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Case Report

Severe Staphylococcal Scalded Skin Syndrome Following Purulent Conjunctivitis

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Abstract

Staphylococcal Scalded Skin Syndrome (SSSS) is an uncommon toxin-mediated disease which causes blistering and desquamation of the skin and induced by *Staphylococcus aureus* (S. aureus). SSSS in neonates is a rare condition and most commonly seen in children aged 6 months to 5 years, with the highest probability of disease being between 2-3 years. This report presents a 35-day-old infant who developed exfoliation and peeling of the skin after purulent conjunctivitis.

Introduction

Staphylococcal Scalded Skin Syndrome (SSSS) is the most common bacterial skin disease seen in childhood, which was caused by *Staphylococcus aureus* (S. Aureus) [1]. SSSS was first described in neonates and characterized by the appearance of bullae and the separation of extended areas of epidermis after infection by exfoliation toxin producing S. Aureus. A pathological process in SSSS is between stratum granulosum and stratum lucidum layers. SSSS often heals without scarring when treated appropriately [1]. Because of this SSSS was admitted to pediatric critical care units for multidisciplinary management.

In this case report, we present a case of SSSS in a neonate. Furthermore, we tried to highlight the importance of early diagnosis and treatment. Early initiation of treatment and protection from infection are the most important steps in the management of SSSS.

Case

A 3000 g mature infant was born at 38 weeks gestation spontaneous vaginal delivery. Postnatal thirty-fifth day he had applied to a hospital because of intractable seizures. Cranial Magnetic Resonance (MR) imaging revealed cystic encephalomalacia than phenobarbital and levetiracetam treatments were started. Metabolic screening tests had revealed normal results. Due to refractory seizures, he was referred to our hospital. Pancytopenia was detected in his laboratory tests and initially, we thought that antiepileptics caused this. But then we also thought that it is not medicinal because it was also in the old laboratory tests. Bone marrow aspiration was performed for the patient by a pediatric hematologist. Bone marrow aspiration revealed hypocellular bone marrow and granulocyte suppression was remarkable. The patient was hospitalized to the pediatric neurology clinic with the diagnosis of cystic encephalomalacia, epilepsy, and pancytopenia. His antiepileptic treatment was replaced to phenytoin and levetiracetam. Because of sudden respiratory failure during EEG monitoring, he was transferred to the pediatric intensive care unit. He was intubated and connected to a mechanical ventilator. The overall condition was worse, the axillary body temperature was 37.7°C, heart rate was 150/min, arterial blood pressure was 86/65 and there was no spontaneous respiration. On physical examination, there was widespread erythema and peeling in his body and purulent conjunctivitis. Laboratory investigations revealed: leukocyte: 1700/mm^3, absolute neutrophil count: 100/mm3, hemoglobin: 7.2 g/dL, platelet count: 18000/mm^3, sedimentation rate was 25 mm/h, sodium: 135 mmol/L, potassium: 3.2 mmol/L, calcium: 7.8 mg/dL, phosphorus: 3,4 mg/dL, aspartate aminotransferase: 34 U/L, alanine aminotransferase: 198 U/L, total protein: 3.5 g/dL, albumin: 2.3 g/dL, urea: 27.3 mg/dL, creatinine: 0.29 mg/dL, C-reactive protein: 67,1 mg/L, prothrombin time: 17.3 seconds, active partial thromboplastin time: 35.5 seconds, pH: 7.42 pCO2: 30.3 mmHg, pO2: 85 mmHg, HCO3: 23.4 mmol/L. Bullous lesions began to form on the flexor faces of the extremities of the patient, which was a common erythematous mass in his body. The epidermis were separated from the dermis, deeply peeled off and Nikolsky sign was positive in the regions where compression was applied.

The differential diagnosis of this patient included drug hypersensitivity, viral exanthemas, scarlet fever, Epidermolysis bullosa, bullous impetigo, Stevens-Johnson Syndrome (SJS), Toxic Epidermal Necrolysis (TEN), and SSSS.Systemic symptoms, such as fever, fussiness, ill appearance,

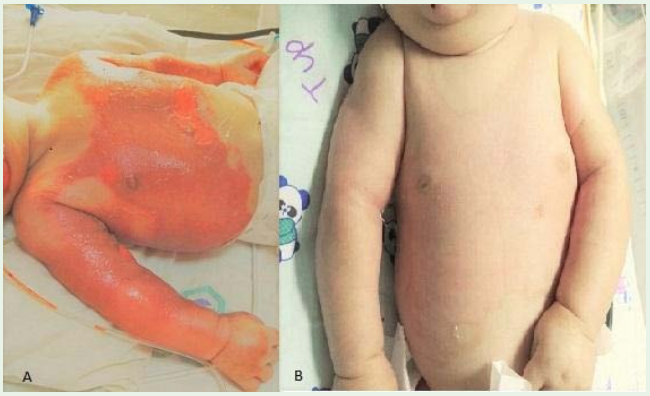

Figure 1: Skin lesions were totally improved after 2 weeks of treatment.

and diffuse skin involvement, are associated with SJS, TEN, and SSSS. Epidermolysis bullosa, a congenital blistering disease, is often present at the birth or during the newborn period. Bullous impetigo rashes are seen on exposed parts of the body, primarily the face and extremities. In bullous impetigo, blisters form at the initial site of infection and not in other areas of the body (localized disease). Scarlet fever is a disorder that causes a diffuse non-bullouseruption, bacterial pharyngitis, and Pastia's lines in flexural areas of the body. Mucous membrane involvement is seen in SJS, TEN, and in more severe forms of Epidermolysis bullosa, but not in SSSS. Mucous membranes and skin were not involved together in our patient. Because of all of these, the main preliminary diagnosis was SSSS in this patient. A skin biopsy and cultures from the lesion, conjunctival swab, and blood were obtained. Intravenous vancomycin, clindamycin, and meropenem treatment was started empirically. Due to some technical insufficiencies, we could not identify exfoliative A and B toxins. Intravenous immunoglobulin (0.5 g/kg) was administered due to severe SSSS and was given for four days. Because of hypoalbuminemia and the presence of edema albumin was also replaced. Serious contact measures were taken and attention was paid to local care guidelines for protection from secondary infections. The wound sites were covered with an antiseptic tulle bandage and wound care was regularly performed on the erythematous areas. Cultures from conjunctiva grew coagulase-positive S. aureus, whereas the bloodstream culture and cultures from lesion sites were sterile. A skin biopsy revealed decomposition in the granular layer of the epidermis, which was compatible with SSSS. It was seen that in the skin biopsy, there was a decomposition in the granular layer of the epidermis, which was in accordance with SSSS. Skin lesions were totally improved after 2 weeks of treatment (Figure 1).

Discussion

Bullous skin problems can be divided into congenital (congenital Epidermolysis bullosa), immunologic (SJS, TEN) and infectious disorders (SSSS). Symptoms of these diseases can be similar to each other [1].

Epidermolysis bullosa is a rare group of inherited disorders that manifest as blistering or erosion of the skin. It is often present at the birth or during the newborn period. Congenital localized absence of skin is known to be a phenotypic pattern that may demonstrate at birth [2]. SJS and TEN are severe type IV hypersensitivity reaction. They usually induced by medications and typically involves the skin and the mucous membranes. SJS and TEN are variants of the same condition and SJS is a minor form of TEN. Skin detachment <10% of body surface area in SJS and >30% in TEN. In SJS and TEN deep blisters and erosions develop at the bottom of the epidermis and mucous membranes which does not occur in SSSS. Although many drugs can cause SJS and TENS, antibiotics, anti-inflammatory, and antiepileptics are associated with these diseases [3,4].

SSSS and TEN are rare, life-threatening conditions with a high mortality rate. It sometimes may be difficult to separate each other because of the similarity of the skin findings. These diseases can clinically appear similar but the important point is that approach of treatment is different. TEN have been usually drug induced and presents with flaccid bullae and extensive erosion. In contrast to SSSS, TEN is characterized by epidermal necrosis and epidermal detachment leading to a dermo-epidermal split and healing with scarring. Both diseases display positive Nikolsky sign. Treatment of SSSS is directed towards the eradication of S. aureus. TEN is usually treated conservatively [5,6].

Bacterial infection of the skin is a relatively common condition in the pediatric patients. SSSS is also one of the major skin infections. In SSSS, large parts of the body get peeled off and appear like burned skin [7]. SSSS is most commonly seen in children aged 6 months to 5 years, with the highest probability of disease being between 2-3 years [8]. SSSS is diagnosed mainly based on the determination of the characteristic symptoms and clinical evaluation. The clinical features of SSSS vary from localized blisters to severe exfoliation and characteristic fragile, thin-roofed, flaccid bullae are formed [9]. In SSSS, large parts of the skin surface may be affected, but the mucous membranes are generally protected [7]. Cultures can be taken from areas that mucosal surfaces, (nares, pharynx, or conjunctiva) to confirm the presence of S. aureus, not be taken from intact or denuded bullae. In neonates with a diffuse desquamating and blistering rash, viral or drug-induced SJS/TEN and epidermolysis bullosa also remain high on the differential diagnosis. In these instances, skin biopsy for histologic examination is extremely helpful [10].

Purulent conjunctivitis was present before the occurrence of body erythema in our case. A large, loose bulla appeared on the skin, peeling off easily with a deep light touch. SSSS usually follows a localized infection of the upper respiratory tract, inner ear, conjunctiva, or umbilical stump [11]. Development of SSSS after several conditions, including nasal septum abscess and tooth eruption has been reported [12,13]. Oyake et al. [14] reported a case of SSSS after purulent conjunctivitis in an adult female patient as in our case. To the best of our knowledge, this is the first pediatric case report of SSSS due to purulent conjunctivitis reported in the literature.

Treatment of SSSS includes eradication of the staphylococcal infection and local wound care. The affected site is sterilized and treated with topical application of ointments that contain antibiotics or petrolatum. The empiric choice of antibiotics should include a penicillinase-resistant penicillin, cephalosporin, clindamycin, andin areas with a high prevalence of methicillin-resistant *S. aureus* infection, vancomycin should be considered.Emollients and non-adherent dressings should be applied to the skin [15]. Supportive care including management of dehydration, temperature regulation,

and infection control is very important in these patients [16]. Many specific pharmacological therapies are cited in the literatüre, but a general consensus and evidence base is lacking. However, topical wound care is an integral part of the overall management of this condition. The areas of epidermal loss can be compared to the wound of a partial-thickness burn and thus many different wound dressings are available. At present, there is no general agreement as to what is the most efficacious and appropriate wound care material. There are a number of new skin substitutes (epidermal, synthetic, one-time application wound and burn dressings) that are used in the treatment of dermal wounds. It is composed of a synthetic copolymer of polylactide, trimethylene carbonate, and e-caprolactone and has been used in partial-thickness burns as well as in split-thickness skin graft donor sites. It is reported to reduce pain in both burn and donor site wounds and also to reduce exudation of donor sites compared to a conventional open method. There are some reports of the use of it in SSSS [17] and TEN in a young infant [18].

In conclusion, patients with SSSS have poor temperature control, can lose extensive amounts of fluids, and may develop secondary infections. Such secondary complications significantly contribute to mortality in neonates and young children. It is important to pay close attention to local care guidelines for the protection of secondary infections. Because of this skin substitute scan be used to treat wounds in SSSS patients. Development of SSSS should be kept in mind in children with local infections such as purulent conjunctivitis.

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