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Acute Simultaneous Thrombosis of the Hepatic Veins and the Portal System in a 30-Year-Old Female with *JAK2*-Positive Essential Thrombocythemia

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Aim: to present a clinical case of simultaneous thrombosis of the hepatic veins and total thrombosis of the vessels of the portal system in a myeloproliferative neoplasm.
Key points. The patient was admitted to the gastroenterology department with complaints of abdominal distension, abdominal pain syndrome, and hyperthermia. An examination revealed widespread thrombosis of the hepatic veins and the portal system. Conservative therapy with diuretics, low-molecular-weight heparins, and albumin preparations proved ineffective. The patient was referred to a federal clinic for verification of the diagnosis of myeloproliferative neoplasm. As a result of the examination, a diagnosis of essential thrombocythemia was established. Against the background of comprehensive conservative therapy, a regression of refractory ascites was achieved. The patient is under dynamic observation of a hematologist, a hepatologist, hepatobiliary surgeon, and a transplantologist.
Keywords: extrahepatic portal hypertension, Budd — Chiari syndrome, hepatic vein thrombosis, portal vascular thrombosis, ascites, essential thrombocythemia
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Острый одномоментный (сочетанный) тромбоз печеночных вен и сосудов портальной системы при *JAK2*+ эссенциальной тромбоцитемии у пациентки 30 лет

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Цель: представить клиническое наблюдение одномоментного тромбоза печеночных вен и тотального тромбоза сосудов портальной системы при миелопролиферативном заболевании.
Основные положения. Пациентка госпитализирована в гастроэнтерологическое отделение с жалобами на увеличение объема живота, абдоминальный болевой синдром и гипертермию. При обследовании выявлен распространенный тромбоз печеночных вен и сосудов портальной системы. Консервативная терапия диуретиками, низкомолекулярными гепаринами и препаратами альбумина оказалась неэффективной. Пациентка направлена в федеральную клинику для верификации диагноза миелопролиферативного новообразования. В результате обследования установлен диагноз эссенциальной тромбоцитемии. На фоне комплексной консервативной терапии удалось добиться регресса резистентного асцита. Пациентка находится

под динамическим наблюдением гематолога, гепатолога, специалиста в области портальной гипертензии и трансплантолога.
Ключевые слова: внепеченочная портальная гипертензия, синдром Бадда — Киари, тромбоз печеночных вен, тромбоз сосудов портальной системы, асцит, эссенциальная тромбоцитемия
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Introduction

Portal hypertension is a frequent complication of various diseases and represents a multidisciplinary interest. The nature of the clinical manifestations depends on its form (subhepatic, intrahepatic, or suprahepatic), the rate of development, and the possibility of eliminating the etiological factor.

Most commonly, portal hypertension occurs in liver cirrhosis. Its clinical manifestations include ascites, bleeding from esophageal and gastric varices, hepatic encephalopathy, as well as splenomegaly with hypersplenism. Extrahepatic portal hypertension, which includes Budd–Chiari syndrome or thrombosis of the portal system, often accompanies the clinical course of myeloproliferative diseases and, in some cases, determines the severity of the patient's condition.

Essential thrombocythemia is a prognostically favorable type of myeloproliferative diseases with an incidence rate of 1 case per 100,000 population. The disease is characterized by uncontrolled megakaryocyte proliferation, an increased number of large and giant megakaryocytes in the bone marrow, as well as thrombocytosis in the peripheral blood (>450 × 10^9/L) associated with a high rate of thrombotic complications and bleeding [1]. Thrombosis of various localizations occurs with a frequency of up to 20 %, whereas venous thrombosis develops in less than 10 % of cases [2–4].

Budd–Chiari syndrome requires a stepwise approach depending on the response to therapy and can include both conservative treatment methods and endovascular or surgical techniques [4]. Since extrahepatic portal hypertension is not a standalone disease but often serves as a consequence of congenital or acquired thrombophilia, we present a clinical case of a 30-year-old female patient with acute-onset refractory ascites and febrile fever against the background of essential thrombocythemia complicated by Budd–Chiari syndrome and thrombosis of the portal system.

Case report

Patient V., a 30-year-old female, was admitted to the gastroenterology department of the Stavropol Regional Clinical Hospital on May 30, 2024, with complaints of severe weakness, increased abdominal volume, abdominal pain syndrome, decreased appetite, and a daily body temperature rise to 38–39 °C.

Medical history: previously, the patient had not sought medical attention for chronic diseases and was not registered for regular medical follow-up. There was no history of pregnancies or oral contraceptive use. Over the past 6 months, she had not undertaken air travel or long-distance trips, nor had she undergone immobilization. The family history was unremarkable for gastroenterological pathology and hematological diseases. Her paternal grandfather had varicose vein disease of the lower extremities complicated by deep vein thrombosis.

In late April, she outpatiently contracted a mild acute intestinal infection. The patient experienced abdominal pain, diarrhea up to 3–5 times a day for several days, rare vomiting, and low-grade fever. Similar symptoms were observed in her mother and husband. Two weeks after the resolution of the infection, intermittent abdominal pain emerged, and weakness began to progress. Febrile fever up to 38–38.5 °C daily in the evening hours joined the symptom complex. The patient's condition progressively deteriorated, and her abdominal volume increased.

An outpatient examination was initiated. According to the abdominal ultrasound, ascites was detected, and transient elastography of the liver revealed a stiffness of 36 kPa. The patient was referred to a gastroenterologist at the regional hospital and urgently hospitalized with a diagnosis of "Portal hypertension of unspecified etiology". On physical examination, an abdomen increased in size due to ascites and slightly tense drew attention. On palpation, moderate tenderness was noted in all quadrants. Liver dimensions according to Kurlov's method were 15–12–10 cm, with a firm and smooth margin. The spleen was not palpable. The patient's height was 170 cm, and her weight was 68 kg (body weight prior to the onset of the disease was 57–60 kg). No pathological changes were detected in other organs and systems.

Laboratory test results as of May 30, 2024

Complete blood count (CBC): white blood cells (WBC) — 11.0 × 10^9/L (without a left shift in the differential WBC count), red blood cells (RBC) — 5.98 × 10^{12}/L, hemoglobin (Hb) — 140 g/L, hematocrit (Ht) — 42.1 %, platelets (PLT) — 339 × 10^9/L (by microscopy — 342 × 10^9/L).

Coagulation profile: prothrombin index (PTI) — 62 %, activated partial thromboplastin time (APTT) — 47.8 s, international normalized ratio (INR) — 1.88, D-dimer — 8.93 μg/mL, antithrombin III — 98 %, protein C activity — 92 %.

Blood biochemistry: ALT — 2,064 U/L, AST — 1,596 U/L, LDH — 1,355 U/L, ALP — 383 U/L, GGT — 126 U/L, albumin — 39.2 g/L, total protein — 69 g/L, creatinine — 70 μmol/L, urea — 5.42 mmol/L, potassium — 3.88 mmol/L, CRP — 216.6 mg/L, amylase — 24 U/L.

According to the abdominal ultrasound with duplex scanning, blood flow through the hepatic veins was not visualized, and the evaluation of portal blood flow was significantly impeded due to severe ascites and meteorism. At the same time, diffuse changes in the liver parenchyma and free fluid in the abdominal cavity with a fluid level up to 140 mm were noted.

Due to the clinical manifestations of acute hepatic vein thrombosis, anticoagulant therapy (enoxaparin sodium 60 mg subcutaneously twice daily, or 1 mg/kg body weight), antisecretory therapy (esomeprazole 20 mg/day for gastroprotection to reduce the risk of erosive and ulcerative lesions of the upper gastrointestinal tract during anticoagulant therapy), and diuretic therapy (furosemide 40 mg/day and spironolactone 100 mg/day as part of the complex management of ascites) were initiated. For the management of abdominal pain syndrome, tramadol 2 mL intramuscularly twice daily was administered. Additionally, ademetionine 800 mg/day was prescribed via intravenous drip infusion. To prevent spontaneous bacterial peritonitis, ceftriaxone 1.0 g intravenously once daily was administered.

An examination was performed to rule out autoimmune liver diseases (antinuclear factor on HEp-2, anti-liver-kidney microsomal antibodies [anti-LKM], anti-smooth muscle antibodies [anti-SMA], serum IgG, antimitochondrial antibodies [AMA-M2], M2-3E, Sp100, LC-1, SLA/LP, and SS-A/Ro-52 were negative), and viral hepatitis (anti-HAV IgM, anti-HCV, HBsAg, HBV DNA, HCV RNA, anti-HEV IgM, IgM antibodies to cytomegalovirus and Epstein–Barr virus, as well as the DNA of these viruses in blood serum were not detected). Wilson's disease (ceruloplasmin — 260 mg/L, 24-hour urinary copper excretion — 35 μg/day) and primary hemochromatosis (ferritin — 321 μg/L, transferrin saturation — 34 %, *HFE* gene mutation: H63D/H63D) were excluded. The patient was screened for paroxysmal nocturnal hemoglobinuria (PNH clone was not detected by flow cytometry) and evaluated for antiphospholipid syndrome (normal serum levels of lupus anticoagulant, IgG/IgM anticardiolipin antibodies, and anti-β2-glycoprotein 1 antibodies were verified).

During the first two days of treatment, a subjective improvement in the patient's condition was noted: abdominal pain decreased, and body temperature did not exceed 37.5 °C. Positive laboratory dynamics were also observed, with a reduction in cytolysis markers (ALT — 874 U/L, AST — 354 U/L).

On June 3, 2025, contrast-enhanced computed tomography of the abdominal organs was performed (Figs. 1, 2), confirming Budd–Chiari syndrome. Concurrently, acute thrombosis of the portal vein with the onset of collateral blood flow formation was detected. The absence of pronounced cavernous

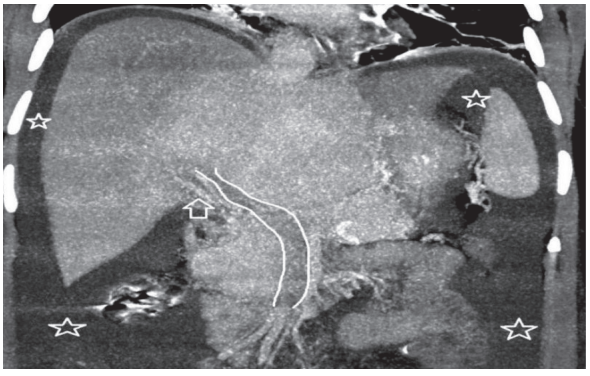

Figure 1. Contrast-enhanced computed tomography of the abdominal organs, coronal reconstruction, venous phase. Thrombosis of the distal superior mesenteric vein and the portal vein trunk (indicated by lines). Isolated small venous collaterals along the common bile duct in the stage of portal cavernoma formation (indicated by an arrow). Free fluid in the abdominal cavity (indicated by asterisks)

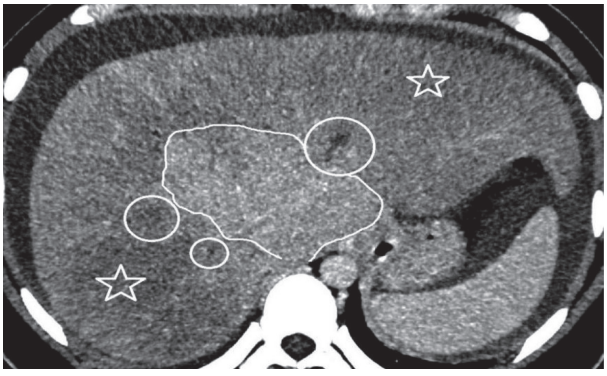

Figure 2. Computed tomography of the abdominal organs, axial reconstruction, venous phase. Pronounced diffuse changes in the liver parenchyma (indicated by asterisks). A zone of preserved perfusion in the central liver (boundaries outlined by a white line). Thrombosed main hepatic veins (indicated by circles)

transformation and the presence of only the first isolated collaterals at the time of the examination indicated the acute nature of the portal vein thrombosis.

Despite the positive laboratory dynamics, against the background of the ongoing conservative therapy, regression of ascites was not achieved, and the abdominal circumference continued to increase. It was decided to perform a diagnostic and therapeutic paracentesis with fractional evacuation of ascitic fluid and adequate replacement of the circulating blood volume deficit with a 20 % albumin solution. Initially, 6 L of fluid was removed, followed by 1.5–2 L daily via a drain. A routine ascitic fluid analysis and its cytological examination were performed: a serous character of the fluid was verified without signs of spontaneous bacterial peritonitis or atypical cells.

By the end of the first week of therapy, pronounced positive dynamics were noted, with a reduction in cytolysis markers: ALT — 464 U/L, AST — 128 U/L, as well as the CRP level — 156 mg/L. According to the CBC: white blood cells — 11.9 × 10^9/L (without a left shift in the differential count), red blood cells — 5.32 × 10^{12}/L, hemoglobin — 118 g/L, hematocrit (Ht) — 36.4 %, platelets — 387 × 10^9/L (by microscopy — 401 × 10^9/L). Coagulation profile: PTI — 68 %, APTT — 62.8 s, INR — 1.57, D-dimer — 7.36 μg/mL.

On the 12th day of enoxaparin sodium therapy, pruritus and erythema developed at the injection site. After another 12 hours, a dark cherry skin discoloration was noted in the injection area, and tenderness on palpation appeared. Evacuation of the hematoma, its sanitization, and necrectomy were performed. Treatment adjustment was made: enoxaparin was replaced with rivaroxaban at a daily dose of 20 mg for long-term use.

According to the thrombophilia marker screening results, a heterozygous C/T mutation was detected in the *ITGB3* gene (β3-integrin), which is not a clinically significant thrombophilia marker. Mutations in the *F1* (fibrinogen), *F2* (prothrombin), factor V (Leiden), *F7* (factor VII), *F13* (factor XIII), *ITGA2* (alpha-2-integrin), and *PAI-1* (plasminogen activator inhibitor-1) genes were not detected. The homocysteine level did not exceed the normal range. In the blood serum, a *JAK2* V617F mutation was detected with a 3.18 % allele burden, which led to the suspicion of a myeloproliferative disease.

One month after the initiation of treatment, abdominal pain syndrome and body temperature elevation were not observed. The ascitic syndrome persisted, requiring the evacuation of up to 2 L of fluid daily. Attempts to slowly titrate up the diuretic doses at this stage of treatment led to limb cramps despite the absence of electrolyte shifts upon laboratory examination. In view of the persistent ascites despite the therapy, the patient was consulted by vascular and endovascular surgeons. A consultation with a hepatobiliary surgeon at the A.K. Eramishantsev City Clinical Hospital was recommended to determine the surgical management strategy.

Laboratory test results as of June 30, 2024: ALT — 26 U/L, AST — 68 U/L, albumin — 31 g/L, total protein — 56.8 g/L, total bilirubin — 19.24 μmol/L, creatinine — 66 μmol/L, urea — 5.28 mmol/L, CRP — 25.5 mg/L. According to the CBC: white blood cells — 10.0 × 10^9/L, red blood cells — 4.92 × 10^{12}/L, hemoglobin — 110 g/L, hematocrit (Ht) — 30.8 %, platelets — 179 × 10^9/L (by microscopy — 214 × 10^9/L). Coagulation profile: PTI — 33 %, APTT — 84.8 s, INR — 3.06, D-dimer — 9.29 μg/mL.

The patient was discharged for further outpatient treatment and referred to a hematologist at the federal center, as well as to a portal hypertension specialist at the A.K. Eramishantsev City Clinical Hospital. The prescribed therapy included: torasemide 5 mg in the morning, spironolactone 100 mg in the morning (diuretic therapy); potassium and magnesium asparaginase, 1 tablet 175 mg 3 times daily (electrolyte deficiency replenishment during diuretic therapy); carvedilol 3.125 mg twice daily (to reduce pressure in the portal vein system); rivaroxaban 20 mg daily (anticoagulant therapy for thrombosis); ursodeoxycholic acid (UDCA) 250 mg 3 times daily (for membrane-stabilizing and choleretic purposes).

On July 22, 2024, a bone marrow trephine biopsy was performed, and the peritoneal drain was replaced due to its unsatisfactory function. The morphological pattern of the bone marrow histological examination was consistent with a myeloproliferative disease. On August 3, 2024, cytoreductive therapy with hydroxycarbamide at a dose of 500 mg/day was initiated.

On August 14, 2024, the patient was hospitalized at the A.K. Eramishantsev City Clinical Hospital. The examination revealed grade 1 esophageal varices, signs of cavernous transformation of the portal vein, thrombosis of the splenic and superior mesenteric veins, occlusion of the hepatic veins, and ascites. In the CBC, thrombocytosis — 645 × 10^9/L — drew attention for the first time. The coagulation profile and blood biochemistry parameters were as follows: INR — 1.6, PTI — 46 %, APTT — 38.1 s, D-dimer — 5.77 μg/mL, total protein — 59.4 g/L, albumin — 28.3 g/L, ALT — 24 U/L, AST — 51.3 U/L, and CRP — 13.6 mg/L. Diuretic therapy was adjusted (spironolactone 200 mg/day, torasemide 5–10 mg/day under urine output control). Rivaroxaban was replaced with apixaban 10 mg twice daily. Anticoagulant therapy was reinforced with antiplatelet therapy (ticagrelor 60 mg twice daily). Laboratory monitoring and dynamic observation were recommended to determine indications for recanalization and angioplasty.

On September 23, 2024, a follow-up visit to the A.K. Eramishantsev City Clinical Hospital took place. Contrast-enhanced computed tomography of the abdominal organs was performed (Figs. 3, 4). A decrease in ascites was noted, the sizes of the liver

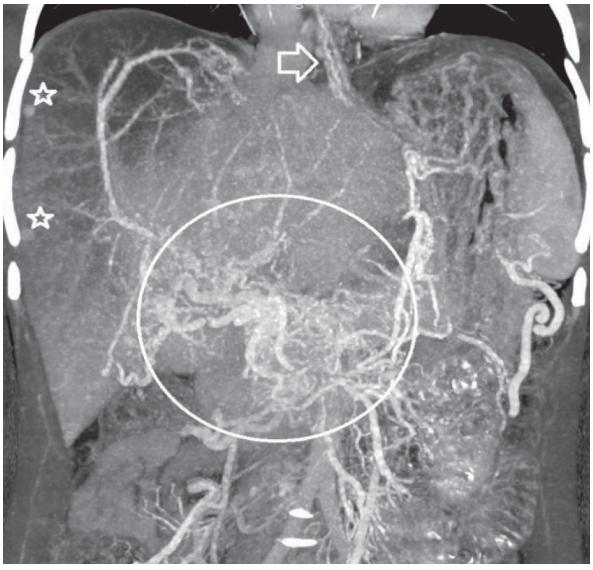

Figure 3. Computed tomography of the abdominal organs, coronal reconstruction, venous phase. Pronounced cavernous transformation of the portal vein has formed as multiple collaterals in the hepatopancreatoduodenal region. The trunk of the superior mesenteric and portal veins is occluded (indicated by a circle). Esophageal varices (indicated by an arrow). Isolated foci of focal nodular hyperplasia in the right liver lobe (indicated by asterisks)

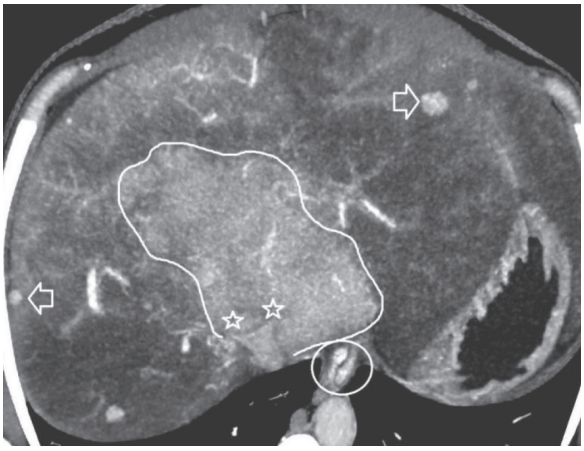

Figure 4. Computed tomography of the abdominal organs, multiplanar reconstruction, venous phase. Pronounced diffuse parenchymal changes with predominant perfusion of the central regions (outlined by a line). Several foci of focal nodular hyperplasia as part of the reparative process (indicated by arrows). Occlusion of the right and left hepatic veins (indicated by asterisks). Esophageal varices (indicated by a circle)

and spleen slightly increased, and pronounced cavernous transformation of the portal vein developed. Pronounced diffuse parenchymal changes persisted due to permanent impairment of hepatic venous outflow. Esophagogastroduodenoscopy (EGD) revealed grade 1 esophageal varices, with no changes in dynamics.

On October 3, 2024, transient elastography of the liver was performed, and the liver stiffness was 29.2 kPa. This parameter did not change significantly over time due to the lack of recanalization of the hepatic veins.

On December 12, 2024, the patient was consulted at the National Medical Research Center for Hematology of the Ministry of Health of the Russian Federation, where clinical and hematological compensation was noted. Following a review of the bone marrow trephine biopsy specimens, a definitive diagnosis of essential thrombocythemia was established.

By January 2025, a decrease in the D-dimer level and platelet count was observed. The total protein and albumin levels increased significantly after the resolution of refractory ascites and the removal of the drain. The patient is in satisfactory condition under the dynamic observation of a hematologist, a hepatologist, and a hepatobiliary surgeon (Table).

Discussion

Myeloproliferative diseases are one of the frequent causes of extrahepatic portal hypertension. According to current data, abdominal thrombosis often serves as the first (manifest) presentation of latently proceeding myeloproliferative diseases, masking classical changes in the complete blood count, which significantly impedes the timely verification of the hematological diagnosis.

Myeloproliferative diseases are the most common cause of Budd–Chiari syndrome and account for up to 50 % of all such thrombosis cases. This pathology is by no means always accompanied by a high platelet count. Sometimes their level at the onset is within the normal range, as in our case [5]. This is probably caused by consumption thrombocytopenia due to massive acute thrombosis. Subsequently, when adequate conservative therapy was selected and the thrombotic process was stabilized, the platelet count increased. When an acute abdominal thrombosis of unspecified genesis is detected in a patient, it is necessary to exclude a number of hypercoagulable disorders, such as PNH, protein C and S deficiencies, antiphospholipid syndrome, and congenital thrombophilia [6].

In the presented observation, the severity of the condition and the complexity of treatment were explained by simultaneous hepatic vein thrombosis and total thrombosis of the portal system vessels, the trigger factor of which could have been, among other

Table. Dynamics of laboratory tests from May to December 2024

Parameter	May 30	June 1	June 6	June 30	August 14	December 5	Normal range
RBC, $\times 10^{12}$/L	5.98	5.67	5.32	4.92	6.35	4.58	4.0–5.2
Hemoglobin, g/L	140	128	118	110	141	129	118–152
WBC, $\times 10^{9}$/L	11.0	10.5	11.9	10.0	7.45	7.11	3.7–9.2
Platelets, $\times 10^{9}$/L	339	341	387	179	645	238	164–380
Platelets (by microscopy), $\times 10^{9}$/L	342	-	399	-	-	-	-
D-dimer, μg/mL	8.93	8.21	7.36	9.29	5.77	2.19	0–2.43
INR	1.88	2.0	1.57	3.06	1.66	2.7	0.9–1.2
PTI, %	62	56	68	33	46	30	70–130
Antithrombin III, %	105	-	-	-	89	148	80–120
APTT, s	47.8	56.4	62.8	84.8	38.1	47.0	25.1–36.5
ALT, U/L	2064.2	874.0	464.6	26.0	24.7	53.4	0–35
AST, U/L	1596.7	354.1	128.4	68.7	51.3	42.8	0–35
GGT, U/L	126.5	104.0	134.0	112.1	142.2	188.9	0–32
ALP, U/L	383.6	362.0	301.2	241.3	93.5	105.0	30–120
Total bilirubin, μmol/L	18.1	15.2	16.72	19.24	21.5	18.8	5.0–21.0
CRP, mg/L	216.6	182.3	156	25.5	13.6	7.3	0–5
Total protein, g/L	69.0	66.3	60.1	56.8	59.4	74.6	66.0–83.0
Albumin, g/L	39.0	39.1	36.2	31	28.3	38.7	35–52
Creatinine, μmol/L	70.4	72.5	68.3	66.0	56.1	58.9	62–115
Urea, mmol/L	5.42	5.8	6.13	5.28	4.8	4.93	2.5–7.3

Note: *RBC — red blood cells; WBC — white blood cells; INR — international normalized ratio; PTI — prothrombin index; APTT — activated partial thromboplastin time; ALT — alanine aminotransferase; AST — aspartate aminotransferase; GGT — gamma-glutamyltransferase; ALP — alkaline phosphatase; CRP — C-reactive protein.*

things, the suffered acute intestinal infection. The statistics of combined thrombosis described in the world literature are scarce. According to various authors, this association occurs with a frequency ranging from 0.9 % to 18 %. However, no descriptions of a vascular lesion of a similar massive scale were found by us in the available literature [7–9].

Severe pain syndrome at the disease onset, which required the use of tramadol, was consistent with the classic clinical presentation of acute massive thrombosis of the involved vessels. A specific feature of this case is the disease onset with abdominal thrombosis at a young age, which, according to current data, frequently serves as the first manifestation of latently proceeding essential thrombocythemia. Classic factors for a high risk of thrombosis in essential thrombocythemia include an age over 60 years, the presence of a mutation in the *JAK2* gene, and a history of thrombosis at any localization. Depending on the presence of molecular genetic driver mutations, patients under 60 years of age without a history of thrombosis are classified into the low or very low risk group for thrombus formation in this pathology [10].

The patient underwent a long diagnostic and treatment journey, which began in the gastroenterology department at her place of residence and continued with multiple consultations by hematologists, hepatobiliary surgeon, and hepatologists at federal clinics. In the presented observation, the time to establish the definitive diagnosis and initiate cytoreductive therapy was more than 6 months.

Essential thrombocythemia is a disease with a high risk of thrombohemorrhagic complications [6]. In this case, long-term combined anticoagulant and antiplatelet therapy was required to achieve compensation for the ascitic syndrome. An additional difficulty was the intolerance to low-molecular-weight heparins. In a myeloproliferative disease presenting with abdominal thrombosis, lifelong anticoagulant therapy is clinically justified [6, 11].

In refractory ascitic syndrome, endovascular interventions are indicated: in the acute period — angioplasty and stenting of the hepatic veins; if they cannot be performed — transjugular intrahepatic portosystemic shunting.

One of the problems in providing care to this patient group is the low availability of endovascular technologies. There are data on a positive experience with the use of antithrombin III in the treatment of thrombotic complications of chronic myeloproliferative diseases; however, this requires further study [12].

Conclusion

In this clinical case, it was possible to achieve stabilization of the patient's condition within the framework of conservative management, without the use of open surgical interventions. However, it should be noted that in refractory cases, such patients are considered candidates for liver transplantation with the formation of a "bridge" to it in the form of transjugular intrahepatic portosystemic shunting [5].

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