Case report

Epidermolysis bullosa with clinical manifestations of sepsis and pneumonia: A case report

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ABSTRACT

Background: Epidermolysis bullosa is an inherited disease that causes bleeding blisters on the skin tissues and mucosal membranes. This study reports a case of epidermolysis bullosa with clinical manifestations of sepsis and pneumonia.

Case report: A 17-month-old female with epidermolysis bullosa presented with clinical manifestations of sepsis and pneumonia, including increased body temperature, anorexia, and wheezing. Blood test results included the following: white blood cell (WBC) count, 24,000/μl; hemoglobin level, 7.9 mg/dl; erythrocyte sedimentation rate (ESR), 99 mm/h; and C-reactive protein level, +2. The patient was given ceftriaxone, vancomycin, hydrocortisone, and acetaminophen. The patient’s fever resolved on the third day of hospitalization, and the ESR and WBC count reached 25 mm/h and 9900/μl, respectively.

Conclusions: According to the report, it is recommended to consider an increase in body temperature, WBC count, and ESR, and perform the required attempts in patients with epidermolysis bullosa.

1. Introduction

Epidermolysis bullosa (EB) is a genetic skin defect that causes skin fragility in which the slightest friction or damage separates the skin layers, causing blisters and open wounds [1] and, in some cases, damages the mucosal membranes and organs. Children with EB typically have skin as fragile as a butterfly wing. Of every one million live childbirths, 50 are diagnosed with epidermolysis bullosa, and there is an estimated 500,000 patients with EB worldwide [2]. The probable prevalence of this disease is the same in all countries and races. The major types of EB include simple, connective, and dystrophic EB. It is diagnosed by genetic testing, prior-to-birth testing, and biopsy or skin sampling for immunofluorescent mapping [3]. Treatments include blister care, daily skin washing, daily dressing using therapeutic products, cooling, an appropriate dietary regimen, pain and itching control, anti-infectious medications, surgery (esophageal stenosis removal, gastrostomy tube placement for skin grafting), and physiotherapy. Esophageal blisters and wounds, stenosis, dysphagia, and sepsis are among the complications of this disease [4].

2. Case presentation

A 17-month-old female with a chief complaint of fever, anorexia, and wheezing was hospitalized in the pediatric ward of the Khatam-ol-anbia Hospital at Gonbad Kavous. She was referred to a dermatologist who had 10 years of experience. She had no history of tuberculosis, genetic disease, and pharmaceutical or dietary allergies. She had fever, weakness, fatigue, anorexia, and nausea. She was hospitalized two months prior due to fever and infection. She was delivered full-term through cesarean section. The first child in the patient’s family was healthy, and her parents had no other family relationships. Some blisters, which were diagnosed at the early stages of EB, on the hand and foot were observed at birth (Fig. 1).

At initial assessment, the patient’s body temperature was 39°C, and the hand and foot blisters were covered with Mepilex dressing and fixed with a Tubifast bandage. Small bleeding wounds around the mouth were
also observed. The patient’s mother had applied Sudocrem and Bepanthen on the blisters and dry and itchy skin areas, respectively. Aquaphor was used to repair and boost the skin of the entire body. Initial laboratory results were as follows: white blood cell (WBC) count, 24,000/μl (reference, 1100–4400/μl); hemoglobin (HB) level, 7.9 mg/dl (reference, 12–16 mg/dl); erythrocyte sedimentation rate (ESR), 99 mm/h (reference, <20 mm/h); C-reactive protein (CRP) level, +2; blood urea nitrogen level, 13.6 mg/dl (reference, 8–25 mg/dl); creatinine level, 0.7 mg/dl (reference, 0.3–0.7 mg/dl); partial thromboplastin time, 12 s; prothrombin time, 12 s; and international normalized ratio, 1. Imaging studies included chest radiography. The patient was diagnosed with sepsis and pneumonia, and was prescribed with 500 mg of ceftriaxone, 100 mg of vancomycin, 2 ml of guaifenesin syrup TID, 25 mg of hydrocortisone, 50 mg of ranitidine BID, 100 mg of acetaminophen, 3–5 ml of hydroxyzine syrup PRN, and 2 drops of nasal NaCl every 4 h. She received 10 ml/kg of fresh frozen plasma. Chest physiotherapy was performed daily for three days. After three days, her laboratory tests were as follows: WBC count, 9900/μl; HB level, 9 mg/dl; ESR, 25 mm/h; CRP level, +1. During the first two days of hospitalization, the patient had a fever of >38 °C, which was controlled by acetaminophen. Her fever and wheezing resolved at the third hospitalization day. She received appropriate nursing and medical care interventions. This work was reported in line with the SCARE 2020 criteria [5].

3. Discussion

Pediatric blister lesions present in a wide spectrum of conditions, including bullous impetigo or toxic epidermal necrolysis, immunologic diseases, such as pemphigus vulgaris or bullous pemphigoid, and hereditary diseases, such as EB. We report a rare case of EB with clinical manifestations of sepsis, including an increase in WBC count and ESR. A 2016 article reported a case of a 19-year-old male presenting with EB along with muscular dystrophy who had skin blisters from birth, bilateral ptosis since the age of 8 years, muscle fatigue and pain during walking since the age of 11 years, depression and aggressive behavior to parents since the age of 15 years, and polyuria since the age of 16 years. EB was diagnosed by muscle dystrophy according to clinical and histopathologic examinations of the patient’s skin and skeletal muscles, and a noted pectin gene mutation [6]. He underwent physiotherapy and psychological care. A 2018 article reported a case of a 28-year-old female with EB and concomitant Duchenne muscular dystrophy and alopecia, despite her parents having unremarkable findings. Mucosal blisters of the skin and mouth appeared at the age of 2 days, and non-scarring alopecia was observed on the scalp. She presented with onychodystrophy, tooth decay, mild dystonia, and severe atrophy. Neurologic examination revealed reduced deep reflexes. The study reported that factors such as stress and nutrition led to a new mutation [7]. Another 2018 article reported a 60-year-old male with unbalanced EB, a history of persistent and incurable wounds, and extensive and persistent wounds along his shoulder, lower abdomen, back thigh, and knee. He had a family history of tuberculosis, and he complained of unintended weight loss. He used topical lidocaine and some oral medications, such as acetaminophen and methadone, for pain control. He responded positively to caring crème for the wounds and dietary supplements for several months. EB was diagnosed based on clinical and histopathological tests of the skin and skeletal muscles [8].

4. Conclusions

These patients were exposed to various types of infections, including sepsis. Therefore, an increase in body temperature should be considered along with other diagnoses. According to the above findings, timely diagnosis and treatment of such patients are critical to reduce risk of infection.

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Ethical approval

N/A.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Dr. Samira Khanmohammadi, Raheleh Yousefzadeh, Maryam Rashidian has designed the concept of the study, literature review, Data Collection and analysis. Dr. Azam HajiBeglo, Karvan Bekmaz has contribution in study concept design, treatment of the patient and manuscript writing.

Registration of research studies

Not applicable.

Guarantor

Karvan Bekmaz.

Fig. 1. Blisters observed among patients.
Declaration of competing interest

The authors report no declarations of interest.

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