



Case Report

Splenic Artery Rupture During Pregnancy Concealed by a Pancreatic Lymphangioma: A Rare Co-Occurrence

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A pregnant woman presented to a local hospital with abdominal pain and hemorrhagic shock. Emergency caesarean section ruled out an obstetric cause and revealed a large mass, interpreted as a hematoma, with active bleeding of unknown origin. Because of her poor clinical condition, the patient was admitted to our hospital. Computed tomographic findings were suspicious for bleeding originating from the splenic artery. Laparotomy confirmed the presence of a ruptured splenic artery. A splenic artery aneurysm—a relatively well known entity during pregnancy—was absent. Hemostasis was achieved by clipping the artery. A large pancreatic cystic mass, which was misinterpreted earlier as a hematoma, was surgically removed. The pathologic examination revealed a pancreatic lymphangioma, an uncommon benign tumor. The ruptured splenic artery was presumably related to the pancreatic lymphangioma and vascular changes caused by pregnancy. A splenic artery rupture in co-occurrence of a pancreatic lymphangioma is a unique presentation which has not been reported previously.

Severe and prolonged hemorrhagic shock resulting in tissue hypoxia can be life-threatening. Persistent hypoxia can lead to severe complications and death.¹ During pregnancy and in the postpartum period, patients can present with hemorrhagic shock, usually related to gynecologic, obstetric, and/or coagulation disorders.^{2–6} However, other less common intra-abdominal causes that necessitate different treatment options should also be considered. In these cases, apart from an obstetrician/gynecologist, the interference of a surgeon,

radiologist, and/or intensive care physician is required.

We describe a case of a healthy pregnant woman presenting with acute abdominal pain and severe hemorrhagic shock caused by a ruptured splenic artery (SA). During laparotomy, a large retroperitoneal cyst, which matched the pathologic diagnosis of a pancreatic lymphangioma, was discovered and surgically removed. The rupture was possibly caused by mechanical pressure and traction related to the large lymphangioma combined with vascular changes occurring during pregnancy. Cases of a SA rupture caused by aneurysmatic changes have in particular been described among pregnant women.^{4–7} No such aneurysm, however, was observed in our patient. The co-occurrence of both uncommon findings with potential life-threatening consequences has, to our knowledge, never been reported before.

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CASE REPORT

A 30-year-old woman, gravida 2 para 1 (37 weeks' pregnant), was transferred from a local hospital and admitted

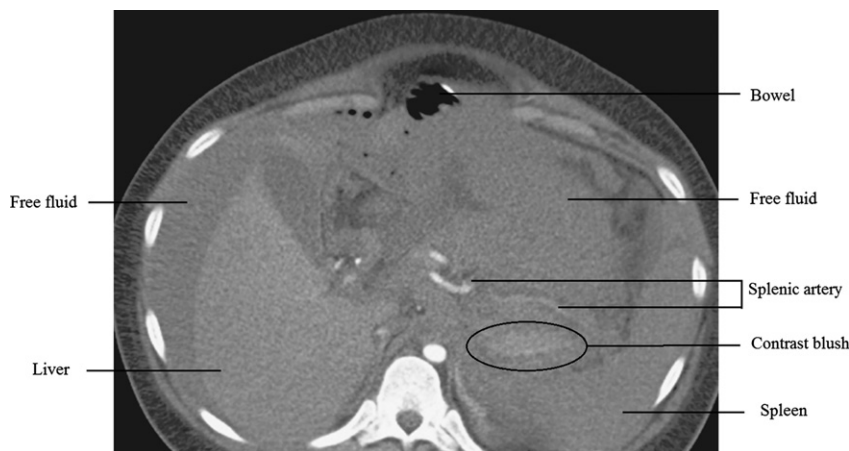


Fig. 1. Contrast-enhanced abdominal computed tomographic angiography (transverse view) showing free fluid around the liver and retroperitoneal space with active contrast blush originating from the splenic artery, representing arterial extravasation.

to our hospital because of severe hemorrhagic shock. A few weeks earlier, the patient endured a transient period of abdominal pain for which no obstetric cause was detected. Earlier that day, she underwent a regular obstetric examination, the results of which were unremarkable. Later that day, the patient was admitted to a local hospital because of recurrent severe abdominal pain and a loss of consciousness. Medical investigation revealed the presence of shock. Because the fetus appeared to be in distress and a placental abruption was suspected, a caesarean section was emergently performed. Also, a large localized retroperitoneal mass with concurrent active bleeding was observed and interpreted as a hematoma. A placental abruption or uterine rupture was not observed. The origin of the bleeding could not be localized. Because of this, the patient remained hemodynamically unstable despite large volume transfusions and the correction of coagulation disturbances. Despite all efforts, the neonate remained in a poor condition. Both mother and child were transferred to the intensive care units (ICUs) of different large hospitals.

At the time of admission to our hospital, the mother's condition was still life-threatening. She was intubated and mechanically ventilated. The intra-arterial blood pressure was 20/10 mm Hg; she had been hemodynamically stable during transport. The hemoglobin level was 3.3 mmol/L, lactate 7.8 mmol/L in the presence of severe acidosis (pH, 6.77) and persistent coagulation disturbances. A neurologic investigation revealed an eye opening, best motor response, and best verbal response (EMV) score of 1-1-Tube. Subsequent computed tomographic (CT) angiography of the abdomen revealed bleeding that was probably originating from the SA and the giant retroperitoneal cystic mass (18.5 × 16.8 cm) of unknown origin in the left quadrant (Figs. 1 and 2). Radiologic intervention by means of embolization was not considered because of her state of shock and the time needed to prepare for the procedure. Therefore,

emergency laparotomy was performed, and afterward the patient was transferred to the ICU. The bleeding was stopped by positioning a clip on the SA, which showed no signs of an aneurysmatic dilatation, and a splenectomy was performed. During laparotomy, the giant tumor seen on the CT scan (Fig. 1) appeared to be a large pancreatic cystic mass. Deroofing of the cyst wall was undertaken. Large quantities of packed red blood cells, fresh frozen plasma units, and platelet concentrate in addition to Ringer's lactate and epinephrine were infused in the pre- and perioperative phase. During surgery, the patient's condition and hemodynamics gradually stabilized. The following day, relaparotomy was performed to remove temporary abdominal packing and to search for active bleeding, which was not noticed. Sedative medication was stopped, whereupon the patient regained consciousness and was successfully extubated. Surprisingly, no neurologic deficits were present. Acidosis was also corrected and inotropic agents were stopped. A histopathologic investigation of the pancreatic mass revealed a benign cyst that matched the diagnosis of a pancreatic lymphangioma (CD31⁺, CD34⁻). A malignant cause was excluded. Seven days after laparotomy, the patient was discharged in good clinical condition. Unfortunately, her newborn son did not survive.

DISCUSSION

Defects of the SA are rare and can cause life-threatening situations if a rupture occurs, such as hemorrhagic shock and death. Although uncommon, aneurysms are an important cause of spontaneous ruptures of the SA.^{3-5,7} In contrast, a rupture in absence of an aneurysm has rarely been reported. The mortality rate of a ruptured splenic aneurysm is high, ranging from 25-70%

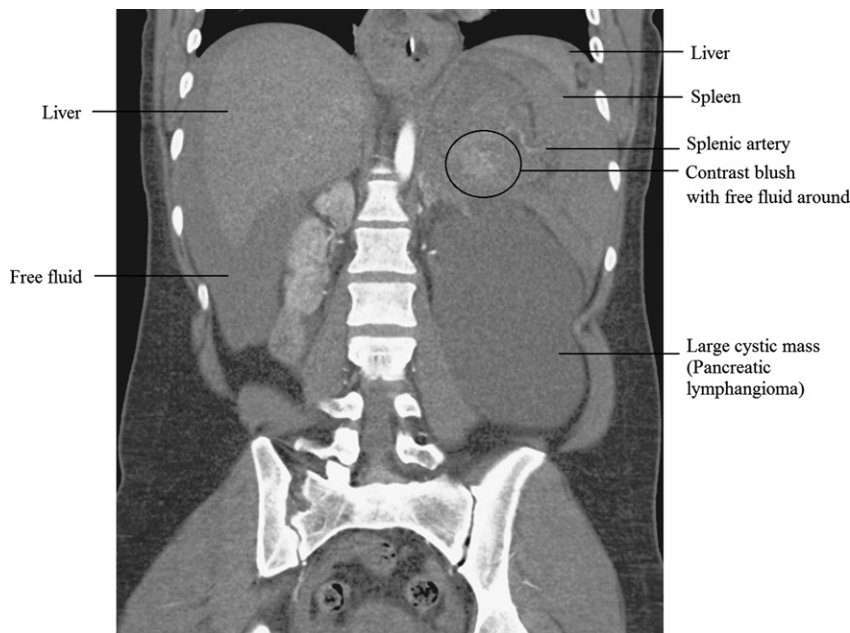


Fig. 2. Contrast-enhanced abdominal computed tomographic angiography (coronal view) showing free fluid around the liver and retroperitoneal space, active contrast blush from the splenic artery, and a large cystic mass of the lower abdomen.

depending on its underlying cause. However, its exact pathogenesis remains unclear.^{3-5,7} Several congenital and acquired risk factors, like pregnancy, are attributed to its formation. Weakness of the arterial wall and increases in blood pressure are proposed mechanisms to play an important role in the pathogenesis.^{3-5,7} Pregnant women and women of child-bearing age are also at risk, accompanied by both high fetal (95%) and maternal (75%) mortality rates.³⁻⁵ Apart from physiologic changes during pregnancy, hormonal influences on the arterial wall related to estrogen, progesterone, and relaxin, resulting in histologic wall alteration, are also thought to play an important role in its formation.^{4-6,8} The risk of a rupture varies between 2–10%, depending on the diameter of the aneurysm and can either develop suddenly or in 2 stages over the course of a few hours to several weeks.^{3,5-8} The latter is characterized by containment of the initial rupture within the lesser sac by either omentum or blood clots that block the foramen of Winslow and, if blood pressure increases further, followed by free rupture into the abdominal cavity.^{3,5,8}

Lymphangiomas are slow growing benign cystic tumors that probably result from a congenital lymphatic malformation and which cause obstruction of the lymphatic flow, resulting in lymphangiectasis. They can originate in almost any organ but most commonly occur in the neck, axillary regions, and mediastinum.⁹⁻¹³ In rare cases, these

benign tumors develop in the pancreas (1%). The available literature is limited to case reports.^{10,12,14} The diameter of (cystic) pancreatic lymphangiomas can vary between 3 cm to even ≥ 30 cm and can mimic pancreatic cancer. A thorough diagnostic evaluation is therefore essential.^{10-12,14} Pancreatic lymphangiomas are mostly asymptomatic, but can also cause symptoms varying from an acute presentation because of rupture, volvulus, torsion, hemorrhage or intestinal obstruction to chronic complaints of abdominal discomfort.^{11,12,14} The initial investigation is typically ultrasonography. CT scanning or magnetic resonance imaging are used for further analysis.^{9-11,14,15} Complete excision of the cyst by laparotomy is the final diagnostic step and choice of treatment.^{9,10,12,14} Histologic examination supported by specific immunohistochemical findings of the lymphatic endothelium (showing reactivity to factor VIII-R antigen, CD31 and, less commonly, CD34) can reliably confirm the diagnosis.¹²

In this case, the patient's transient abdominal pain a few weeks earlier was probably caused by a small contained rupture of the SA or traction or pressure caused by the pancreatic lymphangioma; no obstetric conditions could be detected. More common obstetric differential diagnostic considerations that should always be monitored among pregnant woman are uterine rupture, placental abruption, amniotic fluid embolism, and ruptured ectopic pregnancy.²⁻⁴ The large pancreatic lymphangioma could have created a tamponade effect

of the ruptured SA. A few weeks later, the patient developed severe abdominal pain followed by sudden hemorrhagic shock as a result of an initial arterial rupture or as the second phase in the 2-phase SA aneurysm rupture, leading to free bleeding into the intra-abdominal cavity. During the first emergency surgery, the lymphangioma was misinterpreted as hematoma with active bleeding of an unknown origin. The final diagnosis was based on CT angiographic findings and the second extensive surgical intervention. The rarity of both conditions contributed to the delayed diagnosis and treatment, which led to the prolonged state of shock and poorer prognosis. Despite this, the patient recovered surprisingly well.

The rupture of the SA was probably caused by traction on the arterial wall related to the pancreatic lymphangioma and increased intra-abdominal pressure related to both the large lymphangioma and pregnancy. Although an aneurysm was not diagnosed by CT scan or perioperatively, because of the urgent situation the coexistence of a small aneurysm could not be fully excluded. It is important to note that the patient was especially at risk for splenic aneurysm formation because she was pregnant. Vascular changes associated with hormonal and physiologic alterations during pregnancy could have also contributed to increased weakness of the arterial wall structure and vulnerability to mechanical stresses like traction and pressure, although likely to a lesser extent.

To the best of our knowledge, this is the first case report in which a large pancreatic cystic lymphangioma concealed the coexistence of a ruptured SA, which is another uncommon and potential life-threatening condition. The cause of the rupture was probably related to the lymphangioma and pregnancy. This diagnosis can be easily missed in pregnant women—as can a ruptured SA aneurysm—because the presentation is unclear and other obstetric emergencies seem more likely. To prevent unnecessary maternal and fetal mortality a ruptured

SA or aneurysm should always be considered in the differential diagnosis in pregnant patients presenting with (unexplained) hemodynamic instability accompanied with acute abdominal pain. Immediate surgical and radiologic evaluation is warranted in these cases.

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