Congenital Cutaneous Defects as Complications in Surviving Co-twins

Aplasia Cutis Congenita and Neonatal Volkmann Ischemic Contracture of the Forearm

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Background: During twin pregnancies, several complications may result in the death of a co-twin depending on the date of death. We describe herein 2 infant survivors of monozygotic twin pairs with 2 distinct possible complications: aplasia cutis congenita and Volkmann ischemic contracture.

Observations: One infant had extensive aplasia cutis congenita with an associated monozygotic co-twin who died at 3 months of gestation, and the other child had a localized arm defect due to Volkmann ischemic contracture and brain damage, with a co-twin who died at approximately 6 weeks of gestation.

Conclusions: Congenital cutaneous defects may result in the death of a co-twin. The most common of these defects is aplasia cutis congenita associated with a fetus papyraceus or a dead fetus related to ischemic/thrombotic events in the placenta and fetus. Volkmann ischemic contracture is rare in the newborn but can cause neonatal cutaneous defects. The cause of Volkmann ischemic contracture in newborns is unknown; however, our second observation suggests the possible role of a dead fetus.

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Routine ultrasonographic studies have shown that two thirds of twin pregnancies diagnosed at 10 weeks of gestation result in a singleton by the time of birth.1,2 The death of 1 twin in the first trimester of gestation leads to the resorption of the dead fetus. Early diagnosis using ultrasonography is important to properly interpret this finding. When death occurs during the second trimester, a fetus papyraceus is found at delivery. In the last trimester, the death of 1 twin leads to the mummification of the dead fetus. In most cases, the reason why the fetus dies is unknown.

Several complications may result in the death of a co-twin, depending on the date of death.1-7 Early in gestation, ischemia in a circumscribed area may result in absence of structure, atresia, or hypoplasia. When the co-twin dies later in gestation, clinical features are supposedly related to multiple emboli with disseminated intravascular coagulation. We describe herein a typical case of aplasia cutis congenita (ACC) in a child with a fetus papyraceus and a case of neonatal Volkmann ischemic contracture (VIC) of the forearm in a neonate survivor of a dead co-twin fetus.

Patient 1 was a female neonate born at term (birth weight, 2000 g) after a twin pregnancy. There was no consanguinity in the family and the first child was healthy. Ultrasound follow-up indicated that the second twin died at 3 months of gestation, and findings from an examination of the placenta at delivery confirmed single-chorion placentation with a double amnion and a fetus papyraceus. Results from an examination of the infant’s skin at birth revealed the following features (Figure 1): (1) bilateral absence of skin on the lower limbs from the buttocks to the middle aspect of the legs, and to a lesser extent, on the extensor surfaces of both forearms and the midline of the scalp; (2) reticulated and hypopigmented lesions of the trunk; and (3) perilesional bullae of the forearms and thighs, without other signs of increased skin or mucous membrane fragility. A complete spontaneous coverage of the lesions without contractures occurred within 6 weeks under hydrocolloid dressings (Figure 2).

Findings from an extensive workup for associated malformations were negative. Milia were found at 4 months at the periphery of aplastic lesions. Results from a biopsy specimen taken at 2 months...
to compare normally pigmented and hypomelanotic skin on the trunk showed a complete absence of eccrine glands in the latter and no obvious variations in pigment cell numbers. Findings from subsequent follow-up examinations for 5 years indicated normal growth and development associated with a cosmetically acceptable aspect of the limbs but persistent hypomelanotic skin lesions of the trunk.

Patient 2 was a boy born at term (birth weight, 3550 g) after a twin pregnancy complicated by the death of a co-twin at 6 weeks of gestation determined on ultrasound examination. There was no consanguinity in the family and 5 siblings were healthy. The placenta could not be examined. Results from a physical examination at birth showed a notable unilateral cutaneous defect localized on the left forearm and hand associated with a wrist flexion (Figure 3). Findings from the remainder of the physical examination were normal. A chest x-ray film of the radius showed a double metaphysis line. Healing occurred spontaneously with scarring under hydrocolloid dressings, leaving a persistent underlying muscular defect. Electromyography showed ischemic paralysis of the ulnar and median nerves, leading us to diagnose neonatal VIC and not ACC. During the next 4 months, neurologic development became abnormal. Findings from a physical examination at 7 months revealed a spastic paralysis of the right arm with symmetrically hyperactive myotatic reflexes and bilateral ankle clonus. In addition, the infant developed seizures with deviation of the face and eyes to the right. A computed tomographic scan of the brain showed atrophy of the left hemisphere; the internal capsular area was likely affected because of vascular obliteration of the sylvian artery in early gestation, which was confirmed on cerebral scintigraphy.

**COMMENT**

Aplasia cutis congenita or congenital absence of skin was first recognized by Cordon in 1767. The diagnosis is pri-
marily clinical. The disorder causes neonatal ulcers that may be covered by a thin epithelial membrane at birth and usually heal during the first weeks of life.\textsuperscript{3-15} The histological appearance of ACC varies, but it is typically characterized by the congenital absence of epidermis, dermis, fat, muscle, and sometimes even bone in a well-circumscribed area. Though fairly clinically distinct, this disorder should be distinguished from focal dermal hypoplasia, ankyloblepharon-ectodermal defects—cleft lip and palate syndrome, trisomy 13, disorders of the amniotic band sequence, restrictive dermopathy,\textsuperscript{16} extensive congenital erosions and vesicles healing with reticulate scarring,\textsuperscript{17} and other types of ACC\textsuperscript{10} (associated with epidermolysis bullosa or caused by intrauterine infections).

Frieden\textsuperscript{9} classifies ACC into 9 clinical groups characterized by the location and pattern of the disorder, associated abnormalities, and mode of inheritance. Two types of ACC are reported for singleton gestations: the first involves only a midline scalp defect, and the second is a more extensive form that involves the trunk and extremities and shows symmetrical distribution. Aplasia cutis congenita associated with a fetus papyraceus or placental infarcts has been classified as type V by Frieden. Type V ACC is characterized by sporadic inheritance and can be associated with other abnormalities such as those involving amniotic bands. Numerous causes have been postulated for these defects.\textsuperscript{1,9,10} In 1961, Bernischke\textsuperscript{1} first suggested that some structural defects noted in 1 member of a monozygotic twin pair were caused by an intrauterine vascular accident. Multiple factors are involved in the vascular development of ACC: (1) the existence of placental anastomoses, (2) the date of the death of the co-twin, (3) the maturity of coagulation and fibrinolytic systems, and (4) the proportion of fetal cardiac output through different tissue beds. As a result, various structural defects may develop.\textsuperscript{18}

In monzygotic twin pregnancies, a diamniotic monochorionic placenta is the most common presentation; 85% of diamniotic monochorionic placentas and all monoamniotic monochorionic placentas contain vascular anastomoses in the fetal circulation of the vein-vein, artery-artery, or artery-vein type. The vascular interconnections can also be present in dizygotic twins with conjoined dichorionic placentas. Depending on their nature and shunting effects, complications may affect 1 or both fetuses.\textsuperscript{19,20} The most severe outcome is the death of 1 of the co-twins. Other complications include reverse blood flow with acardiac status in 1 twin during early development, unequal growth due to unequal circulation, vascular disruptions secondary to the death of a co-twin in utero, and structural defects.\textsuperscript{10} These defects are caused by vascular accidents secondary to embolization across placental anastomoses from a deceased co-twin. Clots or necrotic tissue from the dead twin may also enter the circulation of the surviving twin. The proportion of mesodermal and ectodermal tissue supplied by the fetal cardiac output is variable during embryogenesis. During the second month of pregnancy, segmental dorsal arteries irrigate the neural tube and the dorsal thoracic territory. Either mechanism may account for areas of ischemia and disruption with loss of tissue. However, subsequent structural defects vary considerably.\textsuperscript{18}

The maturation of coagulation and fibrinolytic systems may explain at least in part the variability of clinical features according to the time of death of the co-twin. Bleyer and al\textsuperscript{21} note that whole-blood clotting time is infinite before 11 weeks of gestation but becomes comparable to adult clotting times in the ensuing 2 or 3 weeks. The fibrinolytic system follows a parallel development. Thus, as coagulation and fibrinolysis progress, the death of a co-twin poses a higher risk of producing structural defects because of vascular-borne emboli and disseminated intravascular coagulation. Studies of twin-to-twin transfusion syndromes have suggested that during the second trimester, vascularization is essentially related to the iliac and umbilical arteries.\textsuperscript{16} The distribution of ACC over the flanks and upper thighs of our first patient typically indicates fetal death late in the first trimester or early in the second trimester. When ACC is identified at birth, a complete pathologic study of the placenta, membranes, and fetus papyraceus should be performed, as well as a complete physical examination of the affected child. Obstetrical and family history should be obtained to establish a specific diagnosis. Recognizing the vascular origin of ACC type V has practical importance in that the risk of recurrence for similar vascular accidents is negligible. The treatment of ACC depends on the site and extent of the lesions. Spontaneous healing occurs but may lead to hypertrophic or atrophic scarring. Grafting has been reported, but based on our experience, we favor treatment with hydroactive colloidal dressings.

In our second case, the diagnosis of ACC was first proposed. However, muscular and neurologic anomalies associated with radius x-ray film anomalies led us to retrospectively consider the diagnosis of neonatal VIC.\textsuperscript{22} This rare entity is caused by muscular ischemia resulting in increasing pressure within the closed compartmental space of the forearm and myoneural necrosis. Without surgical treatment, muscular fibrosis and contracture appear. There are various causes of VIC: injury from a motor vehicle crash, prolonged external compression, internal bleeding, or fracture. The causal event in a newborn is obscure, since the injury occurs before birth. Some researchers postulate that causes of VIC in neonates are similar to those in adults. External compression of the fetus is possible at various gestational ages, especially after the 37th week, when the volume of amniotic fluid starts to decrease. Another cause might be strangulation by the amniotic bands or umbilical cord. Some children are born with recent injuries that may be confused with neonatal gangrene because of edema and blisters of the forearm, and others present late sequelae with established contractures and skin defects mimicking ACC. Our patient is the first report of VIC in a surviving co-twin, which could be explained by compression by the dead fetus. The newborn presented late sequelae compatible with trauma early in gestation.

Usually neurologic sequelae are associated with late fetal death (late in the second or early in the third trimester), and multiple organ infarction can be found in-
volving the brain, lungs, liver, spleen, and myocardium. Brain infarction can induce porencephaly, hydranencephaly with microcephaly, seizures, and often mental deficiency. Retrospective studies of brain damage in survivors note the death of a co-twin in some cases. In our case, vascular obliteration of the sylvian artery may be due to vascular emboli consecutive to the death of the co-twin or muscular crushing.

In conclusion, both ACC and VIC can cause cutaneous defects in newborns. Aplasia cutis congenita is clearly a complication related to the death of a co-twin, yet the role of a dead fetus in VIC is uncertain. Despite spontaneous healing, the prognosis with VIC is different. While ACC requires only skin dressings, VIC can necessitate early surgery to minimize muscular and nervous sequelae.

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REFERENCES