NOT ALL LINEAR TEARS ARE BIRTH INJURIES: NON-SYNDROMIC APLASIA CUTIS CONGENITA- A CASE REPORT

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INTRODUCTION

Aplasia cutis congenita is a rare disorder presenting with absence of portion of skin over localized or wide spread area. Aplasia cutis congenita affects approximately 1 in 10,000 newborns. The incidence of the nonsyndromic form is unknown. There is no racial or sexual predilection. Most affected babies have a single lesion. The lesions vary in size and can be differently shaped: some are round or oval, others rectangular, and still others starshaped. They usually leave a scar after they heal. When the scalp is involved, there may be an absence of hair growth (alopecia) in the affected area. The exact cause is not known. Most common site is scalp and involve d in almost 85% of cases rest 15% cases involve the non-scalp like forearms, knees, both sides of trunk and neck in order of frequency. About 15-30% of the scalp involved patients have skull defects and defects of the dura. ACC is most often a benign isolated defect. No definite etiology has been described. May occur itself or be associated with other physical syndromes or disorders. Diagnosis is made based on clinical findings. Prognosis depends of the organ malfunction level and size of the lesion. We report you a 1 day old newborn with aplasia cutis congenita.

Skin lesions are typically the only feature of nonsyndromic aplasia cutis congenita, although other skin problems and abnormalities of the bones and other tissues occur rarely. However, the characteristic skin lesions can occur as one of many symptoms in other conditions, including Johanson-Blizzard syndrome and Adams-Oliver syndrome.

KEYWORDS

ACC, Aplasia Cuties, J-B syndrome, A-O syndrome

CASE REPORT

G1P1 with term pregnancy having regular ANC underwent elective LSCS in our hospital. Baby cried immediately after birth on examination. Examination revealed that there is a healing ulcer on the occipital region of the scalp, about 2 cm in diameter, floor was healthy, margin was sloping, and totally absence of hair. Other physical examination was with normal limits. All metabolic and hematological laboratory panels were normal. Skull x-ray and cranial ultrasonogram was within normal limits. Finally we arrived at the diagnosis of Non syndromic Aplasia Cutis Baby was treated conservatively and local antiseptic dressings and reassurance to the mother.

Discussion: ACC is an uncommon condition presented at birth. The most common presentation is solitary lesion over scalp. As for etiology is concerned it is still not clear. The proposed mechanism include genetic factors, teratogens, (like antithyroid drugs), vascular compromise of the skin and trauma.

Maximum tensile force during the development of scalp hair whorl is implicated of the scalp lesion. Early rupture of amniotic membrane forming amniotic bands may be responsible. The probability of genetic component thought because of the occurrence of the condition in family members.

Usually non syndromic ACC has sporadic inheritance and the syndromic ACC has autosomal dominant rarely recessive inheritance. Fridan has provided a classification system for ACC which was nine groups. Classifications is based on the number and location of the lesion and modes of inheritance and associated anomalies.

Cutaneous defects noted at birth may be easily confused with obstetric trauma, or from forceps or fetal scalp electrodes.
DIFFERENTIAL DIAGNOSIS

Localised scalp infection, congenital dermoid cyst, small meningocele, heterotopic brain or glial tissue. The main complications of larger defects include infection, bleeding, and thrombosis which may lead to death.

CONCLUSION

Early diagnosis and appropriate treatment are essential to avoid untoward outcomes. Management is either conservative or surgical depending the extent of lesions. After each delivery every neonate should be searched thoroughly for any congenital defect and communicate properly with the patients' party as well as referred the patient to respective department in due time.

REFERENCES