Aplasia Cutis Congenita: A Simple and Safe Management

Yasir S. Jamal¹, FRCS(C), FICS, Moaath Saggaf², Sabah S. Moshref², FRCS(I),
Mazen O. Kurdi¹, FRCS, and Bayan Ghabashi¹, M.B.B.Ch

Department of Surgery, Division of ¹Pediatric Surgery and ²Plastic Surgery, Faculty of Medicine
King Abdulaziz University, Jeddah, Saudi Arabia

Correspondence

Prof. Yasir S. Jamal
P.O. Box 80215, Jeddah 21589, Saudi Arabia
e.M: ysjamal@kau.edu.sa

Submission: 04 June 2015
Accepted: 09 Nov. 2015

Citation

Jamal YS, Saggaf M, Moshref SS, Kurdi MO,
Ghabashi B. Aplasia cutis congenita: a simple
and safe management. JKAU Med Sci 2016; 23
(1): 1-8. DOI: 10.4197/Med. 23.1.1

Abstract

Aplasia cutis congenita is a rare disorder characterized by absence of the skin and skin appendages. The incidence ranges between 1:10,000 and 2.8:10,000. The management in the literature varies considerably between conservative management and surgery. A retrospective review of all the cases of aplasia cutis congenita was conducted from King Abdulaziz University Hospital, Jeddah, Saudi Arabia. The study period was from January 2004 to December 2014. Data were collected using chart review and the pictures from plastic and pediatric surgery teams. Data were analyzed using simple descriptive statistics. A total of eight cases were recruited to the study. Average follow up was two years. Six lesions were located on the scalp and two were located on the lower limbs. Seven cases were managed conservatively and one case required full thickness skin grafting for a bleeding superior sagittal sinus. Only one death was documented, and was related to a syndrome. The management of aplasia cutis congenita is related to the defect size and depth. When surgery is indicated, skin grafting is a simple, safe and highly recommended choice of surgical management for the coverage of the skin defect and to control bleeding from underlying superior sagittal sinus in deep lesions.

Keywords

Aplasia cutis congenita, Bleeding, conservative, Skin grafting

Introduction

Aplasia cutis congenita (ACC) is a rare disorder characterized by absence of the skin and skin appendages. The incidence ranges between 1:10,000 and 2.8:10,000^[1]. The most common site involved is the scalp^[2]. Aplasia cutis congenita is either isolated or syndromic. Generally, there are two lines of management, conservative or surgical. The conservative management consists of frequent dressings. On the other hand, the surgical management ranges from skin grafting to flap reconstruction. The

management in the literature varies considerably between these two lines.

The mortality rate of ACC ranges from 20% to 50%^[3]. One recent report in the literature observed 3 mortalities due to massive bleeding secondary to large scalp lesions^[4]. The relative paucity in the literature, the high mortality rate and the management controversies all represent a challenge in the management. This study aims to present our center's experience with the disease and explore the management options

advocating the use of patch of full thickness skin graft for the coverage of the skin defect and to control bleeding from underlying superior sagittal sinus in deep lesions.

Methodology

A retrospective review of all the cases of ACC was conducted from King Abdulaziz University Hospital (KAUH), Jeddah, Saudi Arabia. King Abdulaziz University Hospital is an academic tertiary care center that is expected to receive ACC cases. The study period was from January 2004 to December 2014. Data were collected using chart review and the pictures from the plastic and pediatric surgery teams. All pediatric patients with ACC treated in KAUH were included with no exclusion criteria. Data were analyzed using simple descriptive statistics. The Approval of the Research Ethics Committee was obtained prior to conduction the study.

The conservative management was defined by applying frequent honey and antibiotic dressings, and the surgical management in this study was only full thickness skin grafting. The donor site was the skin of the inner aspect of upper arm. The wounds were managed according to the surgeon's preference. We followed the proposed classification of ACC for scalp lesions by Silberstein *et al.*^[4] based on the size and depth of the defect.

Results

Between January 2004 and December 2014, a total of eight cases were recruited to the study. We had a strong female predominance with a 7:1 ratio. Average follow up was about 2 years, and the lesions were located on the scalp (6 out of 8) and the lower limbs (2 out of 8). Among the patients with scalp lesions and based on Silberstein *et al.*^[4] classification, 3 cases were classified as Type I, skin defects with no skull bone defect, 2 cases were classified as Type II, skin and bone defects, and one case was classified as Type III with an exposed sagittal

sinus (Table 1, Fig. 1). Seven cases were managed conservatively with frequent dressings and one case required skin grafting for a bleeding superior sagittal sinus. Seven cases had isolated ACC, and only one case was diagnosed with Trisomy 13. Only one unfortunate death was observed, and was related to Trisomy 13. One case showed a strong familial predisposition to ACC among three generations. Apart from the patient presented with a bleeding superior sagittal sinus, none of the patients had acute complications. Table 1 summarizes the characteristics of the patients.

The Type III ACC case was a 3-day-old baby girl who was born in a private hospital and managed initially conservatively with frequent dressings by the mother. However, during the dressing changes, the eschar was accidentally removed and a fountain of blood was observed. The girl was rushed to our hospital (Fig. 2) where she was resuscitated blood transfusion started and pushed to the operating rooms. A fountain of blood was observed intra-operatively secondary to a bleeding superior sagittal sinus, it was not feasible to suture the edges of the bleeding sinus as passing the suturing needle in the sinus wall will cause damage, and more bleeding so pressure dressing was applied but patient bled severely twice in the ward and needed further blood transfusion twice with a total amount of 1,000 ml until we came with decision of sealing the bleeding site with patch of skin graft without any suturing but under pressure dressing. Skin grafting was taken from the inner aspect of the upper arm and covered with an occlusive dressing insuring proper hemostasis at the operating theater. The baby was kept under close observation, and the dressing was first changed at 5th post-operative day. The graft was taken very well with no evidence of bleeding. She was doing well at her follow up visits.

The two of the lower limbs, one involved the dorsum of the right foot (Fig. 3), second one involved both legs and were healed on conservative treatment only.

Table 1. A summary of the characteristics of the patients with aplasia cutis congenita in the study.

Patient No.	Gender	Location	Type	Syndrome	Treatment	Outcome
1	Female	Scalp	I	No	Conservative	Healed well
2	Female	Scalp	I	No	Conservative	Healed well
3	Female	Scalp	I	No	Conservative	Healed well
4	Female	Scalp	II	No	Conservative	Healed well
5	Female	Scalp	III	No	Skin grafting	Healed well
6	Male	Scalp	I	Yes, Trisomy 13	Conservative	Death related to a syndrome
7	Female	Lower limbs		No	Conservative	Healed well
8	Female	Lower limbs			Conservative	Healed well

Silberstein Type 1.

Silberstein Type 2.

Silberstein Type 3.

Figure 1. Patients with ACC 6 on the scalp and 2 on lower limb.

Silberstein Type 3.

Figure 1. (Continuation) Patients with ACC 6 on the scalp and 2 on lower limb.

A. At presentation

B. In OR first examination showing the bleeding superior sagittal sinus.

Figure 2. The Silberstein Type 3 patient.

C. Pressure occlusive dressing.

D. Skin graft sealing the bleeding superior sagittal sinus.

E. 6 months post operatively

Figure 2. (Continuation) The Silberstein Type 3 patient.

Figure 3. Patient ACC of the right foot. Treated conservatively: **A.** At presentation; **B.** At 4 weeks; **C.** At 6 weeks; **D.** At 2 months

Discussion

The management of ACC varies considerably among different reports in the literature. Silberstein *et al.*^[4] reported 22 cases of ACC with 15 (68%) cases managed conservatively and 7 (32%) cases managed with skin grafting. On the other hand, Maillet-Declerck *et al.*^[5] reported 29 cases of ACC with the majority of the patients treated surgically (83%, 25 out of 29 cases). In another report, all the ACC patients, 11 cases, underwent split thickness skin grafting^[6]. Havery *et al.*^[7] also reported 17 cases of ACC in which 13 cases (76%) were managed conservatively without acute complications. In our experience, the majority of the patients (87.5%, 7 out of 8 cases) were treated conservatively, and only one patient required full thickness skin grafting secondary to a bleeding superior sagittal sinus. Thus,

we advocate the conservative management in types I and II with frequent dressings and patch skin grafting under pressure dressing to control the bleeding sinus without suturing

As ACC is a potentially life threatening disease, the reported mortality in Maillet-Declerck *et al.*^[5] study was 14% (4 out of 29 cases). However, in the Silberstein *et al.*^[4] study, 27% (6 out of 22 cases) died during the infancy or secondary to uncontrollable bleeding. In our study, we only had one death due to an associated Trisomy 13. We relate the mortality to the size, location and depth of the defects.

The complications of ACC include bleeding, wound infections, meningitis, wound healing complications and graft failure^[7,8]. In this study, after the management

was taken, no acute complications were observed. Silberstein *et al.*^[4] reported three mortalities due to uncontrolled bleedings^[4]. One of them died at home, similarly our type III ACC patient give history of sister who died at home due to severe bleeding ACC. In this study, we had this patient of type III ACC referred urgently to our hospital with fountain of blood coming from the wound eschar, the patient had two severe bleeding episodes inward in spite of pressure occlusive dressing so the bleeding could be controlled on 2nd operative session after applying patch of skin grafting under pressure dressing undisturbed for five days. Based on our experience in management of eight patients of ACC. We recommend changing the dressing initially by physicians in the hospital settings to prevent such devastating complications. We also advocate the use of patch of skin grafting under pressure dressing in controlling any bleeding from superior sagittal sinus without suturing of the bleeding sinus or the graft to avoid the needle traumatic damage of the sinus wall, adjacent meninges and brain tissue, which might led to further bleeding points.

The possible choices of wound dressings vary among several reports in the literature. Havery *et al.*^[7] reported several choices of wound dressings. The most common choices were Acticoat™ (Smith and Nephew Pty. Ltd, UK) and Intrasite conformable (Smith and Nephew Pty. Ltd, UK). Silberstein *et al.*^[4] reported the usage of local or systemic antibiotics. In our study, honey and local antibiotics agents were the most commonly used. We found that the wounds eventually heal but we observe some breaks in the skin during the healing process and after in three patients. The patients were sometimes placed on systemic prophylactic antibiotics according to the surgeon's preference when sign of infection observed.

As several classification systems of ACC exists, a recent one proposed by Silberstein *et al.*^[4] for scalp lesion addressed the characteristics of the defects and matched them with possible treatment options. Given the fact that ACC is a rare disease yet potentially lethal, we recommend following their classification for scalp lesions to guide the treatment.

Conclusion and Recommendations

In conclusion, the management of ACC is related to the defect size and depth. Initially, the conservative management is advocated for small superficial defects. When surgery is indicated, skin grafting is a simple, safe and highly recommended choice of surgical

management. We strongly recommend applying patch of skin graft under pressure dressing to control bleeding superior sagittal sinus without any attempt of suturing of the bleeding sinus or fixing the graft. Also we recommend changing the dressing initially only by physicians in hospital setting as this is a potentially lifesaving measure to prevent acute bleeding complications.

Conflict of Interest

The authors have no conflict of interest.

Disclosure

None of the authors received any type of commercial support either in forms of compensation or financial for this study. They have no financial interest in any of the products or devices, or drugs mentioned in this article.

Ethical Approval

Obtained.

References

- [1] Taifour Suliman M, Quazi A. Aplasia cutis congenita of the trunk in a Saudi newborn. *Br J Plast Surg* 2004; 57(6): 582–584.
- [2] Demmel U. Clinical aspects of congenital skin defects: I. Congenital skin defects on the head of the newborn. *Eur J Pediatr* 1975; 121(1): 21–50.
- [3] Wexler A, Harris M, Lesavoy M. Conservative treatment of cutis aplasia. *Plast Reconstr Surg* 1990; 86(6): 1066–1071.
- [4] Silberstein E, Pagkalos VA, Landau D, Berezovsky AB, Krieger Y, Shoham Y, Levy A, Rosenberg L, Silberstein T. Aplasia cutis congenita: clinical management and a new classification system. *Plast Reconstr Surg* 2014; 134(5): 766e–774e.
- [5] Maillat-Declerck M, Vinchon M, Guerreschi P, Pasquesoone L, Dhellemmes P, Duquennoy-Martinot V, Pellerin P. Aplasia cutis congenita: review of 29 cases and proposal of a therapeutic strategy. *Eur J Pediatr Surg* 2013; 23(2): 89–93.
- [6] Tahmeedullah, ul Haq N, Bilal M, Laiq N. Aplasia cutis congenita scalp: management options. *J Postgrad Med Inst* 2010; 24(1): 36–40.
- [7] Harvey G, Solanki NS, Anderson PJ, Carney B, Snell BJ. Management of aplasia cutis congenita of the scalp. *J Craniofac Surg* 2012; 23(6): 1662–1664.
- [8] Browning JC. Aplasia cutis congenita: approach to evaluation and management. *Dermatol Ther* 2013; 26(6): 439–444.

عدم تنسج الجلد الخلقي: معالجة سهلة وآمنة

ياسر صالح جمال^١، ومعاذ محمد السقاف^٢، وصباح صالح مشرف^٢، ومازن عمر كردي^١، وبيان غبشي^١
قسم الجراحة،^١شعبة جراحة الأطفال و^٢شعبة الجراحة التجميلية
كلية الطب، جامعة الملك عبدالعزيز
جدة - المملكة العربية السعودية

المستخلص. عدم تنسج الجلد الخلقي هو اضطراب نادر يتسم بعدم تواجد الجلد ولاحقات الجلد، وتتراوح الإصابة بهذا المرض بين ١ إلى ٢,٨ حالة لكل ١٠,٠٠٠ نسمة. تتراوح أساليب معالجة المرض في البحوث الأخرى ما بين المعالجة المتحفظة والجراحة.

وأجريت المراجعة الاستيعادية لجميع حالات عدم تنسج الجلد الخلقي من مستشفى جامعة الملك عبدالعزيز بجدة، المملكة العربية السعودية. استمرت مدة الدراسة من يناير ٢٠٠٤ حتى ديسمبر ٢٠١٤، وتم تجميع بيانات الدراسة من خلال مراجعة السجلات الطبية والصور التي التقطتها فرق جراحة الأطفال والجراحة التجميلية. تم تحليل البيانات عن طريق استعمال الإحصاء الوصفي البسيط.

شارك مجموع ثمانية حالات في الدراسة، وكان متوسط المدة بين المراجعات سنتين. وقعت ستة من الآفات على فروة الرأس، واثنان منها على الأطراف السفلية. تمت معالجة سبع حالات بالطريقة المتحفظة، بينما احتاجت الحالة الثامنة إلى تطعيم الجلد كامل النخانة بسبب نزيف في الجيب السهمي العلوي. لم تحدث سوى حالة وفاة واحدة، وكانت الوفاة متعلقة بإحدى المتلازمات.

تتعلق معالجة عدم تنسج الجلد الخلقي بحجم وعمق العيب. عندما يتم اقتراح الجراحة كحل، يُعتبر تطعيم الجلد هو خيار المعالجة الجراحية الأسهل والأمن والأكثر اقتراحاً من أجل تغطية العيب الجلدي والتحكم بنزيف الجيب السهمي العلوي الدفين في الآفات العميقة.