Received: 14 April 2020 | Revised: 8 June 2020 | Accepted: 18 June 2020

DOI: 10.1111/dth.13888

SHORT PAPER

DERMATOLOGIC THERAPY WILEY

Type V aplasia cutis congenita in a preterm newborn successfully resolved

Esmeralda Silva Díaz[1] | Maria Odile Molini Menchón[2] | Andrea Estébanez Corrales[1] | Alejandro Garcia-Vázquez[1] | Javier Estañ Capell[2] | Luis Sáez-Martín[1] | Jose Martín Hernández[1]

[1]Department of Dermatology, Clinical University Hospital of Valencia, Valencia, Spain

[2]Department of Pediatrics, Clinical University Hospital of Valencia, Valencia, Spain

Correspondence
Esmeralda Silva Díaz, Department of Dermatology, Clinical University Hospital of Valencia, 17, Blasco Ibañez Avenue, Valencia 46010, Spain.
Email: silvadiaz.esmeralda@gmail.com

Abstract

Aplasia cutis congenita (ACC) associated with fetus papyraceus is a rare subtype of aplasia cutis categorized as type V in Frieden's classification. It is characterized by stellate lesions in a symmetrical distribution over the trunk and proximal extremities. Conservative treatment is recommended, but there is not a well-defined therapeutic protocol. We report the case of a type V ACC in a preterm male newborn with lesions on the trunk and scalp successfully treated with topical 1% silver sulfadiazine and petrolatum gauze with an excellent evolution. This case associates a severe affectation of the scalp which represents a rare variant of type V ACC.

KEYWORDS

aplasia cutis congenita, treatment ACC, type V ACC

1 | INTRODUCTION

Aplasia cutis congenita (ACC) is a rare disorder characterized by congenital absence of all skin layers in a localized or extensive area.[1] ACC is most frequently found as a solitary lesion on the scalp. However, face, trunk, buttocks and extremities might be affected.[1] It may present as an isolated defect, accompanied by other developmental abnormalities or within a syndrome.[2] Frieden classified ACC into nine groups based on the distribution of the affected area, associated anomalies, and mode of inheritance.[3]

1.1 | CASE HISTORY

A 29-week preterm male with a birth weight of 1240 g presented large stellate atrophic skin defects on his trunk and bilateral proximal extremities at birth (Figure 1A,B) associated with a horseshoe-shaped ulceration on the vertex measuring 5 cm × 1 cm (Figure 2A). A transfontanellar ultrasound scan of the newborn did not reveal any alterations in the underlying bone. A fetus papyraceus (FP) was identified in the placental remnants. The mother was a 32-year-old healthy primigravid woman who conceived a monocorionic diamniotic twin pregnancy via artificial insemination. Intrauterine fetal death of one twin occurred at 14 weeks of gestation without other remarkable complications during the rest of the pregnancy. The only medication taken by the mother during pregnancy was paracetamol for odynophagia. She had not undergone invasive techniques and she denied any history of illnesses during pregnancy. There was no family history of ACC, vascular abnormalities, vesiculobullous disease or any other congenital disorders.

The baby was admitted in neonatal intensive care unit and he required orotracheal intubation and phototherapy by hyaline membrane disease with higher bilirubin levels. The remaining physical examination and complementary exams such as electrocardiogram and abdominal ultrasound were normal. Treatment with 2% topical mupirocin on the scalp and with adherent hydrocolloid dressings on the trunk were prescribed with a slight improvement 2 weeks later (Figures 1C,D and 2B) but due to his skin had eroded and the difficulty in changing the adherent cures the treatment was changed. After checking that bilirubin levels, they had been normalized treatment with topical 1% silver sulfadiazine cream covered by no-adherent petrolatum and hydrocolloid gauze (Urgotul) followed by dry gauze once daily was started. Lower decreased hemoglobin without increased bilirubin levels in capillary blood gas sampling was detected. The patient

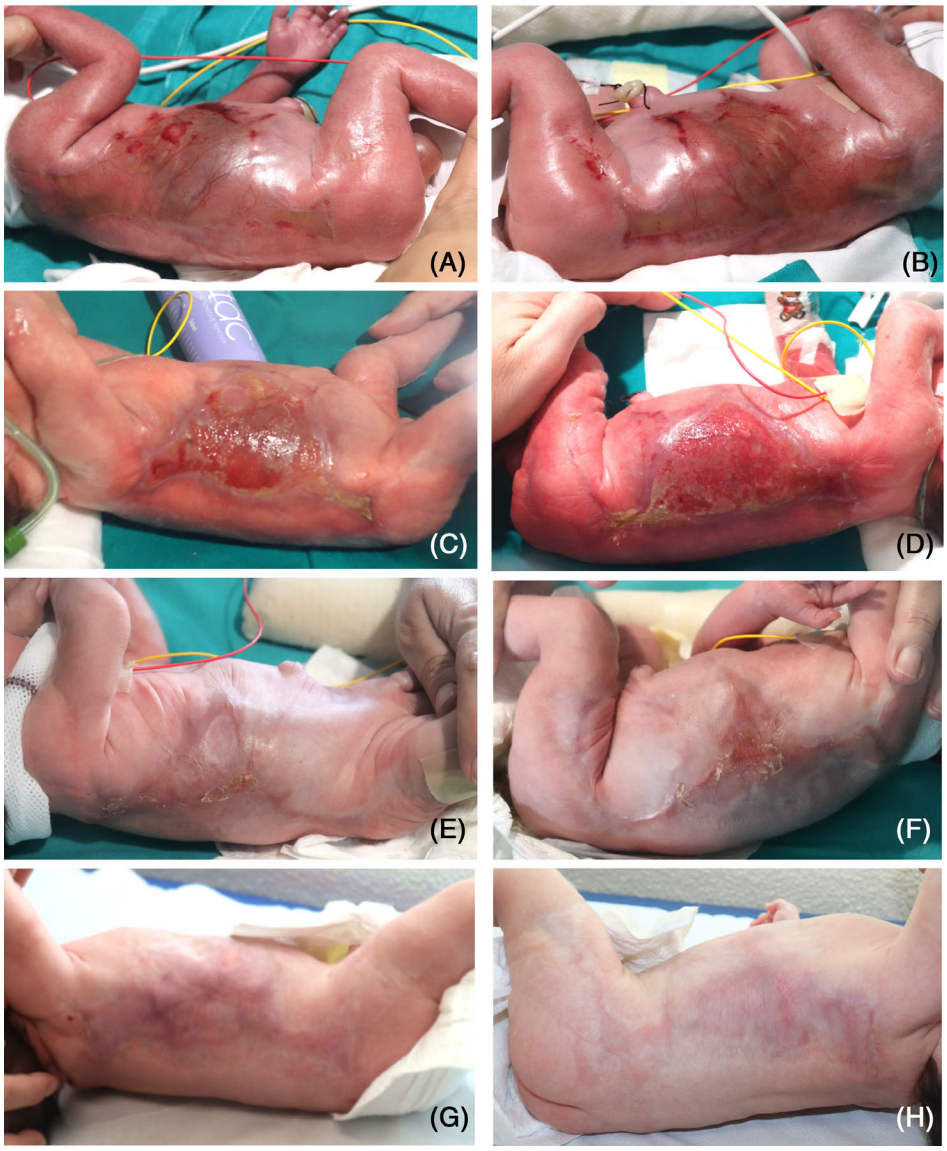

FIGURE 1 Birth: symmetrical bilateral translucent atrophic plaques on the trunk involving the scapular area and proximal area of the lower extremities (A, right side; B, left side). Two-week-old: clinical appearance after cures with hydrocolloid dressings every 72 to 96 hours (C, right side; D, left side). Four-week-old: significant improvement with scars at month after finish treatment with daily cures with 1% topical silver sulfadiazine and petrolatum dressings for 2 weeks (E, right side; F, left side). Twelve-week-old: clinical appearance 12 weeks later after massaging the scars (G, right side; H, left side)

did not develop other complications during the 2 weeks of treatment. At 8-week-old complete healing of the scalp defect associated with a large alopecic area was observed (Figure 2C) as well as total epithelization of the trunk lesions with secondary indurated scars (Figure 1E, F). A massage to soften the scars was done. Twelve weeks after birth the patient had an age-appropriate development with complete healing of the lesions without contractures or mobility limitations (Figure 1G,H). He developed a large scarring alopecic area surrounding the vertex (Figure 2D).

2 | DISCUSSION

ACC associated with FP is a rare subtype categorized as type V in Frieden's classification. It is characterized by stellate lesions in a symmetrical distribution over the trunk and proximal extremities (Snyder, 2019).[2] The presence of scalp lesions in this subtype is rare and it can be an indicator of the involvement of internal organs (Wexler 1990; Pieretti, 2015).[4-7] The ACC etiology is unknown; nevertheless, the use of some toxics (marijuana, cocaine, heroin, and alcohol) during pregnancy might increase the risk of development. Besides this, it has been associated with herpes simplex and varicella zoster viral infections and teratogenic drugs such as misoprostol, valproic acid, angiotensin-converting enzyme inhibitors, benzodiazepines, and specially metimazol.[8]

Type V ACC is associated with twin or multiple pregnancies when one of the twins dies in utero between 12 and 14 weeks of gestation with resultant FP (Snyder 2019).[8] There are different vascular, placental, or ischemic theories about how this fact could alter the correct development of the skin (Snyder, 2019).[6] The differential diagnosis of type V ACC includes congenital focal dermal hypoplasias, amniotic band syndromes, congenital infections, primary bullous diseases, and birth trauma.[8] When a large ACC trunk or low extremity lesion is present, there is a high risk of complications such as infections, fluid, and electrolyte alterations. In addition, there is a risk of local bleeding, thrombosis, or meningitis in cases of extensive scalp lesions that could increase the mortality up to 20% to 30%.[2,6,8]

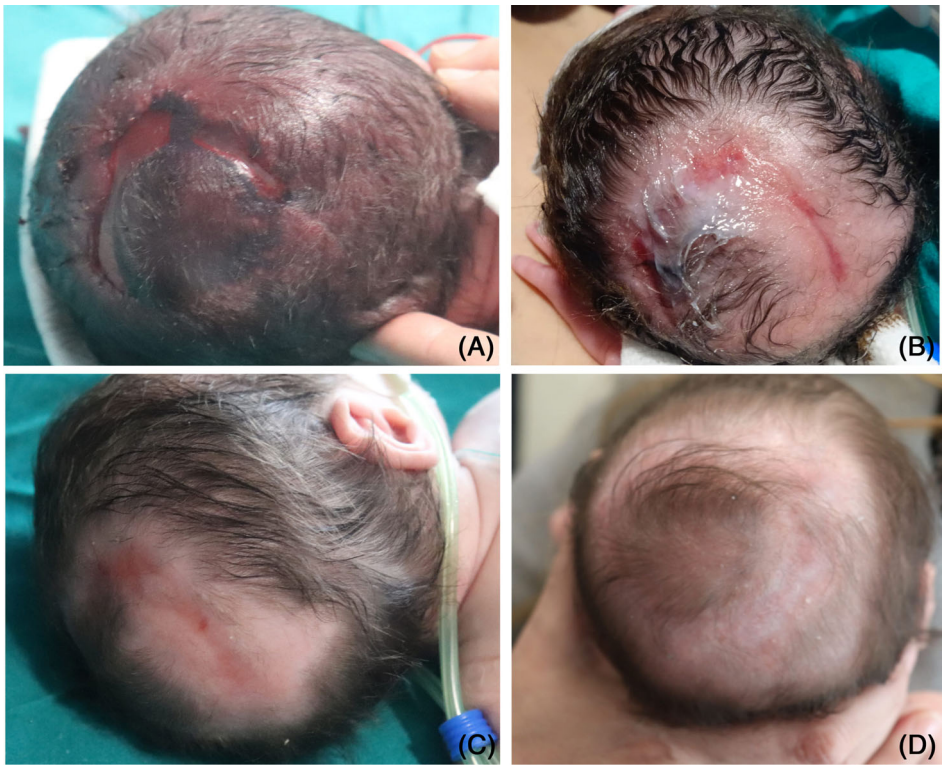

FIGURE 2 Birth: horseshoe-shaped ulceration on the vertex measuring 5 cm × 1 cm, A. Two-week-old: improvement with closure of the defect after 2 weeks of topical treatment with 2% mupirocin, B. Four-week-old: Complete healing with the appearance of an alopecic area after treatment with topical 1% silver sulfadiazine for 2 weeks, C. Twelve-week-old: a large circular alopecic plaque was seen surrounding the vertex, D

Regarding treatment, there is no protocol. Conservative treatment is recommended, and surgery is indicated in those patients who do not improve or those who develop complications (Snyder, 2019; Pieretti, 2015).[1,6,9] As sequelae it can produce scar tissue with contractures of trunk or extremities, in these cases a rigorous massage to soften the tissue is recommended (Snyder, 2019).[6] Exclusive conservative therapy with petrolatum and gauze has been previously used with favorable results.[8-10] However, a recent review of useful treatments in patients with type V ACC recommends to use a topical antimicrobial with near-occlusive dressing to reduce the microbial exposure and the fluid loss.[6] Morrow et al suggested to use the antimicrobial agent silver sulfadiazine twice a day on exposed areas followed by the application of petroleum gauze, dry gauze, and a self-adherent wrap, respectively.[6] Although, there are patients born in a term with extensive type V ACC successfully treated with silver sulfadiazine usage without side effects (Wexler, 1990),[6] the baby must always be monitored due to the risk of potential toxicity and side effects (anemia, renal insufficiency, seizures, increased bilirubin levels, and high liver enzymes).[6,9] Generally, the dressings used in the cures for long term of these patients with extensive lesions are expensive. We used Urgotul which is a no-adherent petrolatum and hydrocolloid gauze that was available in our hospital at the moment. Due to high cost of these type dressings that combine various components, some authors recommend applied each 2 days (Kara M, 2018).[11] In addition, we think that exclusively vaselinated gauzes which are cheaper can be used too.

It is important to recognize type V ACC because multiple pregnancies in the context of assisted reproductive techniques are increasing.[4] Although the affected baby might be at risk of congenital malformations, type V ACC usually has a good evolution with conservative treatment (Snyder, 2019).[4,13]

3 | CONCLUSIONS

We present a case of extensive type V ACC associated with FP in a preterm newborn successfully treated with topical silver sulfadiazine cream with near-occlusive dressing without complications. Although there are mostly born term babies that have been treated with good response and without side effects, the experience in a preterm newborn is limited. In these babies the bilirubin levels and hemoglobin values must be strictly monitored. In addition, this case associates a severe affectation of the scalp and represents a rare variant of type V ACC with few reported cases in the literature to date (Wexler 1990; Pieretti, 2015).[4,5,13,14]

CONFLICT OF INTEREST

The authors declare no potential conflict of interest.

ORCID

Esmeralda Silva Díaz https://orcid.org/0000-0003-2033-2603
Andrea Estébanez Corrales https://orcid.org/0000-0002-5700-6281

REFERENCES

1. Browning JC. Aplasia cutis congenita: approach to evaluation and management. *Dermatol Ther*. 2013;26:439-444.

2. Mesrati H, Amouri M, Chaaben H, Masmoudi A, Boudaya S, Turki H. Aplasia cutis congenita: report of 22 cases. *Int J Dermatol*. 2015;54:1370-1375.
3. Frieden IJ. Aplasia cutis congenita: a clinical review and proposal for classification. *J Am Acad Dermatol*. 1986;14:646-660.
4. Blouin MM, Bernard J, Caron F, Auger I. Aplasia cutis congenita of the trunk and scalp associated with fetus papyraceus. *Int J Dermatol*. 2011;50:733-735.
5. Mannino FL, Jones KL, Benirschke K. Congenital skin defects and fetus papyraceus. *J Pediatr*. 1977;91:559-564.
6. Morrow D, Schelonka R, Krol A, Davies M, Kuang A. Type V aplasia cutis congenita: case report, review of the literature, and proposed treatment algorithm. *Pediatr Dermatol*. 2013;30:e208-e213.
7. Wagner DS, Klein RL, Robinson HB, Novak RW. Placental emboli from a fetus papyraceous. *J Pediatr Surg*. 1990;25:538-542.
8. Tempark T, Shwayder TA. Aplasia cutis congenita with fetus papyraceus: report and review of the literature. *Int J Dermatol*. 2012;51:1419-1426.
9. Ahčan U, Janezic T. Management of aplasia cutis congenita in a non-scalp location. *Br J Plast Surg*. 2002;55:530-532.
10. Starcevic M, Sepec MP, Zah V. A case of extensive aplasia cutis congenita: a conservative approach. *Pediat Dermatol*. 2010;27:540-542.
11. Kara M, Tekgündüz KŞ, Keskin H. A cost-effective treatment model in dystrophic epidermolysis bullosa with congenital absence of skin. *Dermatol Ther*. 2018;31:e12649.
12. Pieretti M, Alcala R, Boggio P, et al. Aplasia cutis congenita associated with fetus papyraceus. *Pediatr Dermatol*. 2015;32:858-861.
13. Snyder ML, Ilyas H. Type V aplasia cutis congenita with fetus papyraceus. *JAAD Case Rep*. 2019;5:3030–305.
14. Wexler A, Harris M, Lesavoy M. Conservative treatment of cutis aplasia. *Plast Reconstr Surg*. 1990;86:1066–1071.

How to cite this article: Silva Díaz E, Molini Menchón MO, Estébanez Corrales A, et al. Type V aplasia cutis congenita in a preterm newborn successfully resolved. *Dermatologic Therapy*. 2020;e13888. https://doi.org/10.1111/dth.13888