and blood cultures, and wound assessment. Imaging studies such as computed tomography (CT scan) and magnetic resonance imaging (MRI) may be helpful in early diagnosis but cannot rule out NF [4].

Hailey-Hailey disease, also known as familial benign chronic pemphigus, is a rare autosomal dominant disorder resulting from a mutation in the ATP2C1 gene and has a prevalence rate of 1:50,000 [5]. It is characterized by the development of blisters and erosions which most often involve skin folds, such as the groin, neck, axillae, and under the breasts. Rarely, lesions may involve the conjunctiva, mucosa, and the vulva [6]. The onset of symptoms usually occurs around middle age and symptoms may be exacerbated by weight gain, trauma, pregnancy, perspiration, infection, and ultraviolet radiation [7]. Tissue biopsy may reveal the abnormal formation of keratin tissue (keratinization) and failure of cell-to-cell adhesion (acantholysis), and the hair follicles are usually spared, typically with acanthosis and dyskeratosis [8].

2. Case Presentation

A 25-year-old primiparous healthy woman who had an uneventful pregnancy presented to the labour ward in spontaneous labour when she...
was 39 weeks pregnant. She had a normal vaginal delivery with a right medio-lateral episiotomy and was discharged home on day 1 post-partum. On the second postpartum day, the women presented to the emergency room with general weakness, severe pain, and swelling at the episiotomy site.

On clinical examination, the woman was found to have tachycardia (pulse rate of 110 BPM) and normal blood pressure (125/80 mmHg). Perineal examination showed ecchymosis, induration, and severe tenderness at the site of the episiotomy, but no crepitus.

Initial investigations showed an elevated white blood count (WBC) of $40 \times 10^9/L$ (normal range: 4.5-11 $\times 10^9/L$) and elevated C-reactive protein (CRP) of 170 mg/L (normal range: less than 5 mg/L). Thereafter, the woman was started on intravenous fluids and a combination of broad-spectrum antibiotics (ceftriaxone, metronidazole, and gentamicin). Additionally, she had urgent exploration of the episiotomy site under general anaesthesia, which showed no obvious hematoma or pus collection, and the tissues in the episiotomy bed were viable. Culture swabs were collected, a surgical drain was sited, and the episiotomy wound edges were approximated with interrupted sutures. Twenty-four hours after exploration, the clinical picture deteriorated rapidly, the woman had tachycardia and hypotension which did not respond to intravenous (IV) fluids resuscitation. Therefore, she was transferred to the intensive care unit (ICU), where blood investigations showed a further rise in CRP and WBC, to 200 mg/L and $60 \times 10^9/L$, respectively.

A pelvic CT scan showed a bulky uterus, pelvic ascites, and no evidence of tissue fluid or gas collection at the episiotomy site. In view of the clinical and biochemical deterioration, surgical consultation was sought.

The joint decision of the obstetrician and the surgeon was to perform a laparoscopy to further explore the ascites and exclude uterine rupture, and to re-explore the episiotomy site. During laparoscopy, there was no evidence of uterine rupture or pelvic hematoma, and a round 100 mL of ascitic fluid was noted, which was drained, and culture swabs were obtained. Perineal re-exploration included an incision lateral to the episiotomy site, which revealed unviable tissue extending from the lateral aspect of the right labia majora to the upper medial aspect of the right thigh. Extensive tissue debridement was performed until healthy viable tissues were reached [Image 1]. The wound was left open, and an iodine-soaked pack was applied. Additionally, the antibiotics were adjusted after consulting the infectious diseases team.

The clinical condition of the woman started to improve, and she had several debridements under anaesthesia until the wound showed viable tissues. Finally, the woman had a V-Y advancement fascial flap to achieve a complete closure of the wound [Images 2, 3]. The woman was discharged in good condition and was followed up in the outpatient department until she had recovered completely. Six months later, she got pregnant, had regular antenatal care, and was delivered by an elective cesarean section at 39 weeks of gestation.

3. Discussion

Necrotizing fasciitis is very rare in obstetrics. As a result, this severely limits the experience of the obstetrics team in the early diagnosis and management. Reported risk factors for NF usually reflect an immunocompromised state and include obesity, malnutrition, anemia, diabetes, systemic lupus erythematosus, renal transplantation, immunosuppressive medications, and intravenous drug abuse [9]. Obstetrical trauma was the only risk factor for NF in our patient. While most patients present with surgical site erythema and swelling, the most consistent finding is pain which is out of proportion to physical findings.

Regarding the causative agents, they could be mono- or polymicrobial and require inoculation of the pathogen into the subcutaneous tissue. This can happen through any break in the integrity of the epithelial or mucosal barriers, or it may be hematogenous [10]. The bacteriological results in our case were polymicrobial: predominantly E. coli, Enterococcus faecalis, and Bacteroides fragilis. Other organisms
that have been associated in NF following a vaginal delivery include Group A and B beta hemolytic Streptococcus, Staphylococcus, Klebsiella, Pseudomonas, Peptostreptococcus, and Peptococcus [10].

Histopathological examination of the debrided tissue revealed acantholysis and dyskeratosis with epidermal hyperplasia, which are features of Hailey-Hailey disease [8]. We believe that the disease was exacerbated in our case because of various factors which included the woman being pregnant, the trauma of the episiotomy, and the infection. The presence of Hailey-Hailey disease may have increased this woman’s risk of having NF.

The woman in this case required several procedures of surgical debridement to achieve viable tissue. This is supported by a report that many patients require multiple procedures of surgical debridement and usually have large, complex wounds that require soft-tissue coverage and a long hospital stay [11]. We believe that early recognition, prompt intervention, and multidisciplinary team management saved the woman’s life.

4. Conclusion

Necrotizing fasciitis is an extremely rare condition which may complicate episiotomies. Obstetric teams should be aware of this very rare complication. A high index of suspicion and prompt intervention which includes resuscitation and surgical intervention by the obstetric and plastic surgery teams is associated with higher survival rates.

Contributors

Heba Abu Saleem was involved in patient care, the literature review and manuscript drafting.
Osama Al-Natour was involved in patient care, the literature review and manuscript drafting.
Ismaiel Abu Mahfouz was involved in the literature review, drafting and revision of the manuscript, and supervised the work.
Mohd Abu Assamen was involved in patient care, the literature review and manuscript drafting.
Said Al-Natour was involved in patient care, the literature review and manuscript drafting.
All authors approved the final submitted manuscript.

Funding

The authors declare that they received no fund from any public, commercial, or not-for-profit organizations.

Patient consent

Written consent was obtained from the woman to use her details and the images included.

Provenance and peer review

This article was not commissioned and was peer reviewed.

Conflict of interest statement

The authors declare that they have no conflict of interest regarding the publication of this case report.

References

[1] A. Giuliano, F. Lewis Jr., K. Hadley, F.W. Blaisdell, Bacteriology of necrotizing fasciitis, Am. J. Surg. 134 (1) (1977 Jul) 52–57, https://doi.org/10.1016/0002-9610(77)90283-5. PMID: 327844.Hakkarainen TW.
[2] N.M. Kopari, T.N. Pham, H.L. Evans, Necrotizing soft tissue infections: review and current concepts in treatment, systems of care, and outcomes, Curr. Probl. Surg. 51 (8) (2014) 344–362, https://doi.org/10.1007/j.cpsurg.2014.06.001.
[3] H. Stephenson, D.J. Dotters, V. Katz, W. Droegemueller, Necrotizing fasciitis of the vulva, Am. J. Obstet. Gynecol. 166 (5) (1992 May) 1324–1327, https://doi.org/10.1016/0002-9378(92)91597-4 (PMID: 1595786).
[4] M. Wronski, M. Slodkowski, W. Cebulski, D. Karkocha, I.W. Krasnodebski, Necrotizing fasciitis: early sonographic diagnosis, J. Clin. Ultrasound 39 (4) (2011) 236–239.
[5] M. Wronski, M. Slodkowski, W. Cebulski, D. Karkocha, I.W. Krasnodebski, Necrotizing fasciitis: early sonographic diagnosis, J. Clin. Ultrasound 39 (4) (2011 May) 236–239, https://doi.org/10.1002/jcu.20766. Epub 2010 Nov 12. PMID: 21480291.
[6] N. Yadav, B. Madke, S. Kar, K. Prasad, N. Gangane, Hailey-Hailey disease. Indian Dermatol Online J. 7 (2) (2016 Mar-Apr) 147–148, https://doi.org/10.4103/2229-5178.178960 (PMID: 27057511; PMCID: PMC4804597).
[7] M. Malan, W. Xuejingzi, J. Si, S.J. Quan, Hailey-Hailey disease: the role of azathioprine an immunomodulator, Pan Afr. Med. J. 32 (2019 Feb 5) 65, https://doi.org/10.11604/pamj.2019.32.65.17877. PMID: 31223357; PMCID: PMC6560993.
[8] A.E. Ortiz, C.B. Zachary, Laser therapy for Hailey-Hailey disease: review of the literature and a case report, Dermatol. Rep. 3 (2011), e28. PMC free article [PubMed] [Google Scholar.
[9] A.J. Headley, Necrotizing soft tissue infections: a primary care review, Am. Fam. Physician 68 (3) (2003 Jul 15) 323–328 (PMID: 12892352).
[10] G.G. Kihiczak, R.A. Schwartz, R. Kapila, Necrotizing fasciitis: a deadly infection, J. Eur. Acad. Dermatol. Venereol. 20 (4) (2006 Apr) 365–369, https://doi.org/10.1111/j.1468-3083.2006.01487.x (PMID: 16643131).
[11] Anaya DA, Dellingner EP. Necrotizing soft-tissue infection: diagnosis and management. Clin. Infect. Dis. 2007 Mar 1; 44(5):705-10. doi: https://doi.org/10.1086/511638. Epub 2007 Jan 22. PMID: 17278065.