ORIGINAL RESEARCH PAPER
Volume - 10 | Issue - 08 | August - 2021 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

NEONATAL APLASIA CUTIS CONGENITA WITH PYOMENINGITIS – A RARE PRESENTATION

Neonatology

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ABSTRACT

Background - Aplasia cutis congenita is a congenital absence of skin in new born. It can be anywhere over body. Aplasia cutis congenita is a rare congenital disorder of skin. **Case report** – We are presenting a case of aplasia cutis congenita in female child associated with pyomeningitis. **Conclusion -** Aplasia cutis congenita, its rarity and unknown etiology is the reason of reporting. It is rarely reported a case of Aplasia cutis congenita with association of pyomeningitis.

KEYWORDS

INTRODUCTION -

Aplasia cutis congenita is a congenital disorder of skin. It is characterized by absence of skin either complete or partially. It may be associated with underlying bone defect [1]. The incidence of aplasia cutis congenita is 0.5 – 3 cases per 10,000 live birth [2,3]. Etiology of aplasia cutis congenita is not clear yet. But various suggested factors are genetics, traumatic mechanism, defective neural tube closure, teratogenic drugs, amniotic defects, vascular compromise ectodermal dysplasia, maternal medication, and intrauterine problems like placental infarcts, infections (herpes and varicella virus), trauma [4,5]. Lesions in Aplasia cutis congenita can be present anywhere over body, but scalp is the most common site and present in more than 80% newborn with aplasia cutis congenita [6,7].

Case report –

A female newborn child admitted in NICU at Rajkiya Mahila Chikitsalaya, Ajmer, Rajasthan with presentation of a scalp lesion over vertex. She was born at our hospital as full term spontaneous normal vaginal delivery. She was a first live birth of first gravida mother. Her birth weight was 1700 gm. Her head circumference was 34.1 cm, and length 47.8 cm. On clinical examination she was dull, poor body movements, and tense anterior fontanelle. There was a lazy triangular shape lesion over vertex. The dimension was 3 cm × 2cm. Scalp bone was not visible through lesion. The lesion was cleaned and dressed with povidine solution. No other associated congenital anomalies were detected.

All base line investigation was normal, only white cell counts were raised. USG brain showed periventricular haziness and mild dilated ventricles. Microbiology of cerebro-spinal fluid (CSF) showed cells contain 120cells/mm3, 80% lymphocyte, 20% polymorphs, sugar 44.6 mg %(RBS at the time of CSF examination is 162mg %), protein 98 mg%, chloride 114 mEq/l. all these features were suggestive of pyomeningitis. Intravenous antibiotics were given for 14 days and she got improved. Gradually she became active and fontanelle became relaxed. USG brain was repeated after seven days and report was normal, no obvious abnormality was detected. On live day 15, she was taking breast feed. She was discharged on live day 16 with weight of 1800gm. Scalp lesion was reducing in size.

Fig. 1 – showing lesion over vertex in reporting case.

DISCUSSION -

Aplasia cutis congenita is a very uncommon congenital skin anomaly. It was the Cordon who described first about it as a lesion on the extremity in 1767 [1]. It is characterized by absent of skin or some time skin defect present with transparent membrane. It is hypothesized that it may be result of in utero degeneration of skin [3]. It was reported that aplasia cutis congenita is comparatively common in children born from consanguineous couples [8,9]. It can be found in association with Johanson-Blizzard syndrome, Adams-Oliver syndrome, trisomy 13 - Patau Syndrome (associated with characteristic facial and cardiac anomalies), Ellis-van Creveld Syndrome, and wolf-hirchhorn syndrome [10,11]. Adams-Oliver syndrome is first described by Adams and Oliver in 1945. It is characterized by presence of congenital scalp defects (aplasia cutis congenita) with limb anomaly [12].

Johanson and Blizzard in 1971 described about Johanson-Blizzard syndrome (JBS). It is a rare autosomal recessive disorder. It is characterized by aplasia cutis congenita, aplastic or hypoplastic nasal alae, exocrine pancreatic insufficiency, mental retardation, hypothyroidism developmental delay, failure to thrive, hearing loss, dental abnormalities, and anomalies in cardiac and genitourinary systems [13].

Frieden classified ACC (Aplasia cutis congenita) in nine groups on the basis of location of the lesions and associated congenital anomalies (table 1). The scalp is the most commonly involved site with lesser involvement of trunk and extremities [14]. Reporting case also have lesion over scalp the common site.

Table 1- showing Frieden Classification

type	Classified description
1	Scalp ACC without multiple anomalies
2	Scalp ACC with associated limb anomalies
3	Scalp ACC with associated epidermal and Organoid nevi
4	ACC overlying embryologic malformation
5	ACC with associated fetus papyraceous or placental infarct
6	ACC associated with epidermolysis bullosa
7	ACC localized to extremities without blistering
8	ACC caused by teratogens
9	ACC associated with malformation syndromes

Aplasia cutis congenita of scalp commonly associated with defect in the underlying bone and dura mater, exposed brain tissue and sagittal sinus [15,16]. In reporting case pyomeningitis is the associated with aplasia cutis congenita of scalp. Treatment of scalp defect either conservative or surgical required preventing further complication [17]. In reporting case pyomeningitis was diagnosed on live day second.

A newer proposed classification for scalp aplasia cutis congenita given by Silberstein E et al. in 2014 according to size and layered involved of scalp and proposed their treatment. [18]. According to this classification our case grouped in type I, <15 cm2 Skin defect, no skull bone defect. It can be management conservatively. We manage conservatively and improved.

CONCLUSION -

Aplasia cutis congenita associated with pyomeningitis, its rarity and unknown etiology is the reason of reporting. It is rarely reported a case of Aplasia cutis congenita with association of pyomeningitis.

Statement of Ethics - The authors confirm that caregivers of their patients were fully informed and they agree to report his case.

Disclosure Statement - The authors have no conflicts of interest to disclose.

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