Purpura fulminans (PF) in the neonate is a rare but potentially disabling illness of acute onset associated with high mortality and long-term morbidity. Its skin manifestations are due to hemorrhagic infarction and gangrenous necrosis caused by intravascular thrombosis. It is most commonly associated with bacterial infections or deficiency of anticoagulants such as protein C and protein S. Neonatal PF is a rare but potentially disabling disorder associated with a high mortality and severe long-term morbidity in those who survive.

Case description: We report a case of a premature infant who developed extensive PF due to late onset group B streptococcus sepsis. Despite early identification and initiation of antibiotic therapy in our patient, PF progressed rapidly, leading to autoamputation of fingers and toes and severe brain injury.

Conclusion: In conclusion, our case highlights the severe sequelae of PF due to late onset GBS sepsis in a premature infant.
and gentamicin 3.5 mg/kg/dose every 24 hours), fluids and inotropic support (dopamine, dobutamine, and epinephrine). CBC revealed total white blood cell count of 2,700/mm³ (5% segments and 12% bands), hemoglobin of 12.9 g/dL, and platelets of 177,000/mm³. Within next 24 hours, the infant developed purplish discoloration of the left upper and lower extremities, which rapidly progressed to gangrenous skin lesions suggestive of PF. Coagulation studies were normal with prothrombin time of 19.2 seconds, partial thromboplastin time of 31 seconds, and fibrinogen of 767 mg/dL. Gram stain of CSF showed numerous gram-positive cocci in chains. Blood, CSF, and urine cultures were reported positive for GBS at 11, 32, and 33 hours of incubation, respectively. The isolate was further identified as serotype Ia (performed at the laboratories of the Centers for Disease Control and Prevention, Atlanta).

After CSF culture results, IV vancomycin was discontinued and IV crystalline penicillin G, 100,000 units/kg/dose every 6 hours, was started and continued for 3 weeks. IV gentamicin was continued for 7 days. Repeat blood and CSF cultures were negative. Over the next 2 weeks, necrosis continued to worsen leading to autoamputation of all fingers of the left hand (► Fig. 1, left lower panel) and all toes of the left leg. On day 29, repeat head ultrasound study showed grade III hemorrhage in the left ventricle. On day 30, the infant developed seizures involving both upper arms, which was confirmed by electroencephalogram. Seizures were managed with midazolam and phenobarbital. Repeat coagulation studies were normal at this time. On day 58, magnetic resonance imaging of brain showed cystic encephalomalacia involving bilateral frontal, parietal, and occipital lobes. Cystic periventricular leukomalacia was also noted around both lateral ventricles (► Fig. 1, upper panel).

The infant was discharged home at 4 months; he subsequently required ventriculoperitoneal shunt for hydrocephalus at 7 months. He had severe developmental delay and died at 9 months because of cardiopulmonary arrest of unknown etiology. The other twin survived and had no evidence of GBS infection. However, he was given a 10-day course of IV penicillin G as a preventive measure.

Discussion

We present a case of premature infant who developed severe PF secondary to late onset GBS sepsis and meningitis leading to autoamputation of digits and severe neurologic injury. Only a few cases of infectious PF in neonates have been reported thus far. Lynn et al., Hon et al., Zenigorlu et al., and Albarrak and Al-Matary have described PF in newborns due to early onset GBS, whereas Issacman et al. have reported PF due to late onset GBS. However, these were cases of full-term infants whereas our case was an extremely premature infant. Stewart et al. reported a preterm neonate with PF presumed to be of infectious origin, but cultures were negative and protein C and protein S were normal. Church et al. reported a late preterm infant with PF secondary to Escherichia coli septicemia.

PF is classified as congenital, acquired, and idiopathic. Congenital causes are protein C and protein S deficiency that can result in thrombosis and PF within 72 hours of birth. Acquired causes are infections, disseminated intravascular coagulation, acute venous thrombosis, warfarin, galactosemia, and congenital heart disease. Idiopathic PF is rare.
and usually reported in older children. The leading cause of acquired PF in neonates is infection, among which GBS sepsis is the most common pathogen, although Staphylococcus aureus, Neisseria meningitidis, Escherichia coli, Enterobacter species, and Pseudomonas species also have been described.

The GBS strain from our patient was identified as serotype Ia, which is one of the most common serotypes in early and late onset neonatal sepsis in the United States.9,10 Martins et al11 found that serotypes Ia and III were the most common isolates for invasive infections in newborns. However, there is no preponderance of a specific serotype causing PF when compared to the overall serotype distribution of invasive GBS isolates. Consistent with findings of other studies, intrapartum GBS chemoprophylaxis did not prevent late onset GBS infection in our patient. Protein C and protein S were not known in our patient, but there was no family history of bleeding or coagulation disorders and the other twin did not have any manifestations. Also, PF occurred much later in life in our index case, which may suggest against congenital protein C and protein S deficiency.

Management of infectious PF is focused on maintaining tissue perfusion and antibiotics, which may be beneficial only to some extent. Although antibiotics were initiated early in our patient, the disease progressed rapidly leading to devastating damage. Issacman et al5 used heparin to treat PF with some improvement. We could not use heparin in our index case because of grade III intraventricular hemorrhage (IVH).

In summary, our case highlights the rare presentation of severe sequelae due to PF secondary to GBS infection in a premature infant.

Conflict of Interest
The authors declare no conflict of interest.

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