



Spontaneous Splenic Vein Rupture with Massive Hemoperitoneum during the Third Trimester of Pregnancy

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Abstract

We describe a case of spontaneous rupture of the splenic vein in a pregnant patient at 33 weeks gestation. For the first time in literature, we report follow-up investigations aimed at understanding the cause of the event.

The woman was admitted to the emergency ward for hypovolemic shock. Maternal and fetal distress prompted an immediate cesarean section. The fetus was delivered stillborn, hemoperitoneum was present and two spontaneous splenic vein's lacerations were found to be the source of the bleeding. The immediate splenectomy and aggressive correction of anemia and coagulopathy determined maternal survival. During the follow-up, no evidence of neither congenital nor acquired causative factors for a splenic vein rupture was found.

The aim of reporting this case is to increase the awareness between clinicians of this condition since it is both rare and with an aspecific clinical picture. It presents itself in apparently low-risk patients and a good maternal-fetal outcome can only be achieved by early diagnosis and prompt treatment.

Keywords

Pregnancy, Hemoperitoneum, Splenic Vein, Follow-up Investigations

Introduction

Spontaneous rupture of the splenic vein is a rare occurrence. It represents a potentially catastrophic event since it leads rapidly to retroperitoneal haemorrhage and hypovolemic shock. The lack of specificity of the presenting symptoms and signs of

this condition makes a prompt diagnosis difficult for the clinician [1].

A number of cases of spontaneous rupture of the splenic vein have been reported to occur in pregnancy [2]. The reason for the higher incidence of this event

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in pregnant women is currently not completely understood. This occurrence seriously jeopardizes the life of the mother and the child, and carries a very high mortality risk for both [3].

We describe a case of spontaneous vein rupture that occurred during the third trimester of pregnancy. It was thought worthwhile to report this case since the rarity of the condition and the non-specific clinical picture pose a diagnostic challenge. Only a prompt diagnosis may prevent an unfavourable prognosis for both the mother and the baby. Moreover, for the first time in literature, it is available a detailed follow up of the patient, by which we demonstrated that she did not suffer from any disease that might be accounted to be the definitive cause of the splenic vein rupture: no congenital aneurism was present, nor an inherited or acquired connective tissue disease, nor an acquired disorder that might have caused an increase in blood pressure in the lineal vessels. Only a gene II mutation was found, this might have played a causative role in this occurrence together with hormonal and hemodynamic pregnancy modifications.

Case Presentation

A 32-year-old woman, gravida II, para I, was admitted to the emergency ward of Santo Stefano Hospital, Prato, Italy, for loss of consciousness and vomiting at 33⁺³ weeks of gestation.

The woman had no past medical history of note. She reported a first uneventful pregnancy. During her second pregnancy, she complained about dyspnoea and was therefore referred to our Hospital's Maternal-Fetal Unit at 23 weeks. Since her weight gain in pregnancy was excessive she had a consultation with a dietitian. Cardiologic examination was performed; thyroid function and haemoglobin were investigated with normal results. Blood pressure monitoring revealed gestational hypertension that was treated with alpha methyl dopamine (250 mg/ die) from the 28th week of gestation.

According to the patient's husband's report, there was no history of trauma. She was collecting her first child from school when suddenly she collapsed. On admission, the woman's examination revealed

lethargy, pale and sweaty skin, loss of bowel control, hypotension. Blood exams showed anemia - Haemoglobin 6.6g/dL - platelet count $287 \times 10^3/\mu\text{L}$, activated partial thromboplastin time (aPTT) 30 s (normal range 25-35), prothrombin activity ratio 1.39 (normal range 0.82-1.19), fibrinogen 216 mg/dL (normal range 200-400), AT III 30% (normal range 80-120). Arterial blood gas test demonstrated metabolic acidosis. Obstetric examination showed a not contracted uterus and no bleeding from the genital tract was present. Ultrasound revealed marked fetal bradycardia.

Due to the maternal shock and fetal distress, the patient was immediately transferred to the Operating Room and the gynecologist performed an emergency cesarean section.

The abdomen was opened through a Pfannenstiel skin incision and a massive hemoperitoneum was found. The uterus was found intact. A transverse incision along the lower uterine segment was made and a stillborn, 2650 grams (98th centile for gestational age), male infant was delivered. After placental extraction, a single-layer closure of the lower uterine segment was performed.

The surgeon then performed a vertical midline skin incision in order to determine and control the source of the haemorrhage. The blood clots were evacuated and the exploration of the abdomen revealed a fresh blood loss in the retroperitoneum, the spleen was found normal with an intact capsule and a massive haemorrhage was seen from two splenic vein's lacerations. To stop the bleeding the splenic vein was ligated. Then, the vessels of the hilum were clamped and ligated and splenectomy was performed.

The patient was transfused with 4 units of red blood cells and 1 unit of frozen fresh plasma to correct anemia and coagulopathy.

At the end of the procedure, the patient was admitted to the Intensive Care Unit, she was extubated, hemodynamically stable, and conscious. Twelve hours later Haemoglobin was 9.7 g/dL, platelet count $100 \times 10^3/\mu\text{L}$, activated partial thromboplastin

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time (aPTT) 24 s (normal range 25-35), prothrombin activity ratio 1.15 (normal range 0.82-1.19), fibrinogen 294 mg/dL (normal range 200-400), AT III 58% (normal range 80-120). The postoperative course was uneventful and the patient was discharged home on the X postoperative day.

The patient suffered no sequelae. Vaccinations against Pneumococci, Haemophilus Influenza Type B, Meningococci, and Influenza virus were administered as recommended for asplenic patients. She was offered psychological support. She underwent several consultations at our outpatient clinic in order to find the possible cause of the spontaneous rupture of the splenic vein. No histopathologic evidence of aneurism was found. A consultation with a rheumatologist did not show any autoimmune disease affecting the connective tissue, nor cirrhosis, nor JAK2 positive myeloproliferative neoplasms. Lastly, she underwent a genetic consultation and analysis failed to show any inherited connective tissue disease. She was proven negative at screening for disorders of haemostasis except for gene II mutation.

Discussion

Retroperitoneal hemorrhage due to the spontaneous rupture of the spleen or of its vessels in pregnancy is a rare and catastrophic event with a very high maternal and fetal mortality rate [3]. Tanchev et al. reported a “splenic emergency syndrome” in pregnancy [2], characterized by violent, spontaneous pain in the left hypochondrium or in the epigastrium followed by a hemorrhagic shock. Other symptoms as nausea and vomiting may be present. The cause of this event has

been reported to be more frequently the rupture of an aneurism of the splenic artery [4] (over 100 cases described in the literature). More rarely this occurrence was related to the spontaneous rupture of the splenic vein, as described for our patient. In the scientific English literature, only a few cases of this condition have been reported [5-13], these cases have been summarized in **Table-1**. Even rarer the cause is the rupture of an aneurism of the splenic vein [14] or the rupture of the spleen itself [2]; a unique case of rupture of venae gastricae breves have been reported [15].

There are two possible etiologies of a spontaneous rupture of a splenic vessel [16]: a congenital disease (e.g a vascular aneurism or an inherited connective tissue disease) or an acute event (e.g. a thrombosis that may be caused by increased pressure in the vessel).

To the best of our knowledge, our case is the first in the English literature in which follow-up investigations are available and a detailed search for the possible cause of the event was performed. With regard to our patient, no congenital aneurism was found, and she did not suffer from an inherited connective tissue disease. As far as a thrombotic event is concerned, possible causes of increased pressure in the lineal vessels were all ruled out: the patient was hepatitis B and C virus-negative, she wasn’t ill with any autoimmune disease and no cirrhosis was present, JAK2 mutations were negative and she did not suffer from any Myeloproliferative neoplasms. The possible etiology in our case is a splenic vein thrombosis that might be due to the pro-thrombotic changes of pregnancy and to the gene II mutation.

Table-1: Summary of available literature about splenic vein rupture in pregnancy

First Author	Year of Publication	GA at onset	Outcome: Mother	Outcome: Fetus
Shepard D [12]	1961	32 weeks	Discharged 9 days after CS	Stillbirth
Eckerling [11]	1962	40 weeks	Discharged 10 days after CS	Stillbirth
Wheelock JB [9]	1982	34 weeks	Discharged 10 days after CS	Stillbirth
Newman B [7]	1963	38 weeks	Demise 1 day after CS	Demise 1 hour after CS
Madhavan P [10]	1998	34 weeks	not reported	Stillbirth
Turan N [13]	2007	20 weeks	Immediate demise	Stillbirth

*CS = Cesarean Section

The possible reason for the higher risk of a spontaneous rupture of splenic vessels in pregnant women is currently not understood. The total blood volume and cardiac output increase in pregnancy and splenic artery outflow raise [17]. Altered levels of reproductive hormones (namely estrogen, progesterone, and relaxin) may affect the elasticity of vascular tissues [3]. Moreover, haemostatic and mechanical factors may also play a role [3].

This occurrence is more frequent (two-thirds of the cases) [4] during the third trimester of pregnancy [4,11,12] when the predisposing factors typical of the pregnant patient are the most likely to cause an issue, as it happened for our patient. However, some cases have been reported during the second trimester [13] or during the post-partum [14] period as well.

The mortality rate has been described as high as 70% for mothers and even higher (almost 95% [4]) for the babies. Early consideration of a diagnosis of a ruptured splenic vessel might increase the likelihood of survival of the woman and the baby. The increase in circulating volume of the pregnant woman might make it harder to recognize hemodynamic instability since signs of hypovolemia only appear when 35% or more of the total circulating volume is lost [17]. On the other hand, fetal distress occurs immediately, because to compensate for the hypovolemia the blood flows to the placenta decreases [3]. Placental abruption is one of the most commonly made misdiagnoses, together with uterine rupture, rupture of other vessel, and pulmonary embolism since they share similar clinical features [3].

The survival of the mother in our case was due to the immediate decision to perform a rapid exploratory laparotomy with the aim of delivering the fetus quickly and control the source of the bleeding. In a pregnant woman with acute severe hypovolemia, performing a cesarean section is indeed part of the haemodynamic stabilization [3]. Unfortunately, it was too late for the baby as in the majority of the cases reported in the literature (the few cases of fetal survival are described in women who were already admitted at the hospital for other reasons [18]). A possible criticism of our management is that a midline incision was probably to

be preferred over a Pfannenstiel incision to create better exposure; however, this would have not improved the fetal outcome.

Conclusion

Spontaneous rupture of the splenic vein may occur during pregnancy. The chances of survival of the patient and the fetus depend considerably on the early detection and rapid surgical intervention. We hope that greater awareness of this condition may lead to faster diagnosis and improved survival rates for mother and fetus. Our follow-up tests demonstrated that no clear predisposing factors can be identified, therefore obstetricians and other frontline health care workers should always consider the rupture of the splenic vein as a possible cause in the case of a pregnant woman presenting with hypovolemic shock.

Conflict of Interests

All authors have read and approved the final version of the manuscript. The authors have no conflicts of interest to declare.

Author Contributions

All Authors helped in conceptualizing and designing the study, drafting the manuscript, reviewing and revising it. All authors approved the final manuscript as submitted and agree to be accountable for all aspects of the work.

References

- [1] Safioleas MC, Moulakakis KG. A rare cause of intra-abdominal haemorrhage: spontaneous rupture of the splenic vein. *Acta Chir Belg.* 2006 Mar-Apr;106(2):237-39. [PMID: 16761488]
- [2] Tanchev S, Popova M, Slavov I. Sindromút "speshnost ot slezkov proizvod" po vreme na bremennost (s p na dva sluchaia) [The "splenic emergency syndrome" during pregnancy (a report of 2 cases)]. *Akush Ginekol (Sofia).* 1992;31(1):32-34. Bulgarian. [PMID: 1342149]
- [3] Kilpatrick CC. Approach to acute abdominal pain in pregnant and postpartum women. *UpToDate.* 2019 Apr 15.
- [4] Sadat U, Dar O, Walsh S, Varty K. Splenic artery aneurysms in pregnancy--a systematic review. *Int J Surg.* 2008 Jun;6(3):261-65. [PMID: 17869597]

Case Report

- [5] RAHN J, STEFFEN FW. [Splenic vein rupture during pregnancy following acute pancreatic necrosis]. *Zentralbl Gynakol.* 1959 Nov 14;81:1823-30. German. [PMID: 14435869]
- [6] Puder H. [Thrombosis and rupture of the splenic vein in "acute obstetrical hypercoagulemic syndrome"]. *Zentralbl Gynakol.* 1961 Aug 26;83:1370-75. German. [PMID: 14489122]
- [7] Newman B, Gaal GZ, Jackson E. Spontaneous rupture of the splenic vein during pregnancy. *South Med J.* 1963;56:11-13.
- [8] Bronstein R, Morin P. Urgence splénique et grossesse [Splenic emergencies and pregnancy]. *J Chir (Paris).* 1975 Jan;109(1):63-74. French. [PMID: 109270]
- [9] Wheelock JB. Spontaneous splenic vein rupture in pregnancy: case report. *Mil Med.* 1982 Jan;147(1):64. [PMID: 6806687]
- [10] Madhavan P, Hegarty P, Akram M, Drumm J. Spontaneous rupture of the splenic vein in pregnancy. *Ir Med J.* 1998 Mar-Apr;91(2):64. [PMID: 9617035]
- [11] Eckerling B, Goldman JA, Yado S. Spontaneous rupture of the splenic vein in pregnancy with massive retro- and intraperitoneal hemorrhage. *Am J Surg.* 1962 May;103:836-39. [PMID: 13889097]
- [12] Shepard D, Foster CE. Spontaneous Rupture of the Splenic Vein During Pregnancy. *Ann Surg.* 1961 Jun;153(6):1029-32. [PMID: 17859665]
- [13] Turan N, Oghan F, Boran T. Spontaneous rupture of splenic vein in a pregnant woman during a religious ritual. *J Forensic Leg Med.* 2007 Oct;14(7):440-43. [PMID: 17720597]
- [14] Parpaglioni R, Metta E, Zagari A, Celleno D. Spontaneous splenic vein aneurysm rupture in the puerperium. *Int J Obstet Anesth.* 2009 Jan;18(1):48-51. [PMID: 18684614]
- [15] Knezovic Z, Stojanovic I, Djakovic I, Vrdoljak DV, Kosec V, Butorac D. Spontaneous Rupture of Venae Gastricae Breves in Pregnancy: Case Report. *Med Arch.* 2019 Oct;73(5):359-61. [PMID: 31819312]
- [16] Renzulli P, Hostettler A, Schoepfer AM, Gloor B, Candinas D. Systematic review of atraumatic splenic rupture. *Br J Surg.* 2009 Oct;96(10):1114-21. [PMID: 19787754]
- [17] Aguree S, Gernand AD. Plasma volume expansion across healthy pregnancy: a systematic review and meta-analysis of longitudinal studies. *BMC Pregnancy Childbirth.* 2019 Dec 19;19(1):508. [PMID: 31856759]
- [18] Al Asfar F, Saber M, Dhar PM, Al Awadhi N. Rupture of splenic artery aneurysm during labor: a case report of maternal and fetal survival. *Med Princ Pract.* 2005 Jan-Feb;14(1):53-54. [PMID: 15608482]