PAEDIATRIC DERMATOLOGY

BULLOUS APLASIA CUTIS CONGENITA

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Background: Bullous aplasia cutis congenita is a rare entity, with an estimated incidence of 3 per 10,000 births. There is a bullous subtype which occurs at birth with very few cases reported in the literature. This could reflect the rare occurrence of the disorder or represent significant underreporting.

Observation: A 1-month-old female infant, with antecedent of being product of the fourth gestate, by vaginal delivery, presented to our Dermatology service with a 3-cm tense blister filled with serous liquid, surrounded by a flat scar and a rim of terminal hairs, located on the left parietal scalp, presented since birth. A skull radiograph showed no bony defect. The patient had no other physical abnormalities. A skin biopsy was performed compatible with aplasia cutis. The diagnosis of bullous aplasia cutis congenita was made.

Key message: Bullous aplasia cutis congenita usually resolves as a scar covered by a thin epithelium. The etiology is unknown, a combination of genetic, teratogenic, pharmacological factors, vascular compromise and trauma has been proposed. Some authors have proposed that bullous aplasia cutis congenita is a form fruste of a neural tube defect and may be derived from a similar embryological defect. The diagnosis is clinical and there is no need to perform a biopsy, its recognition is important to rule out associated defects. Good results are obtained with conservative treatment in a defect up to 7 cm in size. Genetic counseling of the family regarding the risk of recurrence is recommended.