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Effectiveness of Debridement Treatment in Neonatorum Epidermolysis Bullosa: A Case Report

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ABSTRACT

Introduction: Epidermolysis Bullosa (EB) is an autosomal genetic disorder that characterized by blistering of the skin and mucosa following mild trauma such as frictional trauma. There is no definitive treatment has yet to be developed. The goal of surgical debridement is to achieve a clean wound bed in fastest way, reduce bacterial burden, remove as much nonvital tissue, speed up the healing process, which will shorten the length of stay in hospital.

Case Finding: We have 3 days old newborn with clinical appearance similar to a second degree burn lesion bullous lesions and epidermal damage on her both feet, gluteal, and genital regions but without previous history of contact with thermal trauma consulted to patients. General physical examination showed no fever and vital within normal condition. In the perinatal history, the baby was delivered by caesarean section from a 37 years-old G3P2A0 mother with preeclampsia comorbid, normal gestational age, her birth weight was 3800 grams, her birth length was 51 cm, and with APGAR her scores of were 9 and 10 the 1st and 5th minute respectively.

Our newborn patient was looked like 2nd degree burn injury with blisters and epidermolysis but without previous history of contact with thermal trauma. Consulted to plastic surgeon for further evaluation and management. The Baby was discharged day 9 or after 3 days debridement management in stable condition.

Conclusion: The surgical debridement demonstrated greatly effective and efficient in achieving these aims. Observation on day 3 did not reveal any sign of infection, showing controlled symptoms and good healing process. We suggest that performing debridement on Epidermolysis Bullosa (EB) patient would shorten the duration of hospitalization. Therefore, it provides low-risk nosocomial infection and low-cost hospitalization.

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KEYWORDS: Epidermolysis Bullosa, Neonatorum, Debridement, Length of Stay.

INTRODUCTION

Epidermolysis bullosa (EB) is an autosomal genetic disorder that manifests in skin fragility such as blistering of the skin and mucosa after trauma. The exact prevalence of EB is still unclear, but estimated prevalence of mild variants is 1 per 50,000 births and the more severe variants have been estimated to occur in 1 per 500,000 births. The newest consensus has reclassified EB into four subtypes based on the precise location at which separation or blistering occurs, EB simplex (EBS), junctional EB (JEB), and dystrophic EB (DEB), and Kindler EB. Tissue separation of EBS variant occurs at intraepidermal level, whereas the JEB variant occurs at lamina lucida level, and the DEB variant occurs at dense lamina level. Kindler's syndrome is a combination of three other variants of EB in which tissue separation occurs in multiple planes. The varying degrees of skin fragility are caused by mutations in structural proteins involved in basal membrane adhesion.[3] EBS is the most common variant of EB, accounting for 70% of cases and tends to be milder than other variants. In patient with Weber–Cockayne type, a localized EBS variant, bullous lesions may appear as late as the third

decade of life, but presents limited to the hands and feet. While in patient with Köebner type, an intermediate generalized EBS variant, bullous lesions appear scattered all over the body, sometimes accompanied by oral mucosa and nail involvement, and usually begin from infancy. The severe generalized of EBS variant is known as Dowling–Meara type which is commonly found in newborns, presenting with generalized blistering of the skin, oral mucosa bullae, and nail shedding.[4]

If EB is suspected, electron microscopy/ immunofluorescence mapping and molecular genetic diagnosis should be performed at an early stage to determine the precise subtype based on the level of skin separation, improve prognostication, and enable genetic counselling. There is no definitive treatment has yet to be developed, as there are only preventive and supportive therapy to be performed. The management of EB lesions is challenging due the complexity and diversity of its manifestations. Dressing management is specific depends on wound characteristics, presence of infection or colonization, levels of exudate, availability of products and personal preference.[5] The goal of adequate surgical debridement's is to achieve a clean wound bed in fastest way, reduce bacterial burden, remove as much nonvital tissue, speed up the healing process, which will shorten the length of stay in hospital.[6]

CASE FINDING

We have 3 days old newborn with clinical appearance similar to a second degree burn lesion bullous lesions and epidermal damage on her both feet, gluteal, and genital regions but without previous history of contact with thermal trauma consulted to patients.

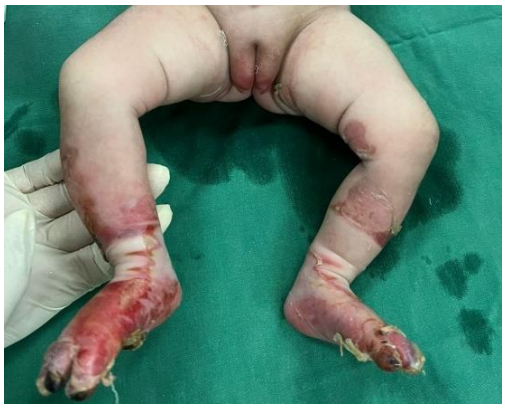

Figure 1. Pre-Debridement

General physical examination showed no fever and vital within normal condition. In the perinatal history, the baby was delivered by caesarean section from a 37 years-old G3P2A0 mother with preeclampsia comorbid, normal gestational age, her birth weight was 3800 grams, her birth length was 51 cm, and with APGAR her scores of were 9 and 10 the 1st and 5th minute respectively. There are no apparent history of bullous disorder in the family. Investigations showed haemoglobin level 13.6 g/dl, leukocytes 4040/mm3, hematocrit 38%, erythrocyte 3.90 10^6/uL, platelet 200000/uL, monocytes 24.8%.

Our newborn was managed by pediatrician team with intravenous antibiotics (ampicillin 200mg/8 hours and gentamicin 16mg/24 hours), and paracetamol drop 0,35ml/8 hours. Plastic surgeon suggested daily wound dressing with sterile moist gauze and in combination with a topical fucilex cream every 12 hours, proper gentle handling was to minimize trauma and prevent more epidermolysis. After 5 days of preparation, plastic surgeon performed surgical debridement.

Figure 2. Post Debridement

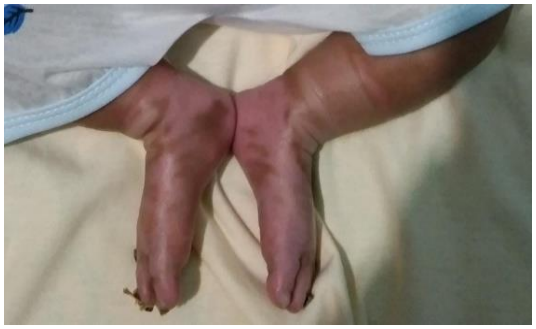

Figure 3. 3 Weeks Post Discharged from Hospital

Our newborn patient was discharged on the 3rd day postoperatively in stable general condition and clean wounds. Parents were trained regarding gentle handling of the child and care for the lesions. Three weeks later, the affected skin on both legs was stable.

DISCUSSION

Epidermolysis bullosa (EB) is a disease that has no cure available to this date, so current treatment aimed to reduce symptoms and provide supportive therapy, prevent forming new blisters and prevent the infection[7]. The primary treatment for Epidermolysis bullosa (EB) is puncture the bullaes with a sterile needle to drain inner fluid and prevent enlargement of the bullaes. It is necessary to cover any open wound with non-adhesive two-layer dressing, a primary one that can be assessed without removal, and secondary dressings that are placed over and can be changed more frequently. Furthermore, suspected infection should be initiate with antibiotics therapy[8]. In this case, we performed additional treatment with debridement as it showed has a promising result[9]. The purpose is to clean the wound, remove devitalized tissue, deroofed blisters and initiate reepithelization[10].

The treatment demonstrated greatly effective and efficient in achieving these aims. Observation on day 3 did not reveal any sign of infection, showing controlled symptoms and good healing process. Patient was discharged on day 3 post debridement due to controlled clinical sign and prevent from risk of nosocomial infection for staying longer in the hospital.

Hospitalized patients are at higher risk of infection, and those with longer length of stay are more likely to develop nosocomial infection. Shorten the length of stay would reduce risk of infection[11]. In general, Epidermolysis bullosa (EB) patients need to stay in hospital for 2-3 weeks until reached controlled disease[12]. Although some of them stayed in hospital for more than 4 weeks[13]. Interestingly, our patient has stable condition no longer than 3 days post debridement. It shows that debridement management in this Epidermolysis bullosa (EB) patient would minimize duration of hospital Length of Stay (LOS), then decrease the possibility of nosocomial infection. In addition, hospital Length of Stay (LOS) is associate with cost hospitalization. Reduced Length of Stay (LOS) in hospital leads to significant savings in hospital costs[14]. Hence, performing debridement on Epidermolysis bullosa (EB) patient result in less risk nosocomial infection and less cost hospitalization.

CONCLUSION

Epidermolysis bullosa (EB) is a rare disease that required supportive treatment. Performing surgical debridement on EB neonate has a preferable outcome that shorten the duration of hospitalization. Thus, it provides low-risk nosocomial infection and low-cost hospitalization.

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DISCLOSURE

The authors have declared that no conflict of interest and no source of funding in this study.

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