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A SINGLE CASE STUDY ON AYURVEDIC MANAGEMENT OF EPIDERMOLYSIS BULLOSA

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ABSTRACT

Epidermolysis bullosa (EB) is a genetic skin disorder characterized clinically by blister formation from mechanical trauma. A mutation in any of at least 18 genes encoding the proteins in the epidermis, basement membrane or dermis causes poor integrity of the skin leading to fragility. EB usually occurs at birth or shortly after. Males and females are equally affected. Occasionally EB may be mild enough at birth not to be apparent, and it is not until the child is older or reaches adulthood before it is detected. The effects that EB has on a child's life varies considerably. Some children have a mild disease that requires taking precautions to prevent injuries. At the other end of the spectrum, EB can cause lifelong disability that needs ongoing medical care. It can be included under *tvak vikara* in *ayurveda*.

KEYWORDS: Epidermolysis bullosa, Tvak vikara.

INTRODUCTION

Epidermolysis bullosa (EB) is the name for a group of rare inherited (Some types are autosomal dominant while others are autosomal recessive) skin disorders that cause the skin to become very fragile.^[1] Any trauma or friction to the skin can cause painful blisters. Blisters can form anywhere on the surface of the skin, within the oral cavity and in more severe forms may also involve the external surface of the eye, as well as the respiratory, gastrointestinal and genitourinary tracts. In some forms of the disease, disfiguring scars and disabling musculoskeletal deformities occur.

EB usually occurs at birth or shortly after. Males and females are equally affected. Occasionally EB may be mild enough at birth not to be apparent, and it is not until the child is older or reaches adulthood before it is detected. the disease is charecterized by symptoms like; skin that blisters easily, blisters on the hands and soles of the feet, thickened skin that may be scarred or change colour over time, thickening of the skin and nails. Currently, there is no cure for EB.^[2] Supportive care includes daily wound care, bandaging, and pain management as needed.^[3] In Ayurveda it can be considered as tvak vikara. Tvak rogas are known by the general term 'Kushtha', derived from the root 'Kushu' which means that it comes out from the inner part to the outer part and destroys the organs of the body, therefore it is called Kushtha.^[4] some of the clinical features

explained are; *Pidakodrama* (Macules), *Sphota* (Eruptions are seen on the hands and feet), *Vaivarnya* (discolouration)^[5] etc. Diagnosis, Prognosis and Treament according to dominance of *dosha* is explained in detail, with usage of both internal administration and external application formulations.^[6]

Case Description

- Recurrent blistering and peeling of skin following mild trauma such as scratching/ hitting since birth
- Persistent mild itching all over the body- >6 yrs.

History

The 8 yr. old male patient was born to non consanguineous parents (LSCS at full term, weighing 2.96 kg at birth) on 31st December 2011 with 10/10 APGAR score. The child was born with absence of skin over right leg from knee joint up to foot. Hence, was shifted to NICU in a private nursing home and underwent treatment for 2 days, but he developed pus in the entire leg. There was also development of blisters at the site of insertion of canula, which burst open later leading to peeling off of skin. So, he was referred to a dermatologist where he underwent cleansing of the wound with antiseptic solution and dressing (with collagen), which was done everyday up to 7 months following which there was skin formation, but while removing the applied collagen the skin used to peel out again. Along with this, blisters filled with blood or watery fluid started to appear following mild trauma



such as scratching. Simultaneously, he also developed itching sensation all over the body. Later recurrent small 3-4 red blisters appeared even inside the mouth, which got cleared off without any medication. Initially they were not washing the affected area with water because of fear of production of pus. But they started consulting a new dermatologist from 8th month onwards, who advised to wash the area with plain water and minimum amount of anti-septic solution followed by application of petroleum jelly or coconut oil and keep the area open. After 3 to 4 days, growth of new healthy tissue was observed and within 9 months, the entire area healed leaving scar marks. Even after this, tendency for recurrence of blistering and exfoliation following a trauma persisted, especially when he used get hit on the affected leg while playing. For above said complaints he consulted physician in SDM hospiital for further treatment.

Personal History

- Diet- vegetarian (smashed and soft food items)
- Appetite-good
- Bowel-constipated(hard stools-passes daily or once in 2days)
- Micturition-regular 4-5 times/day, 1 time/night, no bladder incontinence, burning sensation or discoloration
- Sleep-sound
- Habit- ice cream everyday

Positive Findings

(Rest all the examinations like; Vital Signs, General Examnation, other Systemic Examination and Investigations were said to be within normal limits, no abnormalities were detected)

Integumentory System

- A. SITE OF LESION: Asymmetrical, Right leg from knee joint up to foot, Neck region in left side and Scalp
- B. SIZE OF LESION: Bullae of 1-2cm diameter
- C. PRIMARY LESION: bullous
- D. SECONDARY LESION: Exfoliation
- E. MORPHOLOGY:
- Colour: Bulla: Initially-Yellowish to pink, when it bursts open- site appears pinkish red
- Shape: Irregular
- Border: Well defined
- Discharge: Present
- Odour: absent
- F. Nails, Mucous membrane- no abnormality detected.

Samprapthi Gataka

- Dosha: Pitta Kapha
- Dushya: Rasa, Rakta
- Srotas: Rasavaha & Raktavaha
- Srotodushti: Sangha
- Udbhavasthana: Amashaya

- Sancharasthana: Sarva Sharira
- Vyaktasthana: Tvak
- Rogamarga: Abhyantara
- Vyadhi Prakara: Chirakari

Differential Diagnosis/Sapeksha Nidana

- Linear IgA bullosa dermatosis
- Pemphigus Vulgaris
- Epidermolysis Bullosa
- Visarpa
- Vispota
- Charmadala Kushta

Chikitsa

- Tab Kaishora Guggulu 1 tid (powdered) A/F
- Cap Yashtimadhu Rasayana 2 tid B/F
- Cap Siddhartaka Yoga 2 tid B/F
- *Bhargavaprokta Rasayana* 1/2tsp [morning B/F with warm milk]
- Yashtimadhu taila for E/A

DISCUSSION

As we discussed above regarding the History, Clinical Presentation and a detailed Integumentory examination, which resulted in getting three differntial diagnosis and by further evaluating the case we confirmed the case to be as Epidermolysis bullosa [EB]. EB is a inherited genetic disorder caused by an absence of basement membrane component due to underlying gene mutation. In ayurveda we diagnosed it as Charmadala Kushta due to involvement of *Pitta & Kapha dosha*. Wherein, the main presentation was blister formation [visphota] and itching[kandu], these are due to pitta dosha and kapha dosha respectively⁷. Treatment provided was basically for tvak vikara, and to improve the immunity.

Figures Before and After Treatment



BEFORE TREATMENT



BEFORE TREATMENT



AFTER TREATMENT

CONCLUSION

A marked improvement was seen in reducing of the recurrent blister formation and good healing process of the pre existing peeled skin.

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