Case 12715

Essential thrombocythaemia: abdominal manifestations
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Section: Abdominal imaging
Area of Interest: Portal system / Hepatic veins
Procedure: Diagnostic procedure
Procedure: Thrombolysis
Imaging Technique: CT
Imaging Technique: Ultrasound
Special Focus: Haematologic diseases Case Type: Clinical Cases
Authors: Tonolini Massimo, MD.
Patient: 29 years, female

Clinical History:
A young female patient with unremarkable medical history sought attention at the emergency department due to severe malaise and bilateral lower extremity swelling for two weeks. Physical examination revealed enlarged painless spleen without palpable superficial lymphadenopathies. Laboratory assays disclosed microcytic anaemia (haemoglobin 6.2 g/dL, mean corpuscular volume 65 fL) and increased platelet count (950.000/mmc).

Imaging Findings:
Initially colour Doppler ultrasound (not shown) excluded venous thrombosis in the legs. Requested to assess splenic enlargement, inferior vena cava, deep lymph nodes or masses, multidetector CT (Fig. 1) showed patent portal and superior mesenteric veins, homogeneous splenomegaly with a limited infarct, absent splenic vein, extensive perisplenic, perigastric and periduodenal venous collaterals, and gastric wall varices. Bone marrow biopsy (megakaryocytic hyperplasia without blasts) diagnosed essential thrombocythaemia with positive Janus-Kinase2 gene mutation.

One year later, splenectomy was performed to relieve tender splenomegaly. Eighteen months after surgery, the patient complained of recurrent epigastric pain with persistently elevated (600.000/mmc) platelet count. Ultrasound (Fig. 2) and CT (Fig. 3) showed massive acute thrombosis of portomesenteric system and intrahepatic portal branches with transient geographic attenuation differences in the liver parenchyma, signs of cavernomatous portal transformation and minimal ascites.

After anticoagulation, repeated colour Doppler (Fig. 4) confirmed restored patency of the portal vein with normal hepatopetal flow, and the patient started alfa-interferon therapy.

Discussion:
Uncommon in the general population, essential thrombocythemia (ET) is the commonest (0.77-2.53/100.000 estimated annual incidence) clonal myeloproliferative disorder, which occurs in adults (with a 2:1 female predominance) and is characterized by sustained platelet count elevation (over 600.000/mmc) and increased megakaryocytes at bone marrow biopsy. Janus-Kinase (JAK2) gene mutation is present in approximately half of patients [1-3]. Almost 30% of patients are asymptomatic, others suffer from vasomotor symptoms (headache, dizziness, visual changes, paresthesias, erythromelalgia). Present in 20-30% of cases, splenomegaly is an
independent source of morbidity and detriment to quality of life causing bloating, early satiety, painful episodes of splenic infarction, or even portal hypertension. Required for refractory symptoms or splenic sequestration exacerbating cytopenias, splenectomy does not modify the life expectancy. Transformation into myelofibrosis, myelodysplasia or acute leukemia represents a rare, late event [1-5].

Despite a generally indolent course, ET may be complicated by acute vascular events which represent the leading causes of morbidity and mortality. The rare hemorrhages mostly affect patients with very high platelet counts. Conversely, thrombosis involving the cerebrovascular, coronary and peripheral circulation may manifest at diagnosis or in the preclinical phase (even at relatively low platelet counts) with an overall 6.6% patients/year risk. Particularly after splenectomy, spleno-portal-mesenteric system thrombosis (SPM-T) represents a well-recognized (4% prevalence) complication of ET. Manifesting with nonspecific abdominal pain, SPM-T has a non-negligible mortality and should be promptly recognized and treated with low-molecular-weight heparins followed by long-term anticoagulation. SPM-T may lead to portal hypertension, digestive bleeding or hypersplenism. Haematopoietic stem cell disorders represent the leading (30-40% of cases) cause of splanchnic vein thrombosis [4-6]. Colour Doppler ultrasound may recognize SPM-T with variable echogenicity and absent flow within the thrombosed segment. At CT, acute thrombus in the portal, superior mesenteric, or splenic vein appears as intraluminal hyperattenuation with possible venous enlargement on precontrast images, with corresponding filling defect partially or totally occluding the vessel lumen after intravenous contrast. Arterial-phase transient attenuating differences in the liver parenchyma are commonly noted as accessory signs. Features of portal hypertension including portosystemic collaterals may be present [7-9].

Patients with ET are stratified into high, intermediate and low risk according to age, more or less elevated platelet count, other cardiovascular risk factors or thrombophilia, and history of bleeding or thrombosis. According to risk class, treatment of ET may include antiaggregation (aspirin), platelet-lowering therapies (hydroxyurea, anagrelide and alfa-interferon) or cytoreductive drugs [1, 2].

**Differential Diagnosis List:** Essential thrombocythaemia with portal hypertension. Portomesenteric venous thrombosis after splenectomy., Portal hypertension in chronic liver disease / cirrhosis, Lymphoproliferative disease / Lymphoma, Other myeloproliferative disorders, Sepsis / Intra-abdominal inflammation, Oral contraceptive therapies

**Final Diagnosis:** Essential thrombocythaemia with portal hypertension. Portomesenteric venous thrombosis after splenectomy.

**References:**

Briere JB (2007) Essential thrombocythemia. Orphanet J Rare Dis 2:3 (PMID: 17210076)


Radiographics 22:141-159 (PMID: 11796904)
Description: Axial (a,b in craniocaudal order) and coronal (c) postcontrast images showed homogeneously enhancing splenomegaly, absent splenic vein with enlarged tortuous perigastric venous collaterals (arrows) and gastric wall varices (thin arrows in b). Origin: Tonolini Massimo, Department of Radiology, "Luigi Sacco" University Hospital – Milan (Italy)
Description: Axial (a,b in craniocaudal order) and coronal (c) postcontrast images showed homogeneously enhancing splenomegaly, absent splenic vein with enlarged tortuous perigastric venous collaterals (arrows) and gastric wall varices (thin arrows in b). Origin: Tonolini Massimo, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
**Description:** The enlarged spleen showed a focal hypovascular region (arrowhead) at the upper pole, consistent with limited infarct. Note enlarged tortuous perisplenic, perigastric and periduodenal venous collaterals (arrows). **Origin:** Tonolini Massimo, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
**Description:** Panoramic coronal maximum intensity projection (MIP) reconstruction showed patent portal and superior mesenteric veins, absent splenic vein, extensive venous collaterals (arrows) and gastric wall varices (thin arrows). **Origin:** Tonolini Massimo, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
Description: Axial thick-slab maximum intensity projection (MIP) reconstruction showed patent portal vein, absent splenic vein, venous collaterals (arrow) and gastric wall varices (thin arrows). Origin: Tonolini Massimo, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
Description: Axial (a..c) and coronal (d..f) postcontrast images showed nonenhancing thrombosed lumen of the portal confluence and trunk (arrows), intrahepatic portal branches (thin arrow) and superior mesenteric vein (arrowheads). **Origin:** Tonolini Massimo, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
Description: Note dilated, thrombosed superior mesenteric vein (arrowhead), signs of cavernomatous portal transformation (short arrow), transient geographic attenuation differences in the liver parenchyma (*). Origin: Tonolini Massimo, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
**Description:** Note dilated, thrombosed superior mesenteric vein (arrowhead), transient geographic attenuation differences in the liver parenchyma (*). **Origin:** Tonolini Massimo, Department of Radiology, "Luigi Sacco" University Hospital – Milan (Italy)
Description: Note nonenhancing thrombosed lumen of the portal confluence and trunk (arrow), signs of cavernomatous portal transformation (short arrow), transient geographic attenuation differences in the liver parenchyma (*). Origin: Tonolini Massimo, Department of Radiology, "Luigi Sacco" University Hospital – Milan (Italy)
**Description:** Note nonenhancing thrombosed lumen of the portal vein, intrahepatic branches (thin arrows) and superior mesenteric vein (arrowhead), transient geographic attenuation differences in the liver parenchyma (*). **Origin:** Tonolini Massimo, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
Description: Note nonenhancing thrombosed lumen of the intrahepatic portal branches (thin arrows), transient geographic attenuation differences in the liver parenchyma (*). Origin: Tonolini Massimo, Department of Radiology, ‘Luigi Sacco’ University Hospital – Milan (Italy)
Description: Note nonenhancing thrombosed lumen of the intrahepatic portal branches, and minimal ascites (+). Origin: Tonolini Massimo, Department of Radiology, "Luigi Sacco" University Hospital – Milan (Italy)
Description: Note minimal ascites (+). Origin: Tonolini Massimo, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
Description: B-mode ultrasound showed portal vein and confluence with upper-normal calibre and echoic lumen (TVP) suggesting thrombosis. Origin: Tonolini Massimo, Department of Radiology, "Luigi Sacco" University Hospital – Milan (Italy)
Description: Colour-Doppler examination confirmed echoic portal lumen (+) without flow signals consistent with thrombosis, plus patent tortuous collateral vessels indicating cavernomatous transformation (short arrows). Origin: Tonolini Massimo, Department of Radiology, "Luigi Sacco" University Hospital – Milan (Italy)
Description: After therapeutic anticoagulation, repeated colour Doppler examinations confirmed restored patency of the portal vein with normal hepatopetal flow. Origin: Tonolini Massimo, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
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