

Aplasia Cutis Congenita of Scalp and Back in A Neonate



Medical Science

KEYWORDS : Aplasia Cutis Congenita, Scalp and back, Congenital anomalies

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ABSTRACT

Aplasia cutis congenita (ACC) is a rare congenital absence of skin, most commonly over scalp. No definite etiology is available. But multiple causes have been suggested such as heredity, teratogens, placental infarcts, vascular disruption, intrauterine infections, alcohol or antithyroid drugs, ectodermal dysplasias, etc. It may be associated with multiple congenital anomalies and prognosis depends on the extent of lesion as well as the level of organ dysfunction. A one day old male neonate presented to us with skin defect over scalp and back. He was clinically examined and investigated to rule out associated congenital anomalies.

INTRODUCTION

Aplasia Cutis Congenita (ACC) is a rare anomaly¹. It most commonly presents as a solitary defect over scalp but may also involve the trunk and extremities. These lesions are non inflammatory and well circumscribed. Size may range from less than 0.5cm to more than 10cm. The incidence is 1-3 per 10000 live births. There is no race or sex predilection. It presents at birth but the cause is not clear. Many factors have been implicated in the etiology like genetic factors, compromised vasculature, infections, teratogens, fetus papyraceous⁴, trauma and so on. It may be associated with other physical anomalies like defects of eyes, limbs, GIT, genitourinary system and CNS. Frieden² classified ACC based on etiology and manifestations into 9 groups. Majority of reported cases (upto 85%) belong to group 1.

The main complications of larger defect can be infection, hemorrhage and thrombosis which may be fatal. So prompt diagnosis and appropriate treatment is required to avoid adverse outcome. Management is mostly conservative. Allogenic dermal grafts have been used to reconstruct large defects.

CASE PRESENTATION

A one day old male, full term neonate delivered vaginally presented with skin defects over vertex of skull and back. The lesions were symmetric, stellate appearance with about 3-4 cm in each radiation (figures 1,2).



Figure 1- scalp lesion



Figure 2-lesion over back

There was no other organ abnormality. Radiological examination and ultrasonography of abdomen revealed no abnormalities. Liver and gall bladder were normal in size and echotexture. Routine laboratory data and liver function tests were normal. MRI of brain and whole spine was found to be within normal limits except for the absence of skin over vertex and dorsolumbosacral spine.

The neonate was managed conservatively in the form of Mupirocine ointment application over the lesions and was discharged after one week.

DISCUSSION

Genetic causes, syndromes, teratogens, intrauterine infections, placental infarction, amniotic pathologies are some of the factors that have been implicated in the etiology of ACC but the exact cause is still not clear^{1,3,5,6,7}. A number of cases have been reported without any etiological factor^{5,6}. ACC lesions commonly occur on parietal and occipital areas and are often small and superficial. Our case had a scalp defect along with a lesion over back which is quite unusual. We ruled out other pathologies accompanying the lesion and also carried out a planned diagnostic evaluation.

Management strategies are based on size, presence of underlying skull defect and child's general condition. Small and superficial lesions may heal spontaneously as is expected in our

case. Large lesions with a bone defect require surgical closure to prevent massive hemorrhage, infection and thrombosis. Although surgery is adequate, conservative management is still preferred in large lesions as surgical exposure of brain and sagittal sinus lead to life threatening infections and death. Nearly 22 cases of ACC with agenesis of scalp and skull had been reported till 1956 and 60% died from causes related to the lesion. Mortality is gradually decreasing since then due to upcoming newer treatment modalities. Early surgery has been reported to prevent vital risks, reduce local complications and make final reconstruction easier. More studies are required to establish the cause and adequate management of ACC.

CONCLUSION

Aplasia Cutis Congenita(ACC) is a rare anomaly and a high index of suspicion is required to diagnose this condition. Though multiple lesions may be present, they can mostly be managed conservatively.

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