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CASE REPORT

Spontaneous hemoperitoneum in pregnancy due to rupture of uterine vessels in woman with endometriosis: a case report

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ABSTRACT

Background. Spontaneous rupture of uterine vessels is a rare and life-threating event than can rarely occur during spontaneous and low-risk pregnancies. The definitive association between adverse obstetrical events and pelvic endometriosis is still under evaluation. We report a severe case of spontaneous hemoperitoneum in pregnancy (SHiP) due to a rupture of the uterine vessels related to decidualized endometriosis.

Case presentation. A 38-year-old primigravida woman at 33 weeks of pregnancy with an uncomplicated pregnancy and a history of endometriosis was admitted to the emergency room of our Institution, due to a spontaneous rupture of the right uterine vessels. An alive and vital baby was delivered by hysterotomy. In order to achieve hemostasis, a total hysterectomy with bilateral salpingectomy was performed.

Conclusions. In case of spontaneous rupture of uterine vessels and resulting hemoperitoneum, prompt diagnosis and treatment are the crucial points in order to minimize maternal and fetal/neonatal complications. Further studies are necessary in order to identify endometriosis as a possible cause of spontaneous rupture of uterine vessels in pregnancy.

Key words

Spontaneous rupture of uterine vessels; endometriosis; hemoperitoneum; case report

Introduction

Spontaneous hemoperitoneum in pregnancy (SHiP) is a rare and dramatic complication correlated with a high rate of maternal and fetal/neonatal mortality. Approximately only 100 cases have been

reported in the literature, and since 1950 the maternal mortality rate has been 49.3% [1] [2]. However, nowadays, thanks to medical advances, the mortality rate dropped to 3.6% [3] [4]. Although the etiology of SHiP remains unclear, hemodynamics and hormonal factors have been involved in the pathogenesis [1]. Endometriosis represents a benign chronic disease characterized by the presence of functional endometrial tissue out-side of the uterus. In this sense, the endometriotic lesions undergoing the process of decidualization generate a subsequent inflammatory microenvironment. For all these reasons, endometriosis has been suggested to be involved in the mechanisms of spontaneous rupture of uterine vessels during pregnancy [5]. Because of the increased number of patients with severe endometriosis with the desire for fertility, physicians would need to consider endometriosis-related SHiP among possible causes of hypotension and acute abdomen during the third trimester of pregnancy. Here, we describe a case of spontaneous hemoperitoneum in a healthy woman at 33 weeks of her first spontaneous pregnancy.

Case presentation

A 38-year-old primigravida woman was admitted to the emergency room of our Institution for abdominal pain at week 33 of pregnancy. There was no history of vaginal bleeding, rupture of membranes, abdominal trauma, previous abdominal surgeries, or drug assumptions. The patient had a medical history of deep infiltrating endometriosis treated with progestin-only drugs with good response. The pregnancy was spontaneous, and the antenatal course was uneventful until the admission. On admission, the patient was hypotensive with a blood pressure of 80/50 mmHg, heart rate was 120 beats per minute, respiratory rate was 14 breaths per minute, and body temperature of 36.3°C. The physical examination detected a gravid abdomen, tender at the superior quadrants. No dysuria, vomiting, or diarrhea was reported. The ultrasound assessment confirmed the presence of a singleton cephalic fetus with a normal biophysical profile, a regular placenta, and normal amniotic fluid volume. Moderate maternal abdominal free liquid was detected. Cervical length was 25 mm, and tocography revealed no uterine contractions. Her laboratory tests resulted in a hemoglobin level of 9.0 g/dL and 18.20 x 10^3/uL leukocytes. Metabolic hepatic panel and urinalysis were negative. Intramuscular steroid therapy was submitted in order to induce fetal lung maturation.

After 6 hours, the value of haemoglobin level dropped to 7.9 g/dL, and the free abdomen liquid detected at the ultrasound assessment was significantly increased. Moreover, the patient presented an exacerbation of abdominal pain, despite analgesic infusion.

Ten hours after the arrival at the emergency room, considering the worsening of clinical features and the suspicious diagnosis of hemoperitoneum, the patient underwent urgent laparotomy. About 1.5 L of free blood was aspirated from the abdominal cavity. The surgical exploration revealed a hematoma in the posterior and right uterine walls and active bleeding from the right uterine vessels. Moreover, there were several endometriotic foci in the pelvic peritoneum and severe pelvic adhesions. There was no sign of uterine anomalies, such as arteriovenous malformation and uterine rupture. The surgery proceeded with a low-segment caesarean section and extraction of a male fetus, alive and vital, weighed 1.400 kg, and with Apgar scores of 8 and 9 at the first and fifth minute, respectively. After uterine closure, persistent bleeding appeared from decidualized endometrial lesions on the posterior surface of the uterus and the right parametrium. Because of the difficulty in achieving safe hemostasis without possible damage to the parametrial structures, we decided to proceed with a total hysterectomy. After intraoperative patient's oral consensus, a total hysterectomy with bilateral salpingectomy and ovarian preservation was performed. Successful hemostasis was finally achieved, and the abdominal wall was closed. Estimated blood loss during the hysterectomy was 700 mL, and no intraoperative transfusions were performed. No intraoperative and postoperative blood transfusions were performed. No complications were

reported, and the patient was discharged after 6 days. The infant had an uneventful course and was discharged after few weeks in good condition. On histopathology examination, uterine, tubal, and right parametrial specimens have reported elements suggestive of endometriotic foci, such as haemorrhagic infarction, fibrosis and prominent deciduoid changes.

Discussion

The current study represents a clear case of spontaneous hemoperitoneum in pregnancy (SHiP) as direct consequence of spontaneous rupture of uterine vessels due to endometriotic lesions in an otherwise uncomplicated pregnancy.

Endometriosis has increased in recent decades and is frequently associated with infertility, pelvic pain, and dysmenorrhea. Endometriosis is a very complex condition that could impact sexuality, quality of life and psychology of affected woman. Although these aspects could not be correlated to the severity of disease, they have an important role on psychological wellbeing and interpersonal relationships [6] [7]. Endometriosis can be correlated with pregnancy complications, such as severe preeclampsia, placental abruption, placental abnormalities, premature rupture of membranes, preterm birth, and retained placenta [8] [9].

SHiP is a rare and potentially life-threatening condition that occurs in pregnant women out-of-labor in 61% of cases, of which 39% happened between 33-37 weeks of gestation [10] [11]. According to the International Network of Obstetric Survey Systems (INOSS), SHiP is defined as a non-traumatic intraperitoneal hemorrhage during pregnancy up to 42 days postpartum, excluding ectopic pregnancy, uterine rupture and cesarean section-associated bleeding [12].

In literature, trends regarding parity, age, and length of gestation in patients with SHiP have not been documented [1].

In a review of 25 cases of SHiP, endometriosis has been recognized as the major risk factor and the spontaneous rupture of uterine vessels or direct bleeding of endometriotic lesions were the most common findings .

Moreover, in a recent systematic review by Lier et al., the authors reported that the SHiP was associated with rupturing utero-ovarian vessels in 57% of cases, endometriotic implants in 23% of cases, hemorrhagic nodules in 2% of cases, and a combination of these events in 20% of cases [13].

Furthermore, in almost half of the patients reported in the literature, the diagnosis of endometriosis was misunderstood until the laparotomic visualization of endometriotic lesions and the histological confirmation [10]. Conversely, in our case, the diagnosis of endometriosis was already known at the time of clinical presentation, and the histopathologic examination of the samples confirmed the presence of decidualized ectopic endometrial tissue.

It is well known that the phenomenon of decidualization during the first trimester of pregnancy consists of the loss of pigmentation and fibrosis of endometriotic implants [10]. Recently, it has been supposed that the SHiP is linked to an involution of the decidualization process due to the decrease of progesterone levels and a supposed progesterone resistance. This mechanism causes the production of chemokine, proinflammatory cytokine, metalloproteinases, apoptotic factors, cell death, and bleeding [10].

The incidence of SHiP may be influenced by the use of assisted reproductive techniques (ART), as women with endometriosis could overcome subfertility/infertility problems [14]. The use of ART is linked to a high dosage of progesterone, which can facilitate the process of decidualization. In a recent review of 362 pregnancies reported from 2010 to 2018, *Benaglia et al.* documented that the

frequency of SHiP in women with endometriosis submitted In vitro fertilization is 0.3% [15]. However, in our case, the patient had a spontaneous pregnancy. Besides endometriosis and ART as risk factors, a recent prospective population-based study reported some additional factors associated with SHiP, such as multiple pregnancies, ≥35 years of age in mothers, and previous abdominal surgery [11].

Although the recent evidence, the etiopathogenesis of this condition remains unclear. Increased venous pressure in utero-ovarian circulation due to pregnancy status or muscular activity such as defecation and coughing could be possibly implicated in the pathophysiology of SHiP [16].

In the literature, three factors have been described as explanations for spontaneous rupture of uterine vessels: vessels leakage caused by endometriosis-linked chronic inflammation; adhesions between vessels with relative tensions; decidualization of endometrial foci [5] [17] [18]. Our patient had either a diagnosis of deep infiltrating endometriosis or pelvic adhesions. Indeed, during the surgery**,** adhesiolysis was performed. Moreover, in a few cases, the origin of the bleeding remains unknown, even during laparotomy. During the surgery of our patient, arteries and superficial veins of the posterior surface of the uterus and right parametria have been involved in the bleeding. The surgical visualization of the endometriotic implants' bleeding and the medical history of our patient suggest that the phenomenon of decidualization of endometriotic foci lead to massive and sudden hemoperitoneum in our patient.

In all cases of spontaneous hemoperitoneum in pregnancy, the onset symptoms were acute or subacute abdominal pain, free abdominal fluid, hypovolemic shock, and decreasing values of hemoglobin [10]. A prompt differential diagnosis is a crucial requirement. Placental abruption, uterine rupture, placenta percreta, appendix, hepatic, and splenic ruptures are the most common preoperative misdiagnosis. Vascular sources of hemoperitoneum in pregnancy should be considered as a result of the rupture of a visceral abdominal artery aneurysm such as splenic artery [1].

Our patient's symptoms were similar to clinical presentation described in the literature. Lier et al. reported the most common signs of presentation of SHiP: subacute abdominal pain (94.9%), a decreased level of hemoglobin (62.7%), imaging showing free peritoneal fluid (62.7%) [13]. The sensibility of contrast enhanced computed tomography in the identification of bleeding is documented, but maternal and fetal potential risks from ionizing radiation have to be considered. Ultrasonography could be helpful for the detection and monitoring of abdominal free fluid, but the real diagnosis is often obtained only by laparotomic exploration. In a preterm pregnancy, the decision making should be balanced between risks related to prematurity, delayed diagnosis, and maternal complications. In our case, the worsening symptoms and the hemoglobin drop level guided our decision on laparotomy.

Several questions remain unknown about the management of endometriosis in pregnancy: whether any medical or surgical treatment of endometriosis in the preconception period would add any benefit and prevent pregnancy complications such as SHiP; whether adopting any particular management in pregnancies with a previous diagnosis of endometriosis (in terms of follow-up and mode of delivery); whether to choose any specific flowchart in case of a pregnant woman with acute abdomen and free blood abdominal liquid. Yet, endometriotic lesions should be considered a possible cause of hemoperitoneum during the third trimester of pregnancy [5]. Rapid diagnosis and prompt intervention are essential to correctly manage such complicated cases.

Conclusions

In conclusion, our case represents a rare case of SHiP related to endometriosis confirmed by histologic examination. Moreover, a prompt diagnosis of SHiP was crucial in managing this unique

clinical scenario without either maternal or fetal complications. Exploring the association between the diffusion of endometriosis and the severity of SHiP could be a new challenge.

In consideration of the risk of spontaneous rupture of uterine vessels and SHiP, physicians should be aware that prompt diagnosis and interventions are crucial to minimize maternal and fetal/neonatal morbidity and mortality. More attention to SHiP, with a particular focus on endometriosis as a cause, would help prevent maternal and fetal adverse events.

Abbreviations

SHiP: Spontaneous Hemoperitoneum in Pregnancy; ART: assisted reproductive techniques.

COMPLIANCE WITH ETHICAL STANDARDS

Authors contribution

A.M., F.F.: Conceptualization; G.Z: Writing – original draft; A.M, F.F.: Writing – review & editing. A.M., G.Z., G.P., A.M. F.F.: Data curation, Validation.

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Study registration

Not applicable.

Disclosure of Interests

The authors declare that they have no conflict of interests.

Ethical Approval

Not applicable.

Informed consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Data sharing

The data that support the findings of this study are available from the corresponding author upon reasonable request.

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Fig. 1: The right parametrium and the posterior surface of the uterus are covered by blood clots as site of active bleeding.