JPRAS Open 40 (2024) 215–221

Contents lists available at ScienceDirect

JPRAS Open

journal homepage: www.elsevier.com/locate/jpra

Case Report

Ecthyma gangrenosum: A case report in a child with acute lymphoblastic leukaemia

I Yonjan Lama[a,*], LK Cheung[c], A James[b], M Saghir[b], F Herd[d], J Greenhowe[a]

[a] Royal Aberdeen Children's Hospital, Department of Plastic Surgery, Aberdeen, Scotland, UK
[b] Aberdeen Royal Infirmary, Department of Plastic surgery, Aberdeen, Scotland, UK
[c] Guy's and St Thomas' NHS Foundation Trust, London, England, UK
[d] Royal Aberdeen Children's Hospital, Department of Oncology, Aberdeen, Scotland, UK

ARTICLE INFO

Article history:
Received 4 December 2023
Accepted 18 February 2024
Available online 23 February 2024

Keywords:
Ecthyma gangrenosum
Leukaemia
Neutropenia
Paediatrics
Pseudomonas aeruginosa

ABSTRACT

Aim: We present a case of Ecthyma gangrenosum (EG) affecting left thigh in a child with acute lymphoblastic leukaemia (ALL) with an aim to raise awareness about this condition.
Case presentation: A 7-year-old female child who presented with lethargy, pallor and lumps to inner lip was diagnosed with B-cell precursor ALL. She was started on treatment as per UKALL 2011 guidelines Regime B. On day 28, she developed neutropenic sepsis along with a new lesion in her left thigh. She was started on intravenous Meropenum, Gentamicin and Caspofungin. The clinical diagnosis of EG was made based on lesion progression, positive blood and wound swab & tissue cultures for *Pseudomonas aeruginosa* and patient's immunocompromised status. The wound healed with secondary intention following debridement. We present a series of photographs to demonstrate her remarkable improvement.
Discussion: EG occurs in 1–30% of cases of Pseudomonas sepsis; other bacteria and fungi can be associated with this condition. It is identified more in oncology patients as seen in our patient. A multidisciplinary team approach should be provided in 3 stages with empirical antibiotics, followed by targeted antibiotics or antifungals

* Corresponding author.
E-mail address: indira.yonjan@nhs.scot (I.Y. Lama).

https://doi.org/10.1016/j.jpra.2024.02.010

& surgical debridement. Our patient was treated in similar fashion and made a good recovery.
Conclusion: It is a rare skin condition associated with a high mortality. We suggest all clinicians to be vigilant about this condition to be able to provide accurate diagnosis and prompt treatment to improve the overall prognosis.

Introduction

Ecthyma gangrenosum (EG) is a rare condition, first described by Canadian pathologist Dr Lewellys Baker in 1897.[1] It is defined as a presentation of *Pseudomonas aeruginosa* septicaemia in skin as a rapidly spreading necrotic lesion. The lesion can present as primary Ecthyma gangrenosum without bacteraemia due to localised inosculation of organism or in a classical form with bacteraemia through hematogenous spread.[2,3] The commonly affected areas are anorectal and gluteal areas (57%), extremities (30%) but can involve any part of the body.[4] We present a case of Ecthyma gangrenosum affecting left thigh in a child with acute lymphoblastic leukaemia (ALL) with an aim to raise awareness about this rare condition.

Case presentation

A seven-year-old female child was admitted to paediatric ward with one-week-old history of lethargy, pallor and blistering lesions on her lips. She underwent thorough investigations including full blood count, blood film and bone marrow biopsy. At the time of presentation, her haemoglobin was 92 gm/litre and white cell count was 38×10^9 /L and it raised to 55.5×10^9 /L. The blood film demonstrated normochromic normocytic anaemia, a few nucleated RBCs, neutrophils and monocytes included dysplastic forms along with a significant population of blasts of varying sizes demonstrating the features are of acute leukaemia. Bone marrow biopsy showed uniform infiltrate of blasts which were positive for CD19, CD10, CD34, cCD22, HLA-DR and nTdT makers suggesting acute B-cell precursor variant of ALL. The cytogenetics further revealed abnormal female karyotype with trisomy 21 and three copies of RUNX1 by FISH. Following the discussion in the Managed Service Network (MSN) for Cancer in Children and Young People National Leukaemia Multidisciplinary Team Meeting, she was started on chemotherapy as per UKALL 2011 guidelines Regime B induction escalated to Regime C.

During her treatment, she became confused, agitated, and developed hallucinations for which she was transferred to high dependency unit care. Electroencephalogram (EEG) showed no evidence of clear seizure like activity but there was an excess of slow wave activity suggesting possible chemotherapy related neurotoxicity. Chemotherapy was stopped on day 22. She further developed urinary retention and choreiform movements for which she was started on a midazolam infusion and her symptoms improved dramatically.

On day twenty-eight of her admission, she developed neutropenic sepsis with positive blood cultures for *Pseudomonas aeruginosa*; she was also noted to have a new skin lesion in her left thigh for which she was referred to Plastic surgery team. Clinical examination revealed tender and swollen thigh along with the lesion itself being 2 cm nodule within the necrotic patch of 3 cm diameter with surrounding erythema. She was started on IV Meropenum, Gentamicin and Caspofungin. The lesion was initially managed conservatively with daily dressing till day six following its initial appearance, when it ulcerated with discharge and developed severe tenderness; the wound swab confirmed growth of *Pseudomonas aeruginosa*. The clinical diagnosis of EG was made based on the progression of lesion, positive blood & wound cultures for *Pseudomonas aeruginosa* and patient's immunocompromised status. Debridement of the lesion was carried out under general anaesthesia on day eight; the

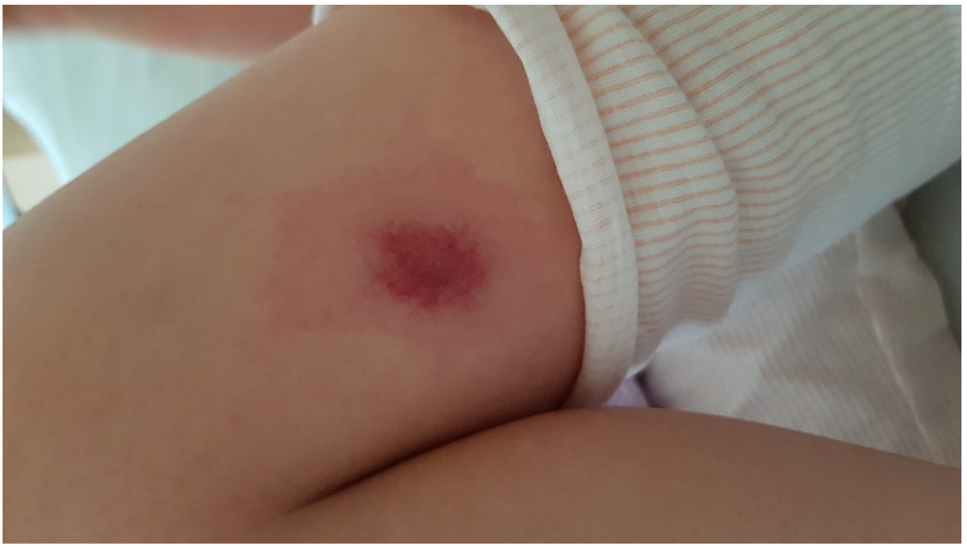

Figure 1. Lesion at day 1.

wound swab & tissue were sent to microbiology, and both confirmed the same organism. The wound continued to be dressed on regular basis. Her chemotherapy regime was restarted on post-operative day 17. She finally got discharged after fifty days of hospital stay; she was advised to continue oral Co-trimoxazole & Ciprofloxacin.

Following this event, the wound healed with secondary intention as demonstrated in the photographs in Figures 1–4. She has successfully completed her chemotherapy after twenty-seven months and has been on regular follow up with 6 weekly clinic review and full blood count, which has remained stable.

Discussion

Ecthyma gangrenosum occurs in 1–30% of cases of the Pseudomonas sepsis[5]; other bacteria such as Klebsiella pneumonia, *Escherichia coli, Morganella morganii, Citrobacter freundi* and fungi such as Fusarium & Candida albicans have been associated with this condition. Vaiman et al. detected *Pseudomonas aeruginosa* in 73.65% cases, other bacteria in 17.35% cases & fungi in 9% cases during their review of 167 cases of EG between 1975 and 2014.[6]

The pathogenesis involves invasion of media and adventitia of vessel walls resulting in vasculitis. The initial invasion causes secondary arteriolar thrombosis via bacterial endo- or exotoxins and / or circulating complexes, therefore, the grey infarcted maculopapular lesion progresses into necrosis.[5] The histopathology demonstrates a rich leucocyte infiltrate, mainly neutrophils.[7]

It can affect all age group ranging from pre-mature baby[8] to elderly patients.[9] It is seen more commonly in oncology patients as in our case; but up to 41% cases can be identified in healthy individuals[6] and these patients need to be investigated to identify any underling immunodeficiencies.

The diagnosis of EG is based on skin biopsy culture and blood cultures. There are various differential diagnoses such as septic emboli, diabetic microangiopathy, warfarin-induced skin necrosis, pyoderma gangrenosum, disseminated intravascular coagulation, etc.

EG has a high mortality rate; Koley et al.[10] describes the mortality rate of 0–28% in primary EG and 38–70% in classical EG. The most important prognosis factor is neutropenia, followed by multiple lesions and delayed treatment.

A multidisciplinary team approach involving oncologists, surgeons, nurses, and microbiologists should be provided to these patients. The treatment should be in three stages: initial empirical an-

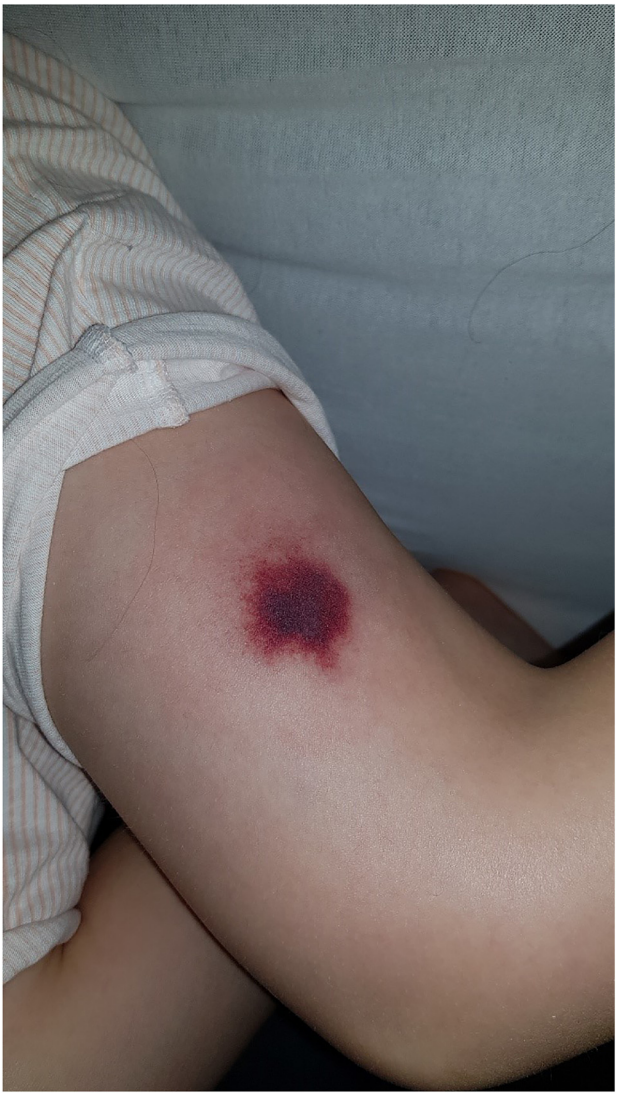

Figure 2. Lesion at day 5.

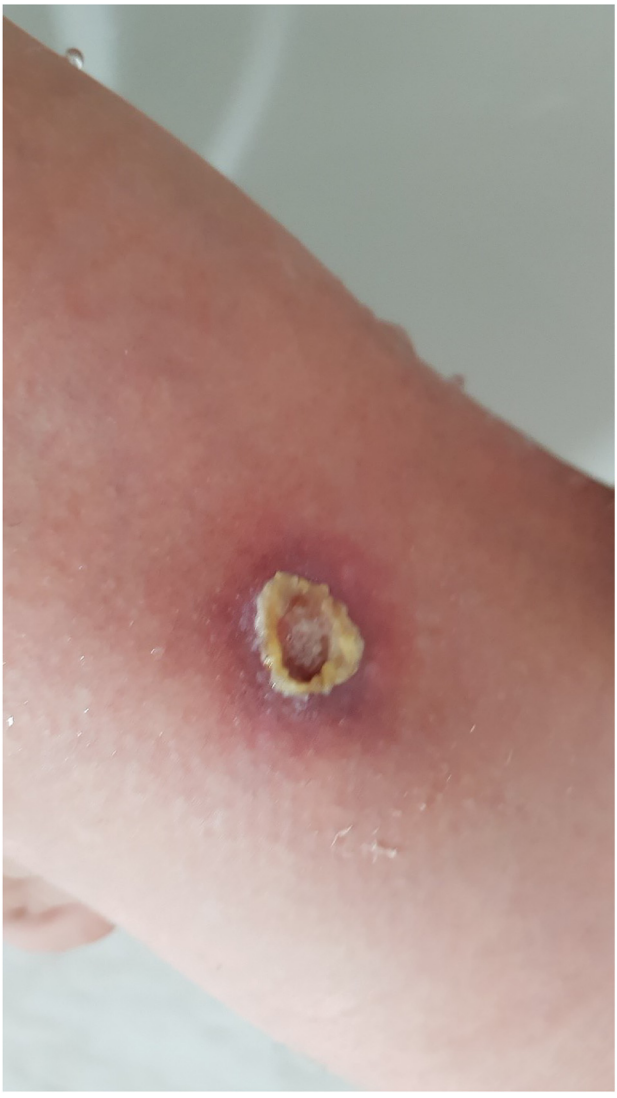

Figure 3. Lesion at day 45.

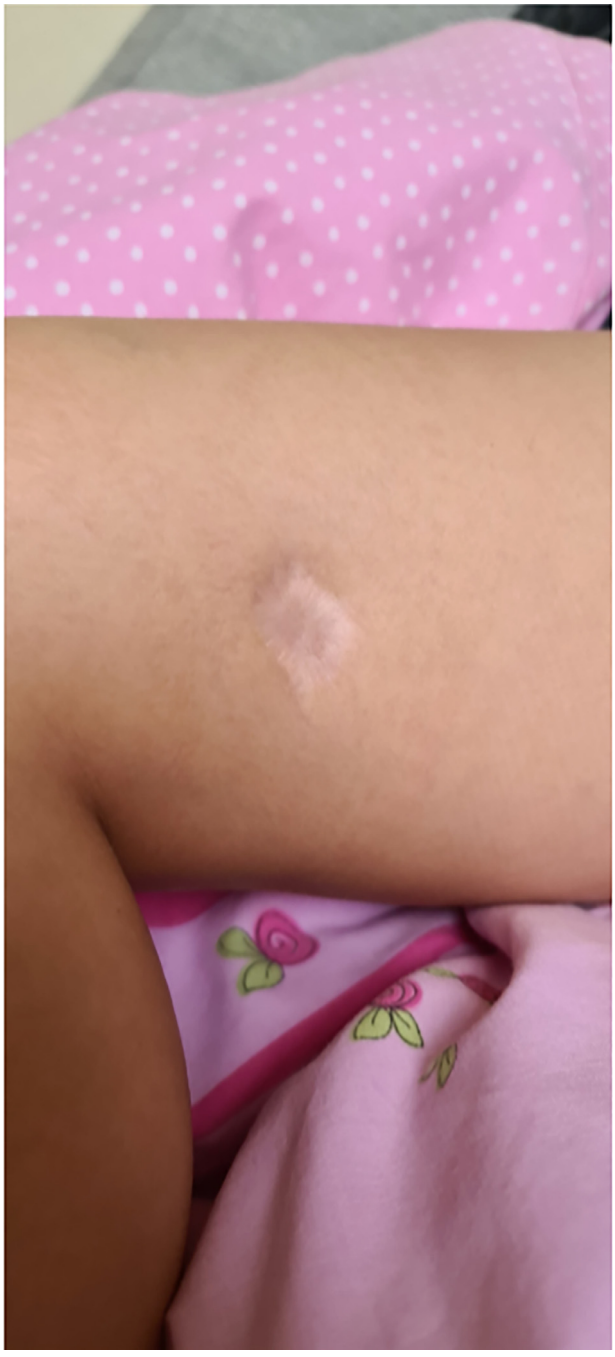

Figure 4. Lesion after 1 year.

tibiotic therapy, followed by targeted antibiotics or antifungals based on organisms, and/or surgical debridement. Our patient was treated in similar fashion and made a good recovery. In the context of our acutely unwell, immunocompromised patient, healing of the residual wound by secondary intention was felt to be most appropriate. Better cosmesis could be achieved by formal excision and direct closure or local flap reconstruction. In the future, cosmesis for our patient could be improved with either complete scar revision or with a combination of lipofilling to improve contour and laser resurfacing for texture and colour.

Conclusion

Ecthyma gangrenosum is a rare skin condition associated with a high mortality rate and predominantly associated with *Pseudomonas aeruginosa*. It calls for expeditious diagnosis and immediate treatment with antibiotics or antifungals, along with surgical debridement when necessary. We suggest all clinicians to be vigilant about this condition to be able to provide accurate diagnosis and prompt treatment to improve the overall prognosis.

Ethical approval

We thank the patient's family for approving the publication of the case report & pictures.

Declaration of competing interest

We declare no conflict of interest in relation with this work.

Funding

No funding or financial support was received.

Acknowledgment

We thank the medical photography department at the University of Aberdeen.

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