Aplasia cutis congenita as a result of interstitial laser therapy for fetal reduction in monochorionic twins: Conservative approach and outcome

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A R T I C L E   I N F O

Article history:
Received 23 August 2017
Received in revised form 2 October 2017
Accepted 2 October 2017
Available online 10 October 2017

Keywords:
Case report
Extreme prematurity
Interstitial laser therapy
Fetal reduction
Aplasia Cutis Congenita
Outcome

A B S T R A C T

Monochorionic (MC) twin pregnancies are known to carry a high risk of twin-to-twin transfusion syndrome (TTTS) that could lead to miscarriage and perinatal death. Demise of one fetus is frequently associated with co-fetal death. Fetal reduction by interstitial laser therapy is an effective procedure to prevent this outcome, but it may be associated with significant risks for both mother and fetus. Aplasia Cutis Congenita (ACC) may occur in up to 8% cases of fetal reduction by laser therapy. We report ACC in a preterm infant, a survivor of interstitial laser therapy for fetal reduction in MC pregnancy. Despite of massive skin lesions we were able to manage this case conservatively. Follow-up at 5 years of age revealed minimal scarring and no motor function limitations.

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1. Background

Aplasia Cutis Congenita (ACC) is not a common condition. It occurs with an incidence rate of 1–3 cases per 10 000 live births, regardless of sex or race. Most of the lesions present on the scalp, and congenital skin aplasia on the lower limb is a rare finding. The cause of ACC lesions is heterogeneous. Cases of familial ACC have been described, as well as association with genetic disorders such as Adams-Oliver syndrome or Bart syndrome. The common mechanisms of ACC include vascular disruption, trauma, teratogens, or certain maternal medications during pregnancy (methimazole, aza-thioprine, valproic acid).

Monochorionic (MC) twin pregnancies are known to carry a high risk of twin-to-twin transfusion syndrome (TTTS) that could lead to miscarriage and perinatal death. Demise of one fetus is associated with co-fetal death in 12–25% of cases [1]. Fetal reduction is the only procedure that could prevent these outcomes. Interstitial laser therapy is commonly used, and considered an effective and safe procedure but it still carries risks for both mother and fetus [2]. O’Donoghue et al. reported that interstitial laser therapy may be associated with Aplasia Cutis Congenita (ACC) in up to 8% of fetuses [3].

We report ACC in a preterm infant, a survivor of interstitial laser therapy for fetal reduction in MC pregnancy. This work has been reported in accordance with the SCARE guidelines criteria [4].

2. Case report

A healthy 27-year-old primigravida mother underwent intrauterine laser ablation at 26 weeks for demise of a twin due to severe IUGR, bladder agenesis and reversed diastolic flow secondary to Twin–Twin Transfusion Syndrome (TTTS). Follow up ultrasound and Doppler studies of the surviving twin revealed no abnormalities. Upon birth at 27 weeks of GA, two small 0.5 mm lesions were found on the right thigh of infant (Fig. 1). A large (2 cm by 7 cm) lesion covered with granulated tissue, along with a 1 cm lesion were seen on the left leg (Fig. 2).

These lesions were diagnosed as Aplasia Cutis Congenita, a rare potentially serious skin defect involving absence of all skin layers. Dermatology and plastic surgery were consulted; they suggested complete emollient therapy to prevent desiccation of the defect. No other therapeutic interventions were used. Gradual epithelization was noted by the end of 6th week of life (Fig. 3). Even though ACC was found in extremely preterm infant, skin lesions healed with no ulcerations or erosions, with formation of a hairless, atrophic scar, which became less prominent with age (Fig. 4).
3. Discussion

ACC is a result of abnormal ectodermal development that could be triggered by mechanical, vascular, teratogenic, or genetic factors. Etiologic mechanism of ACC in multiple gestational pregnancies is different, and usually related to embolic phenomena due to TTTs [1]. Lesions of the scalp are more common (70–90% of all cases), and usually associated with serious complications (hemorrhage, sagittal sinus thrombosis, central nervous system infection) [5]. Less commonly ACC present on trunk, extremities, or abdomen; frequently these lesions are bilateral and symmetric. Mortality depends on size and location of lesions, presence of associated defects, and degree of prematurity; for example, reported mortality for large ACC of the scalp in term infants ranging from 20% to 50%, and in preterm infants it could reach 95% [6,7].

Management of ACC depends on extend of the defect. Treatment choices include use of non-adherent and adherent dressings, skin graft cultured allografts, autografts with acellular dermis, delayed flap reconstructions [6,8]. Conservative approach with isolated emollient therapy is not common choice for management of large lesions [6,9]. Available literature supports aggressive treatment of large defects to prevent development of hypertrophic scars and patchy contractures [6,7,9]. At the same time, emollient therapy reported to increase sub-epidermal inflammatory cells, decrease necrosis, and support regeneration of epidermis [10].

We believe that ACC lesions in this patient are most likely caused by direct burns from the laser therapy. Typical appearance and asymmetric location of ACC lesions on lateral sides of legs support this theory. Extreme prematurity considered as a possible contraindication for aggressive treatment; thus, emollient therapy was used as a first line treatment. Despite of extensive size of lesions and extreme prematurity of patient conservative management of ACC was successful, and not associated with long term complications.

Conflicts of interest

No conflict of interest.

Funding

No funding.

Ethical approval

As it is case report, no ethical approval requested. Parental consent obtained.
Consent

Written informed consent was obtained from the patient’s parents for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

1 Dr. Veronica Mugarab Samed: direct patient care, data collection, manuscript preparation.
2 Dr. Lodha: direct patient care over the first week of life, collaboration with plastic surgeon, obtained first images of lesions.
3 Dr. ElSharkawy: direct patient care and follow up of patient up to 5 years of age.
4 Dr. Al Awad: direct patient care from day 8 of life till discharge, collaborate with plastic surgery and dermatology, proposed conservative approach for treatment, obtained consent for publication and images at 6 weeks of life.

Guarantor

Dr. Veronica Mugarab Samed.
Dr. Essa Al Awad.

Grant information

Not applicable.

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