Journal of Pakistan Association of Dermatologists. 2021;**31(4)**:774-776.

Case Report

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

Samira Khanmohammadi, Raheleh Yousefzadeh* , Maryam Rashidan , Azam Hajibeglo, Karvan Bekmaz**[†]

Department of Nursing, Ali Abad Katoul Branch, Islamic Azad University, Ali Abad Katoul, Iran.
* Department of Nursing & Midwifery, Bojnurd University Medical Science. Khorasan Somali, Iran.
** School of Nursing & Midwifery, Shahid Beheshti University of Medical Sciences, Tehran, Iran.
[†] Ziaeian Hospital, Tehran University of Medical Sciences, Tehran, Iran.

Abstract Epidermolysis bullosa is an inherited disease of skin tissues that causes bleeding blisters on the skin and mucosal membrane. This report presents a girl aged 17 months with epidermolysis bullosa with clinical manifestations of sepsis and pneumonia with increased body temperature, anorexia and wheezing. Blood tests include: WBC: 24000 µl, HB: 7.9 mg/dl, ESR: 99, CRP: +2. The patient underwent treatment with ceftriaxone, vancomycin, Hydrocortisone, and paracetamol. Fever of the patient was stopped at the third day of hospitalization, and the ESR reaches 25 and WC reaches 9900 µl. According to the report, it is recommended to consider increase in body temperature and increase in WBC and ESR, and perform required attempts in patients with epidermolysis bullosa.

Key words
Epidermolysis bullosa, Sepsis.

Introduction

Epidermolysis bullosa is a skin genetic defect which causes fragility of skin and in some cases damages to mucosal membrane and organs. Skin fragility is such that the slightest friction or damage separates the skin layers and causes blisters and open wounds.[1] Children with epidermolysis bullosa is a term used for patients with young ages, the reason for this nomination is that skin of children is as fragile butterfly wings. Out of every one million living child birth, 50 neonates are diagnosed with epidermolysis bullosa. It is estimated there are 500000 patients with EB worldwide.[2] Percentage of prevalence probable of this disease in all countries and all races are the same. The major types of EB include: simple EB, connective EB, and dystrophic EB. It is diagnosed by genetic test, prior-to-birth test, and biopsy or skin sampling to immunofluorescent mapping.[3] Treatments include: caring of blisters, daily washing of skin, and daily dressing of blisters of foot and hand, using therapeutic products in dressing, cooling, appropriate dietary regimen. Medications are for controlling pain, itching and infection, and surgery (removal of esophageal stenosis, placement gastrostomy tube for skin grafting) and physiotherapy. Blister and wound in esophagus, esophageal stenosis and subsequently dysphagia and sepsis are among complications of the disease.[4]

Address for correspondence
Karvan Bekmaz
MS Nursing, Ziaeian Hospital,
Tehran University of Medical Sciences,
Tehran, Iran.
Email: nosratollahmas@gmail.com

Case report

The patient is a child aged 17 months with the chief complaint of fever, anorexia, and wheezing hospitalized in pediatric ward of hospital of

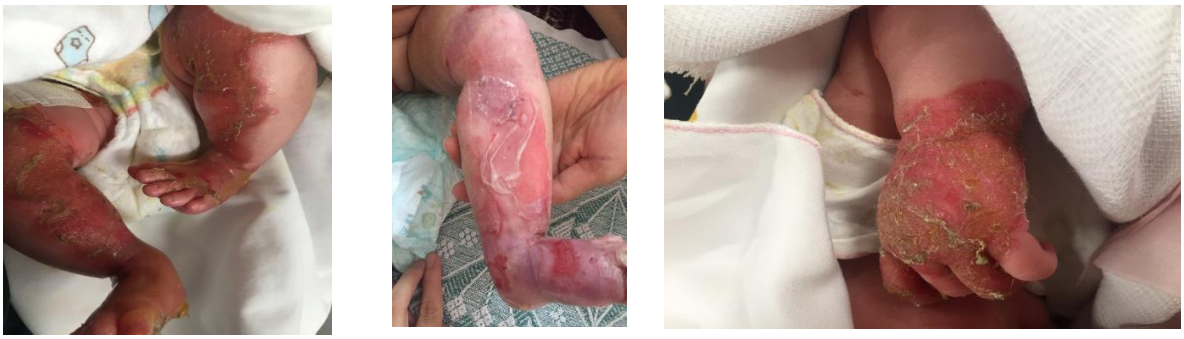

Figure 1 Blisters observed among patients.

Khatam ol anbia at Gonbad Kavous. The patient was affected by fever, weakness and fatigue, anorexia and nausea. Her mother reported a history of fever and infection at two months ago and subsequent hospitalization for her child. She does not report history of pharmaceutical and dietary allergies. The child is born through cesarean section as a term neonate. The mother does not report history of genetic disease. The first child of the family is healthy. Parents have no family relationship. Some blisters are observed on hand and foot at birth, which are diagnosed at the early stages of EB **(Figure 1)**.

At first assessments, body temperature was 39 degree and some blisters were observed in foot and hand of the patient, which these blisters were covered by mepilex and fixed by Tubi fast. In addition, small bleeding wounds were observed around the mouth. Mother of the child uses sudocrem for blisters and bepanthol for areas of skin which is dried and itching. Aquaphor was used to repair and boost skin of entire body. First tests include blood tests such as: WBS: 24000 µl, normal: 1100-4400 µl, Hemoglubin (HB): 7.9 mg/dl, Normal: 12-16 mg/dl, ESR: 99. Normal: less than 20, CRP: +2, Urea (BUN): 13.6 mg/dl, normal: 8-25 mg/dl, creatinine (Cr): 0.7 mg/dl, Normal: 0.3-0.7 mg/dl, PTT: 12 Seconds, PT: 12 seconds, INR: 1. Graphies performed for patients include CXR. The patient diagnosed with sepsis and pneumonia underwent treatment with Ceftriaxone 500 mg/tds, Vancomycin 100 mg/tds, Hydrocortisone 25 mg/bd, ranitidine 50 mg/bd, Acetaminophene 100mg/prn, Guaifenesin syrup 2cc/tds, hydroxyzine syrup 3-5cc/prn, nasal Nacl drop 2ggt/q4h. She received 10cc/pkgr FFP. Physiotherapy of chest was performed daily for three days. The later three days, laboratory tests were checked again for WBC, HB, ESR, CRP: WBC: 9900 µl, HB: 9 mg/dl, ESR: 25, CRP:+1. During first two days of hospitalization, the patient had a fever more than 38 degree which was controlled by Acetaminophene. Patient's fever was stopped from the day three. In addition, wheezing was resolved. Anorexia was attenuated.

Discussion

Blister lesions in childhood present wide spectrum including Bullous Impetigo or Toxic Epidermal Necrolysis and immunologic diseases such as pemphigus or bolus emphigoide and hereditary diseases such as EB. One rare case was introduced in this report for EB with clinical manifestations of sepsis which was concomitant with increase in WBC and ESR. One case report at 2016 showed that a 19 years old boy was with EB along with muscular dystrophy which had skin blisters from birth, Bilateral ptosis was occurred since age of 8 years, muscle fatigue and pain during walking was manifested at 11 years old which underwent physiotherapy. Depression and aggression to parents was observed at age 15 years and underwent psychological caring. Frequent urination was

occurred since the age of 16 years old. In this study, EB is diagnosed by muscle dystrophy according to clinical, histopathologic)skin and skeletal muscles) tests and mutation of pectin gene.[5] One case report at 2018 was reported that the patient was a woman aged 28 years old, with EB concomitant with Duchenne Muscular dystrophy and alopecia that her parents were healthy. Mucosal blisters of skin and mouth were occurred at age of 2 days, non-scar alopecia was observed on skin of head. Anicodystrophy mostly affect organs at every 20 nails, tooth decay, mild dystonia and severe athrophy. Neural examination showed reduced deep reflexes. In this study, factors such as stress and nutrition lead to new mutation.[6] One case report was performed at 2018. The 60-year-old man with unbalanced EB, history of persistent and incurable wounds, and extensive wounds covered almost a third of the patient's body. In addition, wound was existed constantly on the shoulder, lower abdomen and back thigh and knee. The patient has family history with tuberculosis, and complained on un-intended weight loss. The patient used topical lidocaine to control pain and some oral medications such as Acetaminophene and methadone. Patient showed positive response to caring crème for wound and dietary supplement during several months, in this study, EB was diagnosed based on clinical and histopathological tests (skin and skeletal muscle).[7] These patients are exposed to types of infections including sepsis. Therefore, along with other diagnoses, increase in body temperature should be considered According to the above tips, timely diagnosis and treatment of such patients in order to reduce risk of infection are critically important.

References

1. Rezniczek GA, Walko G, Wiche G. Plectin gene defects lead to various forms of epidermolysis bullosa simplex. *Dermatologic clinics*.2010;**28(1)**:33-41.
2. Bruckner AL, Losow M, Wisk J, Patel N, Reha A, Lagast H, *et al*. The challenges of living with and managing epidermolysis bullosa: insights from patients and caregivers. *Orphanet J Rare Dis*. 2020;**15(1)**:1.
3. Intong LR, Murrell DF. Inherited epidermolysis bullosa: new diagnostic criteria and classification. *Clin Dermatol*. 2012;**30(1)**:70-7.
4. Has C, Liu L, Bolling M, Charlesworth AV, Hachem ME, Escámez MJ, *et al*. Clinical practice guidelines for epidermolysis bullosa laboratory diagnosis. *Br J Dermatol*.2020; **182(3)**:574-592.
5. Kyrova J, Kopeckova L, Buckova H, Mrazova L, Vesely K, Hermanova M, *et al*. Epidermolysis bullosa simplex with muscular dystrophy. Review of the literature and a case report. *J Dermatol Case Rep*.2016;**10(3)**:39.
6. Argyropoulou Z, Liu L, Ozoemena L, Branco CC, Senra R, Reis-Rego Â, *et al*. A novel PLEC nonsense homozygous mutation (c. 7159G> T; p. Glu2387*) causes epidermolysis bullosa simplex with muscular dystrophy and diffuse alopecia: a case report. *BMC Dermatology*.2018; **18(1)**:1.
7. Fife CE, Yaakov RA, Serena TE. Epidermolysis bullosa: a case report. *Chronic Wound Care Manag Res*.2018; **5**:17-21.