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Case Report

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Aplasia cutis congenita in a newborn

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ABSTRACT

Aplasia cutis congenita (ACC) is a localized congenital absence of skin with a reported incidence of 3 per 10000 live births. Most common location is the scalp. The diagnosis is made clinically. The management of the lesion depends on the size and most of them are managed conservatively but larger lesions need surgical closure. Although aplasia cutis congenita is rare, it is very important for the general pediatrician to recognize this and consider it in the differential diagnosis of skin lesions specially the lesions on the scalp. We present a newborn infant with scalp lesion which was clinically diagnosed as aplasia cutis congenita and was managed conservatively.

Keywords: Aplasia cutis congenita, Newborn

INTRODUCTION

Aplasia cutis congenita (ACC) is localized congenital absence of skin with reported Incidence of 3 cases per 10,000 live births.[1,2] Approximately 85% of the lesions occur on the scalp and about 15% occur on the trunk and extremities.[2,4,6] ACC is usually isolated but may rarely be associated with various syndromes. Males and Females are equally affected. Most cases are sporadic but rarely familial cases with both autosomal dominant and autosomal recessive transmission have been reported.[1-5]

We present a case of a newborn with scalp lesion which was clinically diagnosed as aplasia cutis and will review literature.

CASE REPORT

A female infant born at full term was noted at birth to have a round, ulcerated lesion on the scalp. The lesion was about 1 centimeter in size and is as shown in the Figure 1 and 2. Bone tissue could be palpated at the base of the lesion. No other lesions or anomalies were noted. Rest of the physical examination was within normal limits. The same lesion on day 2 of life is as shown in the Figure 3 and 4.

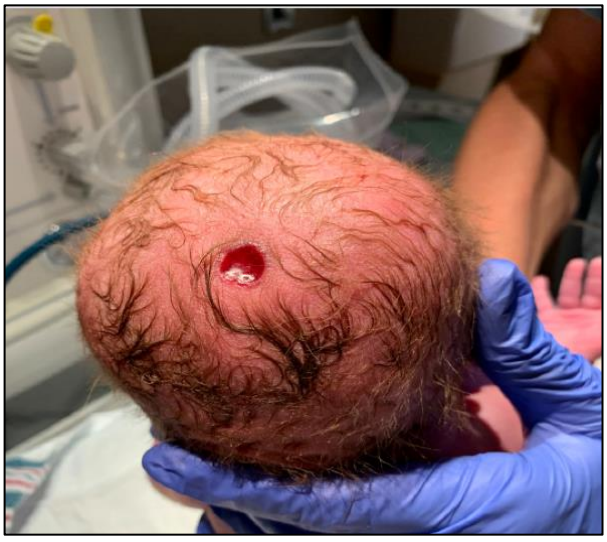

Figure 1: Ulcerated lesion on the scalp.

Pregnancy was uncomplicated with no febrile illnesses, rashes or genital lesions. Mother denies using any medications except for prenatal vitamins.

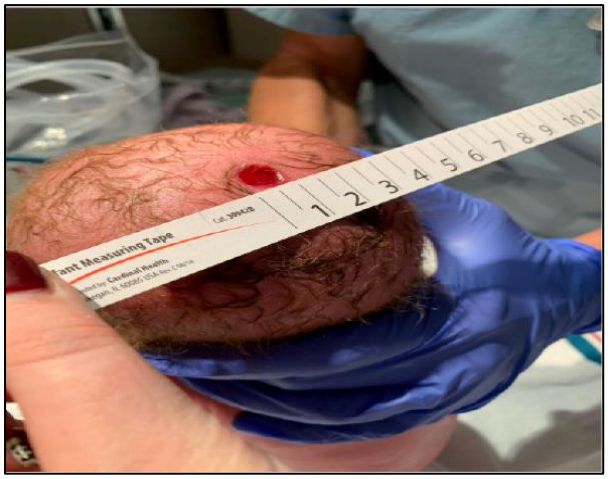

Figure 2: Size of ulcerated lesion on the scalp.

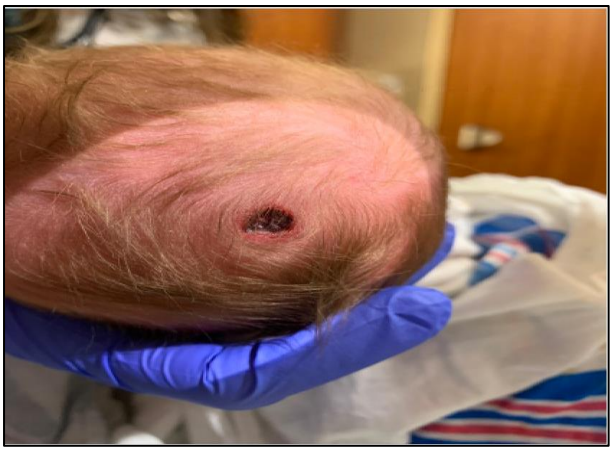

Figure 3: Ulcerated lesion on day 2.

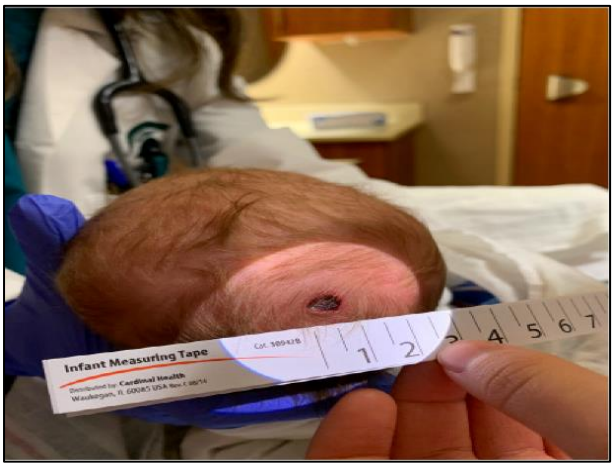

Figure 4: Size of ulcerated lesion on day 2.

No family history of similar skin lesions were reported. Infant was born by spontaneous vaginal delivery and did not require fetal scalp leads or instrumentation during delivery.

Most lesions range from 0.5 to 2 centimeters but some can be larger. Large scalp defects may be associated with underlying abnormalities of the skull, meninges and associated with higher risk of complications. Based on the clinical presentation a diagnosis of aplasia cutis congenita was made.

DISCUSSION

About 85% of the lesions appear on the scalp and are usually solitary.[1,4,6] Lesions on the scalp are in close proximity to the hair whorl (see figure). The remaining 15% of the lesions occur on trunk and extremities. Most lesions range from 0.5 to 2 centimeters but some can be larger.[4] Large scalp defects may be associated with underlying abnormalities of the skull, meninges and are associated with higher risk of complications. Lesions may look ulcerated, eroded or scarred at birth.

The two main types of scalp lesions are membranous and non-membranous ACC.[3,7,8]

Non membranous or irregular ACC have large, irregular, star shaped defects which tend to heal with scar. Non membranous lesions may be common in familial type of ACC. Impaired blood vessel development or vascular insult have been implicated as causes.[3]

Membranous or bullous ACC is round or oval and sharply demarcated It is thought to result from abnormalities in closure of ectodermal fusion lines.[9,10] It can rarely be associated with mild neural tube defects Hair collar sign is a ring of terminal hair surrounding the defect and is specific for defects of neural tube defects.[9,10] Lesions on the trunk and extremities usually are larger than the scalp defects but tend to heal faster.

Differential diagnosis

Membranous ACC on the scalp can be confused with birth trauma, meningocele or encephalocele or focal dermal hypoplasia. Focal dermal hypoplasia may have atrophic or hypoplastic areas of skin but have a unique pattern of skin involvement which follow the lines of Blashko. Neonatal herpes usually presents as multiple vesicles on an erythematous base but sometimes can present as an ulcerated or eroded are of the skin. These can usually be clinically differentiated but if there is any doubt cultures, imaging and biopsies can be helpful

Treatment

Treatment is usually conservative. Local wound care with topical antibiotics and bandages are all that are required. Small lesions measuring less than 4 centimeters even those with associated bone defects typically heal with formation of hairless scar over several weeks. Large lesions may

need excision and surgical closure because of the risk of hemorrhage and infection.[4] Mortality rate for these large defects can range up to 25%.[11]

CONCLUSION

Although Aplasia cutis congenital is rare, it is very important for the general pediatrician to recognize this and consider it in the differential diagnosis of skin lesions specially the lesions on the scalp. Most of the lesions are managed conservatively but a few require surgical closure.

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