International Journal of Neurosurgery
2018; 2(2): 35-38
http://www.sciencepublishinggroup.com/j/ijn
doi: 10.11648/j.ijn.20180202.14
ISSN: 2640-1940 (Print); ISSN: 2640-1959 (Online)

A Rare Case of Scalp Aplasia Cutis Congenita in a Zimbabwean Child

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To cite this article:

Brighton Valentine Nyamapfene, Tafadzwa Moreen Dube, Killion Madondoro, Aaron Musara, Kazadi Kaluile Kalangu. A Rare Case of Scalp Aplasia Cutis Congenita in a Zimbabwean Child. *International Journal of Neurosurgery*. Vol. 2, No. 2, 2018, pp. 35-38.
doi: 10.11648/j.ijn.20180202.14

Received: October 23, 2018; **Accepted**: November 8, 2018; **Published**: December 4, 2018

Abstract: Skull and scalp defects can be a challenge in the pediatric population. They predispose to meningitis and at times catastrophic exsanguination. Aplasia Cutis Congenita is a very rare condition resulting in a variable degree of absence of epidermis and dermis. It can be associated with skull defects especially the parietal vertex. We report a case managed at our hospital of a 2 week old Zimbabwean baby with a scalp and bi-parietal bone defect present at birth. He had no other congenital anomalies. Our case was managed by rotational pedicled flap to cover the skull defect. Various methods to close the skull defects have been highlighted in literature. These may include either split skin graft or full thickness graft especially if the pericranium is viable for nutritional support. Subgaleal scoring with flap advancement can also be employed by making longitudinal incisions perpendicular to the direction of advancement. Rotational flaps are especially useful in parieto-occipital lesions like our case. Patient factors for example size of defect, location and associated skull defects determine the optimal option to be used. Aplasia Cutis Congenita is therefore a rare condition which needs a multimodal approach. Lesions above 2 cm in greatest diameter and associated skull defects should have early surgery to avoid complications like infections and hemorrhage.

Keywords: Aplasia Cutis Congenita, Skull Defect, Scalp Reconstruction

1. Introduction

Aplasia cutis congenita is a rare condition which results in the absence of epidermis, dermis and varying degrees of connective tissue including bone of the scalp vertex [1]. It has an incidence between 0,5-1 per 10 000 births [1]. We report a case of aplasia cutis congenita in a Zimbabwean infant. The pathophysiology has not been fully elucidated though the highest risk has been with the anti- thyroid drug, methimazole [2]. Management should be individualised with special consideration of size, presence of skull defect and comorbidities [3]. Conservative methods have excellent results for scalp defects less than 2 cm in diameter but have hypertrophic scars and keloids as their main complications. [4] Larger defects are associated with risk for meningitis, sinus thrombosis and hemorrhage hence early surgery is advised as shown in our report.

2. Case Presentation

A 2 week old neonate presented to our hospital as a referral from the local clinic with a scalp and bone defect present at birth. She had no other complaints and her immunization status was up to date as per our national protocol. The mother was a 33 year old P3 who was of sober habits who had pregnancy induced hypertension managed with Methyldopa and Nifedipine. This was a booked and planned pregnancy without any associated antenatal infections. She had taken iron and folate supplements and no sonography had been done due to financial reasons. This was a normal vertex delivery with Apgar scores 7/10 -8/10 with a birth weight of 2 020 grams and her head circumference was 31 cm which was within the 2 standard deviation.

On examination the patient was small for gestational age

and not in respiratory distress. She had low set ears and short stubby fingers and toes. He had a cranial defect involving the bilateral posterior parietal bone measuring about 8 cm in its greatest diameter as shown in Fig 1. He had exposed meninges as evidenced in Fig 2. He was moving his limbs normally. His respiratory and cardiovascular systems were normal.

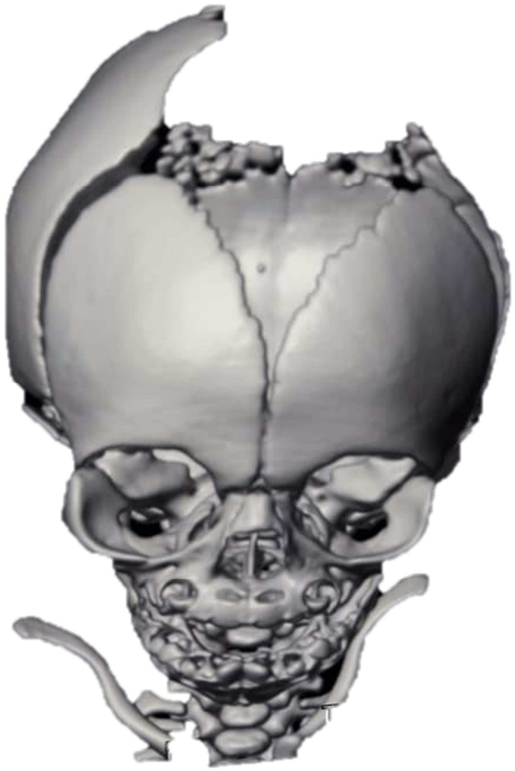

Figure 1. *3D CT reconstruction of Skull defect.*

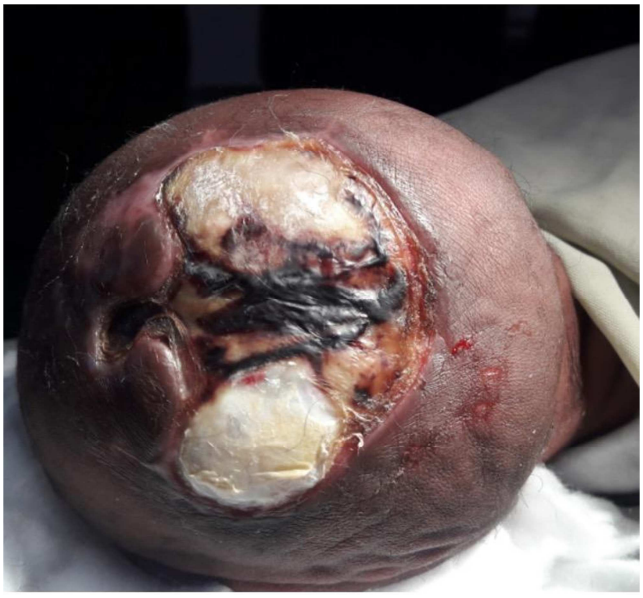

Figure 2. *Parietal scalp defect and intraoperative positioning.*

Patient had a rotational skin flap as shown in figure 3 to cover the scalp defect. He was scheduled for a delayed cranioplasty. Local wound care was achieved through saline dressings and topical antibiotics.

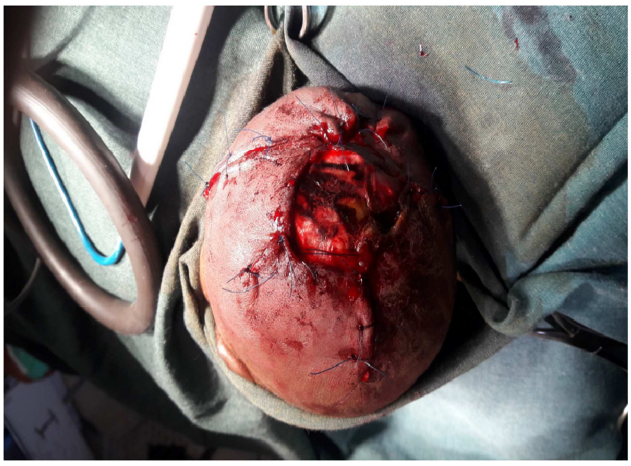

Figure 3. *Rotational skin flap done with reduction in area of defect.*

3. Discussion

Aplasia cutis congenita is a rare condition where *there* is a localized or widespread absence of skin. Most of the cases reported have been outside Africa [5]. To the best of our knowledge, this is the first case report of such a condition in Zimbabwe although the first case in the world was reported by Campbell in 1827 [6]. Localized or widespread cases have been reported with an incidence between 0,5 -1 per 10 000 births. Up to 90 % of Aplasia Cutis Congenita involve the scalp [7]. In 30 % of such cases a concomitant cranial defect is found [7]. This makes our case a unique presentation as it had the dual pathology. Involvement of the torso can also occur [5]

The exact pathophysiology of the condition remains unclear with the possibility of either in utero skin disintegration or total lack of skin development. A positive family history has been noted to be a risk factor as has been published in a Saudi case report by Maglia etal in which a sibling had the same condition [8]. Interestingly consanguinity has been reported to have a risk of 7% relative to the 2 % of non consanguinity [8]. In our case there was absence of known antenatal teratogenic drug ingestion, intrauterine infections which form part of the known risk factors. The high incidence of lesions at the skull vertex may be due to the increased tension in utero around 18 weeks of gestation as there is rapid brain growth. Ultrasound can predict the likelihood of having an aplasia cutis in utero though it is user dependent.[9] There was no measurement of serum alpha fetoprotein in our case although Aplasia cutis congenita is known to be a rare cause of antenatal alpha feto-protein rise [10].

The presentation is of variable spectrum with the bullous and membranous type [11]. The skin epithelium goes through changes hence becomes atrophic and will scar as presented in our case. This is the most common stage of presentations to clinicians. Other cases present with a dark collar hair sign around the atrophic scar indicative of the underlying neural tube defect as in our case. The membranous type is thought

as an incomplete neural tube defect by some authors as the thin membranous covering looks identical to that of encephaloceles and meningoceles both clinically and histologically. Varying degrees of epidermal and dermal loss can be demonstrated on histology with different amounts of connective tissue. Associations with other congenital lesions like hemangioma and nevi has been reported in literature. The presentation is clinical although biopsy can be done to exclude other differentials including brain heterotopia, localized infection, cutaneous meningioma and other dermatosis [4]. Our case was straight forward and no biopsy was deemed necessary.

The management of this rare condition remains largely controversial [12]. Multi- disciplinary approach should be employed for optimal results. [13] Following a thorough obstetric and and family history, genetic counseling to the family members must be sought. It is agreed that lesions up to 4cm X 7 cm can be managed conservatively. The experience of Martinez etal in the Spanish pediatric population highlighted that lesions less than 2 cm in greatest diameter without osseous involvement had good cosmetic outcome with conservative treatment [14]. Application of Silver Sulphadiazine can be done or topical antibiotic ointment for example mupirocin as partly done in our case. Large defects which affect the integrity of the pericrania and skull can present a significant risk for infection, dural sinus thrombosis and even catastrophic exsanguination from a bleeding sinus. With the possibility of all these complications in our case, a surgical option was instituted. Such large defects if untreated surgically can lead to increased mortality estimated to be about 50% in the series of Maillet- Declerck etal [15]. Therefore lesions above 2 cm in greatest diameter with skull defects, are to be treated surgically to abort these complications and longer hospital stay associated with conservative management. Complications of conservative treatment include cicatricial alopecia, hypertrophic scars and keloids.

Flaps have been used with success in Aplasia Cutis Congenita [16]. In our case we used a pedicled rotational flap to repair the defect. Other methods include split skin graft and full thickness graft [17]. Acellular dermis and cultured keratinocyte grafts have been in use [18]. Although bone defects can self generate, some authors advocate for simultaneous skull and scalp repair on the same sitting [19]. Although surgery comes with complications for large defects namely bleeding and infections, the risk is way less than that of non –operative option [20]. Each case must be individualized and managed accordingly [21].

4. Conclusion

Aplasia Cutis Congenita is a rare condition which needs a multimodal approach. Lesions above 2 cm in greatest diameter associated with skull defects should have early surgery to avoid complications like infection and hemorrhage. The surgery includes pedicled flaps with subgaleal scoring, flap advancement and split skin graft. Each case should be therefore individualised with consideration of size, location and nature of defect.

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