International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

A Rare Case of Congenital Purpura Fulminans in Neonate

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Abstract: ***Purpura fulminans also known as purpuragangrenosa. It is an acute often fatal thrombotic disorder which manifests as blood spots, bruising and discoloration of the skin resulting from coagulation in small blood vessels within the skin and rapidly leads to skin necrosis and dissimated intravascular coagulation. Incidence is 1 in 5, 00000 to 1 in 1000000, equal in both sexes.***

Keywords: Congenital purpura fulminans, protein C&S deficiency, purpuric lesions over body

1. Case Report

A term one hour old male newborn weight 3.2 kg born to G4P3L1A1D1 mother through normal vaginal delivery parents noticed purpuric rash over the face which progressed to all over the body within 6 hours of life brought with complaints of purpuric rash all over the body For which complaints they brought baby to SVRRGGH. On examination baby was sick looking with purpuric rash all over the body with cleft lip and cleft palate, presenting with shock at admission. We worked up for sepsis which was negative. Laboratory values were: Hct = 24.6%; WBC count = 22.9 × 10^9/L; platelet count = 168 × 10^9/L; PT = 22.27 s (11–15 s); PTT 61.866 s (25–35 s); FDP > 20 mcg/ml (normal up to 5); D - dimer 35.13 µg/ml (normal 0–0.5 µg/ml); Fibrinogen = 90 µg/L (normal: 180–350 µg/L); protein C 8%, very low (normal 70–140%); Protein S 103% (normal 70–140%); antithrombin III 64% (normal 75–125%). Other laboratory tests including serum electrolytes, renal functions, liver functions, urine and CSF analysis were in normal ranges. blood, urine and CSF cultures did not show any growth.

2. Discussion

Congenital Purpura fulminans is due to defects/ deficiency of protein C anticoagulant pathway. It is a autosomal dominant variant. Mutations of protein c gene (PROC) have been identified.

Diagnosis:
Done by detailed history and physical examination and relevant investigations.

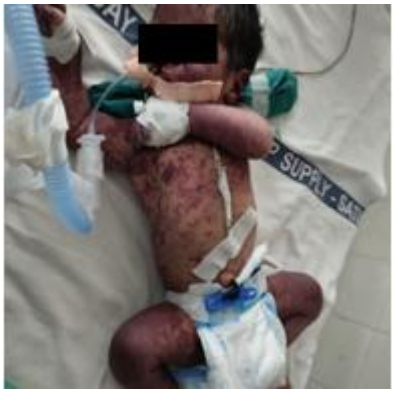

Image 1: Baby skin showing purpuric lesions all over the body

3. Conclusion

Supportive management with fluids oxygenation and antibiotics, replacing protein C deficiency with FFP Purpura fulminans with disseminated intravascular coagulation should urgently treated with fresh frozen plasma (10–20 mL/kg every 8–12 hours) and/or protein C concentrate to replace pro - coagulant and anticoagulant plasma proteins that have been depleted by the disseminated intravascular coagulation process.

References

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