Spontaneous Splenic Hemorrhage and Portal Hypertension in Pregnancy: A Combined Catheter Based and Surgical Approach

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Abstract

Objectives: To review the management of portal hypertension and splenic hemorrhage in pregnancy.

Methods: A retrospective case review.

Results: A 22-year-old female, with past history of portal hypertension, variceal hemorrhage and thrombocytopenia presented at 22 weeks of pregnancy with spontaneous splenic hemorrhage. Her portal hypertension was a consequence of intra-hepatic sclerosis following treatment of acute cell lymphoma in childhood. Because of the risk of variceal hemorrhage as pregnancy progressed, Transjugular Hepatic Portosystemic Shunt (TIPS) was performed first, followed a few days later by splenic artery embolization and splenectomy in a combined setting. The patient has subsequently delivered a healthy girl.

Conclusion: In the era of hybrid suites, catheter and surgical based approaches can more easily be combined. In this setting, the patient had dual risks as pregnancy advanced: variceal hemorrhage and splenic rupture. The combined approach permitted splenectomy without the need for transfusion.

Keywords: Variceal; Splenic; Hemorrhage; Embolization; TIPS

Introduction

Cirrhosis and portal hypertension, either separately or on combination, are rare in pregnancy [1]. When present, portal hypertension in particular volume is associated with an increased risk of variceal bleeding due to the increased vascular volume associated with later stages of pregnancy [2,3]. Spontaneous splenic hemorrhage is also an uncommon, both in pregnancy and as a complication of portal hypertension [4]. To our knowledge this is the first reported case of spontaneous splenic hemorrhage in the setting of both portal hypertension and pregnancy. Because of the combination of features, we decided to approach it in a hybrid manner, employing both catheter based and surgical approaches.

Case Presentation

A 21-year-old woman, G1P0 at 22 weeks gestation, presented with acute onset of left upper quadrant and left shoulder pain. The pain had started abruptly, with no history of trauma. She was at 22 weeks gestation. Her past medical history was notable for pre-B cell acute lymphoblastic anemia at age 14, treated with AALL0232 PC chemotherapy protocol and she had been in remission since. As a consequence of her therapy, she had developed hepatic venoocclusive disease, and subsequently cirrhosis and portal hypertension. She had experienced splenomegaly; chronic thrombocytopenia (platelet range 51-102) and four years prior to this admission had required endoscopic banding for variceal hemorrhage and had been maintained on propranolol.

On arrival, her heart rate was 100 and blood pressure 111/60 mmHg. Initial lab work was notable for: WBC 17.5, HGB 9.7, Platelet 91, INR 1.2 and normal liver functions. An ultrasound revealed a normal intrauterine pregnancy and new ascites (when compared to prior ultrasounds). Surgery was consulted, and because we could not determine the nature of the ascitic fluid, paracentesis was performed revealing a Serum-Ascites Albumin Gradient (SAAG) of 1.31 and RBC 1,985,000/ul, consistent with hemorrhage [5]. This prompted a Computed Tomographic (CT) scan which
demonstrated hepatosplenomegaly, hemorrhagic ascites and two areas of splenic hemorrhage (Figure 1). Initially, management consisted of bed rest and monitoring. She remained stable, but her platelet count continued to decrease, reaching a nadir of 44. Maternal fetal medicine was concerned that as her pregnancy progressed the associated intravascular volume expansion and thrombocytopenia would be associated with increased portal pressures, and risk of variceal hemorrhage and/or further splenic hemorrhage. With this in mind, we initially performed a TIPPS procedure. The pressures were not markedly elevated but did decrease (from 15 to 8 mmHg). However, while possibly diminishing the risk of variceal hemorrhage, our colleagues were concerned with the risk of further splenic hemorrhage as the pregnancy progressed, and so we prepared for splenectomy. Because of her thrombocytopenia, we felt controlling the splenic artery before any platelet transfusions would be needed would be optimal. We determined to perform the procedure in the hybrid suite, with the plan to embolize the spleen and in same setting perform splenectomy. Part of our rational was that in our experience a number of patients experience severe pain post splenic embolization. We decided on open splenectomy both due to the size of the spleen and concern that there would be dense adhesions due to the peri splenic blood. The procedure was performed two days after the TIPPS, at which point her platelet count had risen to 55. Coil embolization of the distal splenic artery was performed. A large branch supplying the upper pole originated proximal to a branch supplying the pancreas. Because we did not want to eliminate flow to this branch, adjunctive gel embolization of the mass of the spleen was performed as part of the procedure, with coil embolization of the main artery distal to the origin of these vessels (Figure 2). Once this was completed, splenectomy was performed though a left upper quadrant incision. The spleen measured 20.5 cm × 16.3 cm × 8.5 cm and weighed 1194 grams (Figure 3). No transfusions were required during the procedure.

The patient had no other complications and was discharged a few days post procedure. She delivered a healthy baby girl and has had ultrasound follow up of her TIPPS which shows a very limited thrombus but overall widely patent. At last follow up 9 months later her platelet count is 316.

**Discussion**

While cirrhosis is uncommon among pregnant women, the combination of portal hypertension and pregnancy is associated with increased risk of variceal bleeding, postpartum hemorrhage, preterm delivery, low birth weight and neonatal death [3,6]. This is felt to be due to the intravascular fluid expansion that occurs, particularly during the third trimester [6]. The management guidelines are not clear, and while standard medical therapy is generally used, TIPPS has been occasionally used, primarily in the setting of varices. Among 10 reported cases, 9 children were successfully delivered with minimal fetal exposure to radiation [7-12].

Literature regarding spontaneous splenic hemorrhage due to underlying portal hypertension is also sparse, is usually associated with advanced liver disease, and is generally managed successfully with splenectomy [4,5,13]. Spontaneous splenic hemorrhage, as distinct from rupture of splenic artery aneurysm, is also uncommon in pregnancy. It generally occurs in the third trimester, but has been reported in late second [14]. It has been described in the setting of eclampsia, but in the absence of identifiable pathology, the etiology has been attributed to a combination of increased blood volume, changes in circulating estrogen/progesterone levels leading to structural weakness and reduced free space of the peritoneal cavity [15,16]. As in the setting of ruptured splenic aneurysm, there is a very high fetal demise rate and emergent splenectomy is usually required [15].

Hypersplenism has been managed by embolization, splenectomy or both. In one series of seventeen patients with hypersplenism secondary to portal hypertension, all experienced successful increase in platelet count. Four (24%) experienced major complications, including splenic abscess, respiratory distress and pancreatitis. The incidence major complications increased when more than 70% of the
spleenic volume of embolized [17].

Pre-splenectomy embolization has been advocated prior to splenectomy, both laparoscopic and open approaches, in the setting of the “giant spleen”. Reported experience argues that there is significantly less blood loss, transfusion requirement, and conversion from laparoscopic to open splenectomy [18-21]. Embolization has been performed using a variety of tools, including coils and gel foam, but an important aspect is to avoid compromising the arterial supply of the distal pancreas [21] can be associated with significant pain following the procedure, as well as coil migration (if used) [18]. Successful laparoscopic splenectomy has been reported during pregnancy for managing immune thrombocytopenic purpura [22]. However, while laparoscopic and open splenectomy can be performed safely during pregnancy, there is evidence that laparoscopic approaches are associated with a higher blood loss and transfusion requirement [23].

References