A very rare case of splenosis and acquired chronic non-cirrhotic portal vein thrombosis with cavernous transformation

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Abstract
We present a clinical case of a young woman who underwent splenectomy for thrombocytopenic splenomegaly at the age of 7 years. An acute diagnostic picture of splenosis of the left epi gastric region and chronic non-cirrhotic portal vein thrombosis with cavernous transformation was found 20 years later.

Keywords
idiopathic thrombocytopenic purpura, portal hypertension, portal vein thrombosis, splenectomy, splenosis

1 | INTRODUCTION

First mentioned in the literature by Buchbinder and Lipkoff in 1939, splenosis is a condition that occurs in 16%–67% of patients after splenectomy or traumatic rupture of the spleen, with varying frequency. Abdominal splenosis is not a rare disease, as it accounts for 67% of cases after conventional splenectomy and up to 80% of cases after laparoscopic splenectomy. The latent period between the appearance and aggressive stage of splenosis is 5–40 years. The newly formed spleen tissue is morphologically organotypic and functions, to some extent, as the normal spleen. Hematological dysfunction after splenectomy is controversial because it occurs in only 7%–10% of patients. Splenosis is very limited and can cause asymptomatic thrombosis of the portal vein with its subsequent cavernous transformation.

Here, we present a rare clinical case of a young woman who underwent splenectomy at the age of 7 for idiopathic...
thrombocytopenic purpura. After 20 years, she developed thrombocytosis with an acute hemorrhagic picture. On admission to our clinic, splenosis of the left epigastric region was detected along with chronic non-cirrhotic portal vein thrombosis (NCPVT) with cavernous transformation.

2 | CASE PRESENTATION

A 29-year-old woman was admitted to the National Surgical Center, Bishkek, Kyrgyzstan, in a serious condition. On admission, she complained of pain in the epigastric region, nausea, coffee ground vomitus, and black stools. In 1999, she was diagnosed with thrombocytopenic purpura secondary to splenectomy. In 2012, she experienced an episode of esophageal bleeding and was diagnosed with moderate dilation of the esophageal veins. At the age of 13 years, the patient was admitted with a history of thromboembolic events.

Systemic examination results revealed normal findings. Likewise, laboratory examination revealed that blood chemistry and liver function test findings were within the normal range. Evaluation of venous thromboembolism includes the examination of Factor V Leiden, protein C and S deficiency, prothrombin G20210A mutation, antithrombin III deficiency, and lupus anticoagulant, all of which yielded normal results. All indicators of the clinical blood test were within the normal range, except for the findings of thrombocytosis (platelet count: $348 \times 10^3/mm^3$) and lymphocytosis (lymphocyte count: 38%). Blood clotting tests performed on July 6, 2021, also revealed normal findings. Clinical analysis of the urine did not reveal any abnormal pathology.

3 | DIFFERENTIAL DIAGNOSES, INVESTIGATIONS, AND TREATMENT

Ultrasound (US) of the abdominal organs showed no ascites in the abdominal cavity or pleural sinuses. The liver was normal size, with a smooth contour and non-compacted capsule. There was no change in its general echogenicity, and no focal changes were detected. There were no signs of recanalization. The portal vein is represented by a mixed pseudo-formation with dimensions of 9.4 × 3.5 mm (Figure 1). Power Doppler imaging revealed clearly differentiable tubular, avascular, anechoic structures. This was perceived indicating cavernous transformation of the portal vein. The diameter of the portal vein (PV) was 0.9 cm, hepatopetal undulating blood flow was noted, and the blood flow rate was 9–10 cm/s (Figure 2). The hepatic artery had a low-resistance type of blood flow, with a speed ≤0.0 cm/s. The blood flow indicators in the splenic and upper mesenteric veins did not differ from the standard.

Doppler US and computed tomography (CT) of the abdominal cavity showed no signs of free fluid collection. On CT angiography, there was a set of tiny tubular formations around the portal vein (Figure 3). Figure 4 shows CT angiography of the normal anatomical structure of the portal vein. The lymph nodes in the abdominal and retroperitoneal spaces were not enlarged.

Figure 5 shows single small parietal calcifications in the lumen of the portal and upper mesenteric veins, a consequence of thrombosis. The patient was diagnosed with a spleen implant in the left epigastric region using CT and sonography with dimensions of 1.7 × 1.2 cm (Figure 6).

In addition, ultrasound and CT angiography revealed an ectopic focus of the spleen, which was mistakenly perceived as an additional lobule of the spleen (Figure 7). This probably caused the repeated episodes of hematological issues.

The patient consulted with a hematologist and was diagnosed with chronic non-cirrhotic HTP with esophageal varicose veins of the first degree, secondary to protein C and S deficiency. Endoscopy of the upper gastrointestinal tract revealed varicose veins of the esophagus of the first degree (Figure 8). Moderate expansion of the esophageal veins was up to 2.6–2.7 mm (Figure 9, white arrow). Protein C and protein S are natural anticoagulants, and their deficiency has been described as an infrequent but probable cause of PVT, with a frequency of 11%.

Following an endoscopy of the upper gastrointestinal tract, the patient received symptomatic treatment for
nausea and vomiting. She received hemostatic therapy, which included daily aminocaproic acid of 10 mg orally for 5 days, octreotide 25 mcg/h intravenous infusion for 5 days, etamsylate 350 mg per 200 ml of isotonic sodium chloride solution infused for 5 days, and omeprazole 40 mg infusion daily for 2 weeks.

4 | OUTCOME AND FOLLOW-UP

To date, the patient’s condition is clinically stable and she is asymptomatic.

5 | DISCUSSION

The accessory spleen and splenosis should not be interchanged. The morphofunctional states of the accessory spleen have all the structural elements of the main spleen, are of congenital origin, and preserve the arterial blood

FIGURE 2 In the mode of spectral dopplerography, hepatopetal monophasic laminar blood flow with reduced velocity indicators of 9.7 cm/s is determined.

FIGURE 3 On CT angiography, there is a violation of the architectonics of the portal vein (white arrow) in the form of a set of tiny tubular formations around the portal vein (black arrow).

FIGURE 4 For clarity, CT angiography of the normal anatomical structure of the portal vein is shown. Portal vein in the portal phase.

FIGURE 5 Single small parietal calcifications in the lumen of the portal and upper mesenteric veins (white arrows), a consequence of thrombosis.
supply to the splenic artery. However, splenosis receives its vascularization from the surrounding tissue and does not have its own vessels, and sustained hypoxia is associated with poor development of white pulp. Contrast-enhanced CT scan showed the ectopic spleen implant fixed to the parietal peritoneum without feeding vessels. Depending on the location of the ectopic splenic tissue, splenosis can originate from different organs and simulate malignant neoplasms, peritoneal metastases, abdominal lymphadenopathy, and endometrioid heterotopias. The size of splenosis can vary from a few millimeters to 12 cm. In our patient, foci of splenosis were detected in an area measuring 1.7 × 1.2 cm.

Acquired splenosis plays an active role in the prevention and correction of the manifestations of hyposplenism and postsplenectomy syndrome. Splenosis is a normal and functionally necessary condition for the body. Morphologically, the foci of splenosis are characterized by a poorly formed white pulp in the normal state of the red pulp, a weakly expressed capsule, and the absence
of trabeculae and gates. The microstructure of the foci is identical to that of a normal spleen. Patients who have undergone splenectomy have impaired immune function, and they even have foci of splenosis. Despite the relatively acceptable vascularization, the total amount of filtered blood is sharply reduced. The microanatomy of the regenerative nodes of the spleen is probably unable to provide close contact between the antigen, phagocytes, and immune cells that are characteristic of a normal spleen.

The recurrence of hematological symptoms in patients who have undergone splenectomy indicates the presence of splenosis. The hematological symptoms in our patient resumed at the age of 20 years, 13 years after the procedure. The probability that a pseudotumor focus in porta hepatitis or thrombosis of the PV trunk was not detected during a detailed examination using various imaging methods was minimal.

Portal vein thrombosis (PVT) is a serious complication of splenectomy and splenosis. Its frequency ranges from 7% to 10% and is confirmed by duplex US. Portal vein thrombosis risk factors after splenectomy are controversial. The blood reflux along the splenic vein practically disappears, and the pressure and blood flow in the PV decrease by 20%–35%. If the blood flow in the PV decreases sharply in the short term, eddies or other hemodynamic disorders can easily form in the PV, which is more likely to lead to the formation of a blood clot. In addition, after splenectomy, the distal end of the splenic vein becomes a dead end, which contributes to blood retention, and the splenic vein thrombus tends to spread to the PV trunk. Although devascularization blocks collateral blood flow, it is insufficient to compensate for PV blood loss after splenectomy. The blood flow rate in the PV usually decreases to 12.13 ± 2.59 cm/s; in our case, according to Doppler US, it decreased to 9.7 cm/s, causing portal hypertension (PHT).

Portal vein thrombosis is described as a rare but potentially fatal complication after splenectomy, but in some cases, spontaneous resolution of PVT may be observed if patients received anticoagulant therapy. A thorough medical history of our patient’s illness did not confirm the use of anticoagulant therapy.

Acute PVT includes abdominal pain, fever, nausea, vomiting, and ascites; episodes of gastrointestinal bleeding are often observed in patients with the background of chronic PVT. According to some studies, PVT can also be asymptomatic. It is diagnosed when there are complications such as varicose bleeding. Occasionally, bleeding can occur decades after splenectomy and PVT. A blood test can show leukocytosis and thrombocytosis, which practically coincides with the anamnesis of our patient. This was possibly achieved because of the formation of peri-portal collaterals. Cavernous transformation, as a protective reaction, manifests in the form of numerous sinuous vascular cavities of various sizes occupying the bed of the PV, which presumably can eliminate PHT.

In our case, the patient was in the acute stage of the disease from the first consultation. Gastrointestinal bleeding, according to clinical and laboratory findings, describes hypovolemia and anemia.

During repeated consultations with hematologists, surgeons, and radiologists, we decided to focus on the diagnosis of splenosis or postsplenectomy thrombosis of the PV complicated by cavernous transformation of the PV and prehepatic vein, in which the systemic examination revealed normal findings. The exclusivity of this clinical case requires further dynamic observations.

ACKNOWLEDGMENT
The acknowledgment of this study was published with the written consent of the patient.

CONFLICT OF INTEREST
The authors declare no conflicts of interest.

AUTHOR CONTRIBUTIONS
Aliya Kadyrova and Elmira Mamytova contributed to conception, design of the work, clinical management, manuscript preparation, and data acquisition. Iliar Baudinov contributed to conception, manuscript preparation, and data acquisition. Kubat Ibraimov, Tynarbek Arzykulov, Sagynali Mamatov, Yethindra Vityala, and Tugolbai Tagaev contributed to manuscript preparation and data acquisition.
CONSENT
The patient gave her informed consent prior to her inclusion in the study.

DATA AVAILABILITY STATEMENT
Data are available from the corresponding author upon reasonable request.

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