

Pemphigus Vulgaris with Toxic Epidermal Necrolysis : A Case Report

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Abstract

Pemphigus vulgaris (PV) is a chronic autoimmune blistering disorders characterized by painful blister formation of the skin and mucous membranes. Although the prevalence of pemphigus vulgaris in children is unknown [1,2]. Toxic epidermal necrolysis (TEN) is a potentially life-threatening skin disorder characterized by widespread erythema, necrosis, and bullous detachment of the epidermis and mucous membranes, leads to sepsis and/or death [3]. It's a report of a case with Pemphigus vulgaris with Toxic Epidermal Necrolysis. This study describes about a 12-year old male child presented with widespread erosions all over the body to the department of dermatology. At presentation child had multiple erosions over face, neck, trunk, axilla, lower limb, involving almost 30% of the body surface area with oral and genitalia mucosa ulceration. General and systemic examination was performed. All routine blood investigations were done. Then child was underwent pus culture/sensitivity test, based on the findings the child was treated with antibiotics and oral steroids.

Keywords: Pemphigus vulgaris; Toxic epidermal necrolysis; Blistering; Autoimmune.

Introduction

Autoimmune blistering disorders are a group of rare skin diseases. It occur when immune system attacks skin and mucous membranes of mouth, nose, and other parts of body. This causes blisters to form. Pemphigus vulgaris is a group of autoimmune blistering disorders characterized by blister formation on the skin and mucous

membranes. Pemphigus, is a life-threatening autoimmune vesiculobullous disease, characterized by Nikolsky's positive flaccid blisters, painful and persistent erosions and crusts in the skin and mucous membranes. The most frequently affected mucosal site is Oral mucosa, with painful, bleeding erosions and ulcers. Genital, ocular, nasal mucosa are less frequently affected [1]. It is caused by circulating autoantibodies directed against keratinocyte cell surfaces A potentially life-threatening disease, it has a mortality rate of approximately 5-15%. The primary lesion of pemphigus vulgaris is a flaccid blister filled with clear fluid that arises on healthy skin [4]. Toxic epidermal necrolysis (TEN) is a life-threatening dermatologic disorder characterized by widespread erythema, necrosis, detachment of the epidermis and mucous membranes, which

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results in gastrointestinal hemorrhage, respiratory failure, ocular abnormalities, and genitourinary complications. TEN is most commonly drug induced. TEN is idiosyncratic, and its occurrence is not easily predicted. TEN involves more than 30% of the body surface [3].

Case Report

A 12-year old male child presented with widespread erosions all over the body and got admitted in to paediatric dermatology department. As statements collected from the child parents, the child had oral ulcers since 10 months, ulcers all over the body since 30 days, recurrent fluid filled lesions over the body since 30 days. All regular general and systemic examinations were done, findings are normal. Based on the local examinations total BSA involved was 30%, multiple flaccid erosions are present over face, neck, trunk, axilla, lower limb, and genitalia are also present. Multiple flaccid bullae over the thigh, axilla, neck are present. Then oral and genital ulcerations are present. Direct nickolskys sign was negative, marginal nickolskys sign positive. All blood investigations were done like complete blood count and electrolyte test. CBC report showed low haemoglobin level, microcytic hypochromic anaemia and neutropenia. Pus culture and sensitivity test was done, aceinobacter baumannii complex are present, it is sensitive to ampicillin/sulbactam, colistin. Child was diagnosed as Pemphigus Vulgaris with Toxic Epidermal Necrolysis with secondary infection. The oral corticosteroids Started at dose of 1 mg/kg, cyclosporine and empirical antibiotics initially. Cyclosporine was stopped because of decreased urine output. Based on pus c/s report IV antibiotics were started. Aceinobacter baumannii, pseudomas aeroginosa, proteus mirabilis, streptococcus pyogens was isolated and based on sensitivity report inj. Clindamycin and Amikacin started initially along with Tab. Doxycycline, later started on colistin, Tigecyclin, and Amphotericin B. other symptomatic treatment measures were included like Anagesics, Antipyretics, H2 blockers, topical medications. Since the Hb level was low 2 units of PRBC were transfused to the child. Child received colistin for 19 days, Amphotericin B for 8 days. Plan is to continue colistin for 21 days Amphotericin B till the culture becomes sterile. Vitals monitored regularly, electrolytes level replaced with IV fluids, daily dressing was done with aseptic technique, intake output monitored regularly, and strict hydration was maintained.

Discussion

Pemphigus is a rare autoimmune skin disorder characterized by blistering formation on the skin and mucous membranes. The most common type is pemphigus vulgaris, which has involvement of painful sores and blisters on skin or mucous membranes, such as in the mouth or on the genitals. Pemphigus can affect any age people, but it's most commonly affecting the people middle-aged people or older. It tends to be a chronic condition, and some types it might be life-threatening if left untreated. Blisters usually begins from mouth [5]. The presented child also had started oral ulcers too; the child had oral ulcers since 10 months, ulcers all over the body since 30 days, recurrent fluid filled lesions over the body since 30 days. The primary lesion of pemphigus vulgaris is a flaccid blister filled with clear fluid that occurs on healthy skin. The blisters are typically very painful but no itching. Blisters in the mouth or throat may make the children difficult to swallow and eat. Since it is autoimmune disorder, It is mediated by circulating autoantibodies fighting against keratinocyte cell surfaces [4], produced antibodies damages the own cells of our skin and mucous membranes. In most of the cases, unknown causes trigger the disease. Toxic epidermal necrolysis (TEN) is one of the life-threatening dermatologic disorder characterized by widespread, irregular shape erythematous with blistering that occurs on all or part of the macule. The Blisters formation results in detachment of the epidermis and erosions more than 30% of BSA. Mucosal membranes are usually involved [3].

Pemphigus vulgaris is not a contagious disease and cannot be transmitted from one person to another The common symptoms of pemphigus vulgaris includes, painful blisters that start in the mouth or skin areas, skin blisters near the surface of the skin that come and go, oozing, crusting, or peeling at the blister site. The different types of pemphigus are diagnosed based on the blister site. They include Pemphigus vulgaris- Blisters usually first appear in the mouth, Pemphigus foliaceus- blisters first appear on the face and scalp, Pemphigus vegetans-blisters that appear on the groin, underarms, and on the feet [2]. The presented child had started blister in the mouth first. On regular physical examination skin blisters can be noticed, presented child had marginal positive Nikolsky's sign. The main aim of treatment is to reduce the pain and symptoms and to prevent the complications like infection.

Andrea Baratta, *et al.*; said about the combination therapy with prednisone and MMF for pediatric Pemphigus Vulgaris appears to be a safe and effective method of approach that is significantly associated with durable remission [6].

Management modalities include following:

- Corticosteroids and immune-suppressing drugs.
- To revert the side effect of steroids, administer calcium and vitamin D supplements.
- Administer antibiotics, antivirals, and antifungals to minimize infection.
- Administration of Intravenous fluids in order to maintain hydration.
- Plasmapheresis; In very severe cases performed, intended to remove the antibodies attacking the skin from the blood.
- Wound management includes; numbing lozenges for mouth blisters, soothing lotions, wet dressings, pain medications, soft-food diets, avoid spicy or acidic foods that may irritate the blisters, avoid too much sun exposure [2].

Complications of Pemphigus vulgaris include the following:

- Infection of skin.
- Chance of sepsis.
- Malnutrition, because of painful mouth sores.
- Medication side effects, such as high blood pressure and infection.

- Death, if left untreated [5].

Conclusion

Pemphigus in children is very rare due to this the confirmatory diagnosis may get delayed, but high suspicion is necessary for the accurate diagnosis in children. Immediate treatment is very essential, and typically involves the use of corticosteroids. The condition is life threatening if the child left untreated.

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Conflict of Interest: Nil

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