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DIAGNOSTIC CHALLENGES OF A RARE KASABACH–MERRITT SYNDROME IN AN INFANT

The article is devoted to topical issues of the complexity of the diagnosis of a rare pathology – Kazabach–Merritt syndrome in infants. Against the background of progressive bilateral polysegmental pneumonia, the child retained thrombocytopenia and consumptive coagulopathy. Syndrome-by-syndrome treatment was performed in the intensive care units of the perinatal and pediatric centers of the republican hospital. The patient received a large amount of blood replacement therapy, and the pleural and abdominal cavities were repeatedly drained. In this clinical case, there were no external manifestations of the disease in the form of a tumor on the skin. Initially, the formation was not diagnosed on early CT scans. Kaposiform hemangioendothelioma complicated by Kasabach–Merritt syndrome was suspected by the exclusion method, which was confirmed at the Federal Center. Properly prescribed complex chemotherapy led to an improvement in the baby's condition. In clinical practice, there are often situations where the symptoms of the disease can be masked by other conditions, such as infectious processes or other hemorrhagic disorders. This highlights the importance of a multidisciplinary approach in the diagnosis and treatment of this disease.

Keywords: Kasabach–Merritt syndrome, kaposiform hemangioendothelioma, thrombocytopenia, consumptive coagulopathy, retroperitoneal formation, newborn, clinical case

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Introduction. Kaposiform hemangioendothelioma (KHE) is a rare congenital vascular tumor manifesting in infancy, histologically resembling Kaposi's sarcoma but not etiologically associated with herpesvirus type 8. According to the ISSVA classification, this tumor is classified as an intermediate malignancy with infiltrative growth and no metastatic potential. The most severe complication of KHE is the development of Kasabach–Merritt syndrome, which occurs in 80-90% of cases and manifests from birth in 50% of cases. The clinical signs of the syndrome are related to the size of the tumor. Kasabach–Merritt syndrome is a symptom complex characterized by the presence of a vascular tumor and associated consumptive coagulopathy and thrombocytopenia. Currently, due to the lack of randomized clinical trials, there is no standardized clinical protocol for the treatment of this disease.

The syndrome was first described by Kasabach and Merritt in 1940, and since then over 200 cases have been registered worldwide [5]. The incidence in the Russian Federation has not been reliably established [1, 2]. Among all vascular tumors, Kasabach–Merritt syndrome accounts for 1.0% in global pediatric practice [2, 5]. Mortality from this disease is quite high due to heart failure, infections, disseminated intravascular coagulation syndrome, and thrombocytopenia leading to massive bleeding, reaching 10-37.4% [1, 2, 5].

Treatment is primarily conservative, including propranolol, corticosteroids, vincristine, sirolimus [2, 3, 5, 8, 9]. Surgical treatment is often impossible due to coagulopathy, thrombocytopenia, and the infiltrative growth of the lesion [1].

Aim: To demonstrate the diagnostic challenges of a rare clinical case – Kasabach–Merritt syndrome in an infant during the first months of life.

Materials and Methods: We present the medical history of a patient who received inpatient treatment in the ICU of the Perinatal Center and later in the ICU of the Pediatric Center of the republican hospital.

Life History. A boy born in 2024. The child is from the 3rd pregnancy, which proceeded without complications. Pre-

natal screening results: I – 13 weeks pregnancy, marginal placenta previa; II – low-lying placenta, local myometrial tone; III – 26 weeks pregnancy, transverse lie of the fetus.

Childbearing: second, natural, at 39 weeks. Birth weight: 3540 g, height 56 cm. Condition at birth: satisfactory, Apgar score 8/9. Cried immediately. Put to the breast on the 1st day, sucked actively. Umbilical cord stump fell off on the 6th day. Vaccination: Hepatitis B, BCG-M – medical exemption due to thrombocytopenia in the complete blood count (CBC) at birth. Ultrasound screening of the abdomen, hip joints, neurosonography – no echopathology detected; Cardiac ultrasound – ventricular septal defect (VSD), patent foramen ovale.

Clinically, the child developed jaundice from day 2. At birth, CBC showed thrombocytopenia 113-109x10⁹/L, with a gradual decrease in platelet count (95x10⁹/L at discharge). Discharged home due to the mother's refusal of further inpatient observation and examination.

Medical history. The illness began on the 20th day of life. The child had decreased appetite, and the family visited the local central district hospital, where a pediatrician detected shortness of breath with intercostal retractions and general weakness. The child was urgently hospitalized in the ICU of the central district hospital; a pleural puncture was performed, yielding 97 ml of serosanguinous fluid. The child was transported by air ambulance to the republican hospital in the Department of Anesthesiology, Resuscitation and Intensive Care of Newborns at the age of 3 weeks. Upon admission, the condition was assessed as severe due to grade 3 respiratory failure and hemorrhagic syndrome. The patient was intubated and connected to a mechanical ventilator; hemostatic therapy was initiated, with transfusions of fresh frozen plasma (FFP), leukocyte- and platelet-depleted erythrocyte mass (LPDEM), and platelet concentrate. Contrast-enhanced computed tomography (CT) of the chest was performed, revealing bilateral polysegmental pneumonia and bilateral pleural effusion. Based on the mother's ELISA results, the child was diagnosed with congenital TORCH syndrome (mixed infection: mycoplasma, parvovirus, and chlamydia). Antibacterial therapy was prescribed: Cefepime/Sulbactam 50 mg/kg/day every 12 hours, Azithromycin 10 mg/kg/day once daily; antifungal therapy with Fluconazole; and for immunomodulation, Immunoglobulin G (Privigen) 1 g/kg/day intravenously, microstream, daily for 5 days. Biochemical analysis of pleural fluid showed increasing triglycerides up to 3.34 mmol/L; the patient was switched to total parenteral nutrition, and Octreotide was prescribed at a dose of 4 μg/kg/h microstream. With the ongoing treatment, the amount of pleural fluid decreased, and feeding with Alfare formula was started at 10 ml every 3 hours, gradually increasing the volume to 90 ml, without an increase in fluid output.

At the age of 1.5 months, the child was transferred to the Department of Anesthesiology and Resuscitation affiliated with the Surgical Department of the Pediatric Center, where he remained until further transfer to a Federal Center. At 2 months of age, contrast-enhanced CT of the chest and abdomen (Fig.1) and contrast-enhanced MRI of the abdomen (Fig.2) were performed. Findings included: a para-aortic mass at the level of the diaphragm, extending into the retroperitoneal space and into the pelvic cavity with involvement of the lumbar muscles; destruction of thoracic vertebrae Th9-Th12; fluid in the left abdominal cavity, possibly hemorrhagic; enlarged adrenal glands. The child was examined by a pediatric oncologist and a phthisiatrician. Tumor markers were within normal limits, ruling out a specific process. Kaposiform lymphangiomatosis or kaposiform hemangioendothelioma with Kasabach–Merritt syndrome of the lower mediastinum, retroperitoneal space, and pelvis was suspected. Following a remote consultation, emergency hospitalization to the N.N. Blokhin National Medical Research Center of Oncology was planned.

At 2 months of age, the child was transported to the airport for medical evacuation, accompanied by an intensivist. At the airport, the patient's condition was deemed non-transportable – there was an increase in signs of respiratory failure, cyanosis of the skin, and active hemorrhagic discharge of 80 ml from the pleural cavity. After fluid removal, the child's condition stabilized. The dressings were heavily soaked with hemorrhagic discharge. The patient was returned to the ICU of the Pediatric Center in extremely critical condition. Transfusion of leukocyte- and platelet-depleted erythrocyte mass, platelet concentrate, and fresh frozen plasma was performed, stabilizing the patient's condition.

A week later, the child developed symptoms of pseudomembranous enterocolitis, which improved with treatment: Metronidazole 7.5 mg/kg x 3 times/day IV + Vancomycin 40 mg/kg/day, divided into 4 doses per os. In coordination with the D. Rogachev National Medical Research Center of Pediatric Hematology, Oncology and Immunology, the child was prescribed Sirolimus. This is a macrolide obtained from *Streptomyces hygroscopicus*, which blocks calcium-mediated and calcium-independent intracellular signaling upon activation of IL-2 receptors in T-cells, thereby suppressing their activation and causing an immunosuppressive effect, at a dosage of 0.1 mg every 48 hours enterally. The child's condition stabilized on this therapy.

According to laboratory data, the child had anemia, thrombocytopenia, leukopenia, impaired hemostasis of the hypocoagulation type, hypofibrinogenemia, and elevated D-dimer levels. Throughout the treatment, bacterial cultures from the oropharyngeal and nasal mucosa, blood, endotracheal tube, urine, stool, and drainage fluid from both pleural cavities and the abdominal cavity showed no growth.

Summarizing the treatment provided, the patient received 11 individually matched LPDEM transfusions, 42 FFP transfusions, and 26 platelet concentrate transfusions.

At the age of 2.5 months, the child was referred for hospitalization to the N.N. Blokhin National Medical Research Center of Oncology of the Ministry of Health of the Russian Federation, where he was treated for 30 days. Upon admission to the medical center, the child's condition was severe, due to multiple organ failure (respiratory failure grade 2, heart failure 2A, gastrointestinal insufficiency), the course of an infectious process (intrauterine mixed infection, TORCH syndrome: parvovirus, chlamydia, community-acquired bilateral pneumonia), bone marrow suppression syndrome, DIC syndrome, and abdominal compartment syndrome due to a retroperitoneal space-occupying lesion.

At the Federal Scientific Medical Center, additional examinations were performed: tumor markers, myelogram, cytology of pleural and peritoneal fluid, consultations with specialists: geneticist, neonatologist, cardiologist. A review of the CT and MRI studies was conducted. PET CT of the whole body: at the time of the study, no data were obtained on the presence of 18F-FDG-positive tumor tissue in the retroperitoneal mass. A consultation was held jointly with the Federal Scientific Center. Considering the comprehensive examination data, the clinical diagnosis of "Kasabach–Merritt syndrome" against the background of a vascular formation of the posterior mediastinum was established. Given the child's severe condition (persistent respiratory failure, tachypnea, hypocoagulation,

Dynamics of Laboratory Parameters

Clinical blood analysis										
Indicator	15.11.2024	24.11.2024	04.12.2024	14.12.2024	25.12.2024	02.01.2025	05.01.2025	08.01.2025	10.02.2025	Ref. Interval
Erythrocytes (RBC)	3.23	4.57	3.93	5.1	3.99	3.72	3.87	3.52	3.02	(3.90 - 5.90) 10E12/ L
Leukocytes (WBC)	5.2	7.9	6.7	9.9	8.9	3.2	3.3	5.9	5.1	(9.00 - 30.40) 10E9/ L
Hemoglobin (HGB)	98.0	140.0	119	150	119	109	113	100	86	(168.00 - 208.00) g/L
Hematocrit (HCT)	29.3	41.4	34.5	44.9	34.8	32.1	33.4	29.9	25	(41.00 - 65.00) %
Platelets (PLT)	65.00	180	44	39	53	48	115	66	31	(150.00 - 400.00) 10E9/ L
Thrombocrit (PCT)	0.05	0.18	0.05	0.04	0.06	0.04	0.11	0.05	0.03	(0.15 - 0.40) %
Lymphocytes (LYMF) abs	2.2	3.0	3.2	5.4	3.5	1.4	1.6	2.1	1.9	(1.20 - 3.00) 10E9/ L
Monocytes (MON) abs	0.8			1.00			0.4	0.6		(0.09 - 0.60) 10E9/ L
Granulocytes (Gran) abs	2.2	3.8	2.6	3.5		1.3	1.3	3.2	2.6	(0.00 - 0.00) 10E9/ L
Band neutrophils	4	1	4	1	4	3	2		1	(1.0 - 6.0)%
Segmented neutrophils	26	56	36	19	53	41	39		46	(16.0 - 45.0)%
Eosinophils	13	7	10	17	0	1	2		1	(1.0 - 5.0)%
Lymphocytes	42	30	39	55	33	44	47		44	(45.0 - 70.0)%
Monocytes	15	6	11	8	10	11	10		8	(4.0 - 10.0)%
Normoblasts	1	-		1						(0.0 - 0.0)%
Reticulocytes			5							(2,0-12,0) %
Coagulogram										
Antithrombin-III	62.7	70.5	82.1							85.00 - 115.00%
Prothrombin time	17.80	14.8	16.8	17.1	17.4	15.7	16.2	15.4	14.8	9.00 - 12.60 sec
INR	1.57	1.3	1.48	17.1	1.3	1.35	1.39	1.32	1.27	0.81 - 1.13 units
APTT	50.30	29.8	42.3	42.00	41.7	No coagulation	No coagulation	32.9	36.3	23.40 - 35.00 sec
Fibrinogen	1.72	3.04	1.72	0.66	1.03					2.92 - 4.12 g/L
D-dimer	>10000	>5000	>5000							110.00 - 240.00 ng/ml
Prothrombin index	50.20	65.12	54.35	47.00	62.00	53.2	50.7	54.8	58.2	78.00 - 142.00 %

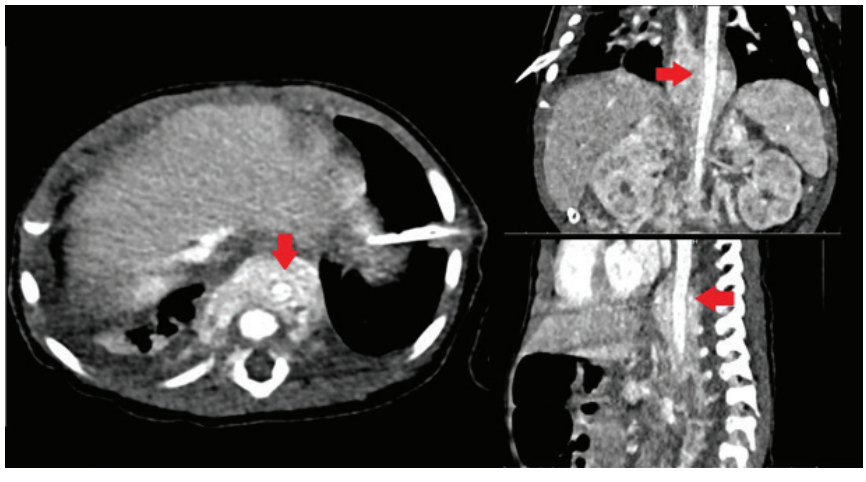

Fig.1. Contrast-enhanced CT of the chest and abdomen – revealed para-aortic mass

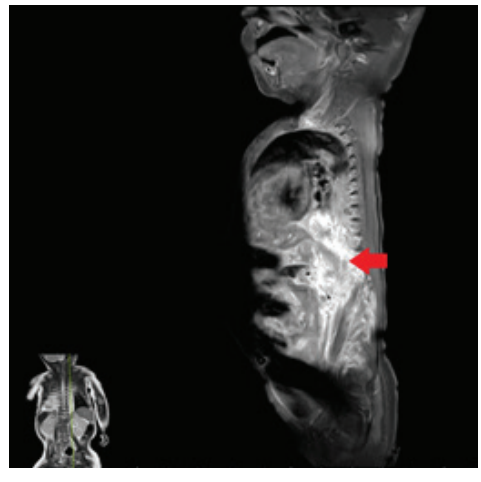

Fig.2. Contrast-enhanced MRI – retroperitoneal mass

thrombocytopenia) and the impossibility of surgical intervention (including biopsy of the mass), a decision was made to initiate special treatment without histological verification of the diagnosis for vital indications, in accordance with the consultation with the Federal Scientific Center. Metronomic therapy was prescribed – Cyclophosphamide, Vinblastine, Rapamycin for 3 courses. Metronomic therapy for 14 days: Cyclophosphamide 50 mg/m²/day, divided into two IV bolus injections over 14 days; Vinblastine 1 mg/m²/day 3 times a week. The course lasts 28 days, with chemotherapy administration from day 1 to day 14 of therapy; Rapamycin at a dose of 0.1 mg every 48 hours (during the 1st week with determination of the drug concentration in the blood, followed by possible dose adjustment). The child was discharged with recommendations for further treatment at the place of residence. The start of the 2nd course of therapy was scheduled for 02/11/2025. A follow-up examination after the 3rd course of metronomic therapy and contrast-enhanced MRI of the abdomen were recommended. The child was hospitalized in the oncology department of the Federal Scientific Center, where the 2nd course of therapy was administered: Rapamycin 0.2 mg/m²/day as 0.1 mg (0.1 ml) x 2 times a day, per os, daily; Vinblastine 1 mg/m² single dose – 0.28 mg IV strictly slowly, 3 times a week; Cyclophosphamide 50 mg/m²/day in 2 doses, single dose 7 mg, daily dose 14 mg, IV strictly slowly, 2 times a day.

The child was discharged with recommendations for further treatment at the place of residence; at the age of 4 months, he was hospitalized in the oncology department of the Pediatric Center of the republican hospital. The 3rd course of metronomic therapy was administered. Magnetic resonance imaging (MRI) of the abdomen and retroperitoneal space was performed, again revealing a pathological paravertebral, para-aortic mass of unclear etiology, with infiltration of the muscles on both sides, the right gluteus medius muscle, and right pleural effusion. Compared to previous studies, the mass was unchanged, and bone marrow edema of the Th9-L5 vertebral bodies was not detected. Clinically, the condition is stable, and the parents report no complaints regarding the child's well-being.

Conclusion. This clinical case demonstrates the difficulties in diagnosing and treating newborns with Kasabach–Merritt syndrome. The disease is characterized by an extremely severe course and high mortality. If a child presents with thrombocytopenia and hypocoagulation in coagulogram tests, it is imperative to perform CT/MRI of the retroperitoneal space, abdominal cavity, and chest organs with contrast to verify the diagnosis. Early treatment will help avoid severe hematological complications and reduce the likelihood of severe functional disorders.

The authors declare no conflict of interest.

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