Abdominal Wall Varices in Pregnancy-A Case Report and Review of the Literature

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Introduction

Pregnancy in patients with portal hypertension (PHT) is an uncommon event [1]. This situation is uncommon due to the numerous changes in liver disease particularly metabolic and hormonal changes leading to amenorrhea and ovulatory cycles. One-third of pregnancies in women with extrahepatic portal vein obstruction result in miscarriage [2]. Furthermore, maternal mortality rates range from 18% to 50% when associated with liver cirrhosis [2].

The risk of variceal bleeding during pregnancy with portal hypertension is another controversial issue, with rates as high as 70% [3]. This reflects a 45% increase in plasma volume during pregnancy potentially leading to an increase in portal pressure increasing the overall risk of variceal bleeding [4].

We present a case of a pregnancy complicated by portal hypertension in order to discuss the approaches and controversies to antenatal care and delivery planning in order to overall optimize maternal and fetal outcomes.

Case

Our patient is a 22 year old primigravida at 33 weeks and 3 days gestation, which was transferred to our care due to an elevation in liver enzymes. Her past medical history is significant for congenital biliary atresia, which necessitated a liver transplant at 8 months of age followed by mesocaval and splenorraphic shunting. At the age of sixteen, the patient experienced an episode of esophageal bleeding secondary to esophageal varices and was treated with endoscopic banding. She subsequently underwent a splenectomy due to recurrent history of esophageal varices. The patient was treated with a combination of immunosuppressive therapy that included Imuran, Prednisone, and Cyclosporine until the age of eighteen.

Her last esophagogastroduodenoscopy (EGD) in 2010 was negative for esophageal varices. Our primary concern was for the possible presence of abdominal wall varices and the inherent risk of hemorrhage during cesarean delivery, if obstetrically indicated. Consultations to the transplant and radiology services were placed for specific recommendations. In an effort to screen for ectopic varices, a Computed Topography (CT) Scan was performed. Varices were noted in the perigastric, mesenteric, and retroperitoneal regions. Although there were multiple small collateral vessels noted along the anterior abdominal and pelvic walls, there were no significant abdominal varices depicted on CT scan.

At 38 weeks and 5 days gestation, the patient was diagnosed with severe preeclampsia. The decision was made to proceed with an induction of labor in addition to initiating magnesium sulfate for seizure prophylaxis. Subsequently during her labor course, the decision was made to proceed with cesarean delivery secondary to arrest of dilatation. During the Cesarean Section, a vertical midline skin incision was performed along with a low transverse uterine incision. Intraoperatively, a Jackson Pratt (JP) drain was placed intraperitoneally secondarily to the presence of approximately one liter of ascitic fluid.

Postoperatively, the patient's hospital course was complicated by continuous ascitic fluid drainage. As per recommendations from the hepatology service, the patient was treated with albumin infusions in addition to the administration of diuretics. Liver enzymes subsequently normalized alongside a reduction in ascitic drainage. The patient was cleared for discharge on postoperative day eight. The JP drain was removed in clinic on postoperative day sixteen.

Discussion

We present a case of pregnancy complicated by a history of portal hypertension and ectopic varices with liver transplantation and its effects with regards to antenatal care and delivery planning in order to optimize both maternal and fetal outcomes.

In patients with portal hypertension, compression of the inferior vena cava by the gravid uterus leads to an increase in collateral flow in theazygous system with the subsequent appearance of transient esophageal varices, most commonly in the second and third trimesters of pregnancy [5,6]. For this reason, we believe that pregnancy might increase the incidence of abdominal wall varices in patients with portal hypertension. Rarely, ectopic varices can affect the peritoneum, ovaries, and the vagina [7].

Awareness of the clinical manifestations of portal hypertension is crucial when providing obstetrical care for at risk patients. Splenic vein thrombosis patients likely develop isolated gastric varices and typically present with hematemesis, melena, or rectal bleeding. Patients with a brisk bleed can quickly develop shock and need to be identified as soon as possible. Patients with portal vein thrombosis are likely to develop ectopic varices. Bleeding from ectopic varices located outside gastroesophageal lesions accounts for 5% of all variceal bleeding [8]. In advanced portal hypertension, the collateral circulation may carry greater than 90% of the blood entering the portal system [9].

By reviewing the current literature, we concluded that cases of severe hemorrhage from ectopic varices are rare but can often be fatal. Di Bisceglie et al. have reported spontaneous rectus muscle hematomas as a result of cirrhosis and portal hypertension [10] Rosenfeld et al. described two cases of postpartum ectopic variceal bleeding. One patient suffered from retroperitoneal hemorrhage after vaginal delivery, and another patient suffered from an abdominal wall...

**Esophageal Surveillance**

The perinatal care of these patients is highly complex and involves different subspecialties. In patients with a history of previous variceal hemorrhage, management of esophageal varices should be performed prior to pregnancy [12]. Prophylactic banding or sclerotherapy is not recommended in the pregnant patient due to the high risk for unpredictable bleeding and no significant overall improvement in overall survival [12].

Seventy-five percent of patients with pre-existing esophageal varices develop variceal hemorrhage during pregnancy [13]. Ideally, an EGD should be performed prior to the initiation of preconception visits to ascertain the severity of the underlying disease process. However, endoscopy should be deferred to the second trimester if the patient presents pregnant [14]. Esophageal varices may recur later on during pregnancy; however, an EGD should only be performed if there are strong indicators, primarily significant acute upper gastrointestinal bleeding [15]. EGD is a safe method of screening when performed with low dose sedative drugs after the first trimester [16]. For acute treatment of upper gastrointestinal bleeding in pregnancy secondary to esophageal varices, the treatment is endoscopic band ligation, which has replaced sclerotherapy as first line treatment [17].

With regards to ectopic varices, there is no consensus as to whether screening should be performed in pregnant women with portal hypertension [3].

Non-selective Beta Blockers have been used to prevent variceal bleeding in patients with portal hypertension by lowering the portal pressure [17] However, the use of Beta Blockers for the prevention of portal hypertension during pregnancy is controversial; but in general, the treatment can be continued with special attention given to the side effects that include neonatal hypoglycemia, neonatal bradycardia, and low birth weight [18].

**Diagnostic Imaging**

With regards to delineating pelvic anatomy and diagnosing pelvic varices, ultrasound should be initially performed, as it is cost-effective and can be performed at the bedside without the use of ionizing radiation. Direct venography can also be used; however, in the pregnant patient, direct venography is contraindicated secondary to the use of ionizing radiation to the pelvis. On the other hand, CT and Magnetic Resonance Imaging (MRI) are superior in depicting specific female pelvic and retroperitoneal anatomy [16].

MRI is specifically useful in diagnosing hemorrhage, intra-abdominal fluid, and masses with high-contrast resolution. However, CT is usually more widely available, faster, and less expensive [19-25]. There have been no effects on the fetus with MRI use during pregnancy due to the lack of use of ionizing radiation, unlike with the use of CT [26]. In addition, contrast agents, primarily gadolinium compounds, should be avoided at all costs due to its ability to cross the placenta and into the fetal bloodstream.

Generally, it has been shown that the porto-systemic collaterals can often be identified on CT scan [19]. In some patients, the vascular venous channels are deep seated and may be only visible by abdominal wall and pelvic MRI [20].

**Intrapartum Management**

The pregnant women with PHT should be managed in a tertiary care center with the ability for interdisciplinary care with hepatologists, anesthesiologists, vascular surgeons, and perinatologists; adequate facilities for blood transfusions; and the availability of an intensive care unit [12].

Even in healthy patients, the contraction of the diaphragm can increase portal pressure three times the normal value [21]. Regarding the effect of the second stage of labor on the incidence of variceal bleeding, it has been shown that intravariceal pressure changes during repetitive Valsalva maneuvers are unpredictable [22]. Furthermore, a vaginal approach can be a potentially safe method of delivery. It is important to note that vaginal delivery can potentially increase intrabdominal pressure leading to an increased portal hypertension, and an eventual higher risk of variceal rupture.

Patients with PHT may rarely also have engorgement and dilation of the extradural veins. This increases the risk of vascular absorption of local anesthetic and the subsequent toxic effects [23]. Toxicity symptoms in patients receiving anesthesia display CNS symptoms of tinnitus, disorientation, and seizures that typically precede the cardiovascular symptoms of hypotension, dysrhythmia, and cardiac arrest. The risk of epidural systemic toxicity is higher than in spinal anesthesia due the intrinsic nature of the anesthetic administered and its dosage requirement for the procedure. Typical risk reduction measures of epidural anesthetic administration include aspirating of the catheter to ensure proper placement of the anesthetic and not within a blood vessel or intradurally, addition of epinephrine to the anesthetic to delay systemic intravascular absorption, test dosing, the use of alternative anesthetic with a lower toxicity profile, and the injection of the anesthetic in incremental, smaller doses instead of bolus injections [27-29]. Forceps or vacuum assisted delivery with extradural anesthesia or pudendal nerve block can help avoid the marked increase in portal pressure during the second stage of labor, decreasing the likelihood of rupturing pelvic varices [23]. Cesarean delivery should be reserved for obstetrical indications only [24].

If necessary, careful planning of a cesarean delivery, primarily the incision, should be considered to prevent hemorrhagic complications. To be considered, the paraumbilical veins course along the falciform ligament connecting the portal venous system to the systemic epigastric veins near the umbilicus [11]. These veins may dilate with the increased portal hypertension and can be easily injured during the initial abdominal wall incisions. Therefore, a vascular surgeon should be consulted in the event of pelvic or abdominal wall collateral hemorrhaging.

Some studies have presented concern surrounding the use of Pitocin during the third stage of labor. The concern relates to the theorization of an increased venous return from contraction of the uterus creating an increased pressure in the inferior vena cava and the esophageal varices, if present. However, there is no solid evidence to support the theory [25].

**Postpartum Complications**

Postpartum Uterine Hemorrhage is a potential source of maternal morbidity and mortality, especially when there is a history of coagulopathy and thrombocytopenia. Blood and coagulation products should be ordered prior to delivery. In addition, oxytocin and other uterine contractile agents should be immediately available. Surgical
therapy using ligation of blood vessels and/or emergency hysterectomy may be additional methods if uterine contractile agents fail. A vascular surgeon or interventional radiologist may need to be consulted, if that is indicated [27].

Acute hemoperitoneum can rarely occur with rupture of intrauterine varices or various vascular vessels, specifically the left ovarian plexus. Rupture of these vessels tend to occur spontaneously and are rarely associated with iatrogenic intervention or trauma to the abdomen or pelvis. Symptoms of an acute hemoperitoneum include intractable abdominal pain, abdominal distention, hypotension, or dizziness, weakness, or fatigue secondary to anemia. Surgical intervention via laparoscopy or laparotomy may be the only way to diagnose by intraabdominal visualization and treat by ligation [3].

To conclude, although this case did not have a complicated postoperative course, pregnancy following liver transplantation has long been a relative contraindication due to the significant physiological changes that come with long-term management of a transplanted organ and the associated increase in mortality rates. It is our view that with proper preconception counseling combined with an interdisciplinary approach, pregnancy can be successfully maintained. Patients with similar conditions need to be managed with EGD to monitor esophageal varices during preconception counseling and the second trimester. Ultimately, MRI provides the clearest picture without the risk of radiation exposure of CT helping to further facilitate the construction of a delivery plan. Under this plan, careful monitoring for postpartum hemorrhage and possible fluid shifts can be managed through effective collaboration with the hepatology service and the administration of diuretics and colloid infusions secondarily to refractory ascitic fluid accumulations.

References