Splenic Pregnancy

A Case Report and Review of the Literature

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• Primary splenic pregnancy is the least common form of extrauterine pregnancy. We report a case of splenic pregnancy occurring in a 29-year-old woman presenting with acute abdomen and hemoperitoneum. Recognition of this rare form of gestation is of critical importance, owing to the risk of exsanguination and death, and should be considered in the differential diagnosis of acute abdomen in women of reproductive age.

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Ectopic pregnancy is defined as implantation of a fertilized ovum anywhere other than within the uterine cavity and occurs with an estimated incidence of 19.7 per 1000 pregnancies.[1] The most common site of ectopic implantation is within the fallopian tube, accounting for 95.5% of all ectopic gestations.[2] Although rare, extratubal pregnancies represent some of the most serious complications of pregnancy. Approximately 1.3% of ectopic pregnancies are abdominal[2] and occur with direct implantation onto the peritoneal surface. Primary abdominal pregnancies have been described in a variety of extrapelvic organs, including omentum, liver, and small and large intestine. Preoperative diagnosis of abdominal pregnancy is difficult, and patients commonly present with signs of shock and hemoperitoneum.

The spleen is one of the rarest sites for ectopic gestation, and to our knowledge, only 9 cases of primary splenic pregnancy have been documented in the English literature to date.[3–11] We report a case of primary splenic pregnancy complicated by hemoperitoneum that, before the postoperative histologic findings, was interpreted clinically and surgically as a ruptured splenic hemangioma. We review the previously published cases to compare the clinical features, as well as discuss the pathologic criteria required for a diagnosis of primary abdominal pregnancy.

REPORT OF A CASE

A 29-year-old, gravida 1, para 1 woman status post uncomplicated caesarean section delivery 3 years earlier presented to a regional hospital emergency room with severe left upper quadrant pain. The patient reported abdominal ''fullness'' that began several weeks before admission. On the morning of admission, the discomfort progressed to severe left upper quadrant pain aggravated by movement and deep breathing. The pain radiated to the left shoulder with deep inspiration. The patient also reported feeling lightheaded and dizzy. At the time of admission, she was expecting her menstrual period and had had unprotected sexual intercourse during the last month. A urine β-human chorionic gonadotropin test was positive.

Physical examination on admission revealed a patient in obvious distress with generalized pallor. She was afebrile but diaphoretic and hypotensive, with a blood pressure of 60/40 mm Hg. Her abdominal examination was significant for diffuse abdominal tenderness with rebound and guarding. Bimanual examination was not performed.

An abdominal ultrasound revealed an enlarged spleen, measuring 17 cm in sagittal length, with an ill-defined heterogeneous focus located centrally within the splenic parenchyma that was interpreted as intraparenchymal hemorrhage. There was a moderate amount of fluid present in Morison pouch. The patient was taken for emergent laparotomy for suspected splenic rupture with hemoperitoneum.

Intraoperatively, 1.5 L of blood and blood clot were found within the peritoneal cavity. The spleen contained a hemorrhagic lesion involving the lower pole, which was actively oozing through a capsular dissection. Further inspection of the pelvic cavity revealed fibrous adhesions around both fallopian tubes and ovaries, and a simple 2-cm cyst of the left ovary. No tubal pregnancy was identified. No other source of bleeding was noted. A simple splenectomy was performed without complication. A serum β-human chorionic gonadotropin level taken postoperatively was 2544 mIU/mL and decreased to 39 mIU/mL 3 weeks after surgery. Two years later, the patient experienced a full-term intrauterine pregnancy.

PATHOLOGIC FINDINGS

The spleen weighed 226 g and measured 11.5 × 8.0 × 4.0 cm. The capsule was generally smooth and had a focal disruption at the inferior pole measuring 4.0 cm in maximum dimension. Adjacent to this capsular defect was a small amount of adherent blood clot and an irregular capsular projection, measuring 3.5 × 3.5 × 2.0 cm. On sectioning, there was a corresponding well-delineated but nonencapsulated spherical nodule of cystic, soft, and hemorrhagic red-brown to focally tan-gray soft tissue. The remainder of the splenic parenchyma was unremarkable. The hilar vessels were intact.

Microscopic examination of the hemorrhagic nodule revealed extensive blood clot with capsular disruption and subcapsular hematoma. Underlying the disruption were numerous chorionic villi and intermediate trophoblasts in-

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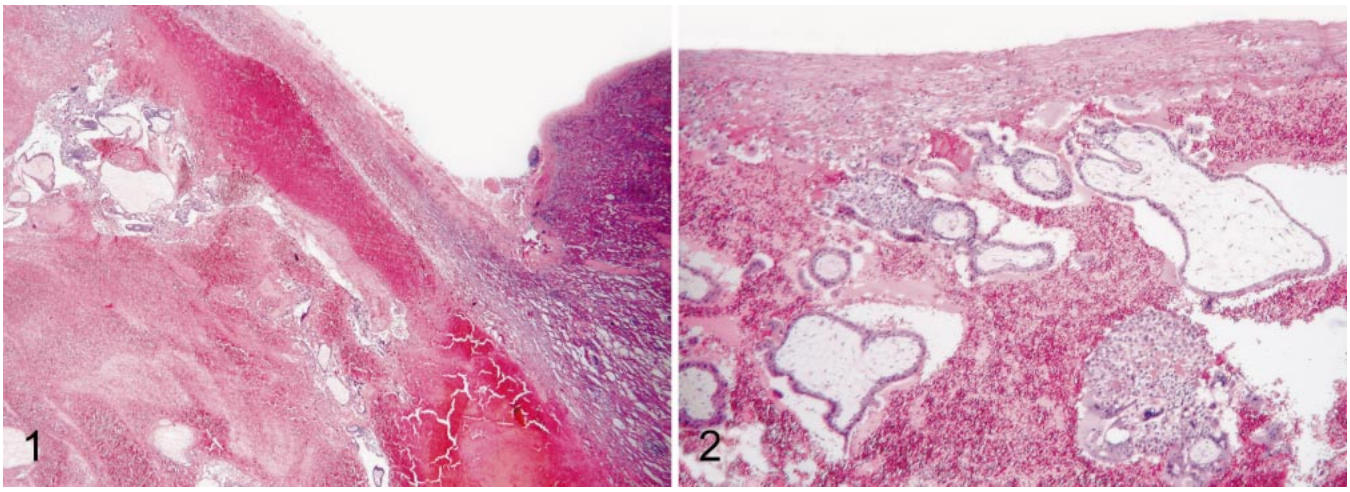

Figure 1. *Section of 3.5-cm subcapsular splenic nodule showing congested splenic parenchyma (right), extensive subcapsular hemorrhage, and immature chorionic villi (left) (hematoxylin-eosin, original magnification ×40).*

Figure 2. *Subcapsular chorionic villi and trophoblastic tissue invading splenic parenchyma (hematoxylin-eosin, original magnification ×200).*

vading the splenic parenchyma (Figures 1 and 2). A diagnosis of primary splenic pregnancy was made. No fetal parts were identified.

COMMENT

Abdominal pregnancies are defined as those occurring within the peritoneal cavity (excluding tubal, ovarian, or intraligamentous pregnancies) and account for 1.3% of ectopic pregnancies.[2] The incidence of abdominal pregnancy is estimated at 1 in 8000 births.[12] Such pregnancies are potentially life-threatening, with a maternal mortality rate of 5.1 per 1000 cases, which is 7.7 times higher than the risk from other ectopic pregnancies and approximately 90 times higher than that associated with intrauterine pregnancy.[13] The risk is particularly critical without an accurate preoperative diagnosis.

Abdominal pregnancies are classified as either primary or secondary; the latter, which is much more common, is associated with tubal rupture followed by implantation at a second site (eg, the peritoneal surface). Primary abdominal pregnancies, which arise from fertilization of an ovum within the peritoneal cavity, are extremely rare. According to Studdiford,[14] the criteria for primary abdominal pregnancy are as follows: (1) fallopian tubes and ovaries are grossly normal and show no evidence of recent injury; (2) no evidence of uteroplacental fistula; and (3) a pregnancy of no more than 12 weeks' gestation with trophoblastic elements related exclusively to a peritoneal surface. The third criterion ensures that the pregnancy is immature enough to exclude the possibility of secondary implantation to the peritoneal cavity after primary tubal pregnancy rupture. In our case, focal adhesions were noted during intraoperative examination of the fallopian tubes, but no other abnormalities were identified. There was no evidence of pregnancy outside the spleen, therefore satisfying the criteria established for primary splenic pregnancy.

Although the most common sites of primary peritoneal pregnancy are the pouch of Douglas and the posterior uterine wall,[13] primary implantation sites have been described in extrapelvic structures, including small and large intestine,[13] omentum,[11] liver,[11] and spleen.[3–11] Risk factors associated with abdominal pregnancies are similar to those of other ectopic pregnancies and include prior history of pelvic inflammatory disease, ectopic gestation, endometriosis, infertility with subsequent in vitro fertilization, and previous tubal surgery.[15]

Our review of previously published reports of primary splenic pregnancies (see Table) revealed a mean age of 27.3 years (range, 23–37 years). As with our case, most patients presented with sudden-onset left upper quadrant abdominal pain that radiated to the left shoulder. Eight of the 9 previously published cases had preoperative diagnoses of ruptured ectopic pregnancy, with the remaining case undiagnosed until the results of histologic examination. All patients survived. A variety of splenic implantation sites have been reported and range from superior to lower pole and hilum. Most gestations manifested as capsular projections and all were subcapsular in location.

Atrash et al[13] showed that abdominal pregnancy-associated deaths occur later in gestation compared to other ectopic pregnancies. Primary splenic pregnancy tends to present earlier than other abdominal pregnancies, presenting with hemoperitoneum occurring at 6 to 8 weeks' gestation. Interestingly, in 9 of 10 splenic gestations the size ranged from 2.0 to 3.5 cm, suggesting that rupture of the splenic capsule occurs when the ectopic gestation exceeds this size. Three of the 10 patients described had a known risk factor for ectopic pregnancy, namely, the presence of an intrauterine contraceptive device. There were no fetal parts identified in any of the reported cases.

The incidence of ectopic pregnancy in the United States has been steadily increasing. Atrash et al[13] found that 3 to 4 women a year in the United States die of abdominal pregnancy. In this study, Atrash et al[13] found that only 1 of 9 women alive at hospital admission had an accurate preoperative diagnosis of abdominal pregnancy and suggested that preventing abdominal pregnancy–related death depends in part on increasing awareness of its clinical characteristics. Although recognition of primary abdominal pregnancy poses a difficult diagnostic challenge, increased detection of ectopic pregnancies is made possible through improved access to ultrasonography and improved sensitivity of urine and serum β-human chorionic gonadotropin tests. Recognition of this rare form of ges-

Primary Splenic Pregnancy: Clinical Details, Management and Outcome*

Source, y	Patient Age, y	Preoperative Diagnosis	Clinical Presentation	β-hCG	Gestational Age	Implant Site	Size, cm	Outcome
Mankodi et al,[3] 1977	27	Ruptured ectopic pregnancy†	1 d of epigastric pain radiating to left shoulder	NR	Unknown	NR	3	A&W
Reddy and Modgill,[4] 1983	24	Ectopic pregnancy†	Sudden-onset lower abdominal pain and dizziness	NR	IUD for 2 mo	Lower pole	2	A&W
Huber et al,[5] 1984	23	Ruptured ectopic pregnancy†	5 h of generalized abdominal pain radiating to both shoulders	+	6 w	Lateral aspect	NR	A&W
Caruso and Hall,[6] 1984	27	Ruptured ectopic pregnancy†	4 h of lower abdominal pain radiating to left shoulder	+	6 w	Superior-posterior	2.8	A&W
Tantachamroon et al,[7] 1986	24	Ectopic pregnancy†	1 d of abdominal pain	NR	IUD for 8 mo	Superior-posterior	2	A&W
Larkin et al,[8] 1988	27	Ectopic pregnancy†	Severe chest and abdominal pain	+	8 wk	Superior pole	2	A&W
Yackel et al,[9] 1988	27	Left adnexal ectopic pregnancy	24 h of sharp shoulder and left upper quadrant pain	NR	~9 wk	Midportion	3	A&W
Kahn et al,[10] 1989	30	Intraperitoneal hemorrhage of unknown etiology	Acute epigastric and periumbilical pain radiating to left shoulder	26 000 IU/L	8 wk	Dorsal side	6	A&W
Cormio et al,[11] 2003	37	Ruptured ectopic pregnancy†	Sudden-onset left upper quadrant pain radiating to left shoulder	9278 mIU/mL	IUD for 2 y	Hilar surface	3	A&W
Present case, 2003	29	Ruptured spleen	3-wk history of worsening left upper quadrant pain	2544 mIU/mL	8 wk	Lower pole	3.5	A&W

* β-hCG indicates β–human chorionic gonadotropin; NR, not reported; IUD, intrauterine device; and A&W, alive and well.
† Preoperative diagnoses of "ectopic pregnancy" presumed by current authors to be primary adnexal gestations.

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