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Case Report / *Olgu Sunumu*

A rare cause of abdominal pain: Portal vein thrombosis

Nadir bir karın ağrısı nedeni: Portal ven trombozu

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ABSTRACT

Portal vein thrombosis (PVT) is the blockage or narrowing of the portal vein by a thrombus, with or without extension to other segments of the splanchnic venous system. It is relatively rare and is linked to the presence of an underlying liver disease or prothrombotic disorders. Currently, PVT is being increasingly diagnosed due to advances in modern imaging techniques. The clinical presentation has a wide range, from an asymptomatic lesion to a potentially life-threatening situation. In this article, we present a young female patient who presented with abdominal symptoms. Imaging revealed the complete occlusion of the portal and splenic veins. The presence of PVT should be considered as a guideline for prothrombotic disorders, liver disease and other local and general factors that need to be investigated carefully. We hope that this case report will help increase awareness of the complexity associated with PVT among the medical community.

Keywords: Abdominal pain; anticoagulant treatment; portal vein thrombosis.

ÖZ

Portal ven trombozu (PVT), splanknik venöz sistemin diğer segmentlerine uzanımı olsun veya olmasın portal venin trombüsle tıkanması veya daralmasıdır. Nispeten nadirdir ve altta yatan bir karaciğer hastalığı veya protrombotik bozuklukların varlığına bağlıdır. Günümüzde, modern görüntüleme tekniklerindeki gelişmeler nedeniyle PVT'ye giderek daha fazla tanı konulmaktadır. Klinik prezentasyonu, asemptomatik bir lezyondan potansiyel olarak yaşamı tehdit eden bir duruma kadar geniş bir yelpazeye sahiptir. Bu yazıda, abdominal semptomlar ile başvuran genç bir kadın hasta sunuldu. Görüntüleme portal ve splenik venin total tıkanıklığını ortaya koydu. Portal ven trombozunun varlığı; protrombotik bozukluklar, karaciğer hastalığı ve dikkatle araştırılması gereken diğer lokal ve genel faktörler için bir kılavuz olarak düşünülmelidir. Bu olgu sunumunun tıp toplumunda PVT ile ilişkili karmaşıklıkla ilgili bilinci artırmaya yardımcı olacağını umuyoruz.

Anahtar sözcükler: Karın ağrısı; antikoagülan tedavi; portal ven trombozu.

Portal vein thrombosis (PVT) is blockage of the portal vein, with or without adding to other segments of the splanchnic venous system (splenic vein or superior mesenteric vein). However, the term does not include isolated thrombosis of splenic or superior mesenteric vein.[1]

Nowadays, PVT is detected more frequently due to improvements in modern imaging techniques. The clinical spectrum varies from asymptomatic presentation to life-threatening conditions. Patients may present with various clinical manifestations ranging from vague abdominal pain to sepsis due to ischemic necrosis- associated perforation.[2] We here report a rare case of PVT, which happened due to factor V Leiden (FVL), methylenetetrahydrofolate reductase (MTHFR) and plasminogen activator inhibitor-1 (PAI-1) mutation.

CASE REPORT

A 32-year-old female presented to the emergency department with abdominal pain, nausea and vomiting. She described the abdominal pain as constant cramping, located on the left upper side of the abdomen with an intensity

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of 6 to 7 out of 10. Associated symptoms reported were nausea and vomiting for one day. She denied any fever, chest pain, shortness of breath, constipation, dizziness, muscle weakness or numbness. The patient's past medical history was free. No surgical history was reported. She was a smoker and often abused alcohol.

Upon physical examination, the patient was afebrile and she had diffuse abdominal tenderness with mild guarding but no overt signs of peritonitis. Bowel sounds were present but hypoactive. The vital signs were stable, except for slight tachycardia with a heart rate of 112. No abnormal coloration was noted in the eyes or on the skin. No murmurs or additional heart sounds were noted. The lungs were clear to auscultation. The abdomen was soft and nondistended but was diffusely tender to palpation, especially in the left upper quadrant. There was 1+ pretibial edema present bilaterally and the pulses were present. Neurological exam was intact. Laboratory findings were as follows: white blood cell count 5,700/mm^3 with normal differential, hemoglobin 10.8 g/dL, and platelet count 276,000/mm^3. Liver function tests, total protein, albumin, globulin, prothrombin time, and activated partial thromboplastin time were within normal limits. Computed tomography (CT) and Doppler ultrasonography (US) revealed PVT, large collateral veins and diffuse peritoneal fluid in the abdominal cavity (Figure 1, 2). The patient was admitted with a diagnosis of portal vein thrombosis and was started on anticoagulation with enoxaparin bridged with warfarin.

A complete hypercoagulability workup was completed. Polymerase chain reaction analysis documented heterozygosity for FVL and MTHFR 677C/T mutation, homozygote for PAI-1. Antithrombin III level protein C and S levels were within normal limits. Liver disease was also excluded, with all the liver disease workups being unremarkable. Patient's pain was relieved with analgesic. Her hospital course was uncomplicated, with significant improvement of her pain and the ability to tolerate a regular diet without any gastrointestinal symptoms, such as nausea, diarrhea or constipation. She was extensively counseled on the need to be compliant with her anticoagulation regimen to prevent the recurrence of thrombosis and was uneventfully discharged with appointments to follow at the medicine and warfarin clinics. She was using warfarin before discharge home with a target international normalized ratio (INR) of 2 to 3. After three weeks, the violence and frequency of the patient's pain and nausea had decreased significantly. She was completely asymptomatic by two-month post discharge.

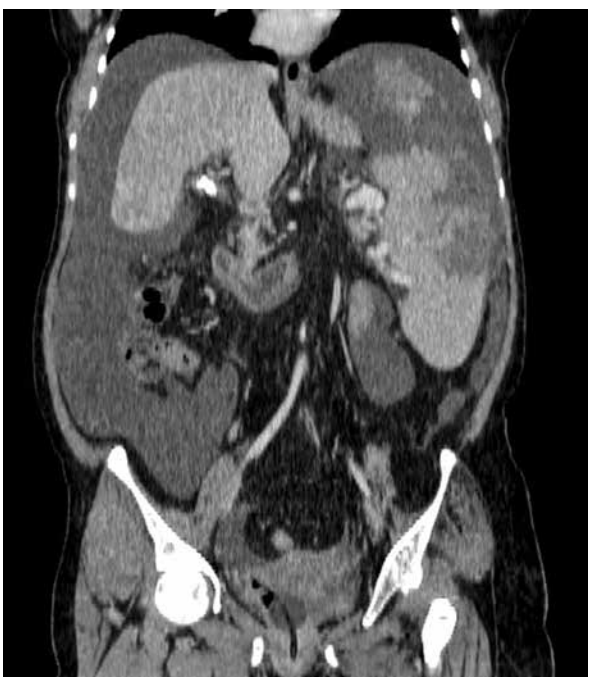

Figure 1. Axial computed tomography image shows portal and splenic thrombosis with diffuse peritoneal fluid.

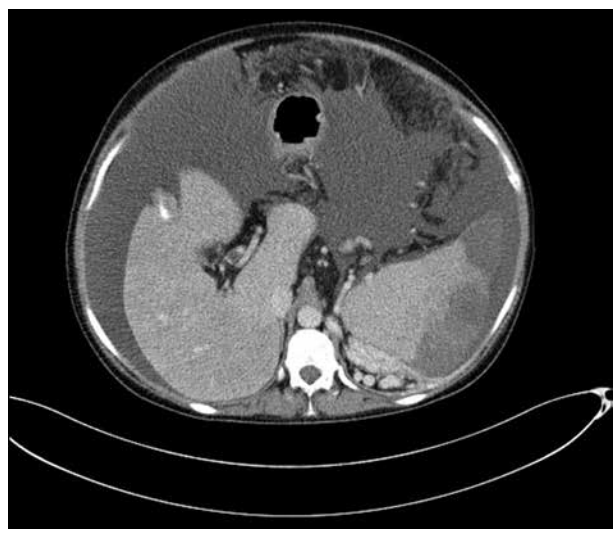

Figure 2. Coronal computed tomography image shows portal and splenic thrombosis with splenic infarction, diffuse peritoneal fluid.

DISCUSSION

Portal vein thrombosis affects about 1% of the general population. According to the severity of cirrhosis in patients this ratio is between 0.6-16% and the incidence is higher in patients with liver cancer.[2] Prognosis is good in non-cirrhotic patients without associated neoplastic disease; with an overall mortality of <10% due to early diagnosis, development of antibiotics, early surgical intervention and use of anticoagulation.[3] Our patient physical examination and laboratory findings were compatible with non-cirrhotic.

Portal vein thrombosis refers to the partial or complete thrombosis of the portal vein trunk, including its right and left intrahepatic branches. The PVT is a rare disease but it is becoming more frequently diagnosed because of the increasing use of high resolution imaging methods such as CT, magnetic resonance imaging and US. Portal vein thrombosis is clinically important because it can be associated with a severe sequelae.[4] There were both splenic and portal vascular thrombosis in our patient's tomographic findings.

Portal vein thrombosis has been declared in poly settings, including inflammatory states, malignancy, trauma, portal hypertension, post splenectomy, mechanical compression such as volvulus or pregnancy, hematological hypercoagulable states and oral contraceptive use.[5] The etiology of portal venous thrombosis is related to the prothrombotic state and other local factors.[6] Protein C and Protein S deficiencies, antithrombin III deficiency, FVL, Factor II gene mutations are hereditary prothrombotic disorders associated with PVT. Acquired prothrombotic disorders include anti-phospholipid syndrome, paroxysmal nocturnal hemoglobinuria, and other inflammatory conditions. Splenectomy, abdominal surgery, abdominal trauma, infection and localized inflammatory lesions such as pancreatitis and cholecystitis are local factors associated with the development of PVT. Malignancy is also 21 to 24% incidence of PVT associated with neoplasm due to prothrombotic changes, direct tumor invasion and local tumor mass effects.[3,6] Our patient's hypercoagulability workup is very rare. Polymerase chain reaction analysis documented heterozygosity for FVL and MTHFR mutation, homozygote for PAI-1.

In our case, FVL heterozygote mutation is a rare cause of portal venous thrombosis and it is a hereditary clotting disorder resulting from substitution of arginine with glutamine at position 506 in the Factor V gene. It is detected in 3-12% of healthy individuals worldwide and in 9% of healthy individuals in our country.[7]

Clinical findings depend on the prevalence of thrombosis, onset rate and the development of compensatory collateral vessels. Patients may be asymptomatic or present with life-threatening complications. Common symptoms include abdominal pain, vomiting, fever, anorexia and nausea, whereas more severe cases may manifest with mesenteric ischemia, infarction and peritonitis.[6] Our patient presented with abdominal symptoms consistent with the literature.

Conclusion

As a result, in non-specific abdominal pain, PVT should be considered as part of the differential diagnosis. The presence of PVT should be considered as a clue in prothrombotic disorders, liver disease and other local and general factors that need to be investigated carefully. It is hoped that this case report will help to raise awareness of the complexity associated with portal venous thrombosis among the medical community.

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