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GAVIN PUBLISHERS

Journal of Gastrointestinal Disorders

Damiris K, et al. J Gastrointest Disord 3: 109.

DOI: 10.29011/JGID-109.100009

Case Report

Chronic Extra-Hepatic Portal Vein Obstruction in a Pregnant Patient Requiring TIPS: A Case Report

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Citation: Damiris K, Ng TG, Seltzer ES, Ahlawat Sk (2020) Chronic Extra-Hepatic Portal Vein Obstruction in a Pregnant Patient Requiring TIPS: A Case Report. J Gastrointest Disord 3: 109. DOI: 10.29011/JGID-109.100009

Received Date: 29 September, 2020; **Accepted Date:** 20 October, 2020; **Published Date**: 26 October, 2020

Abstract

Extra-Hepatic Portal Vein Obstruction (EHPVO) is due to non-cirrhotic or malignant causes and may present with signs and symptoms of chronic portal hypertension. Here we discuss a patient on her fifth pregnancy who was discovered to have EHPVO with cavernous transformation and perisplenic, gastohepatic and retroperitoneal varices after presenting with abdominal pain, splenomegaly, and unremarkable labs. An emergent Transjugular Intrahepatic Portosystemic Shunt (TIPS), although rarely feasible in the presence of cavernous formation, with percutaneous thrombectomy of the superior mesenteric vein, splenic vein, and hepatic portal veins was performed. Unfortunately, post-procedural complications resulted in hypovolemic shock and spontaneous abortion. Genetic testing and bone marrow biopsy confirmed the diagnosis of essential thrombocythemia in the setting of a Janus Kinase2 (JAK2) V617F mutation. Portal vein thrombosis in pregnancy is rare, however, hemodynamic changes of pregnancy may increase the risk of complications due to portal hypertension. Additionally, the diagnosis of primary myeloproliferative disorders is a known risk factor for EHPVO. While prophylaxis and close monitoring is suggested in patients with a history of EHPVO before conception, there is no clear protocol in treating patients who are diagnosed for the first time with chronic EHPVO during pregnancy.

Keywords: JAK2V617F Mutation; Portal vein thrombosis; Pregnancy; TIPS

Abbreviations: EHPVO: Extra-hepatic portal vein obstruction; ET: Essential thrombocythemia; JAK2: Janus Kinase2; MPD: Myeloproliferative disorders; PV: Polycythemia Vera; PVT: Portal vein thrombosis; TIPS: Transjugular intrahepatic portosystemic shunt

Introduction

Extra-hepatic portal vein obstruction (EHPVO) describes either intra- or extra-hepatic Portal Vein Thrombosis (PVT) that is unrelated to cirrhosis or malignant causes [1]. Acutely, EHPVO presents commonly with abdominal pain and may be accompanied by fever or ascites. Chronic EHPVO may present with symptoms of portal hypertension, such as variceal bleeding and hypersplenism, or with the formation of collateral veins known as portal cavernomas [2,3]. Risk factors associated with the development of PVT are not limited to inherited disorders such as Protein C and S deficiency, Factor V Leiden mutation, acquired primary Myeloproliferative Disorders (MPD), pregnancy and postpartum, oral contraceptive use, malignancy, and inflammation [4]. The risk of thrombosis in pregnancy is about six times that of non-pregnant women and even more so in those with concomitant MPD [5]. Here we present a case of a pregnant female with abdominal pain, emesis, and splenomegaly who goes on to develop severe complications of previously undiagnosed EHPVO.

Case Presentation

A 38-year-old gravida 5 para 4 Hispanic female with no past medical history or pregnancy-related complications presented at eight weeks gestation, with a two-week history of right upper quadrant and epigastric pain, complicated by nausea and emesis. The pain was constant without relation to meals. She denied fevers, chills, diarrhea, sick contacts, or recent travel. On physical exam, there was tenderness to palpation in the epigastrium and right upper quadrant, coupled with splenomegaly. Bowel sounds were appreciated throughout. During this time, she remained

hemodynamically stable, and in no acute distress. Her hemoglobin was 12.6 g/dl, platelet count 200 x 10^9/L, prothrombin time 14.6 seconds, partial thromboplastin time 31.3 seconds, with normal liver enzymes. However, a right upper quadrant ultrasound demonstrated complete thrombosis of the main, right, and left portal veins, as well as splenomegaly, with trace perihepatic and splenic ascites (Figure 1). Magnetic resonance imaging demonstrated thrombus extending into the distal superior mesenteric vein and portions of the splenic vein (Figure 2) as well as perisplenic, gastohepatic, and retroperitoneal varices compatible with portal hypertension. Computed tomography demonstrated cavernous transformation of the portal veins (Figure 3).

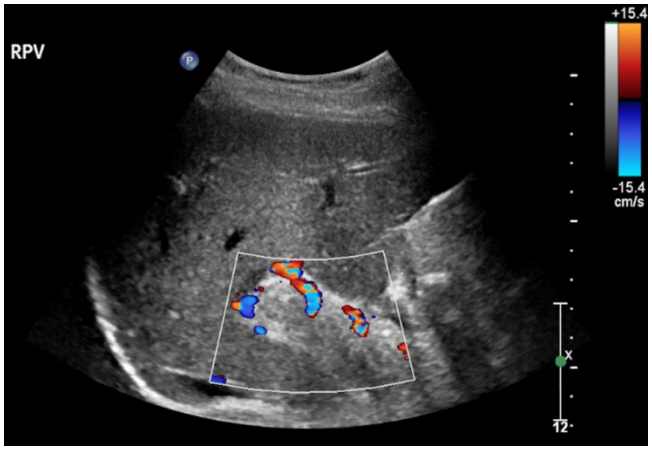

Figure 1: Ultrasound image showing complete thrombosis of the right portal vein.

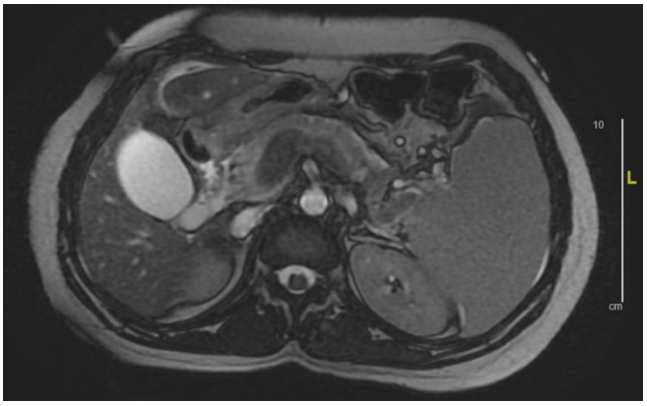

Figure 2: Axial MRI image showing complete thrombosis of the main and intrahepatic portions of the portal vein with thrombus extending retrograde into the distal superior mesenteric vein and into the distal and midportions of the splenic vein. There are perisplenic varices and splenomegaly compatible with portal hypertension.

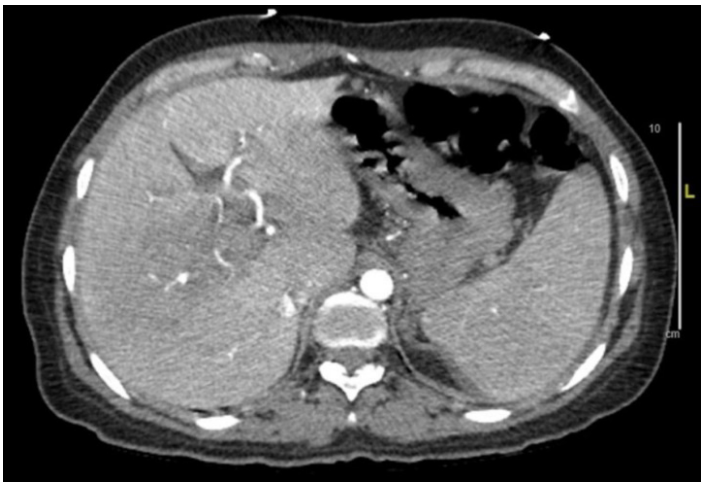

Figure 3: Axial CT scan image showing intrahepatic portal vein thrombosis with cavernous transformation. Splenomegaly is also noted.

Given the severity of her condition and with the patient's permission after an in-depth discussion regarding potential risks to both herself and her fetus, the decision was made by the hepatology, interventional radiology, and obstetrics teams to perform emergent Transjugular Intrahepatic Portosystemic Shunt (TIPS) with percutaneous thrombectomy of the superior mesenteric vein, splenic vein, and hepatic portal veins. She underwent successful thrombectomy and TIPS placement, however her post-procedural course was complicated by cardiac arrest due to hemorrhagic shock from splenic rupture. After the return of spontaneous circulation, she was taken to the operating room for emergent laparotomy with splenectomy and perihepatic packing. Afterwards, she was brought to the surgical intensive care unit, but had persistent intraperitoneal hemorrhage and subcapsular hepatic hemorrhage, requiring percutaneous embolization. Unfortunately, evaluation of the fetus revealed a spontaneous abortion most likely due to the loss of perfusion during the cardiac arrest. Two days after splenectomy and embolization, the patient underwent dilation and curettage. The remainder of the patient's hospital course went without complications and the patient recovered well. Prior to discharge, she was empirically treated for Polycythemia Vera (PV) or Essential Thrombocythemia (ET) given that Janus Kinase 2 (JAK2) V617F mutation was found to be positive. She was discharged on warfarin 7.5 mg daily. Outpatient, she underwent a bone marrow biopsy revealing slightly hypercellular bone marrow with increased atypical megakaryocytes, no increased blasts, and unremarkable myelopoiesis and erythropoiesis. Blood counts at that time revealed thrombocytosis with no increased hemoglobin or leukocytosis. Overall, her features resembled ET and she was started on hydroxyurea for cytoreductive therapy.

Discussion

PVT is rare in pregnancy with incidence up to 4% [6]. In these patients hemodynamic changes that occur physiologically throughout pregnancy are believed to worsen portal hypertension and increase the risk of variceal hemorrhage [6,7]. Hypersplenism, resulting in severe anemia and thrombocytopenia, is a common complication seen in this population, requiring transfusions [2,3,8,9]. While maternal outcomes are favorable even in cases of variceal bleeding, fetal outcomes are at high risk of prematurity, small gestational age, perinatal death, and abortion [1,3,8]. One study found that 44% of pregnancies in women with EHPVO resulted in babies that were small for gestational age [8]. Aggarwal, et al. followed a total of 26 pregnancies in 14 women with EHPVO and found the incidence of abortion to be 20% and preterm delivery to be about 15% [3]. It is recommended that women with EHPVO undergo endoscopic evaluation for varices with treatment by sclerotherapy or band ligation prior to conceiving, followed by periodic surveillance upper endoscopies throughout the pregnancy [1,3,7,8,10]. Prophylaxis with non-selective beta-blockers in this setting is contested due to concerns of neonatal bradycardia, hypoglycemia, and fetal intrauterine growth retardation, which are already known complications [11]. About 40% of patients have gastric varices for which glue injection has been deemed effective, as TIPS is often not feasible in patients with portal cavernomas [1]. Ingraham et al. described five cases of successful TIPS placement in cirrhotic pregnant patients with a high risk of hemorrhage due to gastroesophageal varices, history of or high risk of bleeding varices, and refractory ascites [12]. They estimated the absorbed radiation to the fetus to be an average of 16.3 mGy, a dose at which the fetus feels no radiation effects. Unfortunately, our patient's case was complicated by splenic rupture and abdominal hemorrhage in the days following the TIPS placement, a known risk of the procedure [13].

In this case, the patient's diagnosis of ET was not evident based on the platelet, leukocyte, and hemoglobin levels. Her condition was likely masked by the splenomegaly resulting in hypersplenism. The JAK2 V617F point mutation aids in the diagnosis of MPD especially in EHPVO where hypersplenism due to portal hypertension results in normal blood counts [9]. In 241 cases of splanchnic vein thrombosis, the JAK2V617F was detected in 34% of PVT cases [14]. Less than a fourth of the JAK 2V617F positive patients met any of the blood count cutoffs used for diagnosis of PV or ET, and due to this principle, the WHO classification for MPD changed to include JAK2 mutations as major criteria for the diagnosis of PV and ET [14,15]. Hydroxyurea is the treatment of choice in ET patients with splenomegaly, however, our patient's diagnosis was not made until after splenic rupture had occurred [16]. Still, in this setting hydroxyurea remains the first-line treatment in reducing future thrombotic complications.

Conclusion

Here we faced a rare case of a pregnancy complicated by EHPVO. Pregnancies can be successful in patients with a history of thrombosis with proper treatment starting before conception. In our case, the diagnoses of EHPVO and eventually ET were made during the pregnancy, and due to extensive thrombosis and effects related to portal hypertension, TIPS and thrombectomies were required. The use of JAK2 V617F allowed for the diagnosis of MPD in the setting of normal complete blood counts. While extensive complications ensued, the patient remains alive and well on the proper treatment of her ET.

We confirm that informed patient consent was obtained for publication of the case details

Conflicts of Interest

The authors declare no conflicts of interest

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