APLASIA CUTIS CONGENITA SCALP: MANAGEMENT OPTIONS

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ABSTRACT

Objective: *To document the management of aplasia cutis congenita scalp.*

Material and Methods: *This descriptive study was conducted in the Department of Plastic and Reconstructive Surgery, Hayatabad Medical Complex, Peshawar from February 2000 to September 2007. Eleven patients with aplasia cutis congenita scalp were referred to our unit. A split thickness skin graft from adjacent healthy scalp or thigh was used for coverage of the scalp wound.*

Results: *Among these eleven patients, seven were males and four females. Two patients were observed with associated congenital anomalies. The ages ranged from 05 days to 04 weeks. Mean age was 12.5 days. Split thickness skin grafts were used in all these patients. About 10% graft loss was observed in one patient. One patient developed CSF leakage after wound healing. This patient expired after 03 weeks.*

Conclusion: *Early surgical intervention in the form of split thickness skin graft for wound coverage is the most favorable treatment for Aplasia cutis congenita*

Key words: *Aplasia cutis congenita scalp (ACCS), Split thickness skin graft (SSG)*

INTRODUCTION

Aplasia Cutis congenita is a rare malformation in which there is localized absence of the skin and it can present in any region of the body. Scalp is the commonest location and is involved in almost 85% of cases[1,2]. Fifteen percent of all cases involve the non-scalp areas like forearms, knees, both sides of the trunk and neck in order of frequency[3,4]. The incidence of aplasia cutis congenita scalp has been estimated as 1:10,000 births[5].

It was first described in the extremities by Cordon in 1767 and in the scalp by Campbell in 1862[6,7]. Adams and Oliver reported the association of cutis aplasia with distal limb anomalies in 1945[8,9]. About 15% to 30% of the scalp involved patients have skull defects and defects of the dura matter[9, 10].

The condition is characterized by non-inflammatory, well-demarcated defects of the skin and subcutaneous tissue and more rarely the periosteum, skull and dura, exposing the superior saggital sinus. The more severe forms predispose to multiple potentially life threatening complications including massive haemorrhage, infection and meningitis[11]. Most cases are sporadic but several reports of occurrence in families over several generations suggest a genetic component[10,12]. In the majority of familial cases the mode of inheritance is autosomal dominant with varying degree of penetrance[13-15]. Several theories of aetiology are of historical interest. The possible aetiological factors reported are intrauterine adhesion bands, placental infarcts and mechanical or vascular events during developement[16]. Exposure to teratogens or intrauterine infection has been associated with a number of cases[17].

An increased incidence of aplasia cutis congenita has been observed in offspring of mothers taking carbimazole and methimazole for control of hyperthyroidism[17-19].

Fridens has provided an extensive classification system for description of these defects in groups based on modes of inheritance and associated abnormalities[14]. As Aplasia cutis congenita scalp is an uncommon condition, its management is still a subject of debate for the reason of lack of reliable reconstructive options.

Since there is little understanding of the outcome of different forms of treatment, the management of less severe form is relatively straightforward and the more severe forms are difficult to be taken care of. The mortality rate of all kinds of aplasia cutis congenita is approximately 21%[1,2]. The main objective of this study was to document the management of this rare congenital anomaly which has a very wide spectrum of presentation and there is no consensus on the management protocols.

MATERIAL AND METHODS

This descriptive study was conducted on eleven patients in the department of Plastic and Reconstructive surgery, Hayatabad Medical Complex Peshawar from February 2000 to September 2007. Eleven patients with aplasia cutis congenita scalp were referred to our unit. All the patients were included in this study and there were no exclusion criteria.

All the patients were thoroughly examined regarding the scalp lesion size and associated anomalies and clinical diagnosis of ACCS made. Plain x-ray skulls were performed to rule out any bony defect. The patient particulars like age, sex, site and size of lesion, associated anomalies and the operative details were recorded. These patients were operated under general aesthesia. Split thickness skin grafts harvested with the help of Humby's knife were applied to the defects over the scalp and stabilized with tie over dressings. The dressings were removed five days after the surgery

RESULTS

A total of 11 patients were included in this study. Among them 63.6% (n=7) were male and 36.3% (n=4) were females. The age range was 05 days to 28 days with the mean age 6.5 days.

The maximum scalp defect was 7x9 cm and the minimum was 1x2 cm. The details of all the patients are given in table 1.

In 10 patients (90.9%) we used split thickness skin graft harvested from the thigh while in one patient (9.1%) we harvested the skin graft

Table 1: Comparison of Patients Age, Sex Associated Anomalies Scalp Lesion and Management with Friden's classification

No	Age	Sex	Friden's classificat-on Group	Associated anomalies	Scalp lesion	Management
1	2 weeks	Male	Group 1	Omphalocele	Triangular defect 7x9 cm hypertrichosis in the vicinity of the lesion	SSG comparatively loose tie over dressing
2	5 days	Female	Group 1	Nil	Large scalp defect extendin g from anterior fontanelle to the occiput 6x10 cm	SSG
3	3 weeks	Male	Group 2	Clubfoot	Oval shape scalp lesion 6x6 cm	SSG
4	10 days	Male	Group 1	Nil	Circular scalp defect 5x6 cm	SSG
5	1 week	Male	Group 1	Nil	Oval shape defect 4x5 cm	SSG
6	5 days	Female	Group 1	Nil	Oval shape defect 3x6 cm	SSG
7	2 weeks	Male	Group 1	Nil	C ircular defect 4x5 cm	SSG
8	10 days	Male	Group 1	Nil	S calp defect in right temporal area 2x5 cm	SSG
9	1 week	Female	Group 2	Clubfoot	D efect 5x6cm	SSG
10	4 weeks	Male	Group 1	Nil	D efect 3x3cm	SSG
11	2 weeks	Female	Group 1	Nil	S calp defect 1x2cm	SSG

from adjacent scalp.

Out of eleven patients we observed no complications at donor site while we did face complication of the recipient site in two patients. Out of these two patients there was 10% graft loss in one patient who was treated conservatively. In other patient we observed wetting of the dressing over the graft on 4th post operative day. On the removal of the dressing there was a watery leakage underneath the graft. We consulted the neurosurgeon as well as the radiologist for re-evaluation of the CT scan to know about the probable CSF leakage. The patient was shifted to neurosurgery ICU for management of CSF leakage. Radiologist on reassessment of the scan confirmed a tiny calverial defect and the patient was kept on conservative management but the patient expired after 03 days (7th post operative day). In rest of the patients the dressing was removed on 5th post operative day and in all these patients the grafts take was 100%. In subsequent follow up after 2 weeks, 3 months and 6 months, all the grafts were stable and well healed.

DISCUSSION

In our series nine patients were type 1 and two patients were type 2 according to Friedens classification.

Management of aplasia cutis congenita is controversial and may be conservative, surgical or combination of both[1]. The goal of conservative treatment is to allow granulation tissue and healing by secondary intention while avoiding desiccation and scar formation.

Only two case reports published in national journals were found and in both cases the patients were treated conservatively by the pediatrician and neurosurgeons[20,21]. Our results regarding the graft take and early wound coverage are compatible with the international results[1,9].

Conservative treatment has been described and advocated[22]. But some authors have highlighted the disadvantages of this treatment[23]. The operative management of aplasia cutis congenita ranges from simple closure to scalp flaps and free tissue transfer[24].

Local skin flaps are unreliable in this condition, probably due to the abnormal vascularity of the skin adjacent to these lesions in the form of grossly dilated superficial veins[19]. The choice of immediate split thickness skin graft application would achieve an early wound closure. This would reduce the size of the defect as soon as possible[25,26]. It is evident from various reports that in larger scalp defects there is a greater possibility of skull and dural defects[27-30].We observed skull and dural defect in only one patient.

Reports include saline dressing, continuous saline drips, betadine solutions, bacitracin ointment, silver sulphadiazine dressings and nonadherent dressings (MepitilR)[31-35]. Some authors have suggested conservative treatment of very large defects. The advantage of conservative management is to avoid potential operative risk to the infant. The possible complications of conservative management are wound infection, biochemical abnormalities and fatal haemorrhage. Biochemical abnormalities are due to sulphadiazine dressings. This may be secondary to sodium loss from the scalp defect. Silver sulphadiazine dressing may lead to increased potassium absorption and may cause hyperkalemia[9].

It has been observed that small defects measuring less than 2 cm heal spontaneously with proper care whereas larger defects can be treated in several ways and the overall results are excellent[36-38]. However, large defects pose a very special problem for the reconstructive surgeon. The use of scalp flaps in large defects is associated with an increased risk of partial or total flap failure because of related abnormality of the adjacent skin. Although a delayed flap can be a good option but its execution takes long and survival is not certain[39]. Some patients with ACCS also have calvarial defects so these patients need soft tissue coverage with bone graft for correction of calvarial defects[38,39].We had only one patient with a tiny calvarial defect but that was not diagnosed radiologically by CT scan initially.

Numerous clinical reports have documented the occurrence of bleeding or life threatening hemorrhage[22] and meningitis[40] in cases of aplasia cutis congenita with calvarial defects and the overall mortality that has been reported to be 20 and to 60%[14]. In our series it was approximately 10%.

Recently acellular dermal grafts have been used successfully in the reconstruction of cranial fossa defects[41]. The acellular dermal matrix has a therapeutic role in the reonstruction of full thickness calvarial defects of the immature craniofacial skeleton[42,43].

A split thickness skin graft from adjacent healthy scalp or thigh is a valuable approach for scalp wound closure in neonates. We successfully used split thickness skin grafts in our patients with minimal incidence of complications.

Cosmetic defects may occur by spontaneous healing of small lesions or skin grafted large lesions, both leaving scarred area that is devoid of hair. This may be treated later on with advanced techniques such as, tissue expansion to

provide normal hair bearing scalp or autologous micrograft hair transplantation, to provide hair cover up.

CONCLUSION

In our study, split thickness skin graft appeared to be the most favorable treatment for Aplasia cutis congenita and may be considered for all such cases in our set up. A multi disciplinary approach is advised in cases with complications, as was the case in our study.

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