Successful outcome of kleibsella necrotising fasciitis in an immunocompromised child with conservative measures

Greeshma R.1, Kancharla A.2, Latha M. Sneha3, Agarwal P.4

1Dr. Greeshma Rajeev, Final Year Postgraduate, 2Dr. Adarsh Kancharla, First Year Postgraduate, Department of Pediatrics, 3Dr. Latha M. Sneha, Associate Professor, Division of Pediatric Hemato Oncology, Department of Pediatrics, 4Dr. Prakash Agarwal, Head, Department of Pediatric Surgery, all authors are affiliated with Sri Ramachandra Institute of Higher Education and Research, No.1, Ramachandra Nagar, Porur, Chennai, Tamil Nadu, India.

Corresponding Author: Dr. Latha M. Sneha, Associate Professor, Division of Pediatric Hemato Oncology, Department of Pediatrics, Sri Ramachandra Institute of Higher Education and Research, No.1, Ramachandra Nagar, Porur, Chennai, Tamil Nadu, India. Email: drmslatha@yahoo.com

Abstract

Necrotizing soft tissue infections (NSTIs), are rare, very rapidly progressing disease of superficial fascia and subcutaneous cellular tissue. They are considered as secondary infections as they develop from an initial break down of skin integrity due to trauma or surgery. There is significant local tissue destruction causing grave morbidity and long-term sequelae. NSTIs are rare in children and the predisposing factors are skin damage due to trauma, diabetes, malnutrition and immunosuppression. Broad spectrum antibiotics, wide surgical debridement and supportive care are the standard treatment options. Diagnostic delays are common as the initial signs and symptoms mimics cellulitis or abscess. The diagnostic challenges in immunocompromised host are due to the facts that : i) infections are caused by diverse organisms that are usually not considered pathogens in healthy hosts; ii) infections of soft tissue might be a part of a bigger systemic infection; iii) immune deficiency masks the clinical signs. We report a case of successful outcome with nonsurgical management of necrotizing fasciitis in a boy with newly diagnosed Acute lymphoblastic anemia. He developed Klebsiella Necrotizing Fasciitis (NF) in the first week of induction therapy and his wound healed without surgical debridement or reconstructive measures, due to the high index of suspicion, early diagnosis and adequate intervention.

Keywords: Necrotizing fasciitis, Immunocompromised, Early diagnosis

Introduction

Necrotizing fasciitis, a life-threatening condition with a high mortality rate, is characterized by very rapid progression of the disease with significant local tissue destruction. Caused by toxin producing bacteria, the progression of the disease happens within hours and diagnosis is hindered as cutaneous manifestations does not reflect the rapid disease progression below the surface. Depending on the causative agent, Necrotizing fasciitis is divided into: Type 1 – polymicrobial, caused by a mixture of aerobic, facultative or anaerobic and type 2-monomicrobial, caused by Group A streptococci. Group 2 is a rapid and progressive disease with poor prognosis [1]. Group A B-hemolytic streptococcus was the most frequently isolated causative organism since Meleney’s findings [2]. In children with neutropenia, Pseudomonas was an important causal agent. [3] Radical surgical debridement is the mainstay of treatment and a delay of more than 12 hours has been found to be associated with increased incidence of septic shock and renal failure. With appropriate early treatment, Sudarsky et al reported a significant decrease in mortality from 50% to 0% [4].

Prognosis becomes poorer in the presence of co-morbidities, such as immunocompromised status– wherein they present atypically and cause significant delay in diagnosis.

Case Report

A 14-year-old boy, newly diagnosed with standard risk B cell Acute lymphoblastic leukemia was admitted for chemotherapy and PICC (peripherally inserted central cathether) was inserted in right hand. After first week of chemotherapy, he developed
fever spikes and was started on first line of iv antibiotics with piperacillin and amikacin. One day later, he developed an erythematous induration below the left cubital fossa. The lesion developed into an abscess, the following day. Incision and drainage of the lesion resulted in release of gas, and straw-colored fluid. Suspecting Necrotizing fasciitis, he was started on appropriate antibiotics and regular dressing was continued. The pus culture, PICC line culture and peripheral blood culture grew klebsiella and antibiotics were stepped up as per sensitivity. PICC line was removed. He continued to have fever spikes and the lesion started necrotizing rapidly [Figure 1].

![Fig-1: Gangrenous area with sloughing necrotic tissue in the center.](image1)

Meanwhile, he became severely neutropenic with absolute neutrophil count of 100/cubic mm and had severe thrombocytopenia – 0.4 x 10⁹ per litre. He was managed with regular and meticulous dressing of the wound with antibiotic lock as severe and refractory neutropenia and thrombocytopenia persisted for more than 2 weeks. He also had an episode of septic shock requiring IV fluid bolus and short term inotropes during this period and recovered uneventfully. Both peripheral blood culture and wound specimen were repeatedly positive for klebsiella, in 8 occasions. He also developed pulmonary aspergillosis during this period and an abscess over left lower leg which also grow as klebsiella.

In view of the moribund condition of the patient with refractory neutropenia and thrombocytopenia and lung infection, he could not undergo radical surgical debridement and hence his necrotizing fasciitis was managed by daily wound debridement and broad-spectrum intravenous antibiotics (586eropenem, clindamycin, linezolid, tigacyclin, teicoplanin, colistin, and oral posoconazole) for 4 weeks. Due to the non availability of granulocyte transfusions at our centre, he was managed with daily granulocyte colony stimulating factor injections (GCSF). 3 weeks after the onset of lesion, the wound started healing slowly, counts showed improvement and blood cultures and wound culture became sterile. Wound healed spontaneously with no need for reconstructive measures. [fig 2] We wanted to highlight the positive outcome of a non surgical debridement of necrotizing fasciitis in a boy with multiple risk factors, not needing skin grafts.

![Fig-2: Healed skin lesion with no residual deficit.](image2)
Discussion

Necrotizing Fasciitis is rare in childhood. The predisposing factors are minor injuries, surgical and traumatic wounds, varicella, causing a break in epithelial or mucosal surfaces. Necrotizing soft tissue infections (NSTIs) are commonly seen in the extremities, perineum and genitalia. NSTIs of the extremities typically occurs after extrinsic compromise of skin due to trauma or iv injections. Clinical manifestations of NF start after 1 week of the initiating event. Induration and edema are the presenting features with severe localized pain followed by erythema or purple discoloration 24 to 48 hours later [5].

The presence of gas is highly suggestive of NF. In the largest study of 39 cases of NF among children, the significant risk factor related to fatal outcome was immunosupression [6]. Appropriate antibiotics, wide surgical debridement and supportive care are the treatment measures. Even with optimal treatment, necrotizing soft tissue infections (NSTI) have 25-35% mortality rates and NSTIs has 2 fold higher mortality in immunocompromised individuals [7].

As diagnosis is based on clinical suspicion, the typical skin changes from purple red to dusky blue should be recognized as the first sign, as it is followed by rapid necrosis. Our patient had all the risk factors – active malignancy, steroids, iv chemotherapy and percutaneous catheter insertion.

Early radical surgical debridement to remove the infected and necrotic tissues is the cornerstone of treatment and inadequate and delayed debridement has been found to be associated with increased mortality. The early signs and symptoms of NSTIs are similar to that of an abscess or cellulitis, thereby causing a delay in the diagnosis. But the rapid progression of the lesion even with appropriate iv antibiotics and systemic toxicity should alert the possibility of NSTIs [7].

The diagnosis is hindered due to the fact that the cutaneous manifestations mask the severity of disease and the disease progresses below the surface. With the dissemination of the disease, the patient develops pain and signs of systemic toxicity, which seems disproportionate to skin examination findings. Some of the hard-clinical signs that are more suggestive of NSTIs but occur late in the course of disease are: bullae, skin ecchymosis, presence of gas in tissues, cutaneous anesthesia. Other less specific signs include: pain disproportionate to clinical findings, edema extending beyond skin erythema, systemic toxicity, disease progression despite antibiotic therapy [8].

The presence of liquid necrosis of the superficial fascia during the surgical debridement confirms the diagnosis. Histologically, necrosis of the superficial fascia, leukocytic infiltrates with polymorphonuclear cells predominance in fascia, subcutaneous fat tissue and dermis, arterial and venous fascial thrombosis, angiitis with fibrinid necrosis can be seen [9].

According to skin manifestations, Wang et al defined 3 stages for Necrotising fasciitis: early, intermediate and late. Patients in stage 1 presented with swollen and warm skin, tenderness to palpation, erythema. Stage 2 had blister and bullae formation and skin fluctuance and induration. In stage 3, hemorrhagic bullae, skin anesthesia, crepitus and skin necrosis were noticed [10].

The non specific lab parameters associated with NF are: abnormally high or low WBC counts, elevated BUN and Creatinine values, decreased serum sodium, elevated CRP, elevated serum lactate and elevated serum creatine kinase (CK) [11]. LRINEC score (laboratory risk indicators for necrotizing fasciitis) that incorporates the CRP, White cell count, Hemoglobin, sr. sodium, glucose, creatinine values, have been found to have a higher positive predictive value (94.3%) when the score is >8 [8].

The systemic signs and symptoms are due to toxic process and septicemia. Tissue edema causes depletion in vascular volume and progresses to hemoconcentration, hypotension and shock. Surviving the medical and surgical emergencies, patient might land up in long hospitalizations for multiple reconstructive interventions and intensive rehabilitation measures. Mortality was found to be higher in patients who developed shock and end organ damage, approaching 50-70% [12]. To achieve optimal outcomes, treating physicians should have a high index of suspicion for these rare NSTIs in immunocompromised patients as they have multiple comorbid conditions. A multi-disciplinary team is essential to maintain functional quality of these patients and reduce the risk of long-term morbidity.

Conclusion

A rare clinical identity, NF is a severe multisystemic disorder and early diagnosis and treatment is essential, as symptoms and signs are disproportionate to the clinical findings and the time to antimicrobial therapy and time to debridement decides the outcome.

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