http:// ijp.mums.ac.ir
Case Report (Pages: 16428-16433)

Periorbital **Ecthyma Gangrenosum Due to Pseudomonas Aeruginosa Septicemia in an Infant with Sepsis: A Case Report**

Mohammad Etezad Razavi [1], Seyed Hossein Ghavami Shahri [2], Farid Shekarchian [2]; Sepide Hajmohammadi [3]; *Hamid Reza Heidarzadeh [2]

[1] MD, FICO, Eye Research Center, Mashhad University of Medical Sciences, Mashhad, Iran.

[2] MD, Eye Research Center, Mashhad University of Medical Sciences, Mashhad, Iran.

[3] MD, Department of Pediatrics, Mashhad University of Medical Sciences, Mashhad, Iran.

Abstract

Purpose: This article aimed to report an infant case of congenital neutropenia (CN) with periorbital ecthyma gangrenosum (EG) due to pseudomonas aeruginosa septicemia.

Case report: A previously healthy six-month-old male infant with fever, diarrhea, poor feeding, pancytopenia, periorbital swelling, and eyelid ulcers in the left eye was admitted in pediatric intensive care unit (PICU). He had positive blood and eyelid wound cultures for pseudomonas aeruginosa. He was treated with broad spectrum intravenous antipseudomonal antibiotics and debridement of periorbital lesions and discharged after 18 days with a stable condition. Adequate hematologic workup for pancytopenia revealed the diagnosis of CN.

Conclusion: There is a possibility that EG be developed in patients with immunodeficiency as preseptal cellulitis with isolated typical lesions, and this should be considered in the treatment.

Key Words: Case Report, Congenital Neutropenia, Chronic Neutropenia, Cutaneous Infection, Ecthyma Gangrenosum, Immunodeficiency, Isolated Typical Lesions, Preseptal Cellulitis, Pseudomonas Aeruginosa.

* Please cite this article as: Etezad Razavi M, Ghavami Shahri SH, Shekarchian F; Hajmohammadi S; Heidarzadeh HR. Periorbital Ecthyma Gangrenosum Due to Pseudomonas Aeruginosa Septicemia in an Infant with Sepsis: A Case Report. Int J Pediatr 2022; 10 (7):16428-16433. DOI: **10.22038/ijp.2020. 64385.4887**

Corresponding Author:

Hamid Reza Heidarzadeh, MD, Eye Research Center, Mashhad University of Medical Sciences, Mashhad, Iran. Email: dr.hamidreza.heidarzade@gmail.com

Received date: Mar.13,2022; Accepted date:Jun.25,2022

1- INTRODUCTION

Ecthyma gangrenosum (EG) is a cutaneous infection that mostly occurs in immunocompromised individuals with Pseudomonas aeruginosa (P. aeruginosa) bacteremia (1). It is characterized by a painless gangrenous ulcer with a black eschar (2). EG commonly affects anogenital and axillary areas. Rare cases of periorbital involvement have also been reported (3-8).

Congenital neutropenia (CN) is a rare genetically congenital disorder characterized by a block in neutrophil differentiation in the bone marrow, resulting in chronic neutropenia (absolute neutrophil count (ANC) < 1500 /μL) (9). Neutropenia is commonly defined as a peripheral absolute neutrophil count (ANC) of less than 1500 cells/μL blood. Further categories involve the terms of "mild" (ANC 1000–1500), moderate (ANC 500–1000), severe (ANC 200–500), and very severe (ANC < 200) neutropenia (12). The most frequent preferential sites of infection are the skin and mucosae, the ENT region, and the lungs (9).

Because EG is so rare and it has harvesting complications and morbidity resulting from septicemia and other organ damages, our purpose in this case report is presenting rare Ocular and systemic EG in a congenital neutropenia patient till suitable and early treatment of the disease could prevent devastating and life threatening complications.

In this report, we are going to describe an infant with CN, who was treated with intravenous antibiotic and eyelid ulcer debridement due to EG secondary to P. aeruginosa septicemia.

2- CASE REPORT

A previously healthy six-month-old male infant from Mashhad (Iran) who was fed with breast milk only and had a history of fever, diarrhea, poor feeding, periorbital swelling, and eyelid ulceration of the left eye since 3 days ago, came to the pediatric emergency room. His complete blood counts (CBC) showed pancytopenia (WBC: 3600 /ml (PMN: 27%), Hb: 9.2 g/dl and Platelet: 50000 /ml) and he had abnormal high erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) values (ESR: 96 mm, CRP: 215.9 mg/L). He was admitted in the pediatric intensive care unit (PICU) with a primary diagnosis of sepsis. The pediatrics service requested laboratory tests including blood and wound discharge smear and cultures; and started empirical intravenous antibiotic therapy with meropenem and vancomycin. His biochemical blood tests were in the normal ranges. They consulted with sub-specialists of infectious disease, hematology and immunology pediatrics for further evaluation of septicemia and pancytopenia. His growth and development according to his growth and development card graphs were normal and all graphs were above 50 percentile. Since his ANC (WBC×Neutrophil percent) 3600×27%=972 was under 1500 and also BMA showed maturation arrest at the promyelocyte stage associated with bone marrow monocytosis, there was a possibility of congenital neutropenia.

For ruling out his parent's neutropenia, we have done complete blood count and differentiation that was normal and there was no immunosuppressive disease history in his family.

Ophthalmology service consultant was used to rule out orbital cellulitis. We, then, visited him and he had periorbital swelling and erythema with 2 gangrenous suppurative eyelid ulcers in medial and lateral canthus of the left eye (**Fig. 1**). We requested orbital compound tomography scan (CT-scan). In his orbital CT-scan, we noticed preseptal soft tissue swelling which infiltrated into the orbit in the medial side of the left eye (**Fig. 2**). We considered continuing intravenous

antibiotic therapy and were informed about his wound discharge smear and culture results.

On day 3 after admission, the results of other laboratory tests were prepared. Immunologic tests for human T-cell lymphotropic virus 1 (HTLV1), human immunodeficiency virus (HIV), hepatitis B virus (HBV) and hepatitis C virus (HCV) were negative. The values for CD16, CD4, CD3, CD19, CD8, nitroblue tetrazolium test (NBT), immunoglobulin E (IgE), total IgA, total IgG and total IgM were in normal ranges. His bone marrow aspiration (BMA) was normal at the first time. The nasopharyngeal swab specimen reverse transcription polymerase chain reaction (RT-PCR) test for coronavirus disease 2019 (COVID-19) was negative. The RT-PCR of eyelid discharge for herpes simplex virus (HSV) was negative. The blood culture was positive for P. aeruginosa and it was sensitive to amikacin, ceftazidime, cefepime and ciprofloxacin. They informed us about the result of eyelid wound discharge smear and culture. The smear result was gram negative bacilli and its culture showed P. aeruginosa that was sensitive to amikacin, ceftazidime, cefepime, ciprofloxacin, cefotaxime, meropenem and piperacillin tazobactam. We planned for him to debride the left eye eyelid ulcer. The pediatric service continued intravenous antibiotic therapy with meropenem.

Fig. 1: Medial and lateral canthus gangrenous suppurative ulcers of the left eye secondary to EG

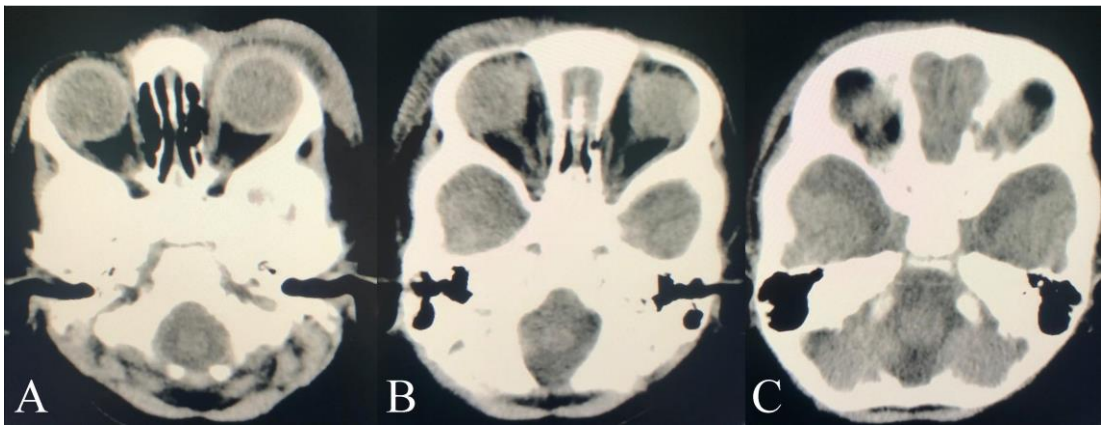

Fig. 2: Multiple sections (A-C) of orbital CT-scan without contrast in axial view show preseptal soft tissue swelling in the left eye that infiltrated into the orbit from the medial side.

Twelve days after admission, he was in a stable condition for eyelid ulcer debridement. Debridement of medial and lateral canthus was done (**Fig. 3**), and he was discharged with oral ciprofloxacin, but he couldn't tolerate oral antibiotic and was admitted for continuing intravenous antibiotic therapy with ciprofloxacin. His CBC values became normal (WBC: 11680 /ml, Hb: 10.5 g/dl and platelet: 581000 /ml) and his eyelid ulcer healed (**Fig. 4**). He was discharged after one week in a stable condition, but after two weeks and in the follow-up visit, his CBC revealed neutropenia again. BMA repeated and showed maturation arrest at the promyelocyte stage associated with bone marrow monocytosis. The hematology service started using granulocyte colony-stimulating factor (G-CSF) (Filgrastim) with oral sulfamethoxazole/trimethoprim combination for antimicrobial chemoprophylaxis. He is under observation for further oculoplastic surgical reconstruction in future.

Fig. 3: A and B show the debridement procedure for the medial and lateral EG lesions. C shows necrotic tissue excited.

Fig. 4: Post operation images show the left eye EG lesion regression.

3- DISCUSSION

In this report, we described a six-month-old infant with no past medical history, who was consulted for his ruling out of orbital cellulitis with a diagnosis of sepsis and pancytopenia. We found 2 gangrenous suppurative ulcers in the eyelid of his left eye. We found P. aeruginosa in his blood and wound cultures, and scheduled him for a debridement of eyelid ulcers with diagnosis of EG. In systemic evaluations, the pediatric service assessed the patient about any causes of immunodeficiency including inborn errors of metabolism, infections and congenital immunodeficiency. Based on the results of the systemic evaluation, we found CN as the cause of septicemia. EG mostly occurs in immunocompromised individuals with P. aeruginosa bacteremia (1). There are limited reports of periorbital involvement with EG (3-8). However, in this case, we showed that EG occured in an infant with CN, presented as isolated unilateral eyelid ulcers.

P. aeruginosa is the most frequent cause of EG. Vaiman et al, reported P. aeruginosa as the cause of 74% of cases (2). Other etiologies include methicillin resistant Staphylococcus aureus (MRSA), Streptococcus pyogenes, Citrobacter freundii, Escherichia coli, Aeromonas hydrophila, Klebsiella pneumoniae, Serratia marcescens, Xanthomonas maltophilia, Morganella morganii, Corynebacterium diphtheriae, Neisseria gonorrhoeae, Yersinia pestis, fungi like the Candida species, and viruses like HSV (1). Kimyai-Asadi et al., reported a man with multiple myeloma and ecthyma-like lesions on the back that HSV was identified in his ulcer (10). In our case, we ruled out HSV with RT-PCR.

Management of EG needs a team work between infectious disease, surgery, hematology and immunology services which we provided for this patient. Initial empiric therapy includes antipseudomonal beta-lactams, cephalosporins, fluoroquinolones, and carbapenems. Combination therapy is recommended in high risk patients with neutropenia and septic shock like our case (11). Other conditions with similar features should be considered like autoimmune vacuities, disseminated intravascular coagulation (DIC), pyoderma gangrenosum, necrolytic migratory erythema, and livedoid vasculopathy (1). For necrotic skin lesions, aggressive debridement may be necessary and grafting may be needed especially in lesions greater than 10 cm (1).

The management of CN needs crucial consideration. It is important to prevent recurrent infections with watchful observation, using antimicrobial chemoprophylaxis, and hematopoietic growth factors. Also, immediate hospitalization is required in case of infection occurrence, and intravenous antibiotic therapy should be started quickly, with a third-generation cephalosporin combined with an aminoside (9).

In general, we reported the case of a previously healthy infant with P. aeruginosa sepsis, and isolated EG lesions of the left eye eyelids in the setting of CN. He was successfully treated with antibiotic therapy and eyelid lesions debridement. The EG lesions were regressed, and he is under observation for CN.

4- AVAILABILITY OF DATA AND MATERIALS

The datasets used during the current study are available from the corresponding author on reasonable request.

5- COMPETING INTERESTS

None.

6- FUNDING

The authors received no funding. It is the authors' work, not funded by the

government or academic institutes.

7- AUTHORS' CONTRIBUTIONS

All the authors contributed significantly to this report, and all authors agree to be accountable for all aspects of the work. All authors read and approved the final manuscript.

8- REFERENCES

1. Shah M, Crane JS. Ecthyma Gangrenosum. (Updated 2021 Jul 18). In: StatPearls (Internet). Treasure Island (FL): StatPearls Publishing; 2021 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK534777.

2. Vaiman M, Lazarovitch T, Heller L, Lotan G. Ecthyma gangrenosum and ecthyma-like lesions: review article. Eur J Clin Microbiol Infect Dis. 2015 Apr; 34(4):633-9. doi: 10.1007/s10096-014-2277-6. Epub 2014 Nov 19. PMID: 25407372.

3. Lin Q, Hu B, Shi J, Shi W, Cao W, Li L. Eyelid and periorbital ecthyma gangrenosum due to Pseudomonas aeruginosa in an infant. Pediatr Investig. 2017; 1(1):47-49. Published 2017 Dec 27. doi:10.1002/ped4.12005.

4. Henshaw EB, Ibanga AA, Obaji DP. Fatal Oculocutaneous Ecthyma Gangrenosum in Human Immunodeficiency Virus/Acquired Immunodeficiency Syndrome: Case Report and Review of the Literature. J Glob Infect Dis. 2019; 11(1):43-46. doi:10.4103/jgid.jgid_54_18.

5. Steinkogler FJ, Huber-Spitzy V. Necrotizing destruction of the ocular adnexa by Pseudomonas aeruginosa. J Craniomaxillofac Surg. 1988 Jan; 16(1):28-30. doi: 10.1016/s1010-5182(88)80010-6. PMID: 3123523.

6. Kim HJ, Grossniklaus HE, Wojno TH. Periorbital ecthyma gangrenosum: a case report and review of the literature. Ophthalmic Plast Reconstr Surg. 2014 Sep-Oct; 30(5):e125-8. doi: 10.1097/IOP.0b013e3182a6504a. PMID: 24317102.

7. Ghosheh FR, Kathuria SS. Bilateral periorbital ecthyma gangrenosum. Ophthalmic Plast Reconstr Surg. 2006 Nov-Dec; 22(6):492-3. doi: 10.1097/01.iop.0000248990.71690.a2. PMID: 17117116.

8. Watson A, Sloan B. Ecthyma gangrenosum arising from Pseudomonas aeruginosa dacryocystitis. Clin Exp Ophthalmol. 2003 Aug; 31(4):366-8. doi: 10.1046/j.1442-9071.2003.00675.x. PMID: 12880468.

9. Donadieu J, Fenneteau O, Beaupain B, Mahlaoui N, Chantelot CB. Congenital neutropenia: diagnosis, molecular bases and patient management. Orphanet J Rare Dis. 2011 May 19; 6:26. doi: 10.1186/1750-1172-6-26. PMID: 21595885; PMCID: PMC3127744.

10. Kimyai-Asadi A, Tausk FA, Nousari HC. Ecthyma secondary to herpes simplex virus infection. Clin Infect Dis. 1999 Aug; 29(2):454-5. doi: 10.1086/520241. PMID: 10476767.

11. Bassetti M, Vena A, Croxatto A, Righi E, Guery B. How to manage Pseudomonas aeruginosa infections. Drugs Context. 2018 May 29; 7:212527. doi: 10.7573/dic.212527. PMID: 29872449; PMCID: PMC5978525.

12. Klein C. Check for Updates Hematology Am Soc Hematol Educ Program (2009) 2009 (1): 344–350. https://doi.org/10.1182/asheducation-2009.1.344.