Primary Splenic Pregnancy-A Rare but Imperative Cause of Hemoperitoneum -Case Report and Review of Literature

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ABSTRACT

Primary splenic pregnancy is an extremely rare form of extratubal ectopic pregnancy. These cases often cause splenic rupture in very early course of their gestation thereby presenting with hemoperitoneum in emergencies. Owing to the higher risk of exsanguination and death caused by hemoperitoneum, it is essential to diagnose these cases for proper management and better prognosis of the patients. We present the case of a 23-year-old female, gravida 2, para 1, live issue 1 presenting to the emergency outpatient department with acute abdomen and hemoperitoneum. There was no history of trauma. The patient had a positive urine pregnancy test and raised beta HCG levels. Emergency laparotomy revealed an otherwise unremarkable fallopian tube and ovary with a hemoperitoneum of 2.5 liters. A tiny splenic laceration was considered to be the source of bleeding and splenectomy was performed. Microscopy was suggestive of a primary ectopic pregnancy, spleen. Since hemoperitoneum in pregnancy is a rare but potentially fatal condition with a high risk of mortality, an accurate preoperative diagnosis is crucial in the management of such patients. The possibility of a ruptured extratubal ectopic pregnancy must be considered as one of the differential diagnoses of acute abdomen with hemoperitoneum in women of childbearing age.

Key Words: Splenic rupture, Hemoperitoneum, Ectopic pregnancy, Spleen, Chorionic gonadotropin

INTRODUCTION

The implantation of a fertilized ovum anywhere outside the uterine cavity is known as ectopic pregnancy. While 95.5 % of the ectopic pregnancies are in the fallopian tube, only 1.3 % are abdominal in location (1). Such rare extratubal pregnancies occur after the direct implantation of the zygote onto the peritoneal surfaces. Although a variety of extra pelvic organs such as the liver, omentum, and small and large intestines have been described in the past, reports from primary pregnancy in the spleen are very rare. Our extensive search of the literature for the same yielded only 11 such cases reported in the past. Since ruptured ectopic pregnancy is a well recognized life-threatening entity in early pregnancy and is also one of the known causes of hemoperitoneum in pregnancy, we present an unusual case of primary splenic pregnancy presenting as acute abdomen with hemoperitoneum being misinterpreted as splenic laceration following blunt trauma to the abdomen in the emergency setting.

CASE REPORT

A 23-year-old, gravida 2, para 1, live issue 1, woman presented to the medical emergency with the chief complaints of abdominal distention along with non passage

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of flatus for one day and left upper quadrant pain, nausea and vomiting since 6 hours. The pain was aggravated on movement and deep breathing but was not radiating to the shoulder. The patient was also feeling weak and dizzy since that morning. There was no history of fever, cough, weight loss, appetite loss or any recent trauma to the chest or abdomen. The obstetric history revealed that the patient had given birth by normal vaginal delivery one year back followed by a medical termination of pregnancy 3 months back in November. Her last menstrual period was due in December at the time of presentation at the emergency and she currently had amenorrhea for 4 weeks and 2 days.

On examination, the patient was afebrile, conscious and oriented. Pallor was present but there was no icterus or pedal edema. Her blood pressure was 80/60 mm Hg and pulse was 100/min. On per abdomen examination, the abdomen was distended and diffuse tenderness was noted. The urine pregnancy test was positive and serum β -human chorionic gonadotropin levels were raised to 6565mIU/ml. Peritoneal tap attempted twice yielded blood only. Emergency transvaginal ultrasound findings were suggestive of free fluid in the pouch of Douglas and an empty uterine cavity. Hemogram findings were indicative of severe anemia with a hemoglobin of 3 gm/dl. The results of all other investigations including PT/PTTK, liver function test and kidney function test were within normal limits.

Based on the above findings, a presumptive diagnosis of ruptured ectopic pregnancy was made and the patient was referred to the gynecology department. She underwent emergency laparotomy for the same. However, intraoperatively it was noted that there was 2.5 liters of blood and blood clots in the peritoneal cavity. The uterus and bilateral adnexa were unremarkable and the source of bleeding was from a laceration in the spleen. Since the spleen was lacerated and not enlarged in size, rupture due to blunt trauma was suspected and medico legal concerns were raised. A splenectomy due to uncontrolled bleeding for a splenic laceration and a dilatation and curettage (D&C) for an incomplete abortion were performed for the patient under general anesthesia. She was given 3 units of blood transfusion postoperatively and was thereby stabilized there after.

Pathological Findings

We received the splenectomy specimen along with some blood clots in one container and an endometrial biopsy (EB) in another. On gross examination, the specimen of spleen measured 10x5.5x3.5 cm and weighed 100 gms. The outer surface was predominantly unremarkable and the capsule was smooth and glistening except at the inferior pole where a small capsular rupture measuring 1 cm in maximum dimensions was seen. The capsule over the inferior pole was also congested. On cut section, a small 1x1 cm welldemarcated nodular hemorrhagic lesion suggesting a blood clot was noted in the subscapular region at the inferior pole. A single focus in this lesion also showed tiny grey-white areas attached to its capsule (Figure 1). The rest of the spleen seemed to be unremarkable grossly. Multiple sections were taken from the spleen and examined histopathologically. The endometrial biopsy and blood clots sent together were also examined microscopically.

On histopathology, the endometrial biopsy revealed the presence of secretory endometrium with extensive decidual change in the stroma. Multiple sections examined did not show any chorionic villi. Section from the nodular lesion at the inferior pole of the spleen showed blood predominantly in the subscapular regions of the inferior pole. Numerous chorionic villi and trophoblastic tissue amongst these hemorrhagic areas were seen invading the splenic parenchyma (Figure 2). The rest of the spleen showed congestion in the splenic sinuses. Based on the above findings, a diagnosis of splenic ectopic pregnancy was made on histopathology.

DISCUSSION

Pregnancies occurring within the peritoneal cavity excluding the fallopian tube, ovaries and ligaments of the uterus are named abdominal pregnancies. These account for 1.3 % of all the ectopic pregnancies with a reported



Figure 1: Gross image of spleen showing gestational sac at inferior pole. The rest of the spleen appears unremarkable grossly.



Figure 2: Low power image of spleen showing products of conception in the parenchyma of the spleen. These products are subcapsular in location and are surrounded by the lymphoid follicles of spleen (H&E; x100).

incidence of 10.9 per 100000 live births in the United States (2,3). The maternal mortality rate of these potentially life-threatening cases is very high, accounting up to 5.1/1000 cases in the United States and as high as 7% in the Indian subcontinent (3,4). The higher incidence in developing countries is attributed to the higher frequency of pelvic inflammatory disease, as in our country (5).

Abdominal pregnancies are classified as primary or secondary depending on the site of fertilization of the ovum. When the fertilization takes place within the peritoneal cavity, the pregnancy is named a primary abdominal pregnancy and when it is associated with a tubal rupture followed by implantation at a secondary site it is named a secondary abdominal pregnancy. Primary abdominal pregnancies are extremely rare when compared to secondary abdominal pregnancies. To accurately diagnose a case with primary abdominal pregnancy, the Studdiford criteria are (1) grossly normal fallopian tubes and ovaries with 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 (6). The third criterion makes it possible to exclude secondary abdominal pregnancies. Since our case fulfilled all these criteria, we labeled it as a primary ectopic pregnancy of the spleen.

Abdominal pregnancy is associated with a very wide range of signs and symptoms due to its variable location. The clinical suspicion index is also low due to the rarity of this condition and the absence of the classical triad of abdominal pain, amenorrhea and vaginal bleeding in these cases. Various locations reported in the past include the small and large intestine, omentum, liver, diaphragm, pancreas, retroperitoneum and spleen (3,7,8). Risk factors associated with these are however similar to other ectopic pregnancies and include a history of PID, ectopic gestation, endometriosis, in vitro fertilization and previous surgeries on the tube such as tubal reconstructive surgeries or tubal recanalization surgeries (9). While there were no such risk factors in our case, 3 of 12 reviewed cases in our study had an intrauterine contraceptive device at presentation (Table I) (1, 10-19).

In accordance with the previous studies, our patient presented with chief complaints of sudden onset left upper quadrant pain and dizziness. On reviewing the previously published reports it was noted the mean age of these patients was 25.3 years, ranging from 23 to 37 years, and in 10 of 12 previously published cases a presumptive diagnosis of ruptured ectopic pregnancy was made preoperatively (Table I). There was only one case where the diagnosis was made only after histologic examination. Although the first diagnosis was ruptured tubo-ovarian ectopic pregnancy in our case, the possibility of missed abortion with splenic rupture due to blunt trauma was considered after exploratory laparotomy. Despite the fact that there was no history of blunt trauma to the abdomen, medico legal concerns were raised in our case. The final diagnosis of splenic ectopic pregnancy in our case was made only after histological examination.

Due to the rarity of such cases and the complexity of the diagnosis, abdominal pregnancies are often missed preoperatively. In a study conducted by Costa et al. it was seen that even ultrasound coupled with clinical evaluation had a success rate of only 50% in the diagnosis (20). The guidelines for the use of USG to diagnose abdominal pregnancy as provided by Allibone et al. (21) still have reported diagnostic errors of 50-90% in various case series. In recent years, only one study quotes the preoperative diagnosis of splenic pregnancy made with transvaginal USG and its successful management by laparoscopy (19). In our case unfortunately the diagnosis of splenic ectopic pregnancy was missed on USG.

Splenic implants have been reported at a variety of sites ranging from the superior pole to the lower pole and hilum. In some cases, the ectopic gestation manifested as capsular projections but in all the cases the subcapsular location was a phenomenon. In our case also, no capsular projection could be identified on gross examination though the gestational sac was seen in the subcapsular region at the lower pole of spleen.

According to the previously reported literature, splenic gestations range in size from 2 to 3.5 cm and present with hemoperitoneum at 6-8 weeks of gestational age. It is hypothesized that the earlier presentation of splenic pregnancies as compared to the other abdominal pregnancies may be because of the rupture of the splenic capsule at a much smaller size of gestational sac than other abdominal pregnancies (3). In accordance with the previous studies our patient presented with hemoperitoneum even earlier at only 4 weeks of gestation with a gestational sac of 1x1 cm only. This could be due to its very superficial location just beneath the capsule in the lower pole of spleen.

In conclusion, since hemoperitoneum in pregnancy is a rare but potentially fatal condition with a high risk of mortality, an accurate preoperative diagnosis is crucial in the management of such patients. The possibility of a ruptured extratubal ectopic pregnancy must be considered as one of the differential diagnoses of acute abdomen with hemoperitoneum in women of childbearing age.

S.no	Source	Age	Preop Diagnosis	Clinical Presentation	β-hCG	G. Sac	Implant Site	Size (cm)	T/t outcome
1	Mankodi et al ¹⁰	27	Ruptured ectopic pregnancy	1 d of epigastric pain radiating to left shoulder	NR	Unknown	NR [#]	3	A&W ^{\$}
2	Reddy and Modgill ¹¹	24	Ectopic pregnancy	Sudden-onset lower abdominal pain and dizziness	NR	Unknown (IUD x 2months)	Lower pole	2	A&W
3	Huber et al ¹²	23	Ruptured ectopic pregnancy	5 h of generalized abdominal pain radiating to both shoulders	+	6 wks	Lateral aspect	NR	A&W
4	Caruso and Hall ¹³	27	Ruptured ectopic pregnancy	4 h of lower abdominal pain radiating to the left shoulder	+	6 wks	Superior- posterior	2.8	A&W
5	Tantachamroon et al ¹⁴	24	Ectopic pregnancy	1 d of abdominal pain	NR	Unknown (IUD for 8 months)	Superior- posterior	2	A&W
6	Larkin et al ¹⁵	27	Ectopic pregnancy	Severe chest and abdominal pain	+	8 wks	Superior pole	2	A&W
7	Yackel et al ¹⁶	27	Left adnexal ectopic pregnancy	24 h of sharp shoulder and left upper quadrant pain	NR	~9 wks	midportion	3	A&W
8	Kahn et al ¹⁷	30	Intraperitoneal hemorrhage of unknown etiology	Acute epigastric and periumbilical pain radiating to left shoulder	26 000 IU/L	8 wks	Dorsal side	6	A&W
9	Cormio et al ⁸	37	Ruptured ectopic pregnancy	Sudden-onset left upper quadrant pain radiating to left shoulder	9278 mIU/ mL	Unknown (IUD x 2 years)	Hilar surface	3	A&W
10	Kalof NA et al¹	29	Ruptured spleen	left upper quadrant pain	2544 mIU/ mL	8 wks	Lower pole	3.5	A&W
11	Siddiqui MN et al ¹⁸	30	Splenic ectopic Pregnancy	left upper quadrant pain	N	Unknown	NR	NA	A&W
12	Gang G et al ¹⁹	32	Splenic ectopic Pregnancy	Pain lower abdomen and spotty bleeding	38,913.3 IU/L	6 wks	NR	4.3 mm	A&W
13	Current study	23	Missed abortion with splenic laceration	Pain abdomen	6565 mIU/mL	4 wks	Subcapsular splenic parenchyma, inferior pole	1.5	A&W

Table I: Comparison of findings in other studies with our report.

 β -hCG: Human chorionic gonadotropin, G.Sac: Gestational Sac, NR: not reported; IUD: Intrauterine device, A&W: Alive and well.

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