Clinical Images and Case Reports Journal

Case Report | Vol 4 Iss 2

Aplasia Cutis Congenita Type VI (Bart Syndrome): Case Report

Hamda Nassereldine* and Hawraa Zgheib

Pediatric Department, Saida Governmental Hospital, Beirut Arab University, Beirut, Lebanon

***Corresponding author:** Hamda Nassereldine, Pediatric Department, Saida Governmental Hospital, Beirut Arab University, Beirut, Lebanon. E-mail: hamda-nasserdine.94@hotmail.com

Received: February 18, 2022; **Accepted:** March 01, 2022; **Published:** March 24, 2022

Abstract

Bart syndrome is classified as type VI aplasia cutis congenita, presented clinically by absence of skin, nail malformation and epidermolysis bullosa [2].

It is a rare familial disease [1]. The exact incidence is unknown but estimated to be 1-2 in 10000 births [8]. Here we present a case of female newborn born with absent skin diagnosed clinically and managed by skin care, well followed up with complete healing.

Keywords: Absence of skin; Epidermolysis bullosa; Nails dystrophy; Skin care

Introduction

Bart syndrome also known as aplasia cutis congenita type VI is one of the extremely rare dermatological diseases [1] characterized by absence of skin. It is first described in 1966 by brucel J. Bart. Clinical manifestations are described by Bart triad: aplasia cutis, epidermolysis bullosa and nails deformities [2].

In this paper we are reporting a case of newborn female born with the classical clinical trial of Bart syndrome without any other systematic involvement.

Case Presentation

We report a case of full term baby, born via normal vaginally delivery for a healthy primigravida mother. Maternal history was negative, no pregnancy complications was mentioned, no drugs or radiation exposure was noticed. The mother denies family history of similar disease. Baby was born pink tonic and crying, no resuscitation was needed upon delivery, she received routine postnatal care, however upon physical examination there was symmetrical bilateral absence of skin over the anteromedial aspect of the legs starting from the knees and extending to the dorsal form of the feet (Figure 1). The lesions were sharply demarcated covered by red ultrathin translucent membrane and vascular structures was visualized. Some finger nails shows dystrophy and no mucocutaneous involvement, otherwise normal physical examination (Figure 2).

The baby was otherwise healthy with normal weight, height and head circumference. He was admitted to neonatal intensive care unit for skin care and further investigations. Screening for any associated anomalies was done including chest X-RAY, KUB, echo cardiography and echo abdominal and pelvis were all normal.

Labs were done and were all within normal ranges (Table 1 and 2).

Table 1: Labs done on admission.

CBCD	
WBC	21100 MM3
RBC	4.48 MIL/MM3
Hemoglobin	15.4 G/DL
Hematocrit	48%
Neutrophils	68%
Lymphocytes	22%
Platelets	341000 CU/MM

Table 2: Labs done on admission.

BIOCHEMISTRY	
Magnesium	1.84 MG/DL
Calcium	9.5MG/DL
Fasting blood sugar	105 MG/DL
Chloride	104 MMOL/L
Potassium	4.58
Sodium	137 MMOL/L
Total bilirubin	8.59 MG/DL
Direct bilirubin	0.49 MG/DL
SGPT	11 U/L
Urea	24 MG/DL
Creatinine	0.5 MG/DL
CRP	45.7MG/L

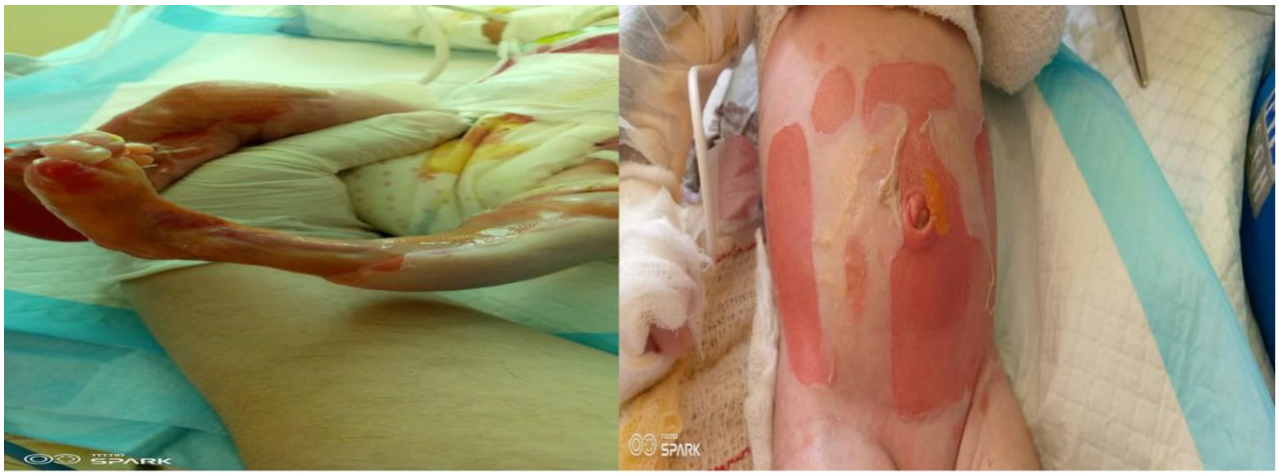

Figure 1: Symmetrical bilateral absence of skin over the anteromedial aspect of the legs starting from the knees and extending to the dorsal part of the feet. The lesions are sharply demarcated covered by red ultrathin translucent membrane and vascular structures.

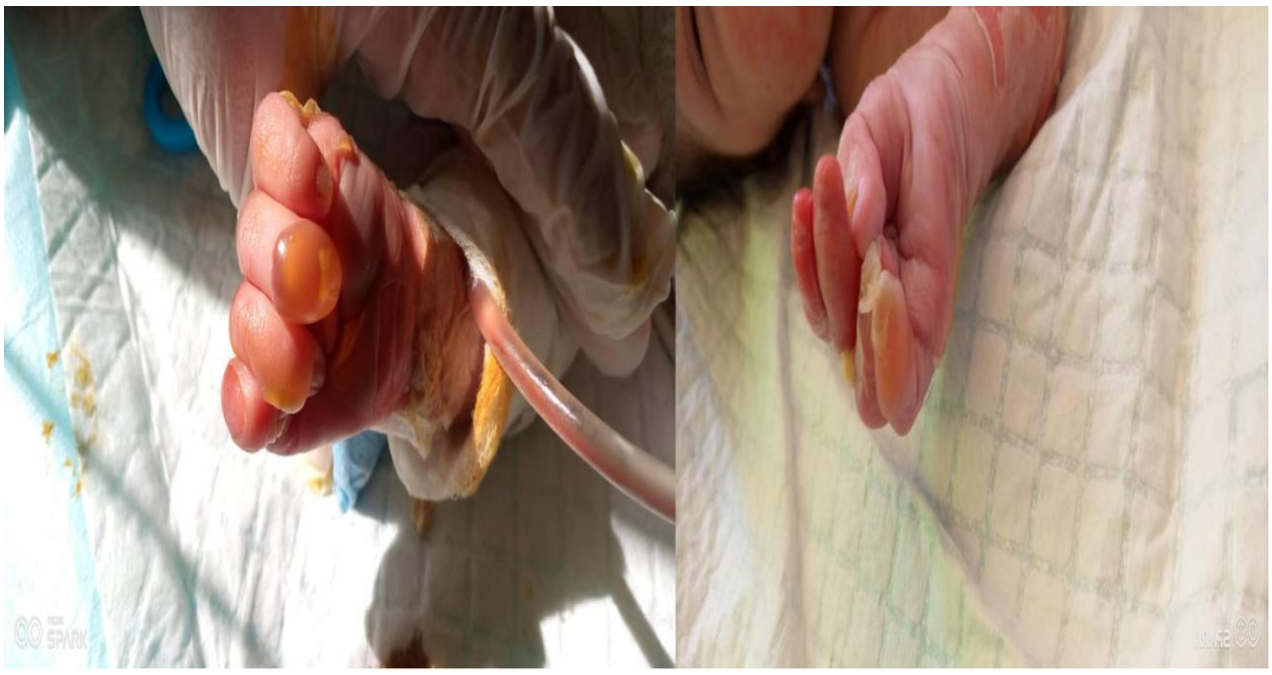

Figure 2: Finger nails dystrophy.

Dermatology team was consulted where they give routine daily skin care using daily dressing with normal saline and antiseptic solution. Mebo cream and fusidic acid were applied twice daily and the wound was closed by vaseline gauze. Systematic intravenous antibiotics was given (ampicillin and cefotaxime) for 7 days. After 1 week of skin care infant was discharged home with detailed instruction about wound care and regular follow up. Patient was followed by dermatology center were a complete healing of skin was observed 6-8 weeks after discharge.

Discussion

Aplasia cutis congenita is one of the rare genetic diseases affecting the mechanobullous system. This disease is described by absence of skin and classified into [9] subgroups differentiated by the number and location of skin lesions and associated systematic congenital malformation [3].

Bart syndrome is type VI of aplasia cutis congenita that is characterized by the triad of aplasia cutis, epidermolysis bullosa and nails malformations [2].

It is an autosomal dominant disease [4], however many sporadic cases were reported as in our case were family history was negative. Causes and underlying pathophysiology is genetic and related to COL7A1 gene on chromosome [3,5-6].

This disease could be isolated to dermatological findings but associated systematic malformation could be found especially with those having junctions epidermolysis bullosa (ureteral stenosis, renal anomalies, pylori atresia, flat nose, wide set eyes, broad nasal root) [4,7]. Our case was isolated to skin lesions only. Diagnosis is made clinically, however skin biopsy and genetic testing is required to confirm the diagnosis [8]. In the reported case above diagnosis was based on clinical triad no skin biopsy was done. Regarding the treatment is conservative treatment including proper wound care and infection control. Specific wound care for Bart syndrome should be given using diluted providing iodine and fusidic acid cream. Lesions should be closed by dexpanthol plus chlorhexidine sterile gauze [8]. But in our case regarding the shortage in medical equipment dressing was done using mebo fusidic acid and Normal saline dressing and it was closed using Vaseline gauze. Rarely surgical treatment is given. Bart syndrome can be associated with other congenital malformations that determine the prognosis, also extension and location of skin lesions is considered a prognostic factor. However, prognosis is good with routine skin Care, infection control and regular follow up [9].

Conclusion

Bart syndrome is one of the subtypes of aplasia cutis congenita that affect the skin, it is a congenital disease that is diagnosed clinically. It can be associated with other congenital malformations. The treatment is based on wound care and infection control. It has a good prognosis while a close follow up is done [9].

REFERENCES

1. Rajpal A, Mishra R, Hajirnis K, et al. Bart’s syndrome. Indian J Dermatol. 2008; 53: 88-908.
2. Duran-McKinster C, Rivera-Franco A, Tamayo L, et al. Bart syndrome: the congenital localized absence of skin may follow the lines of Blaschko: Report of six cases. Pediatr Dermatol. 2000; 17: 179-182.
3. Frieden IJ. Aplasia cutis congenita: A clinical review and proposal for classification. J Am Acad Dermatol. 1986; 14: 646-660.
4. Bart BJ, Lussky RC. Bart syndrome with associated anomalies. Am J Perinatol. 2005; 22: 365-369.
5. Christiano AM, Bart BJ, Epstein EH, et al. Genetic basis of Bart's syndrome: A glycine substitution mutation in the type VII collagen gene. J Invest Dermatol. 1996; 106: 778-780.
6. Zelickson B, Matsumura K, Kist D, et al. Bart's syndrome: ultrastructure and genetic linkage. Arch Dermatol. 1995; 131: 663-668.

7. Kulali F, Bas AY, Kale Y, et al. Type VI aplasia cutis congenita: Bart's syndrome. Case Rep Dermatol Med. 2015; 2015: 549825.
8. Kuvat SV, Bozkurt M. Conservative treatment of a patient with epidermolysis bullosa presenting as Bart syndrome: A case report. Case Rep Med. 2010; 2010: 302345.
9. Kulalı F, Bas AY, Kale Y, et al. Type VI aplasia cutis congenita: Bart's syndrome. Case Rep Dermatol Med. 2015; 2015: 549825.