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Antiphospholipid Syndrome as a Cause of Recurrent Portal Vein Thrombosis in a Man with a Medical History of other Thrombosis

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Aim: to present a clinical case of antiphospholipid syndrome (APS) manifested as recurrent portal vein thrombosis in a man with a medical history of other thromboses.
Key points. APS is a syndrome that includes venous and/or arterial thrombosis, various forms of obstetric pathology, thrombocytopenia, as well as a variety of neurological, skin, cardiovascular and hematological disorders. The article presents a clinical case of a male patient with several episodes of vascular thrombosis, two of which (thrombophlebitis of the veins of the lower extremities and stroke) developed at the age of 39 years, and recurrent thrombosis of the portal vein since 2018. For several years, the patient had the diagnosis of cirrhosis based on the presence of signs of portal hypertension. However, the patient's liver function remained practically intact all the time. During the current examination, APS was suspected, and the result of a blood test for antibodies against phospholipids was positive.
Conclusion. This clinical case of APS reflects the difficulties in diagnosing this disease. It should be remembered that cases of thrombosis at a young age may be due to APS.
Key words: antiphospholipid syndrome, portal hypertension, portal vein thrombosis
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Антифосфолипидный синдром как причина рецидивирующего тромбоза воротной вены у мужчины с другими тромбозами в анамнезе

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Цель исследования: представить клиническое наблюдение антифосфолипидного синдрома (АФС), протекающего как рецидивирующий тромбоз воротной вены у мужчины с другими тромбозами в анамнезе.
Основные положения. АФС — это симптомокомплекс, включающий в себя венозные и/или артериальные тромбозы, различные формы акушерской патологии, тромбоцитопению, а также разнообразные неврологические, кожные, сердечно-сосудистые и гематологические нарушения. В статье приведен клинический случай пациента с несколькими эпизодами сосудистых тромбозов, два из которых (тромбофлебит вен нижних конечностей и острое нарушение мозгового кровообращения) имели место в возрасте 39 лет, а рецидивирующий тромбоз воротной вены беспокоит пациента с 2018 г. В течение нескольких лет пациенту выставляли диагноз «цирроз печени» на основании наличия признаков портальной гипертензии, однако функция печени у пациента все время оставалась практически сохранной. Во время текущего обследования был заподозрен АФС, результат анализа крови на антитела к фосфолипидам был положительным.
Заключение. Представленное клиническое наблюдение отражает трудности диагностики этого заболевания. Необходимо помнить о том, что случаи тромбозов в молодом возрасте могут быть обусловлены АФС.

Ключевые слова: антифосфолипидный синдром, портальная гипертензия, тромбоз воротной вены
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Antiphospholipid syndrome (APS) is a symptom complex that includes venous and / or arterial thrombosis, various forms of pregnancy pathology, thrombocytopenia, as well as a variety of neurological, skin, cardiovascular and hematological disorders. The prevalence of APS is 20 to 50 cases per 100,000 people [1–5].

Patient S., 51 years old, was admitted to the gastroenterology department with complaints of constant pain in the abdomen, mainly in the umbilical region; an increase in the volume of the abdomen; frequent loose stools 4–5 times a day without pathological impurities.

Medial history. In 2018, there were pains in the abdomen for the first time. Magnetic resonance imaging (MRI) was performed, where carcinomatosis of the abdominal cavity was suspected. In the oncological department, diagnostic laparotomy and peritoneal biopsy revealed symptoms of portal hypertension, varicose veins of the esophagus and mesentery of the small intestine, data for peritoneal carcinomatosis were not received. In May 2019, he was admitted to the surgical department with complaints of weakness, nausea, coffee ground vomiting, black feces. The diagnosis was made: "Non-cirrhotic portal hypertension as the outcome of portal vein thrombosis: esophageal varices (EV) of 3rd Grade". An operation was performed: endoscopic azigo-portal separation of esophageal varicose veins. There was a repeated bleeding from EV in November 2019. The patient was hospitalized in the surgical department with the diagnosis: "Cirrhosis: Child-Pugh C class". The last attack occurred in November 2020. There are an increase in the abdomen in volume and frequent loose stools up to 4 times a day with an admixture of mucus. The patient was admitted to the gastroenterological department of the clinical hospital.

Patient's father died at a young age in a car accident. Patient's mother died at the age of 49 from breast cancer. He did not take alcohol for the last two years, but took it 1–2 times a month in small quantities before. He quit smoking two years ago. He has disability due stroke since 2009. Prior to disability, he worked as a carpenter and electrician. The patient had also cholecystectomy for acute calculous cholecystitis in 1987 and thrombophlebitis of the right lower extremities in 2009.

Physical examination. Height was 172 cm, weight was 72 kg, BMI = 24 kg/m^2. The general condition of the patient is satisfactory, consciousness is clear. Skin and visible mucous membranes had physiological color. Peripheral lymph nodes were not enlarged. There was no edema. There is asymmetry of the lower extremities as a result of thrombophlebitis (Fig. 1).

Respiratory system was without features, the frequency of respiratory movements was 17 per minute. Breathing was vesicular, there was no wheezing.

Cardiovascular system. The precardial area was not altered. The boundaries of the heart are within normal limits. Heart sounds was normal. Heart rate was 72 per minute, blood pressure was 120/80 mm Hg.

Digestive system. The tongue was clean. The abdomen was soft, painless, asymmetrical. The liver was enlarged slightly, the size according to Kurlov's method was 11 × 9 × 8 cm. Spleen was enlarged: length was 7 cm, diameter was 8 cm.

The system of urinary organs was without feature.

The following laboratory and instrumental studies were carried out (normal indicators are indicated in parentheses).

Complete blood count. Red blood cells: 4.7 × 10^{12}/l ((3.7–4.7) × 10^{12}/l), hemoglobin: 112 g/l ((120–140) g/l), platelets: 96 × 10^9/l (200–400 × 10^9/l); white blood cells: 5.3 × 10^9 ((4.0–9.0) × 10^9/l), eosinophil: 2 % (0–5 %), band neutrophils:

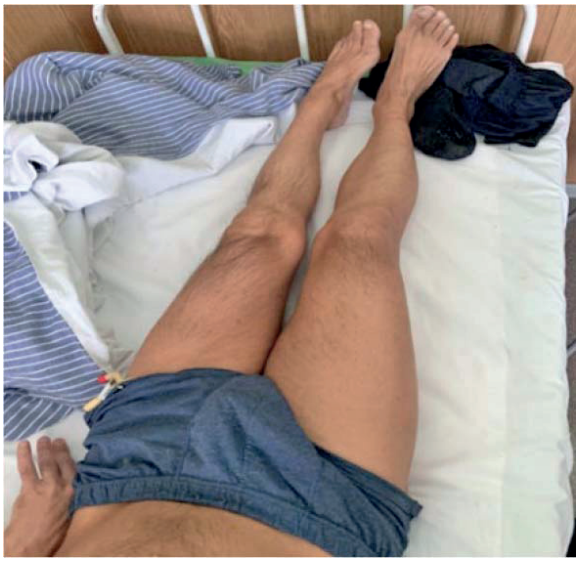

Fig. 1. Asymmetry of the lower extremities of patient S. as a result of thrombophlebitis on the right in 2009

2 % (1–6 %), segmented neutrophils: 80 % (47–72 %), lymphocytes: 8 % (18–38 %), monocytes: 8 % (3–11 %), and ESR: 16 mm/h (2–15 mm/h).

Biochemical blood test. glucose: 5.9 mmol/l (3.5–5.5 mmol/l, total bilirubin: 36.1 umol / l (5–21 umol/l), direct bilirubin: 10.0 umol/l (up to 3.4 umol/l), indirect bilirubin: 26.1 umol/l (1.7–17.0 umol/l), alanine aminotransferase (ALT): 25 U/l (<40 U/l), aspartate aminotransferase (ACT): 24 U/l (<40 U/l), alkaline phosphatase (ALP): 62.9 U/l (30–120 U/l), gamma-glutamyltransferase (GGT): 65.4 U/l (up to 55 U/l), urea: 5.0 mmol/l (2.8–7.2 mmol/l), creatinine: 67 umol/l (59–104 umol/l), amylase: 56.3 U/l (22–80 E/l), total protein: 71.2 g/l (66–83 g/l), albumin: 37.9 g/l (35–52 g/l), calcium ionized: 1.15 mmol/l (1.05–1.3 mmol/l), potassium: 3.80 mmol/l (3.5–5.5 mmol/l), and sodium: 143 mmol/l (135–155 mmol/l).

Clot tests. Activated partial thromboplastin time: 38.3 s. (21–35 s), prothrombin index: 76.5 % (70–130 %), international normalized ratio: 1.15 (0.8–1.2), and fibrinogen: 2.0 g/l (2–4 g/l).

Microreaction for syphilis was negative. No antibodies to HIV were detected. HBsAg and antibodies to HCV had not been detected.

Abdominal ultrasound. Liver: right lobe was 120 mm, left lobe was 65 mm. The contours are smooth. Echogenicity is increased. Echostructure is somewhat heterogeneous. Choledoch is not clearly visualized. Pancreas: not enlarged, contours blurred, echogenicity increased, echostructure diffusely non-homogeneous. Spleen vein: 6 mm. Portal vein: 6 mm, lumen heterogeneous, filled with isoechoic contents. Spleen: dimensions 135×65 mm, contours uneven, structure homogeneous. Free fluid was detected in the small pelvis in a small amount (Fig. 2).

Abdominal computed tomography. Liver with clear smooth contours, homogeneous structure with an expanded portal vein. Intra- and extrahepatic bile ducts are not dilated. The pancreas is not enlarged. The structure is homogeneous. Spleen with clear even contours, enlarged (dimensions 154×73 mm), homogeneous density.

Esophagogastroduodenoscopy: EV of 3 grade.

ECG: sinus rhythm with a heart rate of 70 beats per minute; vertical electrical heart axis; disordered repolarization process in the V3 lead (Fig. 3).

The test for anticardiolipin antibodies was positive.

The clinical diagnosis has been established.

Main disease:

Antiphospholipid syndrome: recurrent thrombosis of the portal vein, thrombophlebitis of the veins of the right lower extremity from 2009, stroke in the basin of the right middle cerebral artery from 2009.

Complications:

Extrahepatic portal hypertension: EV of the 3rd grade with frequent bleeding, ascites, splenomegaly, thrombocytopenia. Edoscopic azigo-portal separation of EV in 2019.

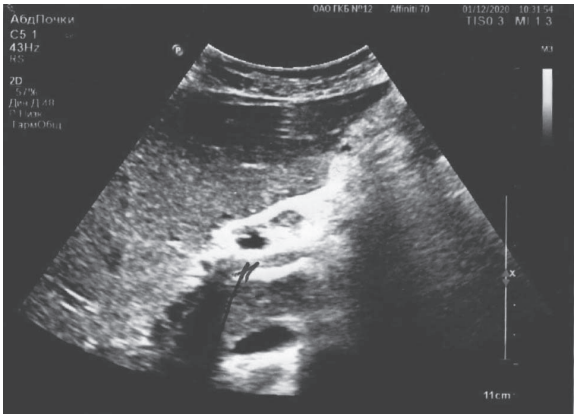

Fig. 2. Abdominal ultrasound of patient S. The lumen of the portal vein is non-homogeneous, filled with isoechoic content (thrombosis)

Post-thrombophlebic syndrome of the veins of the right lower limb.

Concomitant diseases: Condition after cholecystectomy in 1987.

The patient was prescribed anticoagulants for lifetime admission. At follow-up, thrombosis did not recur.

Discussion

This article presents a clinical case of portal vein thrombosis complicated by splenomegaly and thrombocytopenia, with bleeding from EV. Given the fact that the patient at a young age suffered thrombophlebitis of the lower extremities and stroke, the APS was suspected. The patient was advised to take a blood test for antibodies to phospholipids, which turned out to be positive.

According to the literature, the main clinical manifestations of APS are: venous thrombosis, obstetric pathology, hematological complications (thrombocytopenia, hemolytic anemia, false positive Wasserman reaction), skin manifestations (livedo reticularis, lower leg ulcers), heart lesions (valvular pathology, myocardial infarction, intracardiac thrombosis), neurological disorders (stroke) [1–5].

The etiology of this disease still remains unclear [6].

Diagnosis of APS begins with a thorough history taking, it is necessary to clarify the presence of thrombosis and obstetric pathology in the relatives, the presence or absence of acquired risk factors for thrombosis (trauma, surgery, long air travel, etc.). Given the diversity of the clinical picture, the patient's examination should be directed to the diagnosis of signs of the disease associated with ischemia or thrombosis of various organs and systems, the search

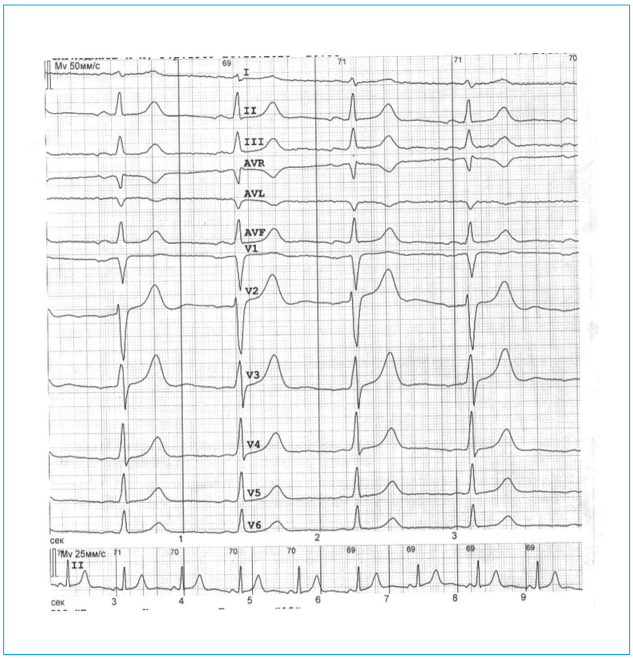

Fig. 3. Electrocardiogram of patient S.

for the underlying disease that can contribute to the development of APS [1, 3, 4].

The diagnosis of APS is established if the patient has at least one clinical (vascular thrombosis and / or patology of pregnancy) and one laboratory criterion (lupus anticoagulant and / or antibody to cardiolipin and/or b2-glycoprotein) [7, 8].

Our patient met these diagnostic criteria for API because vascular thrombosis and antiphospholipid antibodies were diagnosed. This clinical case reflects the difficulties of diagnosing and managing such patients. It must be remembered that cases of thrombosis, both venous and arterial, at a young age can be caused by APS. Our patient had three episodes of vascular thrombosis: two episodes at the age of 39 years (thrombophlebitis of the vessels of the lower extremities and stroke) and the third episode (portal vein thrombosis) occurred in 2020. Several cases of thrombosis, both venous and arterial, at a young age may be due to APS.

A thorough history taking and a comprehensive examination of the patient allows you to make the correct diagnosis, prescribe timely therapy and reduce the risk of complications.

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