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INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)

Article DOI: 10.21474/IJAR01/18213
DOI URL: http://dx.doi.org/10.21474/IJAR01/18213

RESEARCH ARTICLE

ECTHYMA GANGRENOSUM THAT REVEALED AN AGAMMAGLOBULINEMIA: A CASE REPORT

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Manuscript Info

Manuscript History
Received: 19 November 2023
Final Accepted: 29 December 2023
Published: January 2024

Key words:-
Agammaglobulinemia, Echtyma, Pseudomonas Aeruginosa

Abstract

Ecthymagangrenosum is a distinctive skin infection caused by P. aeruginosa bacteremia; it typically occurs in immunocompromised or critically ill patients. However, in previously healthy children with pseudomonas sepsis, the infection may be the first manifestation of underlying pathology, such as agammaglobulinemia. We report a case of echtymagangrenosum in previously healthy boy, leading to the diagnosis of agammaglobulinemia.

Introduction:-

Ecthymagangrenosum (EG) is a relatively uncommon condition, it is a characteristic dermatologic manifestation of severe and invasive infection caused most commonly by Pseudomonas aeruginosa. The clear description of the disease was given by L. Barker in 1897 and the term itself was generally accepted in the 1950s [1, 2]. Up to the 1970s, it was postulated that this condition is pathognomonic of Pseudomonas septicemia (Pseudomonas aeruginosa) and that it should usually be seen in immunocompromised patients, particularly those with underlying malignant disease [3, 4]. However, many cases of Pseudomonas sepsis in previously healthy children have been reported in the literature [5]. We add one more case to this group in an effort to increase clinical awareness of possiblePseudomonas infection in children without previous medical problems.

Observation:-

A 4-month-old male was admitted to emergency with a 5 days of fever reaching 39.9 and two days of a perineal necrotic lesions. The parents noted asymmetrical skin lesions on the patient's perineum. He was previously healthy, and had no known history of drug allergies, recent travel or family history of immunodeficiency.The patient had received age-appropriate live attenuated vaccines and showed nosymptoms of discomfort.

On admission, physical examination showed the patient had met typical developmental milestones. He was lethargic and febrile (39.1°C); pulse rate was 155/min, respiratory rate was 56/min, and blood pressurewas normal.On skin examination: threepurple necrotic lesions surrounded by an intense red areola on the face , scrotum and the left tight , two vesiculobullous skin lesions with mild induration,on the same localizations (figures 1-3)

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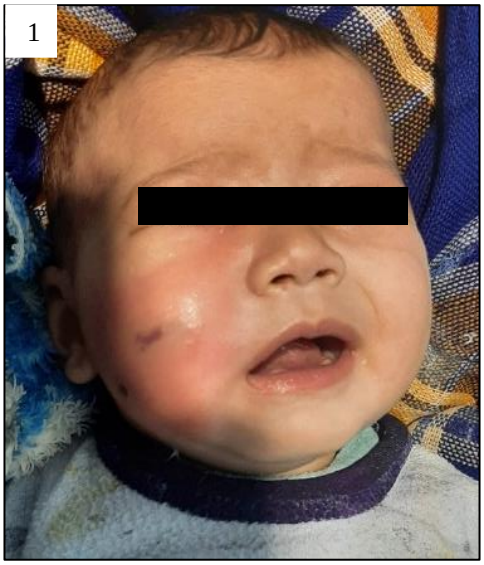

Figure 1:- Edematous and erythematous early lesion of ecthymagangrenosum over the right cheek with asymmetrical aspect.

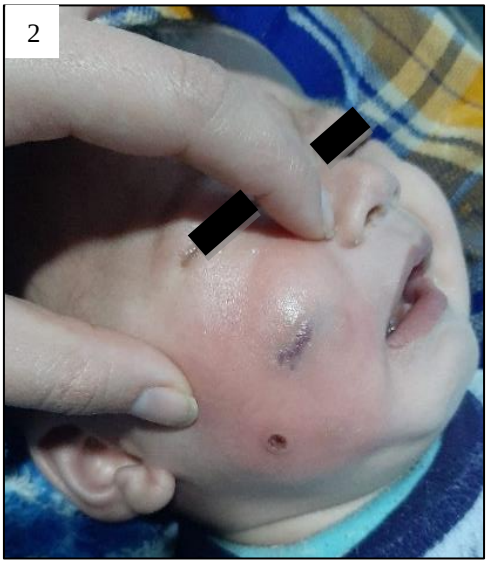

Figure 2:- A mild induration of the right cheek surrounded by a purple necrotic lesion.

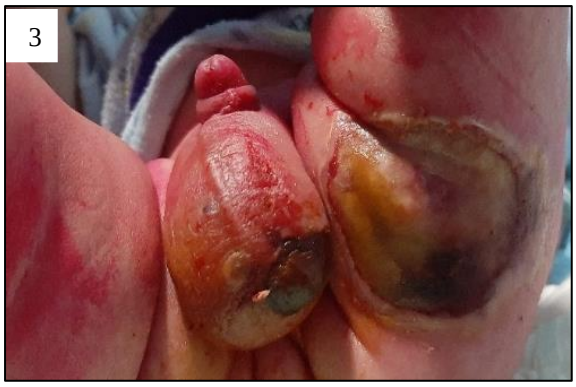

Figure 3:- 2 Large necrotic ulcers with erythematous border over the left tight and scrotum ; bluish bullae as an early lesion of echtymagangrenisum over the scrotum.

The laboratory findings were:peripheral White Blood Cells count 19.2 ×**10^9/L**(normal referencevalues: 4–10 × **10^9/L**) of which 40% were neutrophils, 30% were lymphocytes; C-reactive protein was 190 mg/dL (normal reference value: <8.0).The antibiotic treatment was initiated with intravenous ceftazidime;metronidazole,and gentamicin.During the second day, the infant underwent surgical debridement of the necrotic lesions,the Pseudomonas aeruginosa was isolated from the surgical sample,it was sensitive to ceftazidime and gentamicin.A complete workup for immunodeficiency was made in association:The HIV virologic test was negative.An immune panel was ordered, whichshowed: plasma IgG 3.2 g/L (normal reference: 2.86–16.8 g/L), IgA 0.25 g/L (normal reference: 0.19–1.75 g/L)IgM 0.28 g/L(normal reference: 0.43–1.63 g/L). Complement components (C3, C4, and CH50)were normal and the number of circulating B lymphocytes(CD19, CD20)was less than 1% in peripheral blood (normal reference: 7%-14%).After the empirical use of immunoglobulin (400 mg/kg), he was discharged after 10 days of hospital stay, there were no further appearances of new skin lesions.

Discussion:-

Ecthymagangrenosum (EG) is a severe potentially lethal cutaneous infection that progresses sequentially from maculopapular rash to haemorrhagic bulla and then to necrotic ulceration with surrounding erythema; it commonly caused by Pseudomonas Aeruginusa.

Since the 1980s, more and more data have been accumulated that various bacteria like Escherichia coli, Citrobacterfreundii, Klebsiella pneumonia, various other Pseudomonas species, and Morganellamorganii can be etiologic agents for EG, as well as some fungi (Candida albicans, Fusarium, and others) [6].

While generally accepted, the exact clinical manifestations also have their own unanswered questions. For example, most of the researches agree that the skin lesions usually occur in the gluteal and perineal regions (57 %) or extremities (30 %) [7, 8]. The lesions, however, may appear on the face, chest, arms, neck, and other parts of the body [6]

In individuals with suspected ecthymagangrenosum, prompt treatment with broad-spectrum antibiotics should be initiated after obtaining blood and wound cultures. Once the causative organism and antimicrobial susceptibilities are identified, antimicrobial coverage should be narrowed.

Ecthymagangrenosum affects all age groups and genders. Hence, the presence of this bacterial infection in healthy children is very uncommon. Immunocompromised individuals are particularly susceptible to developing this condition, with up to 62% to 75% of affected individuals having an underlying immunodeficiency.[9][10] EG is also known to occur in otherwise healthy immunocompetent individuals.[11] Common predisposing conditions include neutropenia, leukemia, multiple myeloma, diabetes mellitus, malnutrition, and extensive burn wounds.

In our patient, the diagnosis of brutonagammaglobulinemia was confirmed by his extremely low concentration of all the immunoglobulin isotypes, profound decrease in circulating B-lymphocytes, normal lymphocyte count and normal cell-mediated immunity, and complement.

Conclusion:-

Ecthymagangrenosum (EG) is a cutaneous infection that most commonly occurs in immunocompromised individuals with fulminant bacteremia. EG was first described as a manifestation of Pseudomonas aeruginosa. Although P. aeruginosa remains the most frequent organism identified in EG, other causative pathogens have since been described. In previously healthy children, immunological evaluation is important to rule out an underlying immunodeficiency.

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