Ecthyma gangrenosum leading to fatal outcome in a 1 year old previously healthy boy

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ABSTRACT

Background

Ecthyma gangrenosum is a skin lesion with fatal prognosis, the most common cause being the infection with Pseudomonas aeruginosa. It usually occurs in patients with immunodeficiency or in critically ill patients with chronic diseases. Ecthyma gangrenosum in previously healthy individuals is rare.

Clinical presentation

A1 year old boy was admitted to our paediatric ward with a history of fever, vomiting and diarrhoea for the past 10 days. Necrotic lesions characteristic of Ecthyma gangrenosum developed on the left side of his neck.

Diagnosis

Ecthyma gangrenosum associated with Pseudomonas aeruginosa infection.

Intervention

Empiric treatment was initiated with Intravenous Meropenem (40mg/kg/dose).

Outcome

Severe necrotic and hemorrhagic lesions spread all over the body. The patient's condition deteriorated with multiple organ failure. Unfortunately, the patient died on day 15 of hospital admission.

Lesson

Timely recognition of Ecthyma gangrenosum and appropriate antibiotic therapy might improve the patient outcomes. Also, immunologic evaluation should be performed to rule out and treat if immunodeficiency is present.

Keywords : Ecthyma gangrenosum, ecthyma lesions, Pseudomonas infection, Pseudomonas aeruginosa, healthy infant

1 INTRODUCTION

Ecthyma gangrenosum (EG) is a skin lesion. The most common cause of this skin manifestation is the infection with Pseudomonas aeruginosa. This condition is of clinical significance as it is associated with fatal prognosis. Other rare causes of this skin manifestation are infections with Escherichia coli, Proteus species, and methicillin-resistant Staphylococcus epidermidis. (1,2) The characteristic lesions of EG are hemorrhagic vesicles or pustules that evolve into necrotic ulcers with a tender erythematous border. (3) EG usually occurs in patients with immunodeficiency or in critically ill patients with chronic diseases. EG in previously healthy individuals is rare. (4) Very few case reports of Ecthyma gangrenosum in previously healthy individuals have been mentioned in the literature. We report a case of an immunocompetent patient without any previous chronic disease developing fatal EG.

2 CASE PRESENTATION

A 1 year old boy was admitted to our paediatric ward with a history of fever, vomiting and diarrhoea for the past 10 days.The patient became lethargic with impaired consciousness 2 days prior to the admission. Necrotic lesions developed on the left side of his neck (Figure 1). He had a history of cow milk ingestion and animal contact. His brother died 8 years back due to similar condition. There is no history of chronic medication and the patient had received appropriate immunization.

On admission,the patient appeared to be more toxic.He was irritable,drowsy, and lethargic with a capillary refill time of more than 3 seconds. Physical examination revealed the presence of many black necrotic lesions on his chest, back, thigh and face. The largest lesion was on left side of his neck measuring 6 cm surrounded by an erythematous ring, endured center with blood discharge. The patient presented with a low

blood pressure of 65/34 mm Hg, heart rate of 142 and temperature 38°C. He was tachycardic with no murmurs. On palpation, abdomen was soft with hepatomegaly 3 cm below costal margin. The laboratory investigations were as follows: Haemoglobin: 9g/dl, WBC count: 1.5x103/μL, absolute neutrophil count: 97.5, platelets:109,000/mm3, PT: 15, INR: 1.3, PTT:37, ESR: 48, CRP: 16.4. The cultures of CSF, blood and wound swab were positive for Pseudomonas aeruginosa.

Skin biopsy revealed extravasated red blood cells, scattered mixed inflammatory cells including few neutrophils along with congested capillaries and blood vessels in the superficial and deep dermis. Fibrin thrombi were seen in some of these capillaries and blood vessels. And, vasculopathy in the lower venules and necrosis can be seen pointing to infection. Bilateral infiltration was seen on chest X-ray. Neck ultrasound shows mild diffuse subcutaneous edema associated with enlarged loco-regional lymph nodes.

After collecting the blood, CSF and wound swab samples, empiric treatment was initiated with IV Meropenem (40mg/kg/dose). Next day, the patient suffered respiratory distress and was transferred to pediatric intensive care unit. The patient was intubated and the inotropic support was provided to normalise the low blood pressure. On day 5, severe necrotic and hemorrhagic lesions spread all over the body. The patient's condition deteriorated with multiple organ failure which necessitated transfusion of packed blood cells,platelets and fresh frozen plasma, continuous inotropic support and intensive care management. Despite of the intensive management, the patient died on day 15 of the hospital admission.

3 Discussion

Pseudomonas aeruginosa, being the most common cause of EG often leads to fatal outcomes. EG usually occurs in patients with immunodeficiency or in critically ill patients with chronic diseases. However, EG in previously healthy individuals is rare. (4) Very few case reports are available in the literature reporting EG in previously healthy infants. (5, 6, 7) Most of these patients were female less than 3 years old. The common presenting symptoms were fever, diarrhoea, lethargy, impaired consciousness, respiratory distress and necrotic lesions as seen in our patient. Pseudomonas aeruginosa sepsis was the main etiology of developing Ecthyma gangrenosum in these patients. (5, 6, 7) Among all the cases published on EG till date, Pseudomonas aeruginosa as the causative organism was detected in more than three fourths of the cases followed by Escherichia coli, Staphylococcus aureus, Aeromonas hydrophilia, and Mucor species. (7) In our patient, the cultures of CSF, blood and wound swab were positive for Pseudomonas aeruginosa.

The mortality rate of EG due to Pseudomonas aeruginosa infection reported in the literature is as high as 50%. (5) The mainstay of therapy includes beta lactam antibiotics and aminoglycosides, as a single agent or in combination. (5) Meropenem, a beta-lactam antibiotic was used as an empiric treatment in our patient as per the standard recommendations. Some authors also recommend a combination therapy of beta-lactam with aminoglycosides, as monotherapy may lead to resistance. (7)

Huang et al reported that fever and diarrhoea are the most common presenting symptoms in patients with EG. (8) In our case, the patient presented with a history of fever, vomiting and diarrhoea 10 days before admission.

It is highlighted that the toxins produced by the Pseudomonas results in severe neutropenia (9), which can be seen in our patient. Blood transfusion was performed and our patient was provided with intensive management.

The outcome of EG associated with Pseudomonas aeruginosa infection depends on various risk factors. (7) The presence of unknown immune deficiency and a family history of this condition is the major risk factor. In our case, the patient's brother dies of a similar condition. The next important risk factor is leucopenia. (10) Huang et al reported that around half of the patients with the diagnosis of EG were leukopenic at admission, and 9 out of 10 dead patients had leukopenia. (8) Blood investigations have shown that our patient had low WBC count (1.5x103/μL) on admission. Other risk factors include delayed or inappropriate antibiotic treatment, presence of multiple lesions and septic shock. (8) In our case, the patient condition deteriorated with severe necrotic and hemorrhagic lesions spread all over the body. Thus, the presence of these risk factors in our patient (leucopenia and presence of multiple lesions) might have led to the fatal outcome.

According to Pickard and Llamas, the management of EG needs to be done in an order depending upon the suspicion of EG diagnosis, culture test results and severity of lesions. As soon as the suspicion of EG arises, the patient should be immediately put on the effective empiric therapy. After the confirmation of diagnosis with culture reports, empiric antibiotics should be shifted to specific organism based antimicrobials. In severe cases, surgical removal can be considered. (11)

Even though, appropriate empirical antibiotic treatment was given in our patient, his condition deteriorated rapidly with multi organ failure and finally led to the death.

4 Conclusion

This case report demonstrates that EG is not uncommon in previously healthy individuals. Pseudomonas aeruginosa infection is the cause of EG in our patient. In our case, the major risk factors which might have led to the fatal outcome in the child may be leucopenia and rapid development of multiple lesions. Timely recognition of EG, rapid initiation of empiric antibiotic therapy, and shifting to appropriate antimicrobial agents as soon as the culture reports are available plays important role in improving the patient outcomes. Also, immunologic evaluation should be performed to rule out and treat if immunodeficiency is present. Surgical removal can be considered if antibiotic therapy does not improve the patient's condition.

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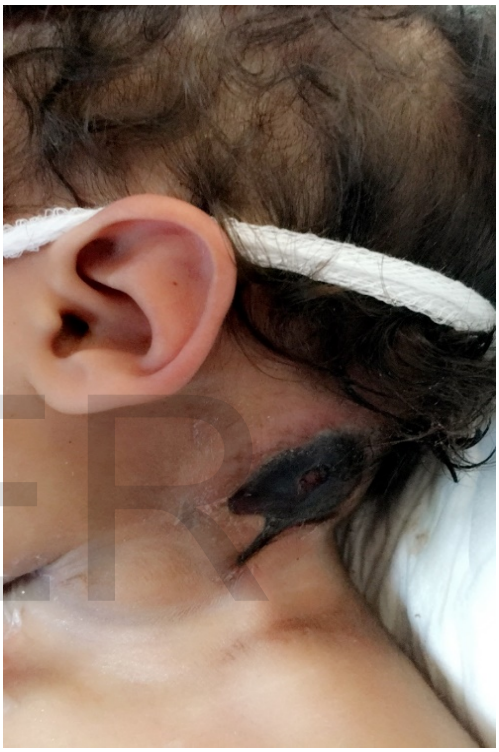

Figure 1. Necrotic lesions developed on the left side of the child's neck