Case Report

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A rare case report of aplasia cutis congenita of the scalp with triplet skin defects in non-syndromic newborn

Naim Sulaiman Abuzarifa*, Wan Azman Wan Sulaiman

Reconstructive Sciences Unit, Universiti Sains Malaysia (USM), Malaysia

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*Correspondence:

Dr. Naim Sulaiman Abuzarifa, E-mail: dr.naim2008@gmail.com

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ABSTRACT

Aplasia cutis congenita is an uncommon localized or widespread congenital skin condition characterized by the absence of the skin, and occasionally underlying tissues occurs in about one every 10.000 birth mostly in the scalp as single or more than one lesion and sometimes occurs in extremities and trunk with an uncertain cause and can be associated with numerous syndromes or can be sporadic which is diagnosed clinically and usually conservatively managed but sometimes surgical intervention needed. In this literature, we present nonsyndromic newborn Malay girl normally delivered with 35 weeks gestational age with triplet skin raw areas at the vertex of the scalp with well-demarcated defect round in shape measured about 0.5 in radius covered with a noninflammatory, necrotic patch. To our knowledge, many works of literatures presented Aplasia cutis congenita at the scalp with solitary single or occasionally more than one skin defect. In contrast, in our case, we present an infrequent rare case of triplet skin defect of aplasia cutis congenita for nonsyndromic newborns.

Keywords: Aplasia cutis congenita, Triplet, Skin, Lesions

INTRODUCTION

Aaplasia cutis congenita (ACC) is a rare localized or widespread congenital defects in the skin or its appendages or underlying structures such as bone or dura membrane. In 1767 Cordon was the first reported ACC lesion on the extremity and in 1826 Campbell presented a case of ACC on the scalp. The ACC incidence is about one to 2.8 in 10,000 live births newborns, with a predilection for girls newborn. ACC can be found at any site in the body but about 86% in the scalp and 14% in nonscalp. Most cases of nonsyndromic aplasia cutis congenital are Sporadic or familial, in addition to a nonsyndromic aplasia cutis congenita can be associated with exposure of the mother to a drug as methimazole or anticonvulsant or trauma during pregnancy, in addition to certain viral infections in a pregnant mother can cause the

baby to be born with ACC. The ACC skin defects predominately in vertex presented as single, double or triple noninflammatory, well-demarcated defects size range from 0.5 cm to 100 cm.¹ defects are frequently roofed with a thin, friable, translucent membrane or necrotic patches.

Histopatholgy of the lesion shows the absence of skin appendages such as hair follicles, sebaceous and sweat glands along with the absence of collagen fibres from the dermal layer.²

This literature presents an extremely rare case of nonsyndromic newborn Malay girl normally delivered with 35 weeks gestational age with triplet skin defect of Aplasia cutis congenita in the scalp managed conservatively with modern dressing.

CASE REPORT

We present patient with day one of life baby girl, Malay, 35 Weeks Gestational age (GA). She has two normal siblings. Mother has a history of epilepsy during pregnancy and she was on Keppra since 24 weeks of GA.

On examination

Baby is healthy feeding well, vitaly stable, heart normal, no congenital anomalies in the trunk or extremities, and average bone size upper and lower limbs. No signs or symptoms of syndromic baby, on examination patient having three skin defects well-demarcated circular in shape. Figure 1 size about 0.5 in diameter covered with a noninflammatory, necrotic patch, no bone defect. Patient managed conservatively with polymem foam dressing.



Figure 1: Aplasia cutis congenita at the scalp with three skin defects lesions.

DISCUSSION

Aplasia cutis congenital is an uncommon skin lesion with an incidence of about 0.5 to 2 /10,000 birth. Pathogenesis of ACC developing is unclear, but recently some factors been implicated, example: chromosomal abnormalities, traumatic mechanism, amniotic defects, intrauterine problems, thrombotic events, vascular alterations. teratogens used in pregnancy: misoprostol, cocaine, methotrexate, angiotensin-converting enzyme inhibitors, methimasol, benzodiazepines, valproic acid.³ Many case reports of ACC, several different clinical patterns arise and are characterized by the site and the form of aplasia cutis congenita, associated congenital abnormalities, and type of inheritance. Frieden et al classify aplasia cutis congenita into nine clinical groups.¹ Group 1: scalp ACC without multiple anomalies, Group 2: scalp ACC with associated limb abnormalities Group 3: Scalp ACC with associated epidermal and organoid 4: ACC overlying embryologic Group malformations Group 5: ACC with associated fetus papyraceus or placental infarcts Group 6: ACC associated

with epidermolysis bullosa Group 7: ACC localized to extremities without blistering Group 8: ACC caused by specific teratogens Group 9: ACC associated with malformation syndromes.1 Evers et al mentioned the vascular theory expected thromboplastin material from a fetus papyraceous or placental changes to be the main cause, as well as Biomechanical trauma on the vertex throughout the embryogenesis, varicella and herpes simplex viral infections, intrauterine hemangioma have all explained to be the causes of ACC.⁴ The scalp is the typical location for aplasia cutis congenita, about 85% of the solitary skin defects lesions taking place there, of these scalp skin lesions, 70% to 75% are single, 20% are double, and less than 8% are triple. The scalp's site is at proximity to, the parietal hair whorl in 80% of small scalp lesions.1 Silberstein et al reported 21 cases of aplasia cutis congenita of the scalp with single or double skin defects lesions.² A triplet areas of aplasia cutis congenital are usually common in infants with trisomy 13.5 The treatment for small skin defects of ACC is conservatively while large defects surgical intervention is preferred.⁶ We present in this literature a case of triplet skin defect areas in a nonsyndromic patient, which is extremely rare mentioned in any other literature reviews.

CONCLUSION

ACC is a rare condition characterized by localized single solitary or widespread congenital defect of the skin or underlying structures. Our literature reported an infrequent rare case of scalp Aplasia cutis congenita with triplet skin defects lesions in nonsyndromic newborn.

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