

# APLASIA CUTIS CONGENITA OF THE SCALP

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## ABSTRACT

*Aplasia cutis congenita is a rare congenital anomaly characterized by localized or widespread areas of absence of skin. Scalp is the most common area involved. No definite etiology is known and multiple factors have been proposed. Our patient was a five days old male neonate who presented with absence of skin over a wide area of scalp with absent scalp bones. Dura formed the base. The lesion was circular and ulcerative with a size of 12 cms x 11 cms on vertex. Regular wound care was provided.*

*Key words*      *Aplasia cutis congenita, Skull defect, Atrophic scar.*

## INTRODUCTION:

Aplasia cutis congenita (ACC) is a disorder first reported by Cordon in 1767. It is characterized by the absence of a portion of skin over a localized or a widespread area.<sup>1</sup> The estimated incidence is approximately 3 in 10,000 births with no sexual or racial predilection. Scalp is involved in 70% of cases and mostly manifests as a solitary defect, though it may present as multiple lesions involving head, the trunk, or the limbs.<sup>2</sup> The lesions are noninflammatory and well demarcated. At birth, the lesions may have already healed and appear as an atrophic, membranous,<sup>3</sup> bullous,<sup>4</sup> or parchment like scar with associated alopecia or may remain as a superficial lesion or erode deeply and occasionally involve the meninges. In this report we present one such case.

## CASE REPORT:

A five days old male neonate reported with absence of skin over a wide area of scalp noticed at birth. The patient was a product of non consanguinous marriage, first in birth order and there was no significant antenatal or family history. On examination, the scalp had a circular, ulcerative lesion of size 12 cms x 11 cms on vertex with dura forming the floor (fig I). There was a distorted area of hair growth along the margins of the lesion. No associated anomalies were found. Imaging by CT scan revealed the absence of bone in the involved areas (fig-II,a,b). The management comprised of wound

care with regular dressings and reconstruction with split-thickness skin grafts.



Fig. I. Large lesion on scalp

## DISCUSSION:

Aplasia cutis congenita is most often an isolated defect,<sup>5</sup> but can be associated with other physical anomalies or malformation syndromes. Based on the number and location of the lesions and the presence or absence of associated malformations, Friedan has devised a classification system for ACC which has nine groups.<sup>1</sup>

As far as etiology is concerned, there is no definite theory that can account for all the lesions of ACC and it is likely that more than one mechanism may be involved.<sup>2</sup> Mechanisms include genetic factors,<sup>6</sup> teratogens,<sup>7</sup> (like methimazole, carbimazole, misoprostol, valproic acid), compromised vasculature of the skin and trauma. Fetus papyraceous has been found to be

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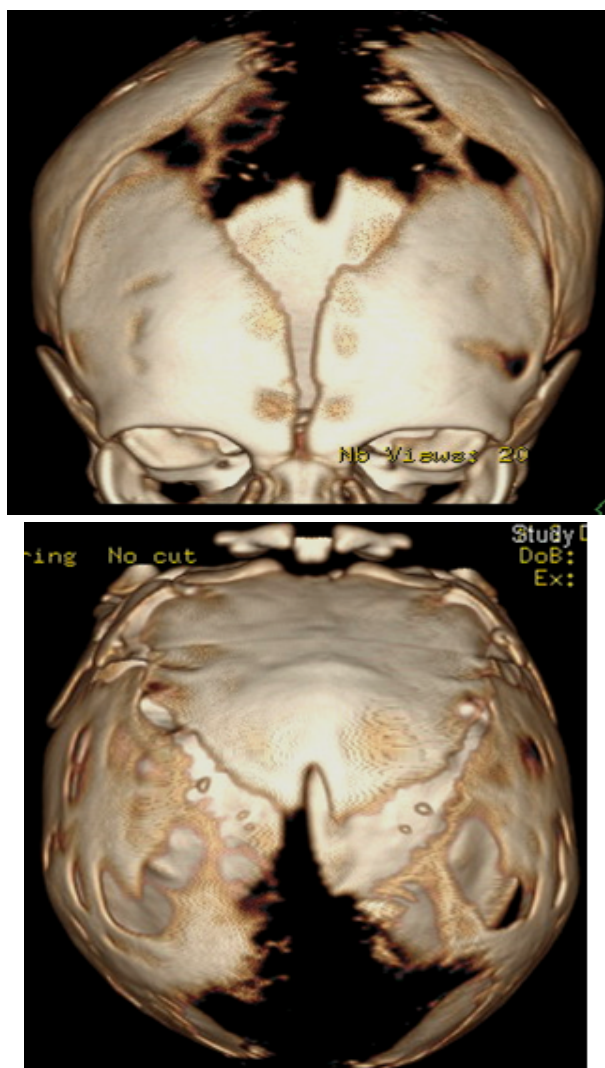


Fig II a and b: Three dimensional CT scan (Coronal View) seen from cranial end showing wide bony defect in skull.

associated with bilaterally symmetric ACC.<sup>8</sup> Differential diagnoses include epidermolysis bullosa,<sup>9</sup> focal dermal hypoplasia syndrome, neonatal herpes, fetal varicella and transient bullous dermolysis of the newborn. These lesions might be mistakenly attributed to birth trauma secondary to vacuum extraction, forceps or fetal scalp monitor electrodes.

The outcome and management of the disorder depends on the extent of the lesion. If the defect is small, recovery is uneventful, with gradual epithelialization over several weeks and formation of a hairless, atrophic scar. Small underlying bony defects usually close spontaneously by the end of first year of life. Large scalp defects may be treated conservatively or might require staged reconstruction, with skin grafts, flaps or tissue expansion.<sup>10</sup>

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