Case Report

Thigh Echymosis in a Newborn

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ABSTRACT

Background and aim: Protein C is one of the important components in haemostasis system and its deficiency could increase the risk of thromboembolic events. This deficiency is seen in 0.2% of the general population. In this case report we present a 3-day old neonate with an echymosis in her left thigh.

Case presentation: A female neonate was visited regards to an echymotic lesion in her left thigh and a lighter one in the right which was present at birth. In her laboratory tests, protein C reported 17% while her parents had normal level. She was treated with anti-thrombotics and level of protein C elevated to 73% at discharge.

Conclusion: Clinical manifestations of protein C deficiency could be severe and life threatening, so quick and careful management is needed.

Key Words: Protein C, hemostasis, deficiency

INTRODUCTION

Protein C and S systems are the major regulatory systems of haemostasis. These are vitamin K dependent proenzymes which are synthesized in the liver. The thrombin thrombomodulin complex on the surface of the endothelial cells is the site for the interaction with protein C and S. After binding to these complexes, protein C becomes activated and protein S acts as a co-factor in this process. Activated protein C inhibits factor VIIIa and factor Va, thus exhibiting its anticoagulant property and it also enhances fibrinolysis through the inhibition of the plasminogen activator inhibitor [1].

Individuals with a mutation in the protein C gene tend to have an increased risk of thromboembolism. Homozygous or compound heterozygous mutants in patients who develop purpura fulminans in the neonatal period are rare [2].

Clinically, the patients with the protein C and protein S deficiencies are at an increased risk for venous thromboembolic disease, occasional arterial thrombosis, neonatal purpura fulminans, childhood stroke and even portal vein thrombosis [1].

The protein C deficiency is present in approximately 0.2% of the total population. This rate includes asymptomatic people and patients with severe thrombotic disease. People with the heterozygous form of the disease can usually live all their life without clinical problems [3].

CASE PRESENTATION

Here, we present a 3-days-old female baby who was delivered by vaginal delivery. She was the second baby from a third pregnancy. The second one was lost in the first trimester. There was a family relationship between the parents (they were cousins). She had no remarkable past medical history except for the beginning of jaundice from the second day of life. Her birth weight was 3200 grams, her present weight was 2900 grams, her head circumference was 34 cm and her height was 51 cm.

A bilirubin of 12.5 mg/dl (Normal range= 0.5-1.5 mg/dl) was reported at first. No poor feeding or abnormal stool or urine were detected. An echymosis of about 4 × 7 cm^2 had been noticed on her left thigh, "which was very light in colour at birth with a hand-size" her mother said. There was also a lighter lesion on the right thigh. Cutaneous and sub-cutaneous necrosis was noticed on her left thigh, plus a mild icter up to the abdomen in her physical examination. Antibiotics (Vancomycin, Ceftazidim and Amikacin) were started from the first day of her life and they were given for 20 days.

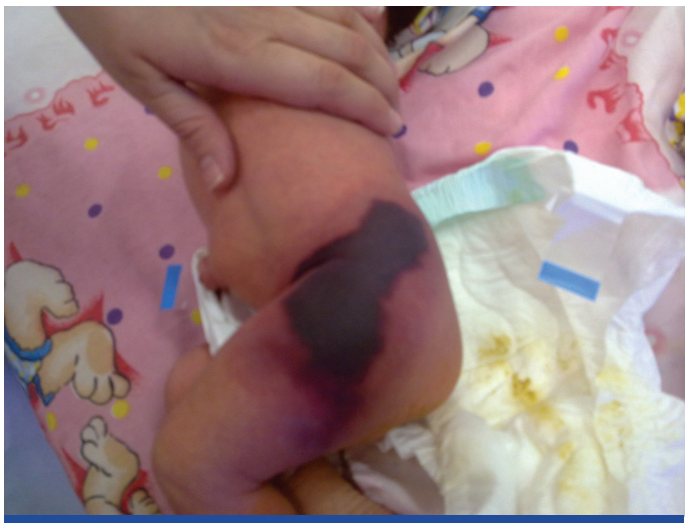
The first laboratory tests were as followings: Platelet=64000, PT=16 sec (PT control=12 sec), INR= 1.7,

PTT=36 Sec (Normal range=30-45 sec), G6PD=sufficient, MBG=B+, NBG=O+

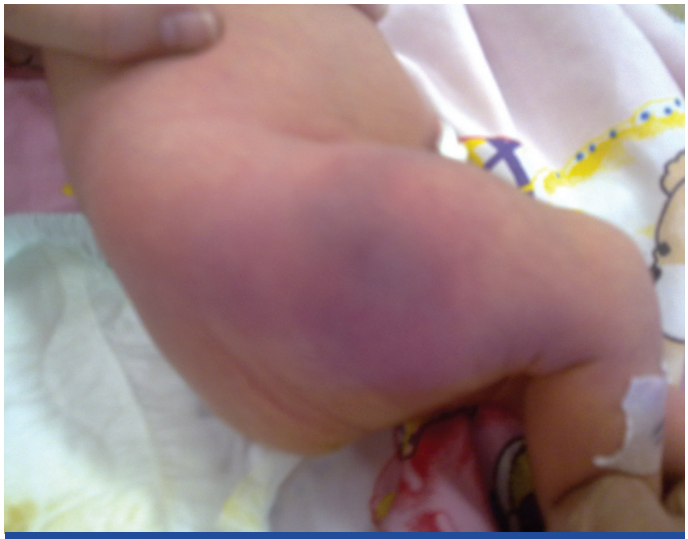
Neonate Protein C=17% and Protein S=40%

Father and Mother: Protein C=98%

A daily transfusion of fresh frozen plasma (FFP) was started from the third day of life for about 2 weeks, and then it was changed to every single day for 2 other weeks. The platelets reached a level



[Table/Fig-1]: Thigh showing the lesion



[Table/Fig-2]: Lesion at discharge

of 151000 after that. Vitamin K was prescribed concurrently. After transferring her to the Oncology Department, the daily Warfarin tablet (0.4 mg/ day) was continued with an increasing dose, which achieved upto 0.8 mg/ day at discharge. Protein C was reported to be 73% at discharge. Debridement of the necrotic area was done a day before discharge and the scar continued to heal.

CONCLUSIONS

The classical neonatal manifestation of homozygous protein C deficiency is a severe form of thrombosis of the large vessels or a purpura fulminans that occurs within a few hours or days of life, causing tissue necrosis and gangrenous and disseminated intravascular coagulation [3].

Abnormalities of antithrombin and the protein C system have been documented in association with spontaneous neonatal thrombotic problems. The protein C levels gradually improve during life. The protein C concentration is 15% of the adult levels during infanthood, 35% of the adult levels in the premature newborn, 80% of the adult levels in adolescents and it improves by 4% every 10 years [3].

Management of these patients with deep vein thrombosis is done with heparin anticoagulation, either conventional or low molecular weight, along with a protein C concentrate or fresh frozen plasma therapy. The patients are advised to avoid dehydration at any cost as a preventive measure [1].

Treatment of a patient with protein C deficiency depends on the individual patient's risk of thromboembolic disease. As a result, the treatment is based on providing a source of Protein C. This can be done with fresh frozen plasma (FFP) or with human plasma protein C concentrate. At this time, there are still no studies that compare the use of the protein C concentrates versus FFP in severe protein C deficiency which is related to thrombosis [3].

REFERENCES

[1] Mondal R, Nandi M, Dhibar T. Protein C and Protein S deficiency presenting as deep venous thrombosis. *Ind Ped* 2010; 47: 188-9.
[2] Iijima K, Nakamura A, Kurokawa H, Monobe S. A patient with homozygous protein C deficiency (Lys 192 del) who developed venous thrombosis for the first time at adulthood. *Thrombosis Research* 2010; 125: 100–1.
[3] Tridapalli E, Stella M, Capretti MG, Faldella G. Neonatal arterial iliac thrombosis in type-I protein C deficiency: a case report. *Italian Journal of Paediatrics* 2010; 36:23.

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