An Unusual Complication of Twin to Twin Transfusion Intrauterine Gangrene of the Lower Extremities

İkizden İkize Transfüzyonun Sıradışı Bir Komplikasyonu Alt Ekstremitelerin İntrauterin Gangreni

Alt Ekstremitelerin İntrauterin Gangreni / Intrauterine Gangrene of the Lower Extremities

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Özet

Yenidoğanın ekstremite gangreni doğumda son derece nadir bir durumdur. Biz okuyucular ile ikizden ikize transfüzyonun beklenmedik bir komplikasyonunu, intrauterin polisitemi ve hiperviskosite sonucu oluşabilecek alt ekstremitelerin gangrenini, paylaşmak istedik.

Anahtar Kelimeler

İntrauterin Transfüzyon; Gangren; Amputasyon

Abstract

Gangrene of the extremities in the newborn is extremely rare condition at birth. We would like to share with the readers an unexpected complication of twin to twin transfusion, gangrene of the lower extremities which may occur as a result of inutero polycytemia and hyperviscosity.

Keywords

Intrauterine Transfusion; Gangrene; Amputation

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Introduction

Gangrene of the extremities in the newborn is extremely rare condition at birth. Less than 100 cases have been reported world-wide. Its etiology and pathogenesis are not clear in many cases. The pathogenesis of intrauterine gangrene can be divided into intrauterine compression or thrombo-embolic phenomena.[1] The compression is generally caused by uterine anomalies, fetal malpresentation with limb prolapse, oligohydramnios, amniotic bands, or umbilical cord entanglement.[1] Intrauterine fetal ischemia caused by thrombosis or emboli has been linked to maternal diabetes, preterm delivery, dehydration, polycythaemia, and twin-to-twin transfusion syndrome.[2] Only 13 of the reported cases of neonatal gangrene have described established areas of necrosis of an extremity noted upon delivery, so-called intrauterine or congenital ischemic necrosis.[3-4] Of these, only 4 cases involve the lower extremity.[2,4] We report a newborn patient with congenital bilateral in utero lower limb gangrene due to twin- twin transfusion.

Case Report

A 42-year-old healthy woman (gravida 9, para 9) was seen at doctors office with complaint of vaginal bleeding at the 27th week of pregnancy. Emergency c-section was performed due to low biophysic score. Twin A was a baby male with a birth weight of 1320 g. The Apgar scores were 7 at 1st minute and 7 at 5th minutes. Twin B was born with 615 gm did not respond the resuscitation efforts and died because of severe respiratory failure. The twin A was intubated at birth and treated for respiratory distress syndrome. At birth, bilateral lower extremity demonstrated marked atrophy and dense blackened necrosis from the superior pole of the patella to the tips of all 5 toes (Fig A). There was no evidence of a congenital amniotic band or evidence suggesting that the extremity had been encircled by the umbilical cord. Higher laboratory results policytemia and hyper viscosity syndrome indicated (Hb: 25, Hct: 76%, WBC: 13.200, PLT: 342.000) an immediate partial exchange transfusion has been performed.

The patient was referred our center for the amputation after



Fig; Clinical photograph of the lower extremity of the patient at birth (A) and discharged (B).

the exchange where we evaluated unremarkable hypercoagulable state evidenced by normal bilateral lower extremity radiography. The infant was hemodynamically stabilized and prophylactically treated with broad spectrum antibiotics. Bilateral knee disarticulation was performed 12 hours after birth due evolving sepsis and a clearly demarcated limb, with no complications. The distal femoral segment was retained, although the cartilage at the distal femoral condyles appeared grayish with questionable viability.

He was hospitalized for 72 days and discharged from the hospital with 2390 gram. The patient was seen after 90 days with normal physical examination and amputated region was with no complication (Fig B).

Discussion

Twin-twin transfusion syndrome presents at 18 to 26 weeks' gestation with marked hydramnios in one sac and a stuck twin with severe oligohydramnios in the other. The natural history is for premature labor to occur, often in association with rupture of the membranes, with almost certain neonatal death from prematurity combined with congestive heart failure or extreme intrauterine growth retardation.

The biochemical balance of the hemostatic system of the neonate is profoundly different from that of an adult. The composition of the hemostatic system of an infant does not resemble that of an adult until approximately 6 months of age.[5] The functional immaturity of the neonate's hemostatic system favors a procoagulant state. Concentrations of antithrombin III, heparin cofactor, protein C, protein S, and plasminogen are reduced by more than 50% of reference adult values. [6-7]

Twin-twin transfusion syndrome results in a relative hypervolemia in the recipient twin, triggering a diuresis response that leads to a polyhydramniotic sac, increased intrauterine pressure, and decreased fetal motion.[4] Polycythemia and the resultant hyperviscosity in the recipient twin are known predisposing risk factors for the development of arterial thrombosis. [2,4]

The diagnosis of twin-twin transfusion was confirmed by severe polycytemia and hypervolemia in laboratory work up, heavy growth retardation determined by weighing and immediate resuscitation need soon after delivery.

A literature review of reported cases suggests that a significant number of the neonates with intrauterine gangrene were born prematurely. It is impossible to determine whether their premature births were a result of the toxic effects of the gangrenous process involving the extremity in utero or whether other factors were involved. We would like to share with the readers an unexpected complication of twin to twin transfusion, gangrene of the lower extremities which may occur as a result of inutero polycytemia and hyperviscosity.

Competing interests

The authors declare that they have no competing interests.

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