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Abstract

Ecthyma gangrenosum (EG) is a rare, necrotising manifestation of cutaneous or systemic infection, most often associated with Pseudomonas aeruginosa. Other causative organisms include: Staphylococcus aureus, Aeromonas hydrophila, and fungi including Candida and Aspergillus species. EG can present as indurated, violaceous, or black lesions and can rapidly progress to frank tissue necrosis. Typically, this condition occurs in immunocompromised patients, such as those with malignancy or severe burns. Early diagnosis is imperative, and management requires to be aggressive, with initiation of high-dose, broad-spectrum antibiotics, providing Pseudomonal cover and urgent surgical debridement. Despite optimal medical and surgical management, mortality rates remain high.

We present a case of Pseudomonas associated EG with photographs in a 17-month old girl with 56% total body surface area (TBSA) partial-thickness scald burns and propose a management algorithm based on review of the literature.

Keywords: Invasive, pseudomonas, ecthyma, gangrenosum, burns, infection, paediatric

BACKGROUND

Approximately 250,000 people suffer burn injuries in the UK each year, with around 5-10% requiring hospital admission [1]. Infection is a major cause of morbidity and mortality, particularly at extremes of age. [2,3] The most commonly isolated microbe from burn wounds is Staphylococcus Aureus. [4] However, multi-resistant organisms, including Pseudomonas species, are the most common infective organisms that cause mortality. [5] Pseudomonas wound infection is typically managed with topical antimicrobial agents. When systemic invasive infection occurs, it is associated with high mortality rates. [3,6,7]

CASE PRESENTATION

A 17-month old girl was transferred to the Regional Paediatric Burns Centre (Shriners Hospital for Children Northern California) 36 hours after sustaining a 56% TBSA area partial-thickness scald to her head, neck, torso, and all four limbs. On arrival, she was appropriately resuscitated, and her wounds were cleaned and dressed with Bacitracin.

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At 72-hours post-injury, her clinical condition deteriorated. The patient developed oliguria, oedema, metabolic acidosis (pH of 6.94) and respiratory failure requiring high frequency mechanical ventilation. Within hours, the patient became leucopenic and round violaceous lesions developed within her burns (Figure 1). A clinical diagnosis of EG was made and she was taken to the operating theatre (Figure 2). The subcutaneous tissue below her wounds was injected with piperacillin, urgently excised and covered with allograft. The causative organism, Pseudomonas *aeruginosa*, and the presence of tissue invasion, was confirmed through histological culture and analysis. She was commenced on intravenous antimicrobial therapy. She required a laparotomy due to developing abdominal compartment syndrome and the open wound was covered with temporary silastic mesh. Vasopressors were required to maintain adequate end-organ perfusion. She had three more procedures on the subsequent three days where her abdomen was re-explored, and the remaining burn wounds were excised and covered with allograft.

The patient continued to deteriorate with increasing ventilatory pressure requirements resulting in bilateral pneumothoraces, renal failure requiring dialysis and bowel ischaemia. Her clinical condition deteriorated to the point where the decision to withdraw care was made following discussion with her family.



Fig1. Widespread violaceous lesions characteristic of Ecthyma Gangrenosum in burn wounds to the head, neck, torso and limbs.



Fig2. Surgical debridement of Ecthyma Gangrenosum with laparotomy and allograft application. a) widespread violaceous lesions of EG, b) tangential excision of infected burn wounds, c) laparotomy demonstrating intraabdominal contents, d) placement of allograft, e) application of Silastic mesh to protect abdominal contents, and f) wound dressing with abdominal window.

Ecthyma gangrenosum (EG) is a rare, necrotising, manifestation of either cutaneous or systemic infection. It presents as indurated, violaceous or black cutaneous lesions most often associated with invasive Pseudomonas aeruginosa infection in an immunocompromised patient, such as those with malignancy or severe burns. [8] Infection occurs either through direct cutaneous invasion or by secondary haematogenous spread. Other causative bacteria include Staphylococcus aureus and Aeromonas hydrophila, which are associated with EG in approximately 17% of cases; fungi including Candida and Aspergillus species are isolated in 9%. [9,10] EG has also been reported in previously healthy patients. [11,12] This may result from a temporary disruption to host defences due to a recent infectious illness or antibiotic therapy.

Pseudomonas aeruginosa, an opportunistic gramnegative bacillus, can originate from both endogenous and exogenous sources. [3,6] Its propensity to cause infection in burns is thought to result from its preference for moist environments and the prolonged use of empirical antibiotics and invasive devices for these patients. [13] Typical Pseudomonas wound colonisation appears yellow or green in colour. [14] This colonisation or infection is often managed with topical antimicrobial agents in a systemically stable patient. [15] Effective local control of burn-wound infection is important in order to reduce the risk of progression. The presence of indurated, black or violaceous lesions can indicate the presence of invasive infection. [14,15] These may progress rapidly (12-24 hours) from small vesicles to necrotic ulceration and black eschar formation. [16] Pyrexia, haemodynamic instability, dyspnoea, and altered consciousness are typical signs that may precede the development of cutaneous lesions. [17] In children, antecedent diarrhoea occurs in around 30% of cases.

Mafenide acetate was developed to prevent this devastating disease. Before its development, 70% of fatal burn wound sepsis involved *Pseudomonas aeruginosa*. The development of this antimicrobial greatly reduced this deadly invasive infection. [18,19] This child presented with superficial wounds, so she was treated with bacitracin which covers only grampositive bacteria. Prophylactic treatment with better gram-negative coverage may have prevented this complication.

The investigation of suspected EG should include sending specimens for microbiology and histopathology to confirm the diagnosis and guide future antimicrobial therapy, but this should not delay expedient management (see Figure 3). [14] Microscopic appearances are of invasion of gramnegative rods into the interstitium with involvement of the adventitia and media of subcutaneous vessels. This results in a haemorrhagic occlusive vasculitis which precipitates development of cutaneous lesions. [8] A high index of suspicion is required to allow timely diagnosis and urgent management. The condition is associated with high mortality rates and should therefore prompt expedient management. [8]



Fig3. Ecthyma Gangrenosum management algorithm. EG = Ecthyma Gangrenosum. Adapted from [16,20,21].

Initial management of EG is aggressive and includes the use of high-dose, broad-spectrum systemic antibiotics until a causative organism(s) and sensitivities are identified. [16] Choice of antimicrobial therapy depends on local guidance; typically, a combination of a beta-lactam penicillin with an aminoglycoside or fluoroquinolone is advised. [20,21] Antifungal therapies may be added dependent upon the clinical suspicion. Predisposing factors for the development of fungal burn wound infection include: increased age and wound size, immunosuppression, poorly controlled diabetes, broad-spectrum antibiotic use and the presence of indwelling catheters. [22] Urgent surgical debridement is indicated, particularly in the context of extensive cutaneous infection to reduce the bacterial load and to reduce chances of progression.

The mortality rate of systemic *Pseudomonas aeruginosa* infection is up to 77%, despite the advent of antimicrobial therapies. [23] In contrast, the mortality associated with localised cutaneous EG is reported to be much lower at 15%. [24] Prompt identification and management of the condition therefore may have a significant impact upon the likelihood of survival.

Burns surgeons should be aware of the clinical signs of EG and recognise that expedient multimodal therapy including resuscitation, antimicrobials and early surgical excision, even in the presence of a partial thickness injury, may be indicated to reduce bacterial load and the associated risk of mortality.

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