Aplasia cutis congenita on lumbosacral area

Piyush Kumar, Ramesh Chandra Gharami[1]

Department of Dermatology, Katihar Medical College, Katihar, Bihar, [1]Department of Dermatology, Medical College and Hospital, Kolkata, India

Aplasia cutis congenita (ACC) is a heterogeneous group of disorders characterized by well-circumscribed focal absence of epidermis, dermis, and occasionally subcutis at birth and a single alopecic lesion on the scalp is the most common form.[1] The exact etiopathogenesis is not known; genetic factors, teratogens (e.g., methimazole, carbimazole, misoprostol, valproic acid), compromised vasculature to the skin, and trauma are believed to play a role. Some authors consider ACC a form fruste of a neural tube closure defect.[1,2]

A 10-day-old female child presented with two ulcers at her buttock since birth. There was no history of intake of alcohol, azathioprine, diclofenac, methimazole, misoprostol, valproic acid, or any other teratogenic drugs by mother. There was no history of varicella or herpes infection during antenatal period. The baby was second issue of a non-consanguineous marriage and first issue, a 5-year-old boy now, was not having similar skin lesions. The documents available with parents indicated that baby was delivered by normal vaginal delivery without any assistance of forceps and the presentation was vertex. Mucocutaneous examination revealed two deep ulcers with sharply demarcated margin, larger one oval in shape, 1.5 cm × 2 cm in size at upper part of right buttock and smaller one irregular in shape, 0.5 cm × 1 cm in size and both the ulcers were surrounded by folds or creases like purse string [Figure 1]. Floors of the ulcers were clean and slightly reddish. Ulcers were healing spontaneously without any medication. Systemic examination revealed no abnormality. X-ray of the lumbosacral region and routine blood, urine and stool examinations showed no abnormality. Diagnosis of ACC was made based on physical findings. Absence of history of trauma and presence since birth ruled out traumatic ulcer. Mother was counseled regarding this condition and was reassured.

Access this article online

Website: www.idoj.in

DOI: 10.4103/2229-5178.126063

Quick Response Code:

Address for correspondence:
Dr. Piyush Kumar, Department of Dermatology, Katihar Medical College, Katihar - 854 105, Bihar, India.
E-mail: docpiyush@gmail.com

DISCUSSION

"ACC" or "congenital absence of skin" is focal failure of development of the skin. Most commonly, lesions of ACC are found on scalp, lateral to midline and are surrounded by an area of distorted hair growth, termed hair collar sign. The lesions may be found on other regions such as on face, trunk, and limbs. The lesion may be solitary or multiple and when multiple, may be found in a symmetrical manner or along Blaschko's lines.[1,2]

It presents as well-demarcated, oval or circular ulcer with variable size from 0.5 cm to 10 cm in diameter. Depth of the ulcer is also varying from just loss of epidermis to involvement of subcutaneous tissue and rarely periosteum. Lesions which develop earlier in gestation may heal before delivery and hence, may appear as atrophic, membranous, bullous lesion or parchment-like scar. Frieden has suggested a classification, consisting of nine groups based on the number and location of the lesions, and the presence or absence of associated malformations. Diagnosis of ACC is essentially clinical. Smaller lesions may heal spontaneously but larger and deep lesions may need skin grafting. Record of this finding is particularly

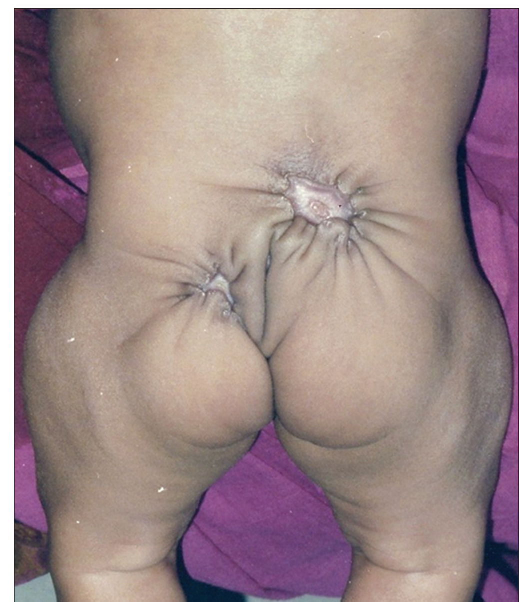

Figure 1: Two sharply demarcated ulcers at lumbosacral area. Note purse-string creases at margin.

important in cases of Battered baby syndrome, a medico-legal condition.[3,4]

ACC in lumbosacral area is uncommon and may be associated with spinal dysraphism, faun tail nevus, meningomyelocoele, and sensorineural deficit in the lower limbs.[5,6] Our patient did not have any of these abnormalities.

REFERENCES

1. Rokunohe D, Akasaka E, Rokunohe A, Kaneko T, Matsuzaki Y, Takiyoshi N, *et al*. Multiple aplasia cutis congenita lesions located along Blaschko's lines in a patient with tetralogy of Fallot-A. J Dermatol Case Rep 2012;6:40-2.
2. Tasin L, Belli S, Chiodini E. Aplasia cutis congenita and methimazole. A case report and literature review. Eur J Pediat Dermatol 2005;15:117-20.
3. Verhelle NA, Heymans O, Deleuze JP, Fabre G, Vranckx JJ, Van den hof B. Abdominal aplasia cutis congenita: Case report and review of the literature. J Pediatr Surg 2004;39:237-9.
4. Skoufi G, Lialios G, Plachouras N, Kutsogiannis D, Mperis A. Aplasia cutis congenita: Successful conservative treatment. Pediatr Int 2006;48:507-9.
5. Chander R, Jain A, Jaykar K, Garg T, Anand R. Faun tail nevus with aplasia cutis congenita. Pediatr Dermatol 2009;26:484-5.
6. Gupta SK, Jasuja DK, Pandit SK. Aplasia cubs congenmta associated with meningomyelocoel. Indian J Dermatol Venereol Leprol 2001;67:329.

Cite this article as: Kumar P, Gharami RC. Aplasia cutis congenita on lumbosacral area. Indian Dermatol Online J 2014;5:103-4.

Source of Support: Nil, **Conflict of Interest:** Nil.