An Analysis of Vulvar Necrotizing Fasciitis in the Unique and Ethnically Diverse Hawaiian Population

John Nakayama MD and Raydeen Busse MD

Abstract

Vulvar necrotizing fasciitis is a surgical emergency with a high rate of morbidity and mortality. Our case series adds seven patients to the literature and presents the first group that is predominantly of Pacific-Islander origin. This study not only confirms traditional risk factors such as diabetes mellitus, obesity and hypertension but investigates ethnicity and socioeconomic status as risk factors. Also presented is a case of recurrent necrotizing fasciitis initially involving the vulva, then the back. In any patient for which there is suspicion of vulvar necrotizing fasciitis, surgical diagnosis remains the gold standard and facilitates rapid debridement of all necrotic tissues. Aggressive surgical debridement with broad spectrum antibiotic coverage is required to minimize mortality.

Introduction

Necrotizing fasciitis is an often fatal bacterial infection that quickly spreads through the subcutaneous tissue and superficial fascia. Although the cutaneous appearance of necrotizing fasciitis is often unimpressive, this is a surgical emergency requiring immediate debridement. If treatment is not initiated in a timely manner, the infection will spread along the fascial plane, liquefying the subcutaneous tissue. Necrotizing fasciitis in any anatomic location can be fatal, but the vulvar and perineal forms are associated with mortality as high as 50%.[1]

There are many predisposing conditions to necrotizing fasciitis including diabetes mellitus, immunocompromise, peripheral vascular disease, increasing age, hypertension, obesity, and radiation exposure. Diabetes mellitus is of particular importance since it not only increases frequency of disease but also mortality.[2] Stephenson et al found diabetes mellitus to be a comorbid condition in 69% of patients.[1] This high comorbidity and mortality is likely due to tissue ischemia secondary to peripheral vascular compromise, decreased phagocytosis and decreased chemotaxis of polymorphonuclear leukocytes, and hyperglycemia promoting bacterial growth. For these reasons, diabetes mellitus is the single greatest risk factor of mortality in necrotizing fasciitis.[2] Another factor important to outcome is developing necrotizing fasciitis postpartum. Mortality can be as high as 50% in this setting.[1,3]

Necrotizing fasciitis is not a single disease entity but manifests as two types. Type I is a synergistic polymicrobial infection of both aerobes and anaerobes that is most commonly seen in the setting of diabetes, peripheral vascular disease and postoperative patients. The most commonly seen bacteria are S. *aureus*, *Streptococci*, *Enterocci*, E. *coli*, B. *fragilis*, and *Clostridia*.[4] Type II is a monomicrobial infection of group A streptococcus (*Streptococcus pyogenes*) and less frequently methicillin-resistant *Staphylococcus aureus* (MRSA) seen in patients without underlying comorbidities. The majority of vulvar necrotizing fasciitis infections are type I.[3, 5] It is believed that an aerobic species is the primary infection, which devitalizes affected tissues creating an oxygen-free environment for anaerobes to infect secondarily.

The primary site of infection is a surgical incision in about 50% of necrotizing fasciitis cases. Most other cases develop in areas where the skin has been damaged by infection or trauma.[2]

The signs and symptoms of necrotizing fasciitis can be divided into early and later manifestations. The early presenting signs include localized edema, induration, and exquisite pain at the site of infection. High fever, leukocytosis, anorexia, and hypocalcemia secondary to fat saponofication are also possible early signs. As the infection spreads, usually 1-2 days after initial presentation, late signs appear. Common features include skin discoloration to a reddish-purple hue, bullae eruption, systemic toxicity, and wound anesthesia. These findings are the result of tissue ischemia leading to denervation and vessel thrombosis.[1] It is important to note that the extent of subcutaneous necrosis is not reflected by the cutaneous appearance. By the time skin necrosis is visible, the subcutaneous infection has extended widely. Therefore, surgical diagnosis is performed to look for devitalized tissues which fail to bleed when incised. The pathognomonic sign of necrotic subcutaneous tissue, which appears as a "gray-brown 'dirty dishwater' fluid," is also found on surgical exploration.[2] Hypotension and shock may be present at any time but are more commonly late signs and are due to massive third spacing. Crepitus is noted 10% of the time and indicates the presence of gas producing organisms such as *Clostridia*.[4]

Early diagnosis is essential given that delayed treatment leads to increased mortality. Stephenson et al found a 75% mortality rate when surgical treatment was delayed by ≥48 hours compared to 12% mortality in those treated in ≤12 hours.[1] Therefore, early surgical intervention is the key to optimizing survival and is needed for histological diagnosis. Although exact diagnostic criteria has not been agreed upon, one set of diagnostic features proposed by Fisher et al, is summarized in Table 1.[6] Recently, CT and MRI scans have been advocated and may have utility in excluding necrotizing fasciitis when the clinical likelihood is low, but MRI in particular has a higher sensitivity than specificity and overestimates deep fascial involvement.[7] Surgical exploration remains the only diagnostic method able to rule out necrotizing fasciitis definitively and should not be delayed for imaging. In addition to surgical diagnosis, blood cultures are often taken but have limited utility. Gallup et al found cultures useful mainly for detecting secondary candidal septicemia and Wong et al had positive blood cultures in only 20% of patients with type I necrotizing fasciitis.[4,8] Diabetic patients with vulvar infection are considered to have vulvar necrotizing fasciitis until proven otherwise.[9]

Table 1.— Necrotizing Fasciitis Diagnostic Criteria
1) Extensive necrosis of the superficial fascia with peripheral undermining of skin
2) Moderate to severe systemic toxicity
3) Absence of muscle involvement
4) No demonstration of clostridia in wound and blood cultures
5) Absence of major vascular occlusion
6) Intensive leukocytic infiltration, necrosis of subcutaneous tissue, and microvascular thrombosis on pathologic examination of debrided tissue

The mainstay of treatment for necrotizing fasciitis remains early aggressive surgical debridement with broad spectrum antibiotic coverage. Optimal surgical treatment removes all necrotic tissue, which is achieved by debriding down to the fascia and outward until bleeding is encountered. Incisions are often left open to facilitate daily re-exploration of the wound with frequent wound changes and additional debridement as warranted. Antibiotic choice is often empiric and is dependent on the type of necrotizing fasciitis. Vulvar necrotizing is primarily type I which is a mixed infection of aerobes and anaerobes. To cover all possible organisms, treatment often consists of a penicillin, clindamycin, and an aminoglycoside (e.g.GAC regimen: gentamicin, ampicillin, and clindamycin) until the patient stabilizes and granulation tissue begins to form.[2]Recently, hyperbaric oxygen therapy added to surgery and antibiotics have been found to decrease morbidity and mortality.[8,10]

Materials and Methods

This retrospective chart analysis includes all patients admitted to the Kapiolani Medical Center for Women and Children from August 1, 1986 through September 1, 2006. Patients were identified using International Classification of Diseases (ICD) 9 coding for the diagnoses of vulvar necrotizing fasciitis and cellulitis. The charts of these patients were reviewed for the following information: age, weight, ethnicity, risk factors, onset of symptoms, appearance of initial lesion, time of diagnosis, time to treatment, procedures performed, level of pain, bacteria identified, length of stay, and outcome. Pathological reports confirming subcutaneous tissue necrosis of the vulva, perineum, pubis or lower abdomen were available for all patients reviewed. Pediatric patients were excluded from this study.

Necrotizing fasciitis is defined in this article as an infectious process of subcutaneous tissue leading to necrosis on pathological section and often resulting in moderate to severe systemic toxicity without major vascular occlusion or Clostridia in wound or blood cultures. Vulvar and perineal necrotizing fasciitis is defined to include patients with vulvar, perineal, pubic, or pelvic involvement.

Results

During the twenty year period studied, seven women were identified to have necrotizing fasciitis that involved the vulva, perineum, pelvic, or pubic areas. Each patient's chart in this study was examined and the major findings of this review are presented in Table 2. Six of the seven patients presented from 2002-2006, and one patient in 1992. Their ages ranged from 28 to 67 years of age with a mean of 41.1 years. The ethnic distribution of our patients consisted of four Pacific-islanders (3 Hawaiians and 1 New Zealander), two Caucasians, and one Filipina. Five patients were uninsured or on public assistance, a group which contained all the Pacific-Islander patients.

Of the seven women, six had at least one risk factor. The most prevalent risk factor was obesity in 71%. Diabetes mellitus type II was seen in 57% and hypertension in 29%. One patient was receiving chemotherapy. There were two puerperal patients. Both had surgical incisions and one had chorioamnionitis as well.

A primary event was noted in six out of seven cases. There were four infected surgical incisions, one recurrent vulvar abscess, one vulvar folliculitis, and one case where no primary event could be identified. Patient number 2 initially presented with recurrent vulvar abscesses which led to vulvar necrotizing fasciitis. After recovering completely, she was readmitted in thirteen days for necrotizing fasciitis of the back which developed from a back abscess. In both cases of necrotizing fasciitis, only MRSA was found in wound cultures.

Table 2.— Clinical Characteristics of Necrotizing Fasciitis Patients

Patient #	Age	Ethnicity	Dx to Tx (in Days)*	Length of Stay (in Days)	Predisposing Factors	Wound Culture	Outcome
1	28	Pacific Islander (Hawaiian/ Filipino)	<1	4†	Infected episiotomy	Coag. neg. staph‡, *B. fragilis, E. coli Peptostreptococcus sp., K. pneumoniae, C. perfringens*	Died
2	44	Pacific Islander (Hawaiian)	<1	3	DMII, Obesity, HTN, Hx of perineal abscesses bilaterally with surgery	MRSA	Recovered
3	35	Pacific Islander (Hawaiian)	<1	1§	Obesity	*S. pyogenes*	Recovered
4	32	Caucasian	<1	51	Chorioamnionitis Status post cesarean section	*S. agalactiae*	Recovered
5	67	Caucasian	<1	44	DMII, Obesity, Chemotherapy, HTN, Status post TAH-BSO	*P. mirabilis, E.coli, Peptostreptcoccus sp., S. agalactiae,* Anaerobic GNR	Recovered
6	41	Filipino	<1	17	DMII, status post TAH	Diphtheriods, Coag. neg. staph, *S. agalactiae, Peptostreptococcus sp.*	Recovered
7	41	Pacific Islander (New Zealand)	<1	29	DMII, Obesity	Coag. neg. staph	Recovered

HTN=hypertension, DMII= Diabetes mellitus Type II, MRSA= methicillin resistant *S. aureus*, GNR=gram negative rods; *Dx to Tx: Time from Diagnosis to treatment in days; † Transferred on day 2 and died at other hospital 2 days later; ‡ Coag. neg. staph.= Coagulase negative staphylococcus; § Transferred to other hospital for recovery

Initial presentation of the patients showed wound erythema in 100% and induration in 86%. Six of seven patients (86%) were in pain. The one patient without pain presented with necrotizing fasciitis arising from an infected incision on post-operative day twenty-five. She had pain earlier but at presentation, the affected area had lost sensation. White blood cell (WBC) counts were > 13,000 in five of seven patients (71%). Of those who had lower WBC counts, one was receiving chemotherapy and the other had a WBC count of 12,900. 71% of the patients were febrile on presentation, and 57% had purulent discharge from the affected area. Three patients (43%) presented with hypotension, and only one patient was noted to have crepitus upon wound palpation (14%).

All patients were taken for debridement within 24 hours of diagnosis. Wounds were debrided to viable tissues, and two women required multiple debridements. Cultures were taken from all patients' wounds, and the results are summarized in Table 2. The most common organisms encountered were Coagulase negative *staphylococcus*, *Peptostreptococcus*, and S. *agalactiae*, which were found in three of the seven patients. Multiple organisms were found in three of the seven patients (43%). Six of the seven women received broad spectrum antibiotics immediately. The patient not put on broad spectrum antibiotics was originally given cefazolin and was changed to cover MRSA after wound cultures returned. Blood cultures were taken from six patients and were all negative. One patient was given hyperbaric oxygen therapy.

There was one mortality in the series. This patient was a 28-year-old G5P1A4 status post vaginal delivery with episiotomy, who developed a fever of 101.4 Fahrenheit on the night of post-operative day two. The fever was associated with perineal pain and swelling, and her WBC count was 37,900 with 67% neutrophils and 19% bands. Initially she was diagnosed with perineal cellulitis and started on penicillin G, gentamicin, and clindamycin. The following morning, it was noted that the cellulitis borders were expanding and her WBC count was worsening. The patient was taken for immediate debridement. All necrotic tissue was removed from the episiotomy site and right vulva. The patient remained febrile and her WBC count increased to a maximum of 134,200 over the next two days. She was transferred to the local trauma center for intensive care, but the patient became septic and died a day after transfer.

Discussion

Vulvar necrotizing fasciitis is rare, as are all types of necrotizing soft tissue infections with an incidence of 0.04 cases per 1,000 person-years, but there is no available incidence for the vulvar type alone.[11] This report uses the features of subcutaneous tissue infection, systemic toxicity, and pathological confirmation of necrosis as the major features of necrotizing fasciitis.

Early treatment is paramount which makes early disease recognition and individual risk assessment crucial. Local symptoms are present in early and late stages of the disease and were found in almost all patients. Systemic symptoms such as fever and WBC count >13,000 were present in 71% of our patients, which is consistent with the Stephenson study.[1] This supports the concept that the majority of patients will present with symptoms that should raises the index of suspicion. Combined with risk factor identification, it is possible to recognize those at high risk of morbidity and mortality. Although there are no established criteria to stratify illness by severity, high risk factors are known. Increased mortality has been associated with delayed treatment, diabetes, vulvar involvement, postpartum onset, WBC >30,000/microL, serum creatinine >2.0 mg/dL, clostridial infection, septic shock, and heart disease on admission.[2,8,12] Diabetes mellitus is particularly important to recognize because these patients have vulvar necrotizing fasciitis until proven otherwise. Prevalence of diabetes mellitus in this group ranges from 62%-97%.[1,8,9] Our study has a similar finding of 57%. The most common co-morbidity noted in our series was obesity at 71% which is similar to the 70%-87% found previously.[1,8]

The one mortality in this series was a 28-year-old woman who developed necrotizing fasciitis at the site of her episiotomy. Despite rapid debridement, the patient remained febrile and her WBC rose to 134,200. Septic shock soon followed and the patient died. This patient highlights that despite being young and healthy, high mortality persist when high risk factors such as vulvar or perineal surgery are present. Overall our mortality was low (14%) which is likely due to all patients receiving debridement within 24 hours of diagnosis and prompt initiation of broad spectrum antibiotics in all but one patient. This is comparable to the three in fourteen and one in eighteen mortality rates reported in recent papers for patients treated within 24 hours of diagnosis.[1,8] Interestingly, none of the diabetic women in this study died considering previous diabetic mortality rates of 35%-50%.[1,9,13] Perhaps the improved mortality in this study is the result of an increased index of suspicion leading to faster surgical debridement.

The findings in this study include the first reporting of Pacific-Islanders and Filipinas with vulvar necrotizing fasciitis in the English literature. 57% of the cases of vulvar and perineal necrotizing fasciitis were found in Pacific-Islanders, an ethnic group that made up just 9% of Hawai'i's population according to the 1995 census. Five of the seven patients, which included all the Pacific–Islanders, were without health insurance or had government provided insurance. This distribution suggests that socioeconomic factors may be a risk factor for vulvar necrotizing fasciitis.

To the best of our knowledge, there are three cases of recurrent necrotizing fasciitis in the English literature.[14,15,16] Our case of vulvar necrotizing fasciitis followed by necrotizing fasciitis of the back is the fourth case overall and the second with vulvar involvement. Only one of the four cases had recurrence in the same location. It would seem logical that if a particular anatomic location is susceptible to infection, the same location would be affected by any recurrent infection. This does not seem to be the case, but the number of cases is small and further study is necessary. Also noted is that with our patient, two of the four patients had MRSA isolated in both the primary and secondary infection. Perhaps a personal history of necrotizing fasciitis, especially with MRSA, predisposes to recurrence.

The limitation of the study is the low number of patients. Despite reviewing the records of necrotizing fasciitis and cellulitis cases in the only women's specialty hospital in the state of Hawai'i for the last 20 years, numbers were low. This is probably due to a lack of awareness of vulvar necrotizing fasciitis as a distinct entity and unclear coding conventions for vulvar and perineal disease until recently. It is also possible that as obesity and diabetes mellitus prevalence, and the number of elderly women increased, necrotizing fasciitis has also increased.

The key points in the management in vulvar and perineal necrotizing fasciitis remain rapid diagnosis, identification of high risk factors and immediate, aggressive debridement of all effected tissues. Broad spectrum antibiotics should be used initially and narrowed as culture results become available to minimize treatment delay. Adherence to this treatment regimen is the best explanation of the low mortality in our series.

Authors' Affiliation:
- OBGYN Resident, Duke University Medical Center, Durham NC. (J.N.)
- Assistant Professor, Department of OBGYN, John A. Burns School of Medicine, University of Hawai'i, Honolulu, HI. (R.B.)

Correspondence to:
John Nakayama MD
4504 Cherry Blossom Circle
Durham, NC
ph: (808) 352-0189
Email: jmn14@notes.duke.edu

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