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BART Syndrome with Varus Foot: A Case Report

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Abstract **Case Report**

Bart'syndrome (BS) is a rare genetic disorder affecting newborns which associate congenital skin aplasia (CCA) with all types of epidermolysis bullosa. A girl was admitted immediately after birth for an ulcerative lesion located on the medial part of the left knee, leg and foot, associated with a foot deformity. We performed a simple of the secretion (no germ was founded), and a biopsy that confirmed the diagnosis of epidermolysis bullosa. She benefits of a local care with honey. Orthopedic treatment of varus foot. After 2 'months of follow-up, she had a good recovery with a complete cutaneous regeneration. Genetic counseling was also recommended to the patient's parents but they refused to do so.

Keywords: Aplasia Cutis Congenital, Bart Syndrome, Foot Deformity, Conservative Treatment, Epidermolysis Bullosa.

INTRODUCTION

Bart'syndrome (BS) is a rare genetic disorder affecting newborns which associate congenital skin aplasia (CCA) with all types of epidermolysis bullosa; its diagnosis is clinical, confirmed by histological examination. It was first described in a large family almost half a century ago [2].

Epidermolysis bullosa is a group of hereditary disorders linked to various genetic mutations.

Many abnormalities can be associated such as nail abnormalities such as congenital absence or dystrophy of the nails [8], foot deformity [7].

We report a case of Bart' syndrome associated with varus foot.

OBSERVATION

A girl was admitted immediately after birth for an ulcerative lesion located on the medial part of the left knee, leg and foot, associated with a foot deformity.

She was the youngest of three children, from a non-consanguineous marriage with similar cases in the family: the elder brother, the father and her aunt. The mother had no particular pathological history.

The pregnancy was well monitored without abnormality, delivery natural way, with good adaptation to extra-uterine life: APGAR at birth 10/10, weight 3000g.

She was in a good general condition, without fever. Local examination showed: an absence of the skin on the medial side of the knee and extended over the antero-medial side of the left leg and foot, with a red lesion non-circumferential covered by a translucent membrane with perilesional erythema and, the presence of a few flaccid bubbles around the periphery of the lesion. There was no exposure of the bones **(figure 1).** She had so we noted a varus foot of the homolateral.

Biological Assessment: Reactive-C-proteine of 37 mg/l without any other abnormalities on the blood tests.

Radiograph was normal.

We performed a simple of the secretion (no germ was founded), and a biopsy that confirmed the diagnosis of epidermolysis bullosa. She benefits of a local care with honey.

Orthopedic treatment of varus foot.

After 2 months of follow-up, she had a good recovery with a complete cutaneous regeneration **(figures 2;3**).

DISCUSSION

BART's syndrome was first described by Bruce J. Bart in 1966 [2], which associate CCA usually on the lower limbs with any type of epidermolysis bullosa, and sometimes associated with malformations.

Hereditary epidermolysis bullosa (HES) is a heterogeneous group of genetic diseases with four clusters **(Table 1)** [5], characterized by excessive fragility of the skin and mucous membranes. This fragility leads to skin or mucosal detachments and bullae formation, usually present at birth or early in the first months of life. Epidermolysis bullosa can be transmitted in a dominant or recessive mode [6].

Congenital skin aplasia (CCA) is a rare congenital disorder in which localized or extensive areas of the skin are absent at birth.

The etiology of CCA is undetermined; genetics plays a role in the pathogenesis, but other factors are included such as intra-uterine trauma, local amniotic adhesions, and exposure to teratogens may be involved [9].

Our clinical presentation is similar to those reported in the literature [6]. Other malformations are also reported.

One hypothesis could explain these associated abnormalities; the cutaneous-mucosal lesion may result in unilateral muscle retraction and consequently in skeletal deformity [4].

In our case, the diagnosis was retained on the history; similar cases in the family, so the clinical and histological characteristics.

We did not performan auto skin graft. We have a very good result with complete regeneration on burned children treated by honey in our institution.

Our management meet with that described in the literature [1-3]. Genetic counseling was also recommended to the patient's parents but they refused to do so.

CONCLUSION

Bart's syndrome is a rare congenital skin condition. Management can be conservative with relatively simple methods for rapid and optimal healing without the need for complex interventions.

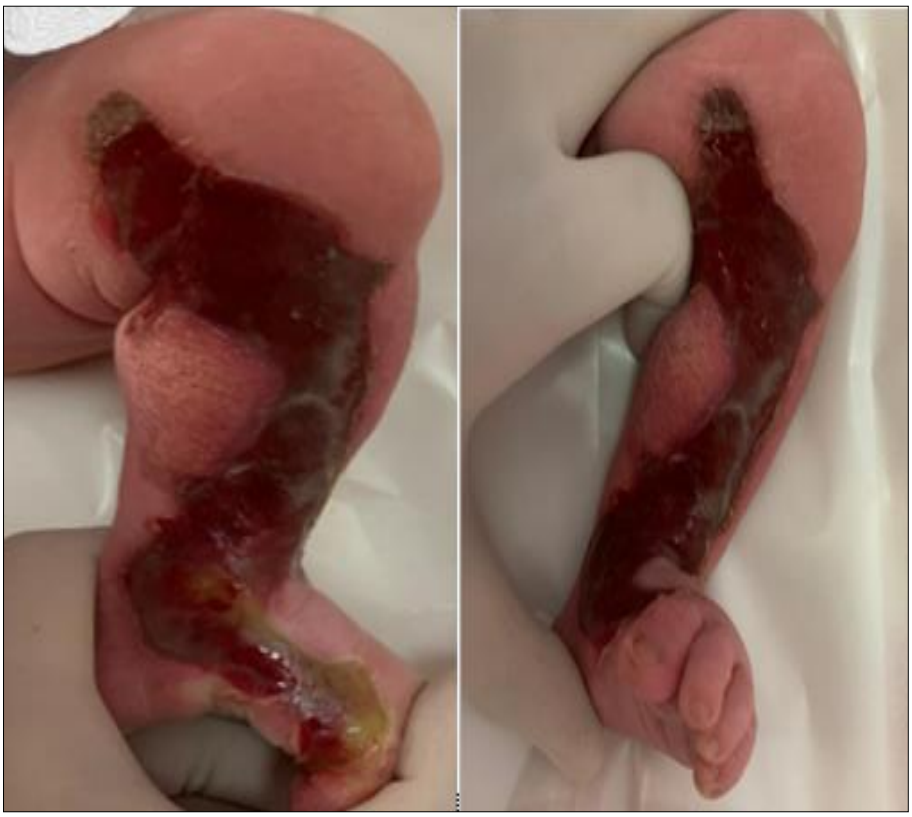

Fig. 1: Clinical image on the first day of life

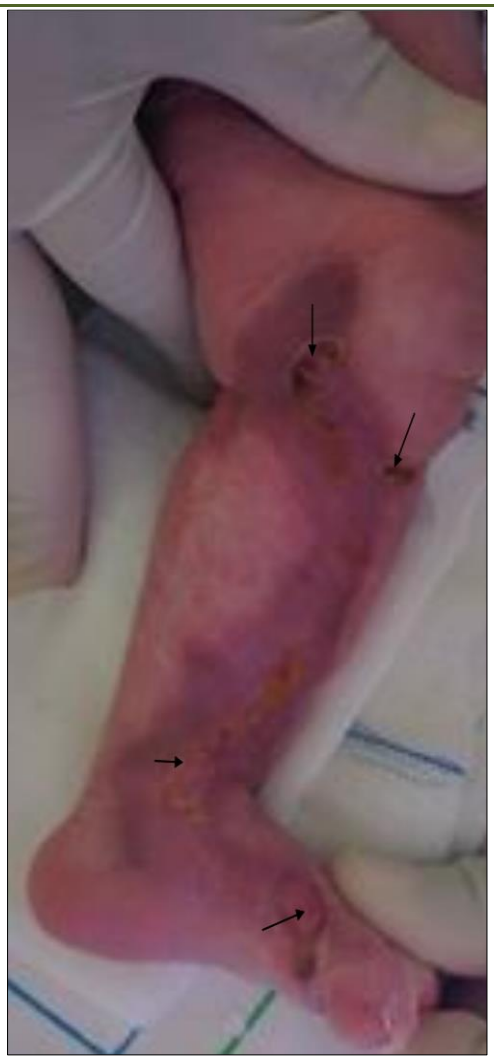

Fig. 2: Clinical picture after 15 days of life, the lesions were completely healed, there were also some bullae healing, as shown by the arrows

Fig. 3: Clinical picture after two months, the lesions were completely healed

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