Nonsurgical approach to congenital scalp and skull defects

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The management of congenital scalp and skull defects, as generally advocated, is surgical. The authors report such a case that was treated conservatively. At her 3-year follow-up review, the patient's scalp and skull defects and other associated cutaneous defects were fully reconstituted. Such a nonoperative approach, while rarely reported, emphasizes the natural course that some of these lesions may follow. The literature on aplasia cutis congenita is briefly reviewed.

KEY WORDS congenital scalp defect • congenital skull defect • congenital defects • aplasia cutis congenita

Congenital defects of the scalp and skull are rare anomalies. The literature regarding these lesions usually suggests that the appropriate treatment is surgical. We have followed a patient with massive cutaneous and skull defects for 3 years. Her cranial vault and cutaneous defects have become fully reconstituted without surgical intervention.

Case Report

The mother of the patient had pelvic disproportion, and had undergone previous Caesarean sections. She had been pregnant three times, with one previous live birth and one abortion. She experienced minor first-trimester bleeding with this pregnancy. At 5 minutes after birth, the child's Apgar score was 9. She weighed 2360 gm. Her head circumference was 32 cm. Abnormal physical findings were restricted to the integument and skull. An 11 × 7-cm scalp and skull defect pulsated freely (Fig. 1). The defect was covered by a hairless, grayish, vascular, very thin parchment-like membrane, which extended over the entire vault and covered an opaque vascularized tissue, which was thought to represent dura mater. There was no visible cerebral cortex or cerebrospinal fluid. The sagittal sinus could not be identified.

Other cutaneous defects were noted extending from each axilla to the abdominal wall and over the buttocks and thighs bilaterally (Fig. 2a and c). The skull defect included the entire vertex (Fig. 1). Initially, some consultants expressed the opinion that these defects should be immediately grafted to avoid severe infection or hemorrhage; however, the patient had no ready source of autogenous grafts and seemed to require full-thickness grafts if possible. It was suggested that a more conservative approach might be feasible.

The defects were kept moist with sterile normal-saline and bacitracin dressings. Reverse isolation was employed for 4 weeks. The surface of the scalp defects gradually epithelialized from the margins. Six weeks later, the patient was discharged with almost complete epithelialization of the scalp and trunk lesions.

At the child's 1 year follow-up examination, a soft, mobile dermis tissue with scattered sparse hair was present. The trunk lesions had healed without contracture (Fig. 2b). The remaining pulsatile defect was only 7 × 6 cm, centered over the anterior fontanel. The bone grew from the edges of the skull defect toward the vertex. At 3 years, there was full skin coverage with a normal skull contour (Fig. 3). The child has developed normally.

Discussion

Aplasia cutis congenita of the scalp was first described in 1826, and since then about 500 cases have been reported. The exact incidence is not known, since minor cases are not reported. It is believed to be slightly more common in females and first-born infants. The scalp
Fig. 1. Scalp and skull defect at birth.  a and b: Top view of scalp showing the extent of the defect. The vascular pattern resembled dura, although no sagittal sinus could be seen. A narrow strip of epidermis can be seen bordering the normal scalp.  c and d: Anteroposterior (left) and lateral (right) views showing the extent of the bone defect in the frontoparietal region.

defect is present without other skin lesions in 60% of cases, and a skull defect is associated with a scalp lesion in about 20%. Trunk and limb skin defects are present in 25% of patients with scalp defects.

The scalp defects are most commonly located over the parietal regions and over the vertex between the anterior and posterior fontanels. Postauricular lesions have been described however, 80% are midline. Usually they are 1 to 2 cm in diameter, but size may vary from small punched-out lesions to those 10 cm in diameter. Two or more lesions may be found over the skull. Similar location and size variability occur in the skull defects.

Pathologically, these defects may be covered with a flat layer of epithelium or a fibrous membrane devoid of either elastic fibers or skin appendages. The edges merge into normal skin. With larger defects that include the skull, the dura may be absent, and the overlying membrane may be covering arachnoid directly. No inflammation or necrosis is present in these lesions at birth.

Associated anomalies may involve the central nervous system (holoprosencephaly, hydrocephalus, myelomeningocele, diffuse cortical atrophy, spastic paraplegia), the skeletal system (absence of the orbital roof or greater wing of the sphenoid, limb deformities), and the renal system (absent kidneys, polycystic kidneys). Dermal (focal hypoplasia, epidermolysis bullosa, a simean
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There is no agreement as to the etiology of congenital scalp and skull defects. Many familial cases with an autosomal dominant preponderance and few with recessive trait have been cited as evidence of a genetic etiology. Some chromosomal abnormalities have been detected in cases of congenital scalp defects associated with other anomalies. Etiologies, such as the failure of early midline ectodermal closure of the neural tube, placental infarcts, or intrauterine amniotic adhesions causing integumental tears, have been proposed. The high incidence of an associated monzygotic fetus papyraceus, mainly in cases of body skin defects with or without scalp lesions, has led some to believe that the dead fetus causes vascular thrombosis in the live twin by passage of thromboplastic material.

Certainly, some of the previously mentioned associated anomalies will turn out to be fortuitous. In addition, the origin of many of these associated anomalies as ectodermal or mesodermal is not fully worked out, and the incidence of each, associated with congenital scalp and skull defects, is poorly reported in the literature. Hence, a uniform hypothesis regarding etiology remains elusive.

The nature of the defect may influence the choice of treatment. Fear of infection has led many authors to advocate early excision and primary repair of small defects. Others have allowed the defects to heal spontaneously. Larger lesions with or without skull defects have been treated by various cumbersome procedures, such as full-thickness skin grafts, scalp or pericranial rotation flaps, and temporary silicone elastomers. A cranioplasty was often recommended at 2 to 3 years of age.

The reported surgical mortality of 20% to 30% compounded by the potential risks of meningitis, hemorrhage, sagittal sinus tear or thrombosis, possible cortical damage, and the limited donor sites available led us in this instance to favor a more conservative approach. We allowed the skin defect in this patient to close spontaneously, using frequent changes of saline and antibiotic-soaked dressings. We did not think that systemic antibiotic prophylaxis was necessary. Careful protection against trauma and infection in the hospital initially led to adequate epithelialization and early dermal coverage.

The patient was allowed to go home under the care of her parents who were trained during her initial period of hospitalization and continued the dressing regimen at home. Sparse hair growth appeared in the area of the scalp defect. Since these defects largely included the full skin thickness, essential baldness will remain. New bone formation gradually reduced the skull defect from the margins. By the end of 3 years, only a small bone defect remains in the region of the anterior fontanel, emphasizing the fact that bone re-
FIG. 3. The patient at the age of 3 years.  a and b: Face and profile pictures of the scalp show complete coverage with a full thickness of scalp free of cicatrix. Sparse hair growth is not evident in the illustration. c and d: Skull x-ray films illustrate the fully reconstituted cranial vault.

pair may also occur spontaneously.

This illustration of a conservative approach to congenital scalp and skull defects has emphasized the natural course that these lesions may follow given adequate medical care and protection. Some small lesions may not close spontaneously and will require excision. But some very large ones may correct themselves with appropriate medical management and the passage of time.

References
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