Picture Story

A case of Bart syndrome

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Background
Bart syndrome (BS) is a rare congenital skin disorder which is characterized by the combination of epidermolysis bullosa (EB) and aplasia cutis (AC). It is also named as aplasia cutis congenita type VI[1]. We present a two month old baby boy with cutaneous blistering lesions and the congenital absence of skin in the lower limbs.

Case Report
A 2 month old baby was referred from a well-baby clinic for further evaluation of skin lesions detected at birth. The baby was delivered at term to a 38 year old mother via normal vaginal delivery with a birth weight of 2.6kg. There were no complications in his antenatal or perinatal periods. The parents were not consanguineous and there was no family history of similar skin lesions.

On physical examination, there were symmetrical well demarcated erosions over the anteromedial aspect of both lower limbs starting from the ankle and extending to the dorsal and lateral plantar aspect of the feet suggestive of cutis aplasia. (Figure 1). In addition, he had blisters on both upper and lower limbs (Figure 2). Later, he developed blistering lesions on the trunk in response to minor trauma or friction suggestive of epidermolysis Bullosa. There was no nail or scalp involvement. Rest of his physical examination was normal.

His basic haematological and laboratory results were within normal range. There were no abnormalities detected in his ophthalmological assessment, ultrasound scans of the abdomen and brain. Diagnosis of Bart syndrome was made clinically based on features suggestive of EB and AC. The baby was managed conservatively with local Fusidic acid cream and mother was advised on minimum handling of the baby.

Discussion
Bart syndrome was first described in 1966 by Bruce J. Bart who published 26 family members with the affected condition[2]. BS consists of a triad of epidermolysis bullosa, aplasia cutis and nail dystrophy. However, there can be cases without nail involvement like the index case. The inheritance pattern of BS is mainly autosomal dominant with few reported cases of new mutations. This baby belongs to sporadic category due to absence of family history.

In addition to mucocutaneous involvement, BS has other associations such as pyloric atresia, microtia, flat nasal bridge and hypertelorism[3]. None of these abnormalities were there in this baby. The diagnosis of BS is usually done on clinical features but the microscopic appearance of the affected skin may aid the diagnosis[4]. The management of BS is mainly based on supportive care. Local application of diluted povidone iodine, fusidic acid and application of non-adhesive bandages impregnated with dexpanthenol and chlorhexidine are recommended forms of wound care and it is not recommended to give systemic antibiotics prophylactically[5]. The prognosis of BS is considered to be good and they have normal life expectancy. However, it is important to protect these children from hypothermia, infections and excessive friction[6].

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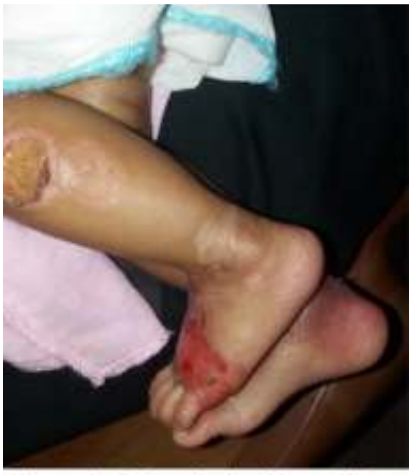

Figure 1: ***Cutis aplasia of lower limbs***

Figure 2: ***Two month old baby with Bart syndrome***

References

1. Duran-McKinster C, Rivera-Franco A, Tamayo L, De La Luz Orozco-Covarrubias M, Ruiz-Maldonado R. Bart syndrome: the congenital localized absence of skin may follow the lines of Blaschko. Report of six cases, *Pediatric Dermatology* 2000; **17**(3): 179–182. https://doi.org/10.1046/j.1525-1470.2000.01747.x PMid: 10886747

2. Bart BJ, Gorlin RJ, Anderson VE, Lynch FW. Congenital localized absence of skin and associated abnormalities resembling epidermolysis bullosa. A new syndrome. *Archives of Dermatology* 1966; **93**:296–304. https://doi.org/10.1001/archderm.1966.01600210032005 PMid: 5910871

3. Bart BJ, Lussky RC. Bart syndrome with associated anomalies. *American Journal of Perinatology* 2005; **22**(7): 365–9. https://doi.org/10.1055/s-2005-871657 PMid: 16215923

4. Kothari C, Doshi N, Avila A, Martin D. Visual diagnosis: newborn with absence of skin. *Pediatrics in Review* 2014; **35**(10): e49–e52. https://doi.org/10.1542/pir.35-10-e49

5. Kuvat SV, Bozkurt M. Conservative treatment of a patient with epidermolysis bullosa presenting as Bart syndrome: a case report. *Case Reports in Medicine* 2010; **2010**:302345.

6. Kulalı F, Bas AY, Kale Y, Celik IH, Demirel N, Apaydın S: Type VI aplasia cutis congenita: Bart's syndrome. *Case Reports in Dermatological Medicine* 2015; **2015**:549825.