CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 28 (2016) 93–96

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Massive upper gastrointestinal bleeding due to splenoportal axis thrombosis in a patient with a tested JAK2 mutation: A case report and review literature

Isabel Macías (PhD)

Department of Surgery, Universitary Hospital Reina Sofia, Córdoba, Spain

ARTICLE INFO

Article history:
Received 8 June 2016
Received in revised form 8 August 2016
Accepted 8 August 2016
Available online 13 August 2016

Keywords:
Portal hypertension
Thrombosis portal
JAK2
Upper bleeding
Primary myeloproliferative disorders
Case report

ABSTRACT

Portal hypertension is a clinical syndrome defined as a portal venous pressure that exceeds 10 mmHg. Cirrhosis is the most common cause of portal hypertension and thrombosis of the splenoportal axis not associated with liver cirrhosis is the second cause of portal hypertension in the Western world. The primary myeloproliferative disorders are the main cause of portal venous thrombosis and somatic mutation of Janus Kinase 2 gene (JAK2 V617F) can be found in approximately 90% of polycythemia vera, 50% of essential thrombocyrosis and 50% primary myelofibrosis. A a 55-year-old man with JAK2 mutation-associated splenoportal axis hypertension and bleeding complications due to oesophageal varices is reported. A massive upper bleeding episode made an emergent surgery to be done immediatelly at seventh day. The patient was discharged home at fifteenth day after surgery.

1. Introduction

Portal hypertension is a clinical syndrome defined as a portal venous pressure that exceeds 10 mmHg. The etiology of portal hypertension can be classified as prehepatic, intrahepatic, or posthepatic (Table 1) [1]. Cirrhosis is the most common cause of portal hypertension and thrombosis of the splenoportal axis not associated with liver cirrhosis is the second cause of portal hypertension in the Western world [2]. There are identified systemics thrombogenic factors in 60% of cases and there are several etiologic factors associated in 15% of them [3].

The primary myeloproliferative disorders (polycythemia vera, essential thrombocytosis and primary myelofibrosis) are clonal disorders arising in a pluripotent hematopoietic stem cell, that causes an unregulated increase in the number of erythrocytes, leukocytes or platelets [4]. They are the main cause of portal venous thrombosis, although changes in blood in a portal hypertension context can make the diagnosis more difficult [5]. Somatic mutation of Janus Kinase 2 gene (JAK2 V617F) can be found in approximately 90% of polycythemia vera, 50% of essential thrombocytosis and 50% primary myelofibrosis [4] (Table 2).

The high incidence of gastrointestinal bleeding in patients with portal vein thrombosis and the association between oesophageal varices and JAK2-related splanchnic vein thromboses should be kept in mind when managing such patients [6].

2. Case report

We present a 55-year-old man with JAK2 mutation-associated splenoportal axis hypertension, splenomegaly and bleeding complications due to oesophageal varices. He's on oral anticoagulants therapy. He had history of cholecystectomy 9 years ago. This patient was referred to our tertiary hospital for evaluation of long duration abdominal pain and hematochezia. He was admitted in the gastrointestinal bleeding unit.

An early endoscopy was performed within 24 h of presentation: stomach fundus was filled with blood clots and there were some isolated fundal gastric varices. (Fig. 1). Blood test revealed leukocytosis (26.000 leukocytos per mm^3) and thrombocytosis (767.000 thrombocytes per mm^3).

A computed tomography angiography (CTA) under fluoroscopic guidance was performed. The procedure was carried out under sedation and local anesthesia. Celiac trunk, splenic artery and upper mesenteric artery were catheterized with a 5 French (Fr) catheter. After vasodilators agents administration, intraoperative control arteriography showed absence of portal and splenic opacification, which supports thrombosis in splenoportal axis (Fig. 2).

The patient had a massive upper bleeding episode at seventh day, with tachycardia, tachypnoea, cool clammy skin, hypotension and confusión and the use of luminal tamponade with

E-mail address: maciaspuerta@gmail.com

http://dx.doi.org/10.1016/j.ijscr.2016.08.013

Table 1
Causes of portal hypertension [1].

ETIOLOGY OF PORTAL HYPERTENSION BY LOCATION		
PREHEPATIC	INTRAHEPATIC	POSTHEPATIC
Portal vein thrombosis	Cirrhosis	Budd-Chiari syndrome
Splenic vein thrombosis	Primary biliary cirrhosis	Congestive heart failure
Congenital thrombosis of portal vein	Infiltrative liver disease	Constritive pericarditis
Arteriovenous fistula	Idiopatic portal hypertension	Tricuspid valve diseases
	Congenital hepatic fibrosis	
	Policystic liver disease	
	Postsinusoidal venooclusive disease	

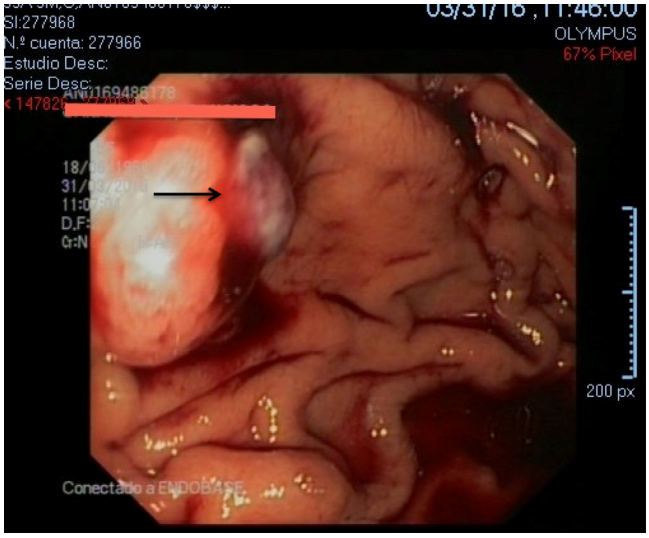

Fig. 1. Early endoscopy shows isolated fundal gastric varices (dark arrow).

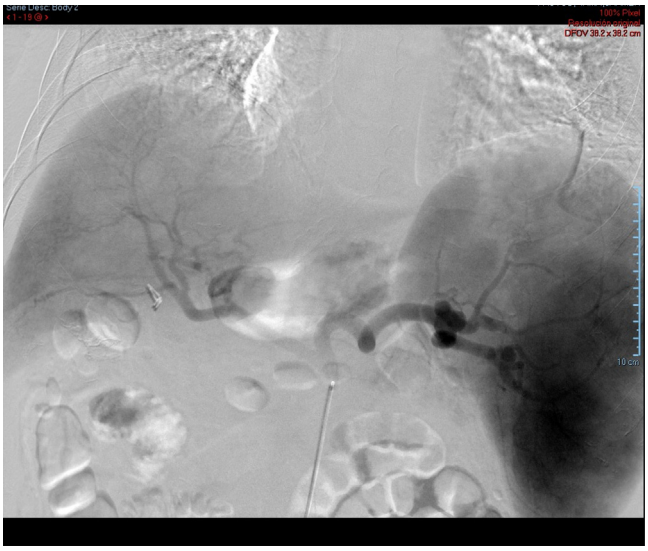

Fig. 2. Angiography under CT guidance. Absence of portal and splenic opacification (thombosis).
Abbreviation: CT, computed tomography.

Table 2
Representative molecular defects in the chronic myeloproliferative disorders [4].

CHRONIC MYELOPROLIFERATIVE DISORDERS	
DISEASE	MOLECULAR DEFECT
Chronic myelogenous leukemia	BCR-ABL
Chronic eosinophilic leukemia	FIP1L1-PDGFRA
Chronic neutrophilic leukemia	BCR-ABL p230
Chronic myelomonocytic leukemia	TEL-PDGFRB
Systemic mastocytosis	KIT D8116V
Polycythemia vera	JAK2 V617F (≈90% positive)
Essential thrombocytosis	JAK2 V617F (≈50% positive)
	MLP W515L/K (≈3% positive)
	MLP K39N
Primary myelofibrosis	JAK2 V617F (≈50%)
	MLP W515L/K (≈14%)

Sengstaken-Blakemore was a life-saving maneuver. Unfortunately, acute bleeding was uncontrolled so an emergent surgery was performed inmediatelly. Transjugular intrahepatic portosystemic shunt (TIPS) is contraindicated because of portal vein thrombosis [7] (see Table 3).

A midline laparotomy was performed: a great stomach filled with blood and blood clots takes up most of left upper cuadrant. Some fundal gastric varices are due to thrombosis in splenoportal axis. Neither esophageal nor other gastric varices were seen. A splenectomy was performed first, providing better exposure for gastric devascularization (Figs. 3 and 4). Acute upper bleeding was successfully controlled and a Sugiura procedure was not required.

The patient was discharged home fifteen days after surgery. Pathologic analysis did not reveal nothing but splenomegaly. Essential thrombocyrosis was diagnosed and he is currently receiving follow-up care by haematologist. No other bleeding episodes were reported.

Table 3
Contraindications for TIPS [7].

Contraindications for Transjugular Intrahepatic Portosystemic Shunt (TIPS)	
Absolute contraindications	Relative contraindications
Severe elevate right heart pressure	Complete hepatic vein obstruction
Severe pulmonary hypertension	Complete portal vein thrombosis
Severe congestive heart failure	Hepatocellular carcinoma
Severe encephalopathy	Severe coagulopathy (INR greater than 5)
Uncorrectable bleeding diathesis	Severe thrombocytopenia (platelet count less than 20,000/cm^3)
Active systemic or hepatic bacterial infection	Advanced liver dysfunction (bilirubin greater than 5 mg/dL or MELD greater than 17)
Unrelieved biliary obstruction	Moderate pulmonary hypertension