CASE REPORT

ECTHYMA GANGRENOSUM: CASE REPORT OF 5-MONTH-OLD FEMALE

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Ecthyma gangrenosum is a rare and severe skin disease that is typically accompanied by a systemic infection caused by Pseudomonas aeruginosa bacteria. It manifests as painful, necrotic lesions surrounded by redness and including a noticeable black eschar in the centre. The majority of ecthyma gangrenosum instances occur in immunocompromised individuals, such as those with significant underlying medical conditions or those who are neutropenic. We present a case of 5-month-old vaccinated female child, treated for dengue fever 2 weeks back presented to the ER at our facility in a critical state with complaints of pus draining from the right ear, right thigh discoloration and swelling, and swelling of the left cheek, along with difficulty in feeding. On examination, she was afebrile, capillary refill test of more than 4 seconds and random blood sugar of 192 mg/dL. On physical examination, her GCS was 8/15, her pupils were reactive to light, and she was responsive to pain, she appeared pale, and her skin and peripheries were cold. Following the detailed history and physical examination findings, a provisional diagnosis of septic shock and Ecthyma Gangrenosum was established.

Keywords: Ecthyma Gangrenosum; Septic shock; Pseudomonas aeruginosa; Necrotic skin lesions; Rare skin infection

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INTRODUCTION

Ecthyma Gangrenosum (EG) is an uncommon cutaneous infection, classically developing in 1.3–13% of patients with pseudomonas aeruginosa sepsis, predominantly occurring in critically ill or immunocompromised individuals.[1,2] Ecthyma Gangrenosum lesions primarily involve axillary and anogenital areas, however, the face, arms, trunk, and legs may also be affected. The classic macroscopic appearance mainly comprises rapidly progressing skin lesions that cause inflammation and necrosis of the skin, thereby, appearing as gangrenous ulcers with erythematous borders.[3] Ecthyma Gangrenosum is notorious for a poor prognosis amongst immunocompromised individuals with neutropenia.[4] Henceforth, we present this case study on the aforementioned clinical condition. An informed consent statement was obtained for this study.

CASE PRESENTATION

A 5-month-old vaccinated female child, weighing 7kg, presented to the ER at our facility in a critical state with complaints of pus draining from the right ear for the past 20 days, right thigh discoloration and swelling for 10 days, and swelling of the left cheek for 1 week, with difficulty in feeding since last night.

Eighteen days back, she was admitted to the PICU at our facility for 3 days with diagnosis of dengue fever with warning signs. Upon discharge, she was advised to follow up in 3 days. Following hospital discharge, she presented to the otorhinolaryngologist with complaints of pus draining from the right ear and discoloration of the right thigh. Despite being advised prompt admission and management with IV antibiotics, her family refused admission. Aural swab was sent.

On day 16, following her previous admission, she presented in the ER in critical state. On examination, she was afebrile, pulse was 174 beats per minute, respiratory rate of 50 breaths per minute, blood pressure of 97/67mmHg, oxygen saturation was 60% with NRB and 98% on 2 liters of oxygen, capillary refill test of more than 4 seconds and random blood sugar of 192 mg/dL. Her GCS was 8/15, pupils were reactive to light, she appeared pale, and her skin and peripheries were cold. The right thigh had a gangrenous skin lesion measuring 5cm x 3cm with purulent discharge from the center of the gangrene (Figure 1). It was associated with purulent discharge from the right ear (Figure 2) and a deep ulcerated lesion on the hard palate. Chest examination revealed increased respiratory effort with an irregular breathing pattern and bilateral harsh vesicular breathing on auscultation. Aural swab results revealed Pseudomonas Aeruginosa and Enterobacter species sensitive to meropenem, tazobactam, and gentamicin. Following the detailed history and physical examination findings, a provisional diagnosis of septic shock and Ecthyma Gangrenosum due to pseudomonas aeruginosa was established. Differential diagnoses of ecthyma

clinically, including conditions such as Necrotizing Fasciitis and Purpura Fulminans were considered but typical skin lesions along with growth of pseudomonas aeruginosa on culture favoured ecthyma gangrenosum. Initial resuscitation was started following the primary assessment, the patient was admitted to the PICU, a cardio-respiratory monitor was attached, she was started on 2 liters of oxygen, an IV line was maintained and three boluses of Inj Normal Saline was given at 20 ml/Kg each. 1st dose of antibiotics; IV tazobactum and Vancomycin was administered as it was fluid refractory shock so inotropic support was also started. She was eventually intubated and kept on a mechanical ventilator with high parameters. The laboratory investigations revealed: Hb - 8.0 g/dl, TLC - 8.6 x 10E9/L with neutrophils 78%, lymphocytes 12%, Platelets - 109 x 10E9/L, Urea - 29 mg/dl, Creatinine - 0.60 Meq/l, sodium - 146 Meq/l, potassium - 3.5 Meq/l, chloride - 99 Meq/l, bicarbonate - 17 Meq/l, CRP - > 200 Mg/L, D-Dimer - 2468 ng/ml FEU, PT - 44, APTT - 64, INR - 4. Dengue serology for IgG and IgM was positive. The blood culture and sensitivity revealed coagulase-negative staphylococci. A multidisciplinary team including ENT specialist, surgeon, infectious disease specialist, was involved in the case. Aural toilet was done pus was sent for CS. As ecthyma gangrenosum has rarely been reported in previously healthy individuals so to rule out any underlying immunodeficiency workup including immunoglobulin levels and flowcytometry was advised but parents refused. Chest X-rays were performed (Figure 3) which showed linear infiltrates more on the left side. On day 3, the patient's condition worsened as the oxygen saturation deteriorated. ABGs were performed which revealed hypoxia. FFPs and PCV were transfused. The antibiotics were changed to meropenem, ceftazidime and antifungal, Fluconazole, was added. Epinephrine and Dobutamine were administered as the patient consistently remained hypotensive. Despite the aggressive intervention, on day 3, the patient collapsed, and efforts to resuscitate and revive the patient failed and she expired.

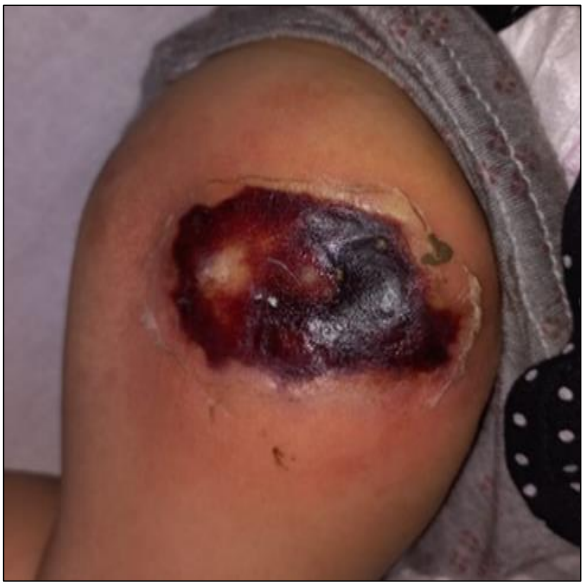


Figure-1: Right thigh gangrene 5cm x 3cm along with purulent discharge from center of gangrene

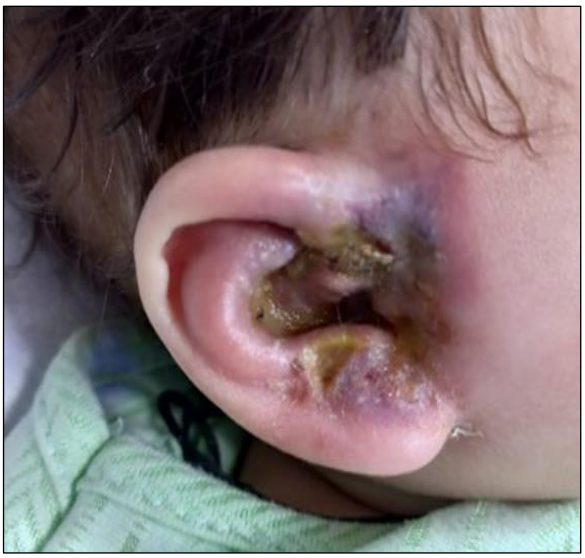
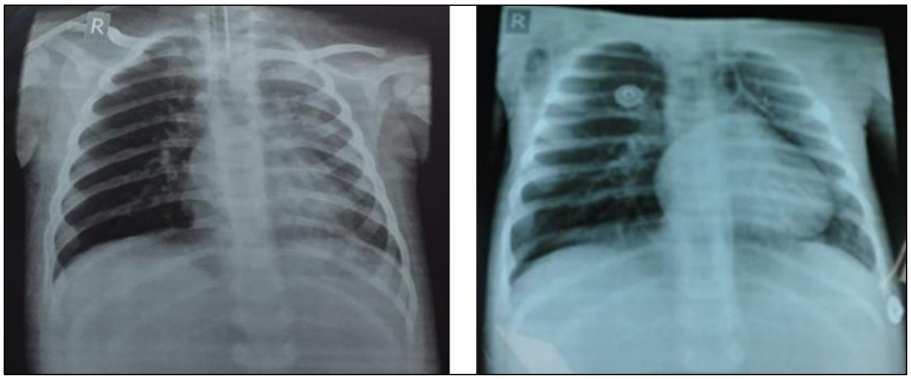


Figure-2: Purulent discharge from right ear.



Chest X-ray at day 1 of admission. **Chest X-ray at day 3 of admission.**

Figure-3: Chest X-Ray, Frontal View showing linear infiltrates that are most prominent on the left side

DISCUSSION

Ecthyma Gangrenosum lesions begin as painless, round erythematous macules and patches that progress to central pustules with surrounding erythema. At the center of the lesion, a haemorrhagic vesicle forms, which then develops into a gangrenous ulcer with a black eschar.[2,5] In as little as 12 hours, early lesions can progress to necrotic ulcers. Lesions can be localized and solitary, or they can be widespread.[6] Although EG can manifest anywhere on the body, it most frequently affects the axilla and anogenital regions. The most frequent location (57%) is the gluteal/perineal region, followed by the extremities (30%), the trunk (6%), and the face (6%).[5] In our patient, the characteristic skin lesion, associated with Ecthyma Gangrenosum, was present on her right thigh and hard palate, and purulent discharge from the right ear. The lesions were not present in the locations commonly involved in EG. The patient presented to the ER after substantial progression of the disease. The lesion on the right thigh had become gangrenous with purulent discharge from the center and the oral lesion had become deeply ulcerated. Detailed history, thorough physical examination, and prompt workup are extremely crucial for early diagnosis of EG.[7] Blood cultures, wound cultures, urinalysis, chest radiographs, complete blood count, comprehensive metabolic panel, C-reactive protein, and HIV testing are all part of the initial evaluation. In the setting of sepsis, procalcitonin and lactate levels are also important.[8] Ecthyma Gangrenosum is classified as bacteremic or non-bacteremic. In comparison to the non-bacteremic type, bacteremic EG is more prevalent. The pathogenic bacterium spreads hematogenously to the capillaries and invades the media and adventitia of arteries and veins in bacteraemia. Perivascular invasion eventually leads to ischemic necrosis of the surrounding skin.[9] Lesions arise at the site of direct inoculation into the epidermis in the non-bacteremic variant. Exotoxin A, elastase, and phospholipase-C are virulence toxins and enzymes generated by Pseudomonas Aeruginosa that damage the skin and vascular system.[10] EG treatment has 3 stages: initial empiric antibiotic therapy is administered as soon as infection is suspected; when the aetiology is established, aggressive antibiotic or antifungal treatment is administered; and finally, surgical excision is often necessary because EG manifests as a necrotizing soft-tissue lesion. Our patient's treatment plan was constructed in accordance with Stage 3. Initially, she was on antibiotics but failed to produce any improvement in the lesion, so we changed her antibiotics and scheduled her for wound debridement, but before that, the patient passed away.

The patient's ear swab culture and sensitivity for purulent discharge revealed Pseudomonas Aeruginosa and Enterobacter as the infectious agents, thereby confirming our suspicion. The underlying immunosuppressive state (e.g., cancer, renal failure, or other) is frequently the most important predictor of clinical outcome. The most significant prognostic factor for mortality is neutropenia at the time of diagnosis.[11] The multi-drug resistance of P. aeruginosa has significantly developed recently, making treatment more difficult. After acquiring blood and wound cultures, urgent therapy with broad-spectrum antibiotics should be started in patients with suspected Ecthyma Gangrenosum.[12] Antipseudomonal beta-lactams (piperacillin/tazobactam), cephalosporins (cefepime), fluoroquinolones (levofloxacin), and carbapenems (imipenem) are among the first-line empiric treatments. In high-risk patients, such as those with neutropenia and septic shock, combination therapy is advised. Antimicrobial coverage should be refined once the etiologic organism and antimicrobial susceptibilities have been determined.[13] Antifungal and antiviral medicines should be initiated in rare cases with a fungal or viral origin. In our patient, we advised prompt admission for targeted combination therapy due to the severe progression of the disease, and owing to the suspicion of immunodeficiency, antifungal treatment was also included. Necrotic lesions or abscesses frequently require surgical excision. For infection source control, abscesses must be cut open and drained. Aggressive debridement and grafting may be required for necrotic skin lesions greater than 10 cm.[14] Due to the high case fatality rate, patients with sepsis or neutropenia should receive treatment at the hospital's intensive care unit. Likewise, we admitted our patient to the intensive care unit to allow for thorough management. EG is a potentially fatal illness that is frequently worsened by septic shock. In individuals with sepsis, the development of EG lesions is associated with a poor prognosis and is fatal in 38–77% of patients.[15] Unfortunately, our patient was a classic example of complicated bacteremic EG without neutropenia. Therefore, despite adequate measures and thorough management, the fatality of the condition could not be overturned and she collapsed on day 3 of admission to the intensive care unit.

CONCLUSION

Ecthyma gangrenosum is an uncommon cutaneous manifestation known for its association with Pseudomonas aeruginosa sepsis. Owing to its rarity, it poses a challenge to clinicians in the early diagnosis and management of the patient. Early recognition of EG plays an important role in providing appropriate empiric antibiotic treatment early in the development of characteristic lesions and improves the outcome. The patient presented to us with septic shock, and

despite providing appropriate antibiotics, the patient collapsed. Efforts to resuscitate and revive the patient failed, and she expired.

However, these lesions present in a predictable pattern, ranging from erythematous nodules to necrotizing ulcers with surrounding erythema. On account of its high morbidity and mortality, to achieve optimal results, prompt clinical recognition and proper antibiotic therapy are required.

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