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Neonatal purpura fulminans manifestation in early-onset group B Streptococcal infection

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Patient: Male, 0
Final Diagnosis: Purpura fulminans
Symptoms: Fever • letargy
Medication: —
Clinical Procedure: —
Specialty: Pediatrics and Neonatology

Objective: Rare disease

Background: Neonatal purpura fulminans (PF) is a rare but frequently fatal disorder associated with high morbidity and mortality. It may be congenital, as a result of protein C and S deficiency, or acquired due to severe infection. Gram-negative organisms and Staphylococcus species are the most common causes of the acute infectious type, and a few cases of causative neonatal group B Streptococcus (GBS) disease have been reported worldwide.

Case Report: We present a full-term male neonate with purpura fulminans secondary to early-onset group B streptococcal (GBS) infection. The mother brought the infant to the emergency department at the age of 43 hours of life, with fever (39.5°C) and lethargy. Neonatal sepsis was suspected, and he was immediately started on intravenous ampicillin and gentamicin. The initial workup revealed disseminated intravascular coagulopathy, and both blood and CSF cultures grew GBS. He had normal levels of protein C and protein S for his age. The infant died 48 hours after admission due to multiorgan system failure despite aggressive neonatal intensive care support.

Conclusions: Neonatal PF secondary to early-onset GBS infection is a fatal condition that should not be missed. Screening pregnant women for GBS colonization and use of protocols for preventing perinatal GBS infection is considered the most important preventive measure of this fatal condition, especially among Saudi women, who have a relatively high rate of GBS infection.

Key words: neonatal • purpura fulminans • GBS • early onset sepsis

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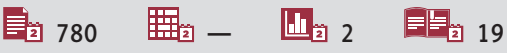

Background

Neonatal purpura fulminans is a rare, life-threatening condition of dermal microvascular thrombosis associated with disseminated intravascular coagulation (DIC) and perivascular hemorrhage that occurs in the newborn period [1]. Gram-negative organisms and Staphylococcus species are the most common causes of the acute infectious type [2], and a few cases of causative neonatal group B Streptococcus (GBS) disease have been reported worldwide [3–6].

Case Report

A full-term male infant was delivered vaginally with Apgar scores of 8 and 9 at 1 and 5 minutes, respectively. He weighed 3100 g and was discharged at the age of 36 hours of life in good condition. The mother presented to the emergency department (ED) in active labor with an unknown GBS status antenatally without prophylactic antibiotics. There was no family history of hematological disorders. The mother brought the infant to the ED at the age of 43 hours of life with fever (39.5°C) and lethargy. Neonatal sepsis was suspected, and he was immediately started on intravenous ampicillin and gentamicin. His condition deteriorated rapidly, aggressive resuscitation was needed, and he was moved to the NICU. A few hours later, a purpuric rash developed over the scrotum and upper and lower extremities, with gangrenous fingers and toes (Figures 1 and 2). The initial workup revealed disseminated intravascular coagulopathy, and both blood and CSF cultures grew GBS. He had normal levels of protein C and protein S for his age. Despite maximum intensive care support and appropriate antibiotic administration, he developed multisystem organ failure and died 48 hours after admission.

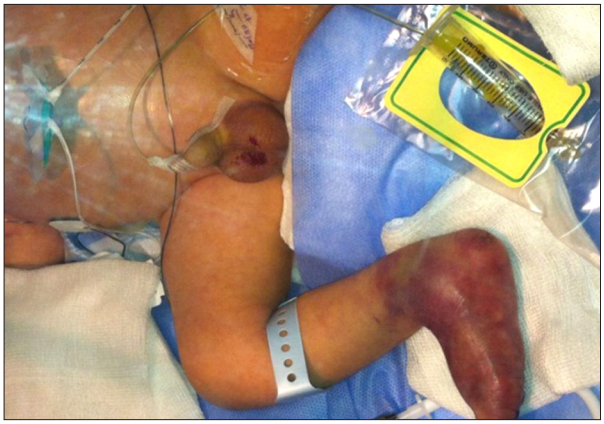

Figure 1. Cutaneous necrosis involving scrotum and foot.

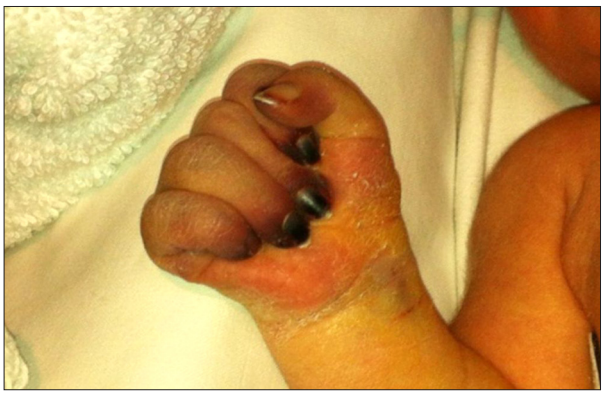

Figure 2. Clearly demarcated cutaneous necrosis involving tip of the fingers and nails.

Discussion

GBS disease continues to be a leading cause of serious neonatal infection despite recent advances in prenatal care [7]. Maternal GBS colonization is one of the most important risk factors for early-onset GBS infection [8]. The transmission rate from colonized mothers to infants is approximately 50% [9]; however, only 1–2% of these infants will develop early-onset GBS disease [10]. Several studies have reported an incidence of GBS colonization in pregnant women of approximately 10% to 30% [11,12]. In the Kingdom of Saudi Arabia (KSA), few data are available in the literature regarding GBS carriage, but most of the available studies show that the incidence of GBS colonization among Saudi women is relatively high [13,14]. El-Kersh et al. reported that 27.6% of Saudi women during the 3rd trimester of pregnancy were GBS carriers [13]. Zamzami et al reported that the overall intrapartum maternal GBS colonization rate was 31.6% at a mean gestational age of 39.2±2.5 weeks [14].

The Centers for Disease Control and Prevention (CDC) recommends that all pregnant women be screened at 35–37 weeks for anogenital GBS colonization and that intrapartum chemoprophylaxis should be offered to all pregnant women identified as GBS carriers [15]. After the universal implementation of the recommended CDC guidelines, the incidence of early-onset GBS disease has declined significantly [16]. Early-onset GBS disease commonly manifests as septicemia, pneumonia, or meningitis occurring within the first 6 days of life [17]. Purpura fulminans is a hematological emergency in which there is skin necrosis and disseminated intravascular coagulation. This may progress rapidly to multi-organ failure caused by thrombotic occlusion of small and medium-sized blood vessels [18]. There are both inherited and acquired causes of neonatal purpura fulminans. Inherited causes are due to a homozygous protein C or S deficiency, and acquired causes are more common and often associated with severe infection causing a consumptive coagulopathy and a relative deficiency of protein C [1]. PF is associated with a more than 50% mortality rate in children and is usually associated with

major long-term morbidity in those who survive [19]. The association between PF and early-onset GBS infection was first reported by Lynn et al. in 1999 [3] in 3 neonates who survived but had markedly compromised neurologic outcomes. In 2009, Hon et al. [4] presented the case of a newborn who died at the age of 9 days with multiorgan system failure despite full intensive care support. In 2010, Zenciroglu et al. [5] reported a 2-day-old neonate with extensive PF due GBS septicemia, who survived but with amputation, cortical blindness, and poor neurologic outcome. Our newborn died at the age of 4 days despite appropriate antibiotic administration and aggressive intensive care support. We are reporting this case to emphasize the importance of implementing the recommended international guidelines for the prevention of perinatal GBS disease [15], especially among Saudi women, who have a relatively high GBS colonization rate [13,14]. Coordination among numerous subspecialties is essential for establishing GBS disease prevention programs.

Conclusions

Neonatal PF secondary to early-onset GBS infection is a fatal condition that should not be missed. Screening pregnant women for GBS colonization and implementing the recommended guidelines are the most important preventative measures.

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