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CASE REPORT

Purpura Fulminans in Neonate: A Case Report

Nishat Jahan[1], Liton Chandra Saha[2], Farhana Bhuiyan[3], Samira Islam Nisha[3]

Introduction

Neonatal Purpura Fulminans (PF) is a clinical condition due to dermal microvascular thrombosis. PF is a rare and often fatal disorder associated with perivascular hemorrhage and disseminated intravascular coagulationNeonatal purpura fulminans was first described in 1962[1].Homozygous type protein C or protein S deficiencies are rare genetic disorder with fatal PF like thrombotic complications.[1] Hereditary (congenital) protein C deficiency is rare, leads to a hypercoagulability state that usually presents at birth with PF and/or severe venous and arterial thrombosis. PF due to congenital protein C deficiency usually presents with a rapid onset of cutaneous purpuric lesions that start few hours to 5days after birth.[2] Acquired protein C deficiency can occur in several other diseases. Sepsis and severe infections especially gram-negative organisms and Staphylococcus species are the most common causes of the acquired type.[2] It is rapidly progressive, is often accompanied by DICand circulatorycollapse.Treatment includes largely supportive management with IVantibiotics to prevent secondary infection, platelet and FFP transfusions to manage DIC.[3] Here we describe a rare congenital protein C deficiency condition presented with a rapid onset of cutaneous purpuric lesions after birth.

Case Report

Ayman a 10 days old term male neonate weight 3100 gms admitted due to jaundice since 3rd day of life, with this complaints baby was treated locally but as the condition did not improve the parents admitted him at Bangladesh Shishu Hospital and institute for better management. patient was admitted with a bilirubin report (Total 21 mg/dl, Indirect 19.5 mg/dl). On examination after admission,he was less active, pink at room air, vitals were normal. Investigation reveals septicemia, blood C/S was sent. Baby was on breast feeding, antibiotics was started. Mother was 23 years old lady was on regular antenatal checkup, no history of taking any offending drug. One day after admission baby developed blackening of tip of fingers of right hand, toes of both feet which were gradually deepening and extend proximally. There was no other bleeding from any site of body, skin over lesion was intact (Fig.-1). Patient was treated locally with heparin gel.

Laboratory values were: Hct = 24.6% (normal 37-47%); WBC count = 17.9 × 10^9/L; platelet count = 232 × 10^9/L; PT = 25 sec (11-15 sec); APTT 58.866 s (25-35 sec); D-dimer >10.13 μg/ml (normal 0-0.5 μg/ml); Factor VIII 351%; Factor IX 128%, protein C 15 %, very low (normal 70-140%); Protein S 67% (normal 70-140%). Other laboratory tests including serum electrolytes, renal functions, liver functions, urine and CSF analysis were in normal ranges. Blood culture revealed growth of klebsiella pneumoniae, urine and CSF cultures did not show any growth.Colordoppler showed normal findings.

Regarding the family history, clinical, laboratory findings, low protein C and growth of gm negative organism, we established a diagnosis of PF due protein C deficiency due to infectious cause. The baby was admitted in NICU and treated antibiotics prophylactically, I/V heparin initially then enoxaparin and FFP. After several days of therapy, the necrotic lesions healed up dramatically (Fig.-2).

1. Assistant Professor, Department of Neonatal Medicice, Bangladesh Shishu Hospital & Institute.
2. Associate Professor, Department of Neonatal Medicice, Bangladesh Shishu Hospital & Institute.
3. MD Phase B Neonatology Resident, Bangladesh Shishu Hospital & Institute.
Correspondence to: Dr. Nishat Jahan, Assistant Professor, Department of Neonatal Medicice, Bangladesh Shishu Hospital & Institute. Cell: 01711977095.
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Fig.-1 *Clinical manifestation*

Fig.-2 *After treatment*

Discussion

Relationship of PF and protein C deficiency was first described in 1983, and was effectively treated with protein C replacement therapy.[1] It presents as a rapid-onset progression of purpuric skin lesions and DIC later on. The spectrum of severity can vary from symptom onset within 12 h after birth, which is the usual scenario, but, there are reports of delayed presentation around 6-12 months.[4] Literature revealed early onset of symptom due to congenital deficiency of Protein C.[2,3] In our case symptom appeared on 13th day of life and patient had culture positive septicemia. Acute infectious purpura fulminans is the most common type and is associated with an acquired deficiency of protein C. The mechanism involves a disruption of the coagulation balance.Bacterial endotoxin triggers consumption of proteins C and S and antithrombin III. This pro-coagulative state leads to thromboses of dermal vessels and is associated with disseminated intravascular coagulation.[5] Protein C and protein S are dependent on Vitamin K and are synthesized in the liver.[4] The diagnosis of homozygous protein C or S deficiency is based on the clinical findings of purpura fulminans, undetectable levels of protein C or protein S, a heterozygous state in the parents, and, if possible, identification of the molecular defect. There may be no family history of thrombosis.[6] Purpura Fulminans is a haematological emergency so early recognition and initiation of immediate treatment to prevent the complications is mandatory. Immediate treatment is the transfusion of FFP to replace procoagulants and anticoagulants that have been consumed. The main concern of transfusing FFP is to replace Protein C and Protein S which are severely deficient.Protein C concentrate is recommended in severe heritable deficiency of Protein C after confirming the diagnosis to minimize the use of FFP . Neonates and children with severe protein C deficiency are at risk of recurrent PF and hence require long-term treatment with antithrombotic drugs are recommended. Replacement of protein C either alone, or in combination with coumarins or low molecular weight heparin is main stay of management.

Conclusion:

Neonatal Purpura Fulminans is a life threatening condition although it is very rare but early diagnosis

and prompt management is crucial to prevent the complications and improve morbidity as well as mortality.

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