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CONGENITAL PROTEIN C DEFICIENCY IN A NEWBORN

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Protein C deficiency is a rare but life-threatening bleeding disorder that may present in the nearest neonatal period. This article presents the case of a newborn girl with acute and progressive neonatal fulminant purpura, thrombosis, DIC-syndrome, intracranial hemorrhage, which developed within 4 days after birth as a manifestation of protein C deficiency. Protein C activity was below 5%. Treatment includes correction of coagulopathy, intensive wound care including negative pressure dressings and skin grafting, and supportive care for central nervous system problems. Long-term follow-up consists of lifelong anticoagulant therapy to avoid recurrence of these complications.
Key words: deficit, protein C, DIC-syndrome, purpura, newborn.

INTRODUCTION

Congenital deficiency of protein C is associated with a tendency to severe thrombotic disorders. Among congenital types of physiological anticoagulant deficiency, such as antithrombin III deficiency, protein C deficiency, protein C deficiency, protein C deficiency is the most common (0.2-0.4% of the population).

Protein C deficiency is inherited in an autosomal dominant manner. The level of protein C in heterozygous carriers is 30–60% of the norm, homozygous carriers have practically no protein C and die in utero or immediately after birth [2,3,4].

Protein C is produced in the liver and inhibits blood clotting by catalyzing active factors V and VIII, which leads to a decrease in the formation of thrombin. Protein C deficiency causes abnormal blood clotting that is difficult to control with anticoagulant therapy. Homozygous states are manifested in early childhood with fulminant purpura of newborns and are often fatal, the level of protein C in such newborns is at an undetectable level [6].

Protein C deficiency is a hereditary or acquired risk factor for thrombophilia, the manifestations of which range from asymptomatic to venous thromboembolism and life-threatening purpura fulminans in newborns. Hereditary protein C deficiency is caused by a mutation in the protein C (PROC) gene located on chromosome 2q14.3. Heterozygous and acquired protein C deficiencies are more common than homozygous deficiencies [1,5].

Severe protein C deficiency is a rare autosomal recessive disease that usually presents in the neonatal period with fulminant purpura and severe syndrome disseminated intravascular coagulation (DIC), often with concomitant venous thromboembolism (VTE) [8,9].

Homozygotes and compound heterozygotes often have a similar severe protein C deficiency phenotype. Mild (i.e., simple heterozygous) protein C deficiency, in contrast, is often asymptomatic, but may include recurrent episodes of VTE, most often caused by clinical risk factors. Coagulopathy in protein C deficiency is due to impaired inactivation of factors Va and VIIIa by activated protein C after the propagation phase of coagulation activation. Mutation analysis of symptomatic patients shows a wide range of genetic mutations [3,5,6].

Other common complications include ophthalmic problems and changes in the central nervous system (CNS).

Treatment includes correction of coagulopathy, intensive wound care including negative pressure dressings and skin grafting, and supportive care for eye and CNS problems. Timely detection and establishment of the causes of protein C deficiency can prevent the occurrence of many pathological processes in the human body. Long-term follow-up consists of lifelong anticoagulant therapy to avoid recurrence of these complications [1].

The purpose of the work is to demonstrate a case of congenital deficiency of protein C in a newborn child.

MATERIALS AND METHODS

This study was conducted on the basis of the JSC Medical University of Karaganda, in the MSI Multidisciplinary Center for Mother and Child in the city of Temirtau. A retrospective analysis of the medical history of patient P, who received inpatient care at the National Scientific Center for Motherhood and Childhood in Astana, was carried out. The child is in the pediatric department under round-the-clock dynamic monitoring of the state of health, a comprehensive examination, consultation of narrow specialists, etiotropic, pathogenetic and symptomatic therapy were carried out.

We present our own clinical observation.

Complaints: neurological symptoms, hemorrhagic manifestations.

Anamnesis morbi: The condition at birth is moderately medium-heavy due to hemorrhagic manifestations, in dynamics with deterioration. Objectively: spontaneous hematomas of both lower extremities, areas of necrosis in the area of the right foot and on the left in the area of the foot and ankle joint were revealed, laboratory data: thrombocytopenia - 34x10/9/l, in the coagulogram: prothrombin time - 21 sec, on the neurosonogram: signs of intraventricular hemorrhage (VZHK) 3 degrees on the right, parenchymal hemorrhage on the left, a preliminary diagnosis was made: Hemorrhagic disease of the newborn, early form. IVH (non-traumatic) of the 3rd and 4th degree in the fetus and newborn. Neonatal convulsions. Competing diagnosis: Coagulopathy.

Consulted by narrow specialists: pediatric surgeon, hematologist, neurologist. Antihemorrhagic therapy was carried out: vit K1 10 mg, anesthesia; locally: a compress with troxevasin (heparin); IV 1 course of antibiotic therapy. On the 4th day, vasoprostan 0.005mcg/kg/min was connected to improve microcirculation. Conducted transfusion of fresh frozen plasma due to clinical and laboratory data, well tolerated. For further examination and treatment on the 4th day of life, she was transferred to the National Scientific Center for Mother and Child (NSCMiD) in Astana, where she was hospitalized for up to 2 months of life.

Objective data at the age of 4 days (in NNSMiD): T – 37.2C, HR-148/min, RB-44/min, SpO2 - 94%. Weight: 3372gr. The condition is severe, due to neurological symptoms, hemorrhagic syndrome against the background of impaired hemostasis. Nursed in the intensive care unit. Responds to examination with reduced motor activity. Scream of medium strength, painful, when touched, anxiety intensifies. Eyes slightly open, photoreaction positive. Pastosity of the face. The head is rounded, head circumference 37cm. Large fontanel 4.0x4.0 cm, not tense, divergence along the sagittal suture 0.6 mm, along the coronal sutures 0.4 mm. The skin is pale pink (described below), visible mucous membranes, pink. Symptom "white spot" up to 2 seconds. Tissue turgor is preserved. Subcutaneous fat is developed enough, evenly distributed. Muscle tone is reduced, reflexes of innate automatism are depressed. The chest is cylindrical. Breathing independent, oral wheezing. Saturation within 90-98%. In the lungs, auscultatory breathing is carried out in all fields, moist and wired rales. Heart sounds are clear, rhythmic. Hemodynamic parameters are stable. The abdomen is soft, palpable, painless. The liver protrudes from under the edge of the costal arch by +1.5 cm, the spleen protrudes from under the edge of the costal arch by +1.5 cm. Stool, gases depart. Peeing. Peripheral catheters are functioning.

Status localis; there is a purple hematoma in the inguinal region with spread to the left large labia with an uneven demarcation line, a pronounced hematoma in the right thigh, occupying the entire back, side and front surface (fig.1), large blisters filled with blood, hemorrhagic impregnation from the entire surface, on the right forearm hemangioma 4.0-4.0 cm. n the area of the lesion, there is an open bladder measuring 0.5 x 1.0 cm. (fig. 2). Hematomas in the area of the heel and ankle joint on the left, 4 x 6 cm in size, with the presence of an unopened bubble of a gray-purple color with a cloudy liquid inside. On the back of the right hand, there is a hematoma with a diameter of 2.0 cm, on the little finger 0.5 cm with an uneven demarcation line. During manipulation, the injured limbs are painful. The range of motion in both lower extremities is preserved, pink fingers are mobile.

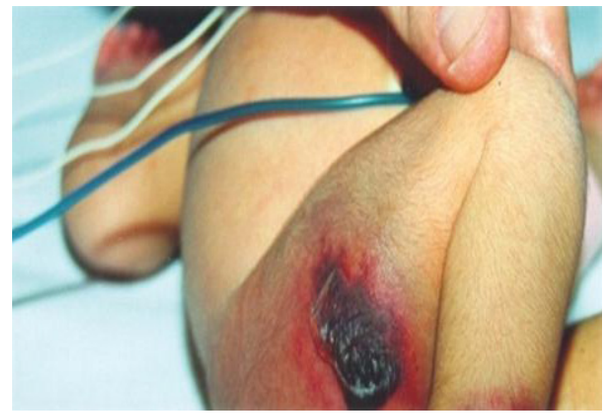

Figure 1 – Purple hematoma on the right thigh

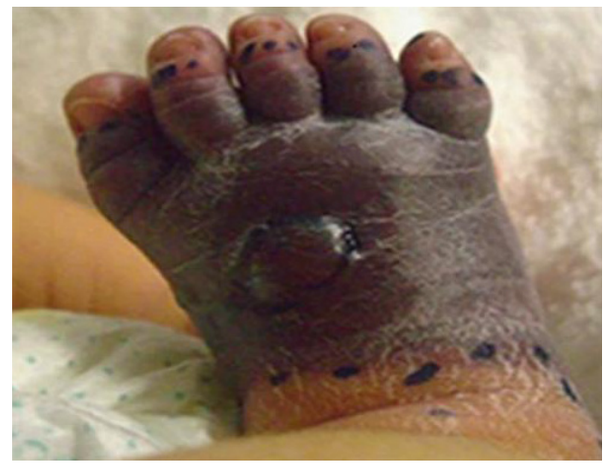

Figure 2 – Purple hematoma with a large bladder on the right foot

Laboratory and diagnostic studies:

Complete blood count from 10/23/2022 00:19: hemoglobin - 101 g/l, erythrocytes - 2.86 × 10/12/l, hematocrit - 28.8%, leukocytes - 19.40 × 10/9/l , platelets (manual counting) - 53 × 10/9 / l, s / i - 61%, p / i - 0%, e - 2%, l - 21%, m - 16%.

Complete blood count in dynamics from 12/09/2022: Hb - 122 g/l, erythrocytes -4.2×10/12/l, hematocrit - 37.3%, platelets-626×10/9/l, leukocytes – 11.43×109/l, neutral. -30.4%, b -0.2%, l. -52.3%, m - 12.1%, e -4.6%.

Biochemical blood test from 10/23/2022 00:21: urea - 5.24 mmol / l, creatinine - 44.06 μmol / l, total bilirubin - 163.3 μmol / l, direct - 11.68 μmol / l, total protein - 49.48 g/l, albumin - 33.08 g/l, AsAT - 28.58 U/l, AlAT - 5.70 U/l, CRB - 6.66 mg/l.

Biochemical blood test 10/24/2022 08:49: CRB - 12.8 mg / l.

Coagulogram 10/27/2022 06:08: APTT-44.4 sec, INR - 1.02, Quick PTI - 99.1%, PTT - 11.4 sec, TV - 15, fibrinogen - 1.1 g / l ↓

Coagulogram in dynamics 12/14/2022: MHO-0.90; APTT-25.6 sec; Prothrombin time - 10.2 sec.; PTI according to Quick - 121.2%, TV 18.2 sec, Fibrinogen - 1.7 g / l.

Analysis of the thromboelastogram 10/28/2022: signs of hypercoagulability, in all links of hemostasis. Given the level of D-dimer 18.97 mg/l, signs of hemorrhagic syndrome, bruising, bleeding, DIC syndrome in the stage of hypercoagulability is not excluded. The level of functional fibrinogen is 3.6 g/l.

Analysis of thromboelastogram 10/31/2022: normocoagulation with a tendency to hypercoagulation. Functional fibrinogen 2.6g/l.

Blood coagulation factors 10/28/2022: Factor V-166%, Factor VII-81.9%, Factor VIII-172.5%, Factor IX-61.9%, Willebrand factor antigen - 153.6%.

Blood clotting factors 11/16/2022: Factor V-132.3%, Factor VII-89.1%, Factor VIII-129.9%, Factor IX-104.5%, Willebrand factor antigen - 103.8%.

Antithrombin III 10/24/2022 -61.8%

Antithrombin III 12/12/2022 - 65.0%

D-dimers 10/24/2022 08:48-18.9 mg/l↑.

D-dimers 10/312022 -35.2 mg/l ↑.

Protein C 11/21/2022 - 3.41% ↓↓↓.

Protein C 28/11/2022 - 2.08% ↓↓↓.

Protein C 12/12/2022 - 10.0%↓↓

General urine analysis 10/26/2022: total protein - 0.08 g / l, light yellow, transparent, blood -, bilirubin -, glucose -, specific gravity - 1.005, pH - 7.5, leukocytes 3-4 in p / sp, erythrocytes unchanged. - 1-2 in p / sp, erythrocytes meas - 0-1 in p / sp.

Coproscopy 11/16/2022- Neutral fats +, Greenish stool color, There is superficial mucus, Leukocytes on mucus in p.z. Not found, Leukocytes in p.z. 3-4, Erythrocytes on mucus in p.z. Not detected.

Instrumental examinations:

Neurosonography 10/27/2022 Echo picture of hemorrhage of the fronto-occipital region of the right hemisphere, fronto-parietal region of the left hemisphere.

Neurosonography in dynamics on 11/21/2022: Echo-signs of periventricular leukomalacia (stage of cystic degeneration), asymmetric ventriculomegaly of 1-2 degrees, dilatation of the subarachnoid space.

Doppler ultrasound of the arteries of the lower extremities 11/25/2022 - Arterial blood flow in the main arteries of the legs and feet on both sides was not changed.

EchoCG on 10/23/2022: ODA 2.0 mm. OOO 4.0 mm. Slight dilatation of the right side of the heart. The thickness of the myocardium of the left ventricle is normal.

CT of the brain 10/23/2022: signs of hemorrhage in the parieto-occipital region of the right and fronto-parietal region of the left hemisphere of the brain, with signs of edema of the right hemisphere with a shift of the median structures to the left. Left intraventricular hemorrhage. Cephalohematoma of the fronto-parietal region on the left.

Consultation of a neonatal surgeon 10/26/2022 - Hematomas in the heel area on both sides and the ankle joint on the left. Recommended: treatment with betadine x 1 r / d.

Consultation of an oncohematologist on 11/01/2022: Diagnosis: Coagulopathy of unknown etiology. Rec-but: 1) Determine coagulation factors (V, VII, VIII, IX, von Willebrand). 2) intravenously drip FFP until complete correction of the coagulogram. 3) Octaplex IV 30 IU/kg* 2r/day.

Consultation of a neurosurgeon 11/30 2022 - a picture of mixed hydrocephalus, atrophy of the brain substance. Recommended: CT scan of the brain in 3 months.

Consultation of a neurologist 11.01.2022: Recommended: 1) tab. Phenobarbital 0.1 mg, 0.005 mg (2 powders) 2 times a day, then dynamic inspection. 2) CT of the brain in dynamics.

Consultation of a hematologist 11/30/2022: Secondary coagulolopathy, protein C deficiency. Recommended: IV Octaplex 50-80 IU/kg* 2 times a day, FFP 10-15 ml/kg x 1-2 times a day under the control of a coagulogram, ultrasound of the abdominal organs., control of CBC, hemostasis in dynamics. The prognosis for life is serious.

Consultation of a neurologist 12/05/2022: Diagnosis: HIE. Benign myoclonus. Epilepsy? Recommended: EEG 3 hours, with sleep. Kepr 30mg/kg/day in 2 divided doses every 12 hours. Repeated consultation with EEG results.

Consultation of an angiosurgeon 11/07/2022: Recommended: Doppler Ultrasound of the arteries and veins of the lower extremities.

Dermatologist consultation 11/11/2022 - Diagnosis: Hemorrhagic purpura of the newborn. Recommended: spray Panthenol x 2 r / d on the wound area.

Treatment performed:

Received complex treatment:

Protective mode. Joint stay with the ward with mother.

Open resuscitation system, monitoring of vital functions.

Enteral nutrition with a mixture of 55-57 ml every 3 hours through a gastric tube.

Antibacterial therapy: tazar 100 mg / kg / 12 hours IV No. 4.

With an antifungal purpose - nofluk 3 mg / kg / 72 hours No. 1.

Antihemorrhagic therapy: Amri-k 0.1 ml IM No. 4.

To replenish protein C: Octaplex 40 U/kg IV every 6 hours.

In order to improve microcirculation, vasoprostan 0.005 mcg/kg/min No. 2. Furosemide 1% IV No. 1.

In order to form intestinal microbots - Linex 5 drops x 2 r / day inside. For anticonvulsant purposes: Kepr 30 mg/kg x 2 r/day orally.

Baxet baby orally. Ferrovit 5 cap/kg orally.

Parenteral nutrition in the composition of 10%, 40% glucose solutions, 10% aminoven, electrolytes, Soluvit No. 3.

Hemotransfusion: PFF No. 1 of 10/25/2022. Endovasal (endovenous) laser coagulation (obliteration) of veins No. 1 dated 10/25/2022.

Conclusion: For 2 months of treatment, the activity of protein C did not reach the control level, she was transferred to inpatient treatment at the place of residence in Temirtau with the final diagnosis: Fulminant purpura of newborns. (D68.2) Hereditary deficiency of protein C. Complication: Disseminated intravascular coagulation in the fetus and newborn.

Competing: P91.6 - Hypoxic ischemic encephalopathy of the newborn. Mixed hydrocephalus, atrophy of the substance of the brain.

Complication: P90 Neonatal convulsions.

Concomitant: other intracranial (non-traumatic) hemorrhages in the fetus and newborn. p29.3 Persistent fetal circulation in newborn. Severe anemia (stopped).

Recommendations were given enteral feeding according to the physiological need for fluid. Control of blood tests, hemostasis. Correction of hemostasis according to indications. CT scan of the brain in dynamics. Observation of a hematologist, a neurologist in dynamics. Symptomatic treatment.

Objective data at the age of 2 months: HR-134 per minute, BR=34 per minute. T=36.2*C. SpO2=98%. Weight: 5110 gr.

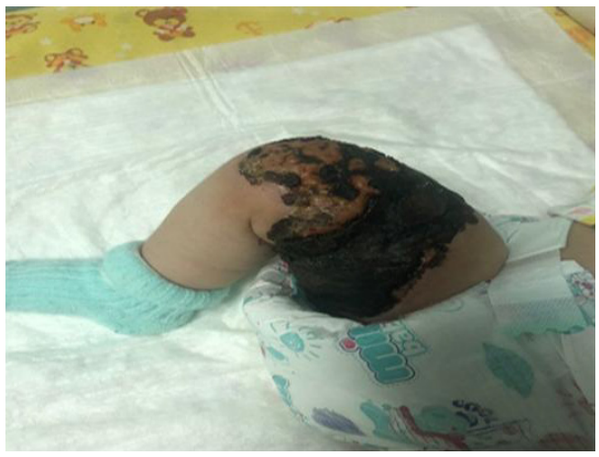

Figure 3 – Hematomas in the right thigh, occupying the entire posterior, lateral and anterior surface of the crust, which began to move away from the edges, oozing purulent discharge

The child's condition is moderate, stable. She is nursed in the ward of joint stay with her mother. Body temperature is within normal limits. He reacts to examination with moderate physical activity, wakes up to feeding. Eyes open, photoreaction positive. Crying is loud and emotional. Receives anticonvulsant therapy, there were no convulsions and convulsive readiness, a slight tremor of the upper and lower extremities. L.F. 4.5x4.0 cm, not tense, divergence along the sagittal suture 0.5 mm, along the coronal lice 0.5 mm. Muscle tone is moderately reduced, reflexes of congenital automatism are reduced. It is fed with a mixture of 120 ml every 3 hours from the horn, supplemented through a tube, assimilates. Didn't burp, didn't vomit. The skin is described below. Breathing is independent. Saturation is within the normal range. In the lungs, auscultatory breathing is carried out in all fields, there are no wheezing. Heart sounds are clear, rhythmic. Hemodynamic parameters are stable. The abdomen is soft, not swollen, palpable. The liver and spleen are not enlarged. The chair for the past day was 3 times independent. Urinating, diuresis is adequate.

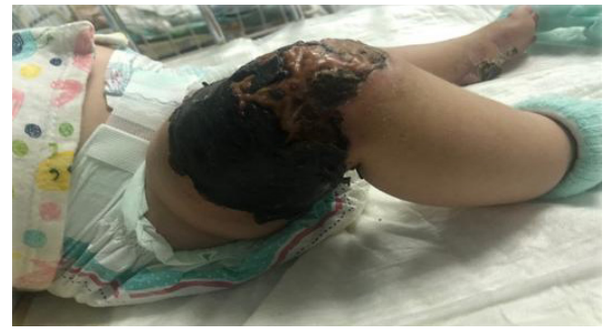

Figure 4 – Hematomas in the area of the right thigh, occupying the entire posterior, lateral and anterior surface of the crust, which began to move away from the edges, oozing purulent discharge

Figure 5 – Residual crust of purpura in the area of the left heel, without signs of inflammation

Figure 6 – The left hand in the place of which crusts formed in the place of the hematoma, without signs of inflammation

Local status: in the area of the anterior abdominal wall on the left, the crusts completely disappeared, the skin underneath is pink, scarred, without signs of inflammation, painless on palpation. In the area of the ankle joint on the left and in the area of the heel, the crust in dynamics moves away, the skin underneath is pink, scarred, without signs of inflammation, painless on palpation. In the area of the outer part of the thigh on the left, there is a superficial hemangioma 2x2 mm in size, not progressing (fig. 3,4,5,6).

Anamnesis vitae:

Child from 3 pregnancies, 2 births.

1st pregnancy - 2020 - urgent delivery, 3300gr, no

2nd pregnancy - spontaneous miscarriage at 7 weeks

3rd pregnancy - this.

The child's mother has been registered since 12 weeks. I half of pregnancy - at 12 weeks threatening abortion, inpatient treatment, at 18 weeks of age chronic cytomegalovirus infection, latent course, mild severity, viferron suppositories, chronic salpingo-oophoritis, colpitis were prescribed. Obstetrical diagnosis: Rapid term labor at 38 weeks + 4 days. Single loose entanglement of the umbilical cord around the neck of the fetus. Birth weight 3070 gr, height at birth 49cm. Head circumference 33cm. Chest circumference 32cm. Apgar score 8/9 points. Family history is not burdened by hemorrhagic syndrome.

Opinion: In this clinical case, we described a case of fulminant purpura of a newborn due to protein C deficiency, complicated by DIC, intracranial lesion, which required urgent diagnostic measures and treatment. Protein C activity was below 5% at a rate of up to 35%. Determination of its level of content was carried out in conjunction with a comprehensive laboratory study of other indicators of the coagulation and anti-coagulation systems of the blood. Against the background of the pathogenetic and symptomatic therapy, the condition stabilized in dynamics, there were periods when new hemorrhagic eruptions appeared against the background of a decrease in fibrinogen. Congenital deficiency of protein C required preventive and therapeutic measures to prevent the development of thrombosis and fatal complications. According to the literature, liver transplantation can be an effective treatment for congenital protein C deficiency [9,10].

Prenatal diagnosis may facilitate preemptive and definitive treatment of severe protein C deficiency with a positive family history of hemorrhagic syndrome. Molecular genetic study of the child and her parents remains relevant for the confirmation or exclusion of hereditary diseases. Given the uncomplicated history of hemorrhagic syndrome, there may have been a mutation in the gene responsible for the synthesis of protein C.

Financing. The research was not sponsored.

Conflict of interest. The authors absence no conflict of interest.

Informed consent: A written voluntary informed consent was received from the patient's parents to publish a description of the clinical case, including the use of his medical data, the publication of his images in a medical journal, including its electronic version (results of examination, treatment and observation) for scientific purposes (date of signature — 12/30/2022).

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ЖАҢА ТУҒАН НӘРЕСТЕДЕГІ ТУА БІТКЕН С ПРОТЕИНІНІҢ ЖЕТІСПЕУШІЛІГІ
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Протеин С жетіспеушілігі- сирек, бірақ өмір үшін кауыпты қанның ұю қызметінің бұзылумен, неонаталды кезеңде пайда болу ықтимал. Бұл мақалада жаңа туылған қыздың жедел және үдемелі, кенет жаңа туылған балалардағы пурпурамен, тромбозбен, диссеминирленген тамыр ішілік қан ұю бұзылыс синдромымен, бас сүйек ішілік қан құюылуымен, протеин С жетіспеушілігінің туылғанан кейін 4 күн ішінде дамуын көрсетеді. Протеин С белсенділігі 5%-дан төмен. Емі коагулопатияны түзеу, теріс қысыммен байлау және теріні ауыстырып отырғызумен қатар(трансплантация), қарқынды түрде күтіммен қарау, сонымен қатар, орталық жүйке жүйесінің зақымдануында күтім терапиясы. Ұзақ уақытты бақылау, қайтіп асқынуларды болдырмау мақсатында өмірлік антикоагулациялық терапиядан тұрады.
Кілт сөздер: жетіспеушілік, протеин С, тамыр ішілік қан ұю бұзылысының синдромы, пурпура, жаңа туылған нәресте

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ВРОЖДЕННЫЙ ДЕФИЦИТ ПРОТЕИНА С У НОВОРОЖДЕННОГО
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Дефицит протеина С является редким, но опасным для жизни нарушением свертываемости крови, которое может проявиться в ближайшем неонатальном периоде. В этой статье представлен случай новорожденной девочки с острой и прогрессирующей молниеносной пурпурой новорожденных, тромбозом, ДВС синдромом, внутричерепным кровоизлиянием, развившиеся, в течение 4 дней после рождения как проявление дефицита протеина С. Активность протеина С была ниже 5%. Лечение включает коррекцию коагулопатии, интенсивный уход за раной, включая повязки с отрицательным давлением и пересадку кожи, а также поддерживающую терапию при проблемах с центральной нервной системой. Долгосрочное наблюдение состоит из пожизненной антикоагулянтной терапии, чтобы избежать повторения этих осложнений.
Ключевые слова: дефицит, протен С, ДВС- синдром, пурпура, новорожденный