ISSN No: -2456-2165 https://doi.org/10.38124/ijisrt/26may1535

Congenital Aplasia Cutis Associated with Anonychia and Hemangioma in a Term Neonate: A Case Report

Dr. Dina Houjjaj[1]; Dr. Sara Mouhmouh[2]; Dr. Sara Yasyn[3]; Dr. Fatima Zahra Azraq[4]; Amal Benbella[5]; Bouchaib Allae Eddine[6]; Abdelhai Adibe Filali[7]; Mohammed Hassan Alami[8]; Rachid Bezad[9]

[1,3,4,5,6,7,8,9]Department of Obstetrics and Gynecology, Les Orangers Maternity Hospital, Mohammed V University, Rabat
[2]Department of Obstetrics and Gynecology, Mohammed VI University, Agadir

Publication Date: 2026/06/10

Abstract:

- ***Introduction:***
  Congenital aplasia cutis (CAC) is a rare malformation characterized by the absence of skin at birth. Its association with congenital anonychia and hemangiomas represents an even more exceptional entity.

- ***Case Report:***
  We report the case of a female newborn, weighing 3,670 g, delivered vaginally at 39 weeks of gestation with an Apgar score of 10, born to a 35-year-old multiparous mother (G5P4) with no notable personal or family history, no consanguinity, and no exposure to medications or medicinal plants. Clinical examination at birth revealed multifocal aplasia cutis lesions (trunk and dorsal region), complete anonychia of the toes, and a large vascular formation consistent with a hemangioma on the right upper limb.

- ***Discussion:***
  CAC may occur in the context of complex syndromes (Adams-Oliver, Bart, Johansson-Blizzard). The association with anonychia and hemangiomas suggests Adams-Oliver syndrome, which requires rigorous multidisciplinary management.

- ***Conclusion:***
  This case illustrates the importance of a precise clinical description and a comprehensive etiological workup in the neonatal period for any newborn presenting with aplasia cutis, in order to establish a prognosis and guide the family.

Keywords**:** *Aplasia Cutis Congenita; Anonychia; Hemangioma; Adams-Oliver Syndrome; Newborn.*

How to Cite : Dr. Dina Houjjaj; Dr. Sara Mouhmouh; Dr. Sara Yasyn; Dr. Fatima Zahra Azraq; Amal Benbella; Bouchaib Allae Eddine; Abdelhai Adibe Filali; Mohammed Hassan Alami; Rachid Bezad. (2026) Congenital Aplasia Cutis Associated with Anonychia and Hemangioma in a Term Neonate: A Case Report. *International Journal of Innovative Science and Research Technology*,11(5), 3950-3953. https://doi.org/10.38124/ijisrt/26may1535

I. INTRODUCTION

Congenital aplasia cutis (CAC) is a rare developmental anomaly of the skin defined by the focal or multifocal absence of skin at birth. Its prevalence is estimated at approximately 1 in 10,000 births. It may present in various clinical forms, ranging from simple superficial cicatricial lesions to deep defects exposing underlying structures. According to Frieden, it is classified into nine groups based on location, morphology, and associated anomalies.

Congenital anonychia—the partial or complete absence of nails at birth—is an even rarer ectodermal anomaly that may be isolated or associated with other malformations. The combination of these two entities with hemangiomas constitutes a clinically rare presentation that may occur within complex genetic syndromes, particularly Adams-Oliver syndrome (AOS).

We report here the case of a newborn presenting with multifocal aplasia cutis, anonychia of the toes, and an

ISSN No: -2456-2165 https://doi.org/10.38124/ijisrt/26may1535

extensive hemangioma of the right upper limb, born at term to a multiparous mother with no notable medical history.

II. CASE REPORT

The patient was a 35-year-old multiparous woman (G5P4) whose four previous pregnancies had been uncomplicated, all delivered vaginally. She had no notable medical or surgical history, no consanguinity, and reported no exposure to teratogenic medications or medicinal plants during pregnancy.

The current pregnancy (G5), estimated at 39 weeks of gestation based on the last menstrual period (LMP), was followed by a general practitioner. Prenatal biometric measurements were normal, and no morphological abnormalities had been detected on prenatal ultrasound.

The patient presented to the maternity ward in advanced labor, with a cervical dilation of 5 cm and complete effacement (100%). Vaginal delivery occurred without instrumental complications. The female newborn weighed 3,670 g, with an Apgar score of 10 at both the first and fifth minutes, and no immediate neonatal distress.

Systematic clinical examination at birth revealed the following abnormalities:

- *Multifocal Congenital Aplasia Cutis*

Two erythematous, well-demarcated lesions with a granulomatous appearance, located on the anterior trunk (thoracoabdominal region) and the dorsal region, with an irregular vascular surface, with no signs of infection or active bleeding.

- *Complete Anonychia of the Toes*

Total absence of the nails on both feet, with erythematous nail beds visible on all toes, and no structural anomalies of the phalanges.

- *Large Vascular Hemangioma*

A well-circumscribed, large, purplish vascular formation localized on the right upper limb (upper brachial region), with a lobulated surface, no pulsatility, and no audible bruit on auscultation.

The remainder of the clinical examination was unremarkable: anthropometric measurements were within normal ranges (head circumference, length); neurological examination was normal (tone preserved, primitive reflexes present); cardiopulmonary, abdominal, and musculoskeletal examinations were normal. The umbilical cord was normal, with the clamp in place.

Initial laboratory workup (complete blood count, C-reactive protein, serum electrolytes, liver function tests) was within normal limits. Dermatological and pediatric surgical consultations, as well as a genetic evaluation, were requested as part of the etiological workup.

III. ICONOGRAPHY

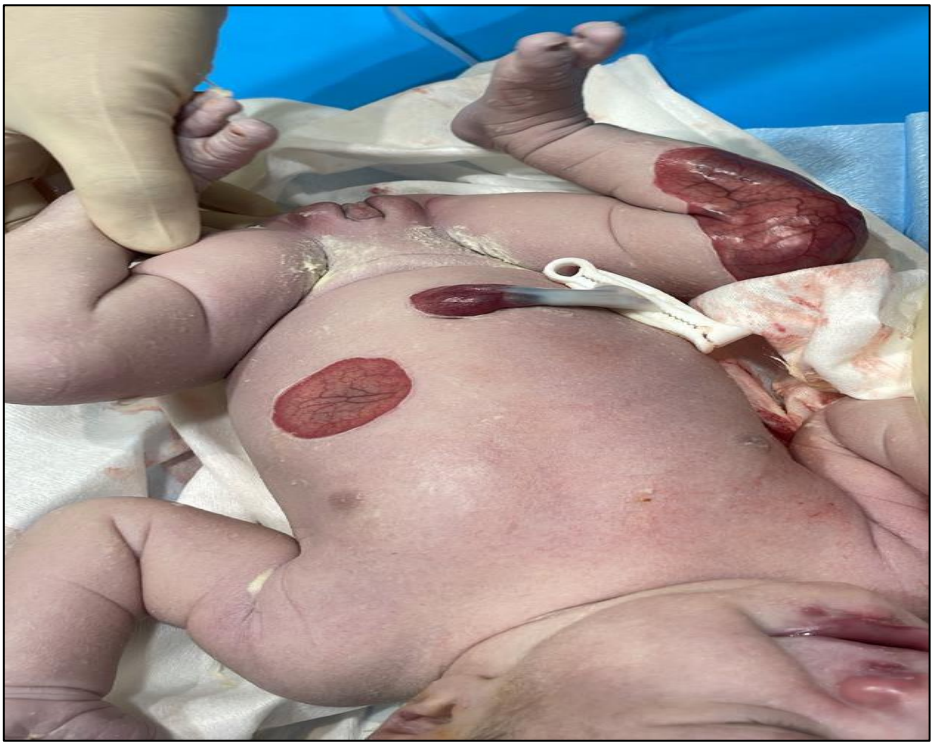

Fig 1 Overall View of the Newborn at Birth. Two Congenital Aplasia Cutis Lesions are Visible: One in the Thoracoabdominal Region and One at the Right Knee. Note the Well-Demarcated, Erythematous, Granulomatous-Appearing Defects with an Irregular Vascular Surface, Without Signs of Infection or Active Bleeding.

ISSN No: -2456-2165 https://doi.org/10.38124/ijisrt/26may1535

Fig 2 Close-Up View of the Newborn's Trunk Highlighting the Multifocal Aplasia Cutis Lesions: The Thoracoabdominal Lesion (Erythematous, Vascular Appearance) and the Hemangioma at the Right Knee, Showing a Well-Circumscribed, Purplish Vascular Formation with a Lobulated Surface.

Fig 3 Complete Anonychia of the Toes. Total Absence of the Nail Plate is Observed Bilaterally, with Erythematous Nail Beds Visible at Each Toe and No Structural Anomaly of the Underlying Phalanges.

IV. DISCUSSION

Congenital aplasia cutis (CAC) is a rare entity first described by Cordon in 1767. Its prevalence is estimated to range between 1/10,000 and 3/10,000 live births. The Frieden classification (1986), still widely used in clinical practice, distinguishes nine groups according to topography and associated malformations. In our case, the trunk-located lesions correspond to Frieden group III, which is often associated with structural or vascular anomalies.

The etiology of CAC is heterogeneous. Genetic factors (mutations in BMP4, ARHGAP31, DOCK6, RBPJ, EOGT, NOTCH1), as well as vascular, mechanical, and infectious factors, have been implicated. In the majority of sporadic cases—particularly in mothers with no relevant history, as in our observation—no specific cause is initially identified.

The association of CAC with anonychia and vascular anomalies (hemangiomas, vascular malformations) strongly suggests Adams-Oliver syndrome (AOS), first described in 1945. This syndrome, with autosomal dominant transmission (or recessive in rare cases), classically combines scalp

ISSN No: -2456-2165 https://doi.org/10.38124/ijisrt/26may1535

(vertex) CAC with hypoplasia or aplasia of the distal extremities (transverse limb defects), and potentially with cardiac, cerebral vascular, or hepatic malformations. In our case, the truncal location of the CAC lesions, complete toe anonychia, and hemangioma constitute an atypical but compatible presentation within the AOS spectrum.

Other syndromes should be considered in the differential diagnosis: Bart syndrome (CAC with congenital epidermolysis bullosa), Johansson-Blizzard syndrome (CAC with exocrine pancreatic insufficiency and dental anomalies), and MIDAS syndrome (Microphthalmia, Dermal Aplasia, Sclerocornea; X-linked). The absence of blisters, the predominantly ungual rather than diffuse ectodermal involvement, and the otherwise normal general examination argue against these diagnoses in our case.

Initial management of CAC lesions relies on rigorous local care (non-adherent moist dressings, infection prevention, monitoring of healing). Superficial lesions, such as those in our patient, generally heal spontaneously over several weeks. Reconstructive surgery is reserved for deep or extensive defects. The hemangioma, depending on its course, may require treatment with oral propranolol or laser therapy if spontaneous regression is insufficient beyond the first few weeks.

The recommended etiological workup includes: genetic analysis (AOS/CAC gene panel), cerebral and cardiac imaging (echocardiography), and ophthalmological and orthopedic evaluations. Multidisciplinary follow-up involving dermatologists, pediatric surgeons, cardiologists, and geneticists is essential.

V. CONCLUSION

Multifocal congenital aplasia cutis associated with complete anonychia and an extensive hemangioma in a term newborn is a clinically very rare association. This case illustrates the importance of a systematic and thorough clinical examination of the newborn from the delivery room onward. The etiological orientation toward Adams-Oliver syndrome should be confirmed by molecular genetic analysis. Early multidisciplinary management is essential to optimize the cutaneous, orthopedic, and vascular prognosis of these newborns.

REFERENCES

[1]. Frieden IJ. Aplasia cutis congenita: a clinical review and proposal for classification. J Am Acad Dermatol. 1986;14(4):646-660.
[2]. Adams FH, Oliver CP. Hereditary deformities in man due to arrested development. J Hered. 1945;36(1):3-7.
[3]. Verdyck P, Blaumeiser B, Fryns JP, Wuyts W. Adams-Oliver syndrome: clinical description of a four-generation family and exclusion of five candidate genomic regions. Clin Genet. 2006;69(2):114-119.
[4]. Snape KM, Ruddy D, Zenker M, et al. The spectra of clinical phenotypes in aplasia cutis congenita and terminal transverse limb defects. Am J Med Genet A. 2009;149A(8):1860-1881.
[5]. Kariminejad A, Bozorgmehr B, Najafi A, et al. Aplasia cutis congenita, epidermal nevus and eye/brain abnormalities in a child with MIDAS syndrome. Pediatr Dermatol. 2014;31(2):193-198.
[6]. Koenig M, Brueton L, Woolfson J, et al. Mutations in ARHGAP31, encoding a RhoA/Rac1 GTPase regulator, cause a novel form of Adams-Oliver syndrome. Am J Hum Genet. 2013;93(2):366-372.
[7]. Patel DG, Bhakta S, Bhatt M. Congenital aplasia cutis congenita: a case series and review of literature. Indian J Dermatol. 2016;61(2):218-221.
[8]. Drolet BA, Esterly NB, Frieden IJ. Hemangiomas in children. N Engl J Med. 1999;341(3):173-181.
[9]. Leauté-Labrèze C, Hoeger P, Mazereeuw-Hautier J, et al. A randomized, controlled trial of oral propranolol in infantile hemangioma. N Engl J Med. 2015;372(8):735-746.
[10]. Swanson AG. Congenital aplasia of the scalp. AMA Arch Dermatol. 1961;83(3):501-502.