Annals of Clinical and Medical Case Reports

ISSN 2639-8109

Case Report

Portosystemic Shunt for Treatment of Symptomatic Varices in Polycythemia Vera Patient: A Case Report

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Volume 4 Issue 5- 2020

Received Date: 12 June 2020

Accepted Date: 25 June 2020

Published Date: 29 June 2020

1. Abstract

Myeloproliferative disorders are commonly associated with portal vein thrombosis, which can lead to extensive and symptomatic variceal disease due to the development of noncirrhotic portal hypertension. In severe cases, a decline in liver synthetic function may indicate the need for liver transplantation. We report the case of a 51 year-old female with polycythemia vera with symptomatic melena, found to have extensive portal vein thromboses and retroperitoneal varices in the setting of normal liver synthetic function. She was treated successfully with splenectomy and creation of a portosystemic shunt between the inferior mesenteric vein and gonadal vein.

2. Key words

Portosystemic shunt; Polycythemia vera; Portal venous thrombosis; Noncirrhotic portal hypertension; Variceal disease

3. Summary

Myeloproliferative disorders are commonly associated with portal vein thrombosis, which can lead to extensive and symptomatic variceal disease due to the development of noncirrhotic portal hypertension. In severe cases, a decline in liver synthetic function may indicate the need for liver transplantation. We report the case of a 51 year-old female with polycythemia vera with symptomatic melena, found to have extensive portal vein thromboses and retroperitoneal varices in the setting of normal liver synthetic function. She was treated successfully with splenectomy and creation of a portosystemic shunt between the inferior mesenteric vein and gonadal vein.

4. Introduction

Myeloproliferative Disorders (MPD) are the main cause of Portal Venous Thrombosis (PVT) involving a JAK2 mutation in 90% of polycythemia vera (PV), 50% of Essential Thrombocytosis (ET) and 50% of primary Myelofibrosis (MF) [7]. Studies have shown that a JAK2 mutation is a risk factor for splanchnic circulation thrombosis, especially in patients who are homozygous for the gene [12]. These disorders exhibit myeloid cell expansion in the peripheral blood and come with a significant risk of thrombogenic and hemorrhagic complications, such as variceal bleeding and ascites. Most patients diagnosed with PV are women with a median age of 54 years [3]. MPDs promote platelet aggregation and increase thrombin generation. When PVT occurs, the liver compensates with increased hepatic arterial flow and preserves normal liver function, but if this goes untreated, it can result in intestinal ischemia or the portal vein can undergo cavernous transformation due to chronic portal hypertension [4]. Some cases have been shown to have only mild increases in liver function enzymes in patients with myeloproliferative disorders who have portal hypertension.

Portal hypertension is defined as a hepatic venous portal pressure gradient greater than 6 mmHg, but usually becomes clinically significant when greater than 10 mmHg. Cirrhosis is the most common etiology followed by thrombosis of the splenoportal axis not associated with cirrhosis [7]. Portal hypertension affects 7- 18% of patients with myeloproliferative disorders, specifically PV and MF have a higher incidence than ET. This is thought to be secondary to thrombosis, splenomegaly, or intrahepatic extramedullary hematopoiesis. Even in the absence of thrombosis, these patients can exhibit noncirrhotic portal hypertension [12]. Mortality is highest in patients with cirrhosis or cancer (26%) when compared to patients without these diseases (8%) [9]. Mortality arises from complications of portal hypertension, which include hepatorenal failure, bacterial peritonitis, variceal bleeding, leukemic transformation, or progression of their myeloproliferative disorder [12].

PVT usually involves the extra-hepatic portal vein, but can include the intrahepatic portal, superior mesenteric and splenic vein. When PVT is seen in patients with noncirrhotic, nonmalignant

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Citation: Doe-Williams S, Portosystemic Shunt for Treatment of Symptomatic Varices in Polycythemia Vera Patient: A Case Report. Annals of Clinical and Medical Case Reports. 2020; 4(5): 1-4.

disease, survival can be considered good, with a five-year survival rate of 90% when treated[5]. Treatment options include anticoagulation and cytoreduction medications, Transjugular Intrahepatic Portosystemic Shunt (TIPS), endoscopic thrombolysis, or surgical portosystemic shunt placement (Figure 1).

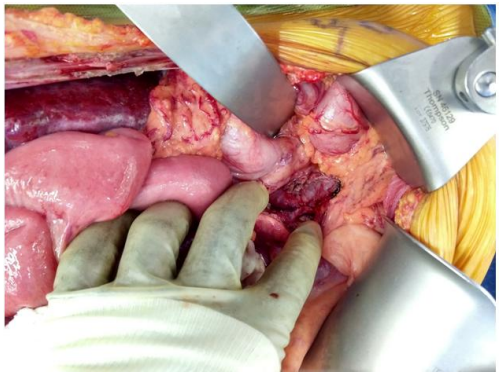

Figure: "Inferior mesenteric vein and gonadal vein anastomosis for portosystemic shunt creation"

5. Case Report

We report a 51 year-old female, diagnosed with polycythemia vera six years prior to our encounter with splenomegaly and chronic portal vein thrombosis with extension into the superior mesenteric vein without cirrhosis. The patient had remained stable on hydroxyurea and warfarin until she presented to an outside hospital with melena, productive cough, abdominal bloating, nausea, and bilious emesis. The patient underwent an EGD showing large varices without active bleeding and the patient was transferred for a possible open TIPS procedure. Physical exam revealed a thin, afebrile female with normotension and without tachycardia. Her abdomen was soft with minimal ascites, non-distended with no hepatomegaly, but splenomegaly was noted. Labs upon admission included creatinine 0.52 mg/dL, hemoglobin 12.2 mg/dL, hematocrit 36 mg/dL, albumin 3.7 g/dL, total protein 5.8 mg/dL, alkaline phosphatase 149 U/L, ALT 32 U/L, AST 34 U/L, total bilirubin 1.6 mg/dL, PT 16.4, and INR 1.4. MRI and CT abdomen showed chronic thrombosis of the portal vein and portal venous confluence with associated cavernous transformation, extensive splenic, gastric and esophageal varices, marked splenomegaly up to 19cm, and moderate to large volume ascites with a large right pleural effusion, felt to be consistent with a hepatic hydrothorax without evidence of cirrhosis.

It was deemed that the open TIPS procedure would not benefit the patient as the portal hypertension was pre-hepatic in etiology. The chronic portal venous thrombosis had progressed to a cavernous transformation, thus making a standard TIPS procedure not possible. The patient's CT also suggested hepatic outflow obstruction that led to the ascites and hepatic hydrothorax. This usually leads to Budd-Chiari syndrome, however the hepatic veins appeared to be patent. The patient had a transjugular liver biopsy that showed no underlying liver disease, such as cirrhosis, with a normal portosystemic pressure gradient. Thus, the transplant surgery service was consulted to perform a splenectomy with a surgical portosystemic shunt placement between the inferior mesenteric vein and gonadal vein to address her variceal disease.

Through a bilateral subcostal incision based on the left side with an epigastric extension, the abdomen was opened. There was 2 L of ascitic fluid encountered, and there were extensive varices noted in the retroperitoneum. A systemic, retroperitoneal venous tributary was identified, draining into the gonadal vein and a tense portal collateral was identified as the inferior mesenteric vein. These were dissected carefully, and side-to-side anastomosis was done with a 1.5 cm ostium with 6-0 Prolene. There was a good thrill and flow, and the pressure in the portal system decreased significantly, as the portal collaterals had decreased in pressure. Finally, the spleen was mobilized from the retroperitoneal space and was carefully removed successfully.

The patient was doing well post-operatively, had mild urinary retention relieved by a urinary catheter. Postoperative labs were notable for AST 48 U/L, ALT 37 U/L, and alkaline phosphatase 184 U/L. She was discharged on post-operative day six with a Lovenox bridge as she was subtherapeutic on warfarin with an INR of 1.5.

On post-operative day 13, the patient was admitted after being seen in the hepatology clinic due to continuing elevation of her LFTs; AST 107 U/L, ALT 132 U/L and alkaline phosphatase 447 U/L, which was concerning for a potential thrombosis in the setting of elevated platelets of 1824 and having a subtherapeutic INR on warfarin, Lovenox and aspirin. A multiphase CT abdomen and pelvis and an abdominal ultrasound were obtained that were remarkable for unchanged diffuse portal venous system thrombosis without a patent main portal vein suggestive of cavernous transformation, but the surgical shunt remained open. An echocardiogram was obtained that showed an ejection fraction of 71% with mildly elevated pulmonary systolic pressure of 33-38mmHg. She was discharged on hospital day two after having the hydroxyurea dose increased, with labs demonstrating improvement: INR 1.3, AST 107 U/L, ALT 133 U/L and alkaline phosphatase 413 U/L.

6. Discussion

Myeloproliferative disorders exhibit myeloid cell expansion in the peripheral blood and come with a significant risk of thrombogenic and hemorrhagic complications. These disorders are the most common cause of noncirrhotic portal vein thrombosis. This case was different in the fact that the patient presented with ascites and hepatic hydrothorax, which is uncommon in patients with PV complicated by PVT as the liver function was normal. This is in contrast to hepatic vein thrombosis, also known as Budd-Chiari syndrome, where findings of hepatic failure and ascites are almost always present [10]. It can cause elevations in liver enzymes greater

than 1000 U/L, however this patient had a maximum AST of 107 U/L, ALT of 133 U/L and alkaline phosphatase of 447 U/L with imaging showing the hepatic vein remaining patent [8].

Diagnosis of portal venous thrombosis is accomplished by using doppler ultrasonography, contrast-enhanced computed tomography, or magnetic resonance imaging to define the occlusion in the vasculature. When there is cavernous transformation of the portal vein, this means that a cluster of varying-sized vessels have replaced the portal vein and are arranged haphazardly within the connective tissue support at the liver hilum. Liver architecture is well preserved [6]. It is seen in patients with chronic PVT in which it functionally replaces the portal vein, as in our patient.

Life-long oral anticoagulation with Vitamin K antagonists (warfarin) is the mainstay treatment for myeloproliferative disorders in the setting of portal vein thrombosis. Hydroxyurea is the first line cytoreductive medication for patients with polycythemia vera. Recurrences of thrombosis may occur in 15-20% of patients. When medication alone fails, other treatment options include TIPS placement, surgical portosystemic shunting, or angiography with intravascular thrombolysis or thrombectomy [7]. Absolute indications for shunt surgery include medically and endoscopically refractory variceal hemorrhage, hypersplenism, severe thrombocytopenia, refractory hepatic encephalopathy, hepatopulmonary syndrome, and portopulmonary hypertension [6].

Achieving a suitable portal branch can be very difficult when creating a TIPS and is successful only 60% of the time, making TIPS a relative contraindication when a PVT is present [1]. Trials studying TIPS versus surgical portosystemic shunt placement versus endoscopic thrombolysis in patients with cirrhotic portal hypertension have found that surgical shunt placement has the lowest bleeding-related mortality among the three options. It was also shown that surgical shunt placements may be the most effective without the increased risk of hepatic encephalopathy versus TIPS [13]. There are no studies found that compare these treatments in patients with noncirrhotic portal hypertension, such as our patient. Conclusive evidence regarding which method is more efficacious in our patient subtype is pending at this time.

In a study of 56 patients with noncirrhotic portal hypertension, 49 patients underwent portosystemic shunt placement and all remained patent 30 days post-operatively, and there were no recurrences compared to patients who underwent thrombolysis. Shunt placement options included mesocaval, distal splenorenal, proximal splenorenal, side-to-side portocaval, paraumbilical-jugular, and portal to right atrial shunts [11]. An interesting part of this surgical case is the anastomosis between the inferior mesenteric vein and gonadal vein, which is usually seen in treatment options for nutcracker syndrome and has not been seen in this type of case [2]. This has proved to be a successful surgical option for patients with PV and PVT complicated by portal hypertension. There was also one case study reported that created an extrahepatic portosystemic shunt via a dilated coronary vein, displaying the feasibility of using any vein to create a shunt when other options are difficult [1].

In conclusion, surgical portosystemic shunting is the best option for patients with a subacute presentation and intact liver function when TIPS is not practical as seen in this patient's case, where her chronic PVT had cavernous transformation [8]. This was a rare indication for surgery in a patient with polycythemia vera and PVT, especially since the patient did not have cirrhosis and her liver function was otherwise normal prior to the operation.

To date, eight months postoperatively, our patient remains stable on hydroxyurea and her liver function and platelet levels normalized. A repeat surveillance EGD performed at this time showed a grade 1 varix in the lower third of the esophagus without stigmata of bleeding. She will be due for follow up imaging to ensure her portosystemic shunt remains patent.

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