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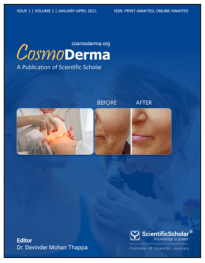

Visual Treats in Dermatology

A case of Bart's syndrome

Arun Somasundaram[1]

[1]Department of Dermatology and STD, Jawaharlal Institute of Postgraduate Medical Education and Research, Puducherry, India.

***Corresponding author:**
Arun Somasundaram, Department of Dermatology and STD, Jawaharlal Institute of Postgraduate Medical Education and Research, Puducherry, India.

arunsomasundaram25@gmail.com

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A 2-day-old neonate presented with areas of absence of skin over extremities and few blisters present since birth. The baby was a full-term firstborn of a nonconsanguineous marriage delivered by normal vaginal delivery and there was no similar history in any family members. The baby was normal in terms of weight, length, head circumference, and vital signs. Cutaneous examination revealed a well-defined area covered with a red glistening membrane over the leg [Figure 1a]. A few discrete erosions were also noted over the forearm [Figure 1b], and nails were dystrophic with an absence of skin over the distal half of the fingers [Figure 1c]. Oral cavity and genital examination did not reveal; any abnormalities. Based on history and clinical examination, a diagnosis of Bart's syndrome was offered – congenital absence of skin with blistering and nail changes. The child was not evaluated with us further as the child was taken to another center. Parents were counseled for genetic testing and we came to know that the child succumbed to death 5 days later due to sepsis.

Bart's syndrome is orchestrated by the congenital absence of skin on the lower leg and blistering of skin, mucous membranes, and nail dystrophy. Bart's syndrome has been classified as Type VI aplasia cutis congenita (ACC) by Frieden, it is a misnomer since ACC indicates failure of the development of skin, while in Bart's syndrome, skin is present initially and lost later.

Cutaneous lesions appear as a well demarcated glistening red moist ulcerations from the dorsal and medial surface of the foot extending to the shin which are usually unilateral. The skin around the nose, oral cavity, and ears can be affected due to friction and trauma. Nail changes include nail dystrophy or anonychia. Associated abnormalities include pyloric atresia, rudimentary ear

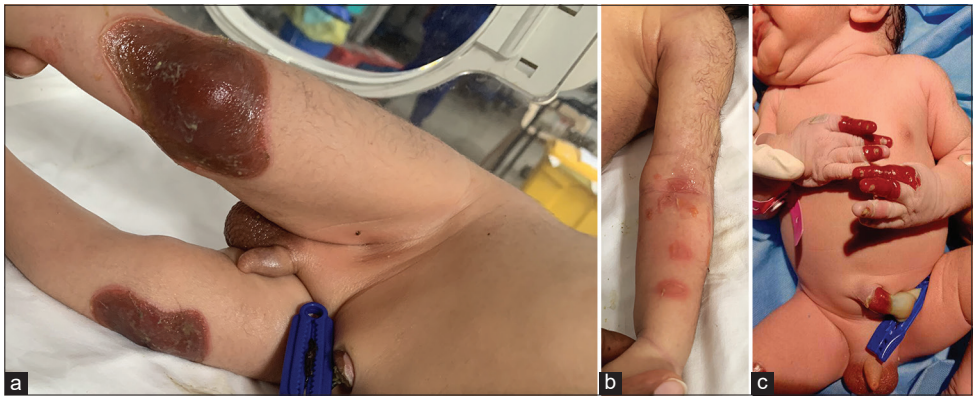

Figure 1: (a) Well-demarcated shiny red glistening membrane over lower leg – the absence of skin. (b) Discrete erosions over the forearm. (c) Nail dystrophy associated with the absence of skin.

development, flattened nose, broad nasal root, and wide-set eyes. The inheritance pattern is autosomal dominant; however, isolated cases have been reported. Abnormalities of anchoring fibrils (Type VII collagen) at the dermoepidermal junction have been described as the cause.[1] Differential diagnosis includes ACC, Epidermolysis bullosa, Adams–Oliver syndrome, and Kindler syndrome. This case highlights us to recognize this entity when a neonate presents with an absence of skin associated with blistering of skin and nail dystrophy. Definitive diagnosis needs to be done with the help of antigen mapping studies and genetic testing.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The author confirms that there was no use of Artificial Intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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1. Trivedi M, Mehta RD, Ghiya BC, Soni P. Bart syndrome: A deceptively scary presentation. Indian Pediatr Case Rep 2023;3:93-5.

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