Cutis aplasia represents congenital absence of all the skin layers, and it may occasionally extend through the bone and dura of the skull (1).

Aplasia cutis congenita is a rare entity. Just over 500 cases have been reported (2). Clinically, the neonate presents one or more sharply circumscribed surface areas where skin has failed to develop. The defect is usually small and solitary, but extensive defects have been reported, resulting in early death. Approximately 80% of these defects occur on the scalp, especially at the vertex, and the remaining 20% over the lower and upper extremities in that order of frequency (3). The lesions may be ulcerated with a red glistening base, eczematous, bullous, or atrophic at birth. They heal spontaneously from the periphery over the course of 1-3 months to leave a smooth, yellowish, hairless, and papery scar (4).

Cordon in 1767 first described the disease in an extremity and later Campbell in 1826 described it in the scalp. In 1828, Billard reported scalp involvement with an underlying skull defect (1). The etiology of aplasia cutis congenita remains uncertain, but at least one factor in the developmental defect is genetic. Most cases appear sporadically (4). There have been reports of clusters occurring in the same family pedigree, the disease being transmitted in an autosomal dominant or recessive manner or on occasion as an X-linked disease. This indicates an undoubted genetic component to the condition (5). Other proposed etiologies include teratogens, an arrest skin in the skin development in embryonic life,

**SUMMARY**

A case of cutis aplasia with a full thickness defect involving scalp and cranium is presented. After a failed surgical procedure healing was obtained conservative local treatment with the use of silvadene cream.

**Key Words:** Aplasia Cutis Congenita, Surgical Treatment, Conservative Treatment.

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**ÖZET**

Aplasia Cutis Congenita

Makalede skalp ve kraniumda tam kat defekt mevcut olan bir “Aplasia cutis congenita” vakası takdim edilmektedir. Başarısız cerrahi tedaviyi takiben uygulanın konservatif lokal tedavi ve silvadene krem kullanılmıyla iyileşine sağlanmıştır.

**Anahtar Kelimeler:** “Aplasia Cutis Congenita”, Cerrahi Tedavi, Konservatif Tedavi.
an intrauterine vascular abnormality (including placental infarcts and solitary umbilical arteries), intrauterine trauma, early rupture of amniotic membranes or amniotic bands (4). Examination of the placenta and the umbilical cord may aid in the diagnosis (4). It is important to recognise this as a sporadic intrauterine event unrelated to birth trauma or forceps delivery (6). Other associated abnormalities include cleft lip and/or palate, abnormal ears, hydrocephalus, brain malformations, microptalmus, meningocele, congenital heart disease, tracheoesophageal fistula, omphalocoea, anorectal malformation, absent or polycystic kidney, duplication of cervix or uterus, vascular anomalies, nail aplasia or dysplasia, absent, fused or accessory digits, simian creases, epidermolysis bullosa like lesions, Golt’s syndrome (focal dermal hypoplasia), trisomy-13, intestinal lymphangiectasia and pyloric atresia. Association with moya moya disease has been reported (4,7).

The first attempt to analyze the histopathologic features of the lesion was made by Gross et al in an extensive review of the literature and a detailed study of their own cases in 1957. The membrane covering the brain consists of a very thin, flattened layer of cuboidal, nucleated cells set in a regular manner in a single layer. This histological study of the skin bordering these lesions revealed a gradual transition from normal skin to a corium that was deficient in elastic fibers. In the intermediate zone, the dermal appendages are found to be absent, rudimentary or malformed, decreasing in numbers centripetally more or less proportionately with the attenuation of elastic fibers. Centripetally also, the epidermis thins out to one or two rows of flattened epithelial cells. The collagen is compact, with little adipose tissue present. Sweat and sebaceous glands as well as the elastic fibers are absent (8).

Case Report

Our patient is a 2900 gram white boy of 38 weeks’ gestation. Pregnancy, labour and delivery courses were uneventful. No abnormal family history was noted. At birth, he was found to have big bullae over the scalp. A large round skull defect at vertex 6x8 cm was detected. The defect including scalp and skull which was represented only by a thin membranous layer exposing underlying brain and sagittal sinus. The thin membranous layer quickly became covered with a dark dry eschar. The infant was otherwise normal, including pediatric neurological examination.

The wound was dressed with aqueous Betadine solution to avoid the possible complication of infection, maceration and subsequent haemorrhage. The eschar became well demarcated and started to become macerated 10 days after birth (Fig. 1). At the 12 days after birth the eschar was removed. We planned to cover the defect by using two temporo-occipital scalp flap after delaying procedure (Fig. 2a-2b). Immediately after delaying procedure a gradual necrosis appeared at the distal edges of the both flaps. We cancelled second stage of the planned operation and decided to go conservatively. After this failed surgical treatment, necrotic areas and defect area were treated with Silvadene and gauze dressing. With this conservative local treatment the wound epithelialized gradually (Fig. 3a-3b-3c). Although there was a great risk of bleeding from the exposed sagittal sinus, it did not occur. And also we did not notice any sign or symptom indicating a possible menegitis or other local or systemic infections. By 45 days the wound had completely epithelialized and contracted (Fig. 4).

Figure 1: The child at 12 days after birth, with a full thickness scalp and scull defect.
Figure 2a: Preoperative planning of two temporo-occipital scalp flaps (posterior view).

Figure 2b: Preoperative planning of two temporo-occipital scalp flaps (lateral view).

Figure 3a: The child at postoperatively 10th day.

Figure 3b: The child at postoperatively 20th day.

Figure 3c: The child at postoperatively 30th day.

Figure 4: The child at 45 days of following conservative therapy.
Discussion

Since Billard’s description of the scalp and skull defects in 1828, multiple treatment regimens for aplasia cutis congenita of this area have been suggested. The stated aims of all these regimens have been to avoid meningitis, bleeding, and trauma to the unprotected brain in these children. Historical mortality figures for aplasia cutis congenita range from 20 to 55 percent. These deaths were primarily a result of associated congenital defects or bleeding from the sagittal sinus secondary to surgical intervention or drying eschar and separation (1,9). Treatment choices may be either conservative or operative.

The superficial lesions heal spontaneously under conservative antibacterial dressing. Conservative approaches involve regular dressing to keep the defect evenly moist, prevent desiccation and allow spontaneous epithelialisation to occur. The operative treatments include primary closure, skin grafting, local scalp flaps with or without tissue expansion, free flaps and cranial vault reconstruction using bone grafts.(1,10)

Both conservative and operative managements are well accepted. The goal of conservative treatment is to allow granulation and healing by secondary intention whilst avoiding desiccation and eschar formation. Reports include saline dressing, continuous saline drips (8), betadine solution (11), bacitracin ointment (12) and silver sulphadiazine (1) dressing. Some authors have suggested conservative treatment in very large defects considered too large for local scalp flap coverage (10). The advantage of conservative management is the avoidance of potential operative risk to a newborn infant (13).

Conservative management may carry potentially fatal risks of infection and sagittal sinus haemorrhage. Therefore, definitive early wound cover is essential. Operative treatments have ranged from simple closure to scalp flaps as the most common reconstruction. Tissue expansion has been described although this technique may not always be possible. Split rib cranioplasty has been used to reconstruct the cranial vault in aplasia cutis congenita (13,14).

Split thickness skin grafting is simple and effective but will not provide adequate brain protection in the long term. Local flaps in the neonatal stage appear to be less reliable than expected. If employed, they should be delayed and transposed only after viability is confirmed. A skin biopsy of the adjacent skin may be helpful to confirm normal histology (15). Full-thickness flap coverage of the defect is critical and homograft application could allow adequate time for properly planning and delaying the flaps. Fluorescein, used to evaluate flap viability is definitely helpful (8).

Unrepaired bone defects proved to be the main source of problems in achieving long-term primary healing. The absence of dural cover and skull support allowed the growing brain to exert expanding forces on the local flaps and the intervening scar tissue causing recurrent breakdown. The weakness of the bony defect was associated with a progressive conehead deformity (13).

Use of a split rib cranioplasty and free latissimus dorsi muscle flap to reconstruct a complex defect of aplasia cutis congenita had been demonstrated. Microvascular free tissue transfer with split rib cranioplasty should be considered for very large defects of aplasia cutis congenita particularly in severe forms where brain is exposed or as a salvage procedure for reconstruction when conservative treatment or local scalp flaps have failed (13).

The philosophy of conservative management is taking advantage of the ability of rapid regeneration in the newborn (16). Silvadene therapy for cutis aplasia provides an antibacterial dressing that resist eschar drying and separation while allowing for epithelization and bony ingrowth of the skull to occur. Furthermore, this conservative method of treatment eliminates the operative risks that may be encountered by the neonate (1).

We believe that the failure of our scalp flap was due to the abnormality of the adjacent abnormal skin features and inadequate blood supply. Scalp flaps in aplasia cutis congenita...
have also partially or totally failed in other reported earlier cases. Local scalp flaps in the neonatal stages appear to be less reliable than expected. If employed they should be delayed and transposed only after viability is confirmed. We believe that conservative treatment of aplasia cutis congenita have less complication rate and more reliability on neonates. Finally conservative management of aplasia cutis congenita may be considered even in large defects.
REFERENCES