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Case Report

Neonatal purpura fulminans: a rare disease with a sinister outcome

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ABSTRACT

Neonatal purpura fulminans is a rare but frequently fatal disorder associated with high morbidity and mortality. Purpura fulminans describes a clinico-pathological entity of dermal microvascular thrombosis associated with disseminated intravascular coagulation (DIC) and perivascular hemorrhage occurring in the newborn period. It is usually congenital as a result of deficiency of protein C and S. it maybe be either idiopathic or acquired due to severe infection by gram negative organisms or *Staphylococcus* species. Here we present the case of a neonate born to an HIV reactive mother who presented to us at 6th day of life with desquamation of skin in periumbilical area which gradually progressed to involve the limbs, trunk as well as face. The neonate was managed with supportive therapy in ICU but despite intensive management, she expired at 10 days of life due to septicaemia and multi-organ failure.

Keywords: Protein C, Staphylococcus, Gangrene, Purpura fulminans, Discoloration, Endotoxin

INTRODUCTION

Protein C is one of the major inhibitors of coagulation system, which has important influence on physiological function of haemostasis to ensure patency of micro-circulation.[1] Activated protein-C specifically inhibits factor (F) Va and F VIIIa which in turn down regulates thrombin generation. Protein C deficiency leads to macro-and micro vascular thrombosis. Gram-negative organisms and *Staphylococcus* species are most common causes of acute infectious type and few cases of causative neonatal group B *Streptococcus* disease have reported worldwide.[2] Purpura fulminans is acute purpuric rash characterized by coagulation of micro-vasculature, which leads to purpuric lesions and skin necrosis. It is rapidly progressive, is often accompanied by DIC and circulatory collapse. Treatment includes largely supportive management with IV antibiotics to prevent secondary infection, platelet and FFP transfusions to manage DIC.

CASE REPORT

An outborn female child with birth weight of 3kg was delivered by LSCS to an HIV reactive mother on ART. The neonate was on formula feeds by katori spoon and was apparently well till day 6 of life when she developed skin desquamtion in peri-umbilical region and on face. She was admitted in neonatal intensive care unit of our hospital at 6th day of life and treatment was started. Routine investigations showed neutrophilic leucocytosis (TLC 30000, 78% neutrophils) and positive C-reactive protein (38 mg/L). She was suspected to have staphylococcal scalded skin syndrome and empirically started on intravenous cefotaxim and fluids. Dermatology consultation was taken and paraffin gauze dressing was done as per the dermatologist opinion. Blood culture and skin swab culture were sent and they came positive for staphylococcus aureus. Supportive treatment was continued. On day 8 of life, the baby developed features of shock and deep bluish discoloration of hands, forearms, feet and legs. This discoloration then rapidly progressed to dry gangrene at the tips of fingers, thigh and back. Inotropic support and fluid boluses were started and antibiotics were upgraded to vancomycin in view of fulminant sepsis. Platelet and fresh frozen plasma transfusions were given in view of low platelet count and deranged coagulation profile. The possibility of neonatal purpura fulminans was considered keeping in mind the

characteristics and extent of lesions. Protein C and S couldn't be done due to financial constraints. Patient developed refractory shock with purpura and gangrene on whole body and despite intensive supportive management, she expired at day 10 of life due to septicaemia and multi-organ failure. The sudden and rapid progression to dry gangrene within two days raised strong possibility of neonatal purpura fulminans.

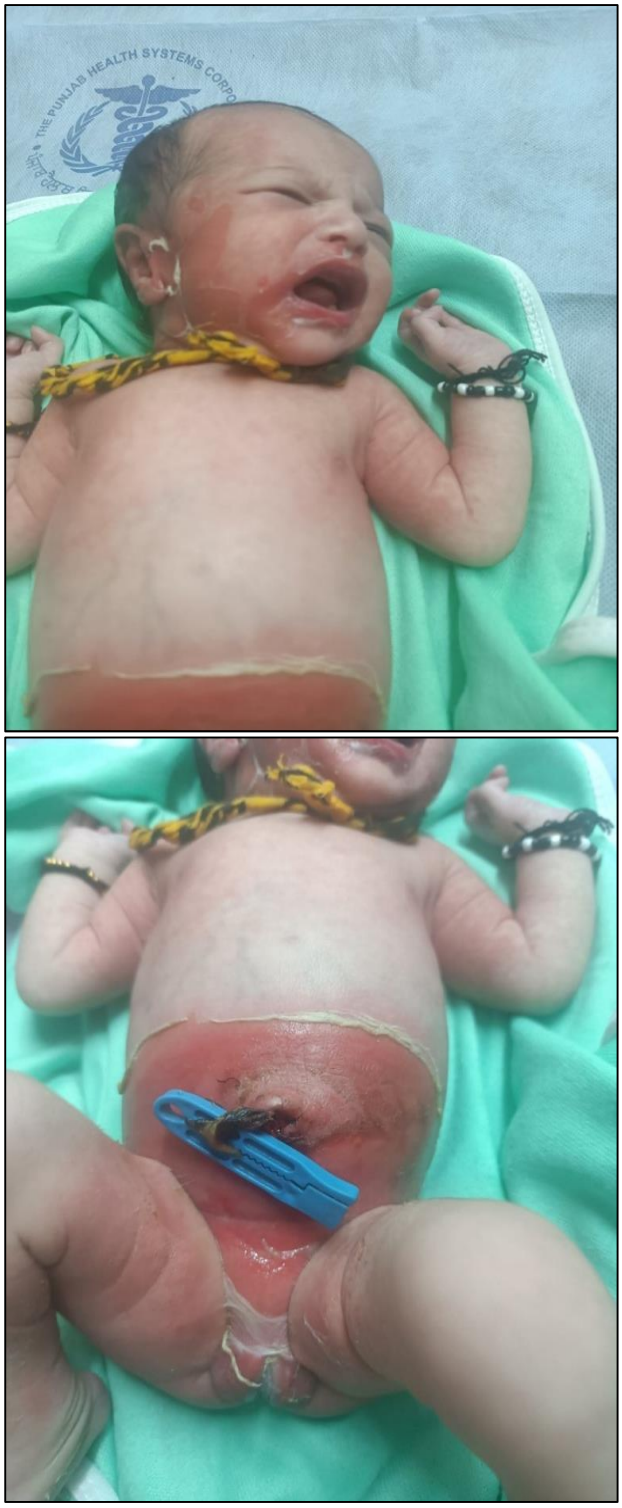

Figure 1: Day 6 of life (on admission).

Figure 2: Day 8 of life.

DISCUSSION

Sespsis associated purpura fulminans is rare but a well described condition. In children, *Neisseria meningitides*

is the most common cause, others being group A and B *streptococci*, *Hemophilus influenza*, *Escherichia coli*, Varicella and *Pneumococcus*. *Staphylococcus aureus* sepsis causing purpura fulminans has been reported in adult population, however it is very rare in children and neonates.[3] A literature search revealed very few cases occurred in neonatal age group. 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.[4] The skin lesions may present early as petechial rashes. These rapidly progress to larger ecchymotic areas. In general, the history and physical exam for purpura fulminans should be aimed at early recognition and determination of the underlying cause. Petechial rash or bruising in a neonate or septic patient should trigger consideration of purpura fulminans. Management is largely supportive, aimed at correcting the fluid and electrolyte imbalance, administering intravenous antibiotics to prevent secondary infection and platelet and FFP transfusions to manage underlying DIC. Sepsis related PF is reported to havre 50% mortality in the presence of septic shock and 90% mortality when patients require skin grafting and amputation. Our case highlights the fact that *Staphylococcus* is an important pathogen implicated in the etiology of purpura fulminans and it has a rapid downhill course.

CONCLUSION

Neonatal PF secondary to *Staphylococcus* infection is a fatal condition that should not be missed. The patient should be screened for congenital and acquired causes. There is a necessity of multidisciplinary approach for better management of this fatal disease.

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REFERENCES

1. Stenflo J. A new vitamin K-dependent protein. Purification from bovine plasma and preliminary characterization. J Biol Chem. 1976;251:355-63/.
2. Gurses N, Ozkan A. Neonatal and childhood purpura fulminans: review of seven cases. Cutis. 1988;41:361-3.
3. Verma P, Pandhi D, Yadav P, Dhawan AK. Neonatal purpura fulminans due to methicillin resistant *Staphylococcus aureus*. Pediatr Dermatol. 2013;30:266-7.
4. Colling ME, Bendapudi PK. Purpura Fulminans: Mechanism and Management of Dysregulated Hemostasis. Transfus Med Rev. 2018;32(2):69-76.

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