Fetal Hydrops in a Twin Pregnancy

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Abstract

A monozygotic twin diagnosed in utero with fetal hydrops of unknown etiology was severely dysmorphic and had no heart sounds upon delivery. The presentation and autopsy results led to suspicion of an acardiac twin. The complications present in the surviving twin may have been prevented with earlier diagnosis and increased awareness among healthcare providers.

Keywords: Twin reversed arterial perfusion sequence; Acardiac aneuploidy

Introduction

Hydrops fetalis is a well known condition with an extensive list of differential diagnoses. The etiology is divided into immune-related and nonimmune-related types, and currently almost 90% of cases are associated with the latter category [1]. Among the various nonimmune-related causes, one of the most common are cardiac aberrations [2]. Prenatal ultrasound serves as the initial diagnostic tool for evaluation of nonimmune hydrops fetalis [3]. We report a case of prenatally diagnosed fetal hydrops in a stillborn twin, the suspected cardiac etiology, and the effects on the surviving twin.

Case Report

An 18 year old gravida 2 para 2 female gave birth to monochorionic, diamniotic twins at 30 weeks and 4 days gestation. Due to Twin A’s growth failure and poor bio-physical profile, delivery was induced. Twin A was born via spontaneous vaginal delivery and Twin B was subsequently delivered by cesarean section after failure to progress during attempted vaginal delivery. Prenatal ultrasound had led to the diagnosis of fetal hydrops of uncertain etiology in Twin B (Figure 1). The pregnancy had been followed closely by a perinatologist as Twin B had begun to exhibit increasing hydrops since 8 weeks of gestation. Neonatology was emergently called to the delivery due to the undetermined etiology of the severe hydrops and the potential outcome.

Figure 1: Perinatal ultrasound of Twin B’s cranium demonstrating enlarged ventricles (vertical arrow). There is also marked edema (horizontal arrows) surrounding the fetus. Twin A does not show these abnormalities.
Discussion

Twin reversed arterial perfusion (TRAP) sequence, also referred to as acardiac anomaly, is a rare congenital abnormality that has been detected in 1% of monochorionic twin pregnancies [4,5]. This unique complication involves the presence of an “acardiac twin” with an absent or nonfunctional heart who is perfused through placental arterial anastomoses by its “pump twin” [5]. The acardiac twin is nonviable, and the perinatal mortality rate for the pump twin is typically over fifty percent if no treatment is provided [5]. Acardius ances is a rare subcategory of this anomaly and refers to an acardiac twin with partial development of cranial features [6].

While the cases of acardius ances are not as frequent as other varieties of this already rare malformation, this subtype has been identified as a possible prognostic indicator of unfavorable obstetrical outcome [7]. In addition, if the weight ratio of the acardiac twin is greater than 70% relative to the weight of the pump twin, the prognosis of the surviving twin is less favorable [8]. In our case, the weight ratio of 122% is consistent with the newborn’s preterm delivery and the consequences of her increased cardiac output to provide blood flow to such a large recipient twin.

Studies of experimental animals have demonstrated that oxygen deficiency during the early stages of embryogenesis can alter the normal development of the head, brain, and heart [9]. With this principle in mind, it is apparent that disruption in oxygen supply due to TRAP sequence may provoke the changes to these organs in acardius ances [9]. It has been proposed that an occlusion of the abdominal and thoracic vessels is the cause of thrombosis in the umbilical vessels of acardiac twins, leading to limited or no organ development of upper abdominal and thoracic organs in the nonviable twin [10].

Early and accurate antenatal diagnosis by ultrasound is very important for survival of the pump twin [11]. Management options to improve the survival of the pump twin by occluding blood flow to the acardiac twin have been successfully investigated, however, a definitive approach to decrease mortality has not yet been identified [12]. This case serves as a reminder that in twin pregnancies with fetal hydrops, an acardiac twin should be considered and appropriate post-mortem studies should be requested when the index of suspicion is high. Appropriate early evaluation, efficient documentation and collaboration among perinatologists and neonatologists are imperative to provide successful management and improved prognosis for the surviving twin.

References

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