ORIGINAL ARTICLE

Managment of Aplasia cutis congenita - early experience

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Abstract:

Background of the study: *Aplasia cutis congenita refers to a heterogenous group of disorders in which localized areas of skin are absent or scarred at birth. No definite etiology is available in literature. Strong genetic predisposition is suspected.*

Objectives: *To overview observed cases and study the rare congenital anomaly.*

Methods: *Total 4 cases that reported in the department of plastic surgery, Dhaka Medical College Hospital included in the study. All the necessary data plotted and analyzed for purpose.*

Results: *None of the patients had positive family history. No parents had history of consanguinity of marriage. None of them had history teratogen exposure or taking drug during pregnancy. No positive history of maternal smoking or alcohol consumption noted. Both legs were most frequently involved 3 cases. Followed by scalp and hand. All the patients were advised for regular follow up. 1 patient died at the age of 30 days in home and lost follow up. Other patients improved after regular dressings.*

Introduction:

Aplasia cutis congenita (ACC) is an uncommon anomaly of absence of skin. It may be localized or widespread. It most commonly presents as a solitary lesion of the scalp and may be as large as 70 percent of that site. Involved areas are well-circumscribed, not inflamed and vary in size from 0.5 to 10 cm or larger. At birth lesions may appear as scars or ulcers[1]. On the scalp they may appear as parchment-like scars with alopecia.

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Although usually benign, they may be associated with other physical abnormalities and syndromes. Frieden classified them into 9 groups based on the number and presence or absence of other anomalies.[2] Nearly 86 percent belong to the first group with a solitary lesion.

Aplasia cutis congenita is an uncommon anomaly of newborns. More than 500 cases have been reported since it was first described, but because of significant underreporting of this generally benign disorder, the precise frequency is unknown. One estimate of incidence is approximately 3 in 10,000 births.[1]

There is no race or sex predilection. It is present from birth.

The cause is not certain as it is the result of more than one disease process. Teratogens, compromised vasculature to the skin, and trauma resulting in early rupture of amniotic membranes are all implicated. Many forms of ACC are inherited.[3]

Maximum tensile force during the development of scalp hair whorl is implicated for the scalp lesion. Early rupture of amniotic membrane forming amniotic bands may also be responsible.

Histological features vary depending on the depth of aplasia and duration. Ulcers are seen at birth. After healing, the epidermis appears flattened with proliferation of fibroblasts within a connective tissue stroma. Total absence of the epidermal appendages remains a characteristic feature.

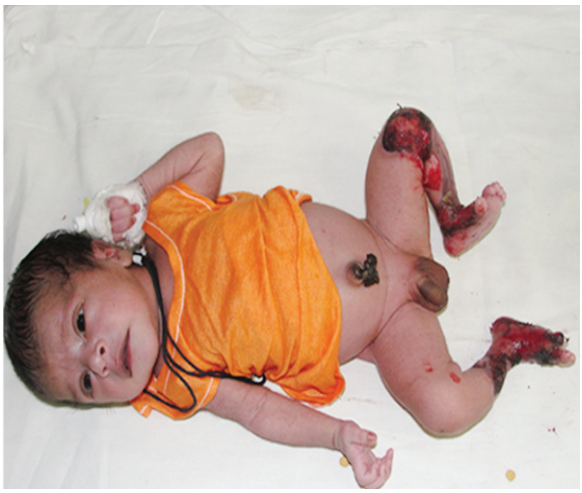

Fig 1: Case 1-loss of skin in legs.

If the defect is small, recovery is uneventful, with gradual epithelialization and formation of a hairless, atrophic scar over several weeks. Small underlying bony defects usually close spontaneously during the first year of life. Surgical repair of large or multiple scalp defects with excision and primary closure if feasible or with the use of tissue expanders and rotation of a flap, may be considered. Truncal and limb defects, despite their large size, usually epithelialize and form atrophic scars which can later be revised if necessary.

Underlying or associated defects may also significantly affect mortality and morbidity. Full-thickness defects of the scalp, skull, and dura are associated with a mortality rate of greater than 50%. Even large defects on areas other than the scalp usually heal well with conservative skin care using silver sulfadiazine ointment. The rare larger scalp defects are prone to complications of hemorrhage and infection; subsequently patients

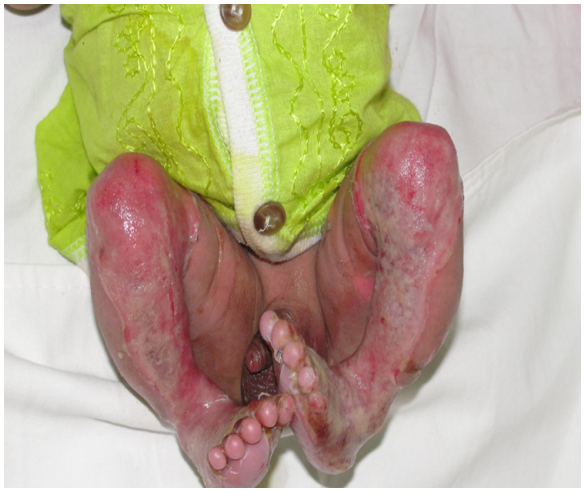

Fig 2: Case 2-loss of skin in legs.

are at risk for death. Extensive aplasia cutis congenita of the scalp may be associated with an increased risk of sagittal sinus thrombosis. For these reasons surgical intervention may be required for large, full-thickness scalp defects.

If the defect is small it can be partially excised and then closed surgically. Usually the only treatment required for aplasia cutis congenital is gentle cleansing of the affected area and the application of a silver sulfadiazine or other ointment to prevent the patch from drying out. Most affected areas will heal on their own over several weeks, resulting in the development of a hairless scar. Surgical repair is not usually needed unless the missing area of skin is large, or there are several areas on the scalp affected.

In the last 3 year, the department of plastic surgery, DMCH managed 4 cases of ACC & hereby the cases are reported.

Case description:

Total 4 patients were treated. Among them 2 (50%) were male and 2 (50%) were female. All the patients had history of absence of skin in different areas of the body since birth. Average age of presentation 3.5 days. None of the patients had positive family history. No parents had history of consanguinity of marriage. Among 4 mothers 3 were under standard antenatal check up and taken vitamin and folic acid regularly. 1 mother was not

Fig 3: Case 2-loss of skin in scalp.

under antenatal care did not take any supportive medicine. None of them had history of teratogen exposure or taking drug during pregnancy. Maternal history of smoking or alcohol was absent. Different area of the body was involved. Both legs were most frequently involved in 3 cases. Followed by scalp and hand. Face was involved in 1 case. Only skin were absent. No case has exposed bone or other vital structure. All the patients treated conservatively by application of silver sulfadiazine and suffratulle dressings. 2 patients were discharged after 15 days of dressings. 1 patient was discharged after 7 days. 1 patient was treated in hospital for 30 days and healed. All the patients were advised for regular follow up. 1 patient died at the age of 30 days in home and lost follow up. Other patients improved after regular dressings.

Discussion:

Aplasia cutis congenita is a rare disorder. This disease is not frequently reported in the world. In our country there is no reported literature. In our case series no sex dominance is noted. No positive family history was noted in our series. Genetic predisposition noted in case report in India[4]. In our series all the patients were referred very early. Various case series reported late presentation also[2-4]. Legs were most frequently effected in our series. Though most frequent effected site was scalp in reported literature[5]. All cases in our series were treated conservatively. 1 patient died after 30 days. Other patients wound healed with scar after regular dressings. No further follow up is continued.

Conclusion:

Aplasia cutis congenita is uncommon disorder present at birth. Diseases like this kind must be reported and further epidemiological study is warranted. Appropriate management by plastic surgeon can decrease mortality and morbidity from the disease. Careful close observation is needed.

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