APLASIA CUTIS CONGENITA

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INTRODUCTION

Aplasia cutis congenital (ACC) is a rare congenital anomaly characterized by partial or complete absence of the skin[1]. This disorder was first described by Cordon in 1767 in the extremities and in scalp by Campbell in 1862[2]. Association of ACC with distal limb anomalies was demonstrated by Adams-Oliver in 1945[3]. Various subtypes are explained on the basis of modes of inheritance, associated malformations, pattern of lesions and their location[4]. The condition has an incidence of 3 per 10000 live births and in 70% of cases occurs as a solitary lesion but may present as multiple lesions with involvement of different body parts[5].

CASE REPORT

We are reporting a case of ACC in a twin delivery in our setup. Twin-1 a baby girl delivered by elective lower segment c-section had a small defect at the left side of the vertex of scalp. The lesion was measuring 1×1 cm circular in shape with clear margins, non-erythematous with underlying bone visible. There were no other congenital malformations, ultrasonography of the brain was normal and the systemic review was unremarkable. Twin-2, a baby girl with similar defect at the left side of the vertex of scalp measuring 2×1.5 cm irregular in shape with clear margins, non-erythematous and underlying bone visible. Systemic examination was unremarkable and ultrasound brain was normal.

Owing to the small size of the defects both patients were managed conservatively with regular follow up presently at two months of age.

DISCUSSION

ACC is a rare disorder having different modes of presentation with multiple theories for its etiology[5]. Various factors thought to be involved in the causation include failure of ectodermal fusion during embryogenesis[6], teratogens[7] and mutations of the BMS-1[8]. The disease can have multiple modes of inheritance including sporadic, autosomal dominant and recessive[9]. The condition was classified into nine different groups based on the number and location of lesions and presence or absence of other malformations by Frieden. Group 1 is scalp ACC with no other anomalies, group 2 is

Twin-1 Twin-2

scalp ACC with limb anomalies, group 3 is scalp ACC with epidermal and sebaceous nevi, group 4 is ACC with a hair collar overlying embryonic malformations, group 5 is ACC with fetus papyraceous, group 6 is ACC with different types of epidermolysis bullosa(EB), group 7 is localized ACC without EB, group 8 ACC due to teratogens and group 9 is ACC with other malformation syndromes[4].

We are reporting the case of ACC in a twin delivery with both female issues being affected in comparison to a similar case in two brothers reported by Ejaz[10].

Management of ACC can be conservative, surgical or a combined approach using both conservative and surgical methods[11]. The conservative treatment allows the granulation tissue to regrow and helps in healing by secondary intention while avoiding the scar formation and dessication. Ejaz and Arif have published reports in local literature where patients have been managed by conservative methods[10,12]. Conservative management options include dressings by paraffin gauze 1, saline

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dressings, bacitracin ointment, betadine solutions and non adherent dressings (MepitilR)[2]. In our case due to smaller size of the defects patients were managed conservatively with recovery of smaller defect at two and the larger defect at three months of age. The larger defects require surgical reconstruction. Various options for the surgical closure include the split thickness skin grafts and the acellular dermal grafts[12,13].

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