

## Spontaneous rupture of splenic artery aneurysm during a multiple pregnancy

### Rotura espontânea de aneurisma da artéria esplênica durante uma gravidez múltipla

Catarina Peixinho<sup>1</sup>, Mafalda Laranjo<sup>1</sup>, Marta Rodrigues<sup>2</sup>, Fátima Soares<sup>2</sup>, Joaquim Silva Mendes<sup>3</sup>  
Hospital Pedro Hispano

#### Abstract

We report a rare case of a 39-year-old woman, twin pregnancy with 32 weeks' gestation, who underwent an emergency caesarean section and subsequent exploratory laparotomy with splenectomy due to hypovolemic shock secondary to splenic artery aneurysm rupture. The patient was admitted for observation due to suspicion of threatened preterm labour. The final diagnosis was not considered due to its rarity. The postoperative course was favourable for the mother, the newborns died. Spontaneous rupture of splenic artery aneurysm during pregnancy is a life-threatening event. This condition is commonly asymptomatic, and diagnosis is often incidental: during an ultrasound, intraoperatively or at autopsy.

**Keywords:** Rupture; Splenic artery aneurysm; Pregnancy

Corson first described the association of splenic artery aneurysm (SAA) with maternal death in 1869, when he documented the sudden and unexpected death of a 29-year-old multigravida at eight months' gestation. The diagnosis was made following post-mortem examination<sup>1</sup>. Since then, SAA has been identified as a rare condition associated with significant maternal and foetal mortality, with rates as high as 75% and 95% respectively<sup>2</sup>. The patient may present without symptoms or with abdominal pain, a frequent symptom in pregnancy which can mimic other more common pregnancy conditions<sup>3</sup>. The condition is commonly asymptomatic and as a result, SAA diagnosis is often incidental: at the time of ultrasound examination, intraoperatively during an emergency c-section or at autopsy<sup>1</sup>. Surgical intervention pre-rupture is rare; most reports describe the identification of the aneurysm post-rupture<sup>2</sup>.

We report a case of maternal collapse in a twin preg-

nancy due to spontaneous rupture of a SAA at 32 weeks' gestation.

#### CASE REPORT

A 39-year-old Caucasian woman presented at the Gynaecology and Obstetrics emergency department at 32 weeks' gestation. It had been an uneventful dichorionic diamniotic twin pregnancy. The patient presented with moderate abdominal pain, mainly on the left side. There was no previous trauma history or other additional symptoms. On physical examination, the patient was conscious and oriented. Both skin and mucosa were of normal colouration; vital signs were within normal range: blood pressure 125/70 mmHg, heartbeat 73 bpm, body temperature 36.4°C. Abdominal palpation revealed subtle abdominal pain on the left side, with no guarding or rebound tenderness. At gynaecological examination a white discharge was present with no evidence of previous or active loss of amniotic fluid; the cervix was soft, 30% effaced and closed. Transvaginal ultrasound showed a 22 mm length cervix without funneling. At transabdominal ultrasound scan both foe-

1. Interna de Ginecologia e Obstetrícia

2. Assistente Hospitalar, Ginecologia e Obstetrícia

3. Assistente Hospitalar, Cirurgia Geral

tuses were cephalic, with normal foetal heart rate and movements, amniotic fluid volume was normal, with no apparent retroplacental hematomas. Cardiotocography was normal by FIGO classification, with irregular uterine contractions.

Blood tests were performed and revealed haemoglobin 9.8g/dL, leucocytes 13.99/  $\mu$ L, C-reactive protein 7.5 mg/L, and normal renal and hepatic functions.

Paracetamol and tramadol were administered for initial pain management. This improved but did not completely resolve symptoms, so the patient was admitted to the Maternal Unit for observation, due to suspicion of threatened preterm labour. Dexamethasone for lung maturity was initiated.

The night was uneventful, and the patient reported a slight improvement when she woke.

Vital signs remained within normal range: blood pressure 106/68 mmHg, heartbeat 71 bpm, body temperature 36.6°C.

Cardiotocography was repeated, which again registered a normal foetal trace, without uterine contractility.

The patient went to the bathroom and after a few minutes was found unconscious and covered in vomit. Skin and mucosa were pale, and she was hypotensive (70/40 mmHg), tachycardic (130 bpm) and O<sub>2</sub> saturation was 95%. Consciousness returned in response to pain stimulus. The internal emergency team was activated. It was very difficult to remove the patient from the bathroom but we had transferred her into the labour ward within about seven minutes from finding her unconscious. On physical exam the abdomen was soft and the uterus was not tender. Gynaecologic examination was similar to the initial evaluation with no vaginal bleeding (excluding active labour and possible placental abruption). An abdominal ultrasound scan revealed both foetuses with normal foetal heart rate and intra-abdominal free fluid. Arterial blood gas (ABG) test showed a high level of lactate 13.60 mmol/L and blood sample investigation revealed a haemoglobin drop from 9.8g/mL to 6.2 g/dL, thrombocytopenia (85.000/uL), prolonged partial thromboplastin time (67.5 seconds), low fibrinogen (1.65 g/L). Maternal stabilization was initiated with fluid therapy and oxygen supplementation. The patient's condition deteriorated rapidly, with blood pressure of 40/20 mmHg, so the patient was immediately transported to the operating room. An emergency C-section under general anaesthesia was performed approximately fifteen minutes after she was initially

found unconscious.

Entering the peritoneal cavity with the uterus still intact, about 2L of fresh blood and blood clots were seen. A lower segment uterine incision was performed and both newborns were delivered with no signs of placental abruption. Umbilical cord blood samples were not collected due to maternal haemodynamic instability. Hysterorrhaphy was performed and after that the abdominal cavity was reviewed with collaboration from general surgery. A midline vertical incision extending to the xiphoid was made to allow for better access. There was peripancreatic haematoma and bleeding stemming from the splenic artery and after proper dissection, SAA rupture was confirmed as the cause of massive bleeding. Proximal ligation with aneurysmectomy and splenectomy were performed, but haemostasis was very difficult due to disseminated intravascular coagulation (DIC). General surgery decided to use abdominal packing to control the bleeding and the abdomen was closed. Intraoperatively, ABG test revealed haemoglobin levels of 3g/dL and the patient received 7 units of packed red blood cells, 5L of crystalloids, 6 units of fresh frozen plasma and 2g of fibrinogen. The patient was transferred to the Intensive Care Unit (ICU) with multiorgan dysfunction. Forty-eight hours after C-section, a new laparotomy was performed to remove the packing and review haemostasis status. The patient remained in ICU for 9 days, followed by 7 days in the surgery unit. She was discharged home in stable condition on day 16.

Regarding the newborns, two live preterm infants were delivered: the first delivered weighing 1920g (appropriated for gestational age-AGA), Apgar Score of 2/6/6 and the second weighing 1939g (AGA), Apgar Score of 6/7/8. The first one was immediately intubated, and the second was initially admitted to the Neonatal Intensive Care Unit (NICU) on spontaneous ventilation. Their clinical condition deteriorated in the first two hours of life. The second infant was also put on mechanical ventilation. Both infants required inotropic support and bicarbonate perfusion concerning to severe metabolic acidosis. Neurological deterioration was also seen, with both presenting mydriasis and bilateral clonic-tonic movements of arms and hands (only subsided after more than two doses of anticonvulsant treatment). Serial transfontanelar ultrasound revealed progressive worsening of diffuse hyperechogenicity from the first 24 hours to 72 hours of life. The infants died 72 hours after birth with multiorgan failure and also with a diagnosis of hypoxic-ischaemic

encephalopathy, based on clinical, analytical and imaging findings.

Final pathology revealed a 173g spleen measuring 11.5 × 6.3 × 5.4 cm with a splenic artery aneurysm and medial artery fibrodysplasia.

## DISCUSSION

Splenic artery aneurysm occurs in approximately 0.1% of all adults. It is more common in women between 5<sup>th</sup> and 6<sup>th</sup> decades of life (mean age 52 years old)<sup>4,5</sup>. It is estimated that 6% to 10% of SAA will rupture and 25% to 40% of those ruptures will occur during pregnancy, especially during the 3<sup>rd</sup> trimester or labor<sup>1</sup>. SAA rupture is a catastrophic event, usually presented as sudden hypovolemic shock or death. The condition is associated with significant maternal and foetal mortality of around 70% and 95%, respectively<sup>1,3</sup>. Although the precise aetiology of SAA remains unclear, risk factors include fibromuscular dysplasia, collagen vascular diseases, atherosclerosis, pancreatic and liver diseases, multiple pregnancies, hormonal and local haemodynamic events in pregnancy and portal hypertension<sup>6</sup>. The mechanisms involved in the formation of this vascular defect are not fully understood. Haemodynamic and hormonal shifts in late stages of pregnancy (oestrogen, progesterone and relaxin) may contribute to increased blood pressure and splenic artery flow, weakening the arterial wall resulting in aneurysmal dilatation<sup>1,7</sup>. In this case, the patient had multiple risk factors: twin pregnancy, 3<sup>rd</sup> trimester and fibromuscular dysplasia which was diagnosed upon histologic examination.

SAA is generally asymptomatic but may present as abdominal left upper quadrant/epigastric pain. Diagnosis is usually made by means of radiological methods<sup>8</sup>. Clinical presentation of SAA rupture is often a constellation of non-specific symptoms and signs such as acute epigastric pain, hypotension, and anaemia. In 25% of cases there is a two-stage rupture: initial rupture occurs into the lesser sac or splenic pedicle, resulting in a tamponade of initial bleeding. Then, after a variable time (6-96 hours) a secondary rupture occurs with exsanguination into the abdominal cavity and patient collapse<sup>2,9,10</sup>. The initial rupture may provide vital time for diagnosis and preparation for intervention but when rupture of both sacs cause haemorrhaging in the peritoneal cavity the development of symptoms are fast<sup>6</sup>.

In pregnancy, abdominal pain is a frequent symptom, especially in the third trimester and can be a sign of threatened preterm labour when uterine contractility associated with shortened cervix is observed. Besides that, abdominal/epigastric pain associated with hypotension, and anaemia can mimic hypovolemic shock caused by placental abruption or uterine rupture<sup>6,11</sup>.

We received a twin pregnancy at the emergency room with left abdominal pain. At the vaginal ultrasound, a 22 mm-long cervix was observed and contractions were recorded in cardiotocography. This clinical presentation, which was interpreted as a possible threatened preterm labour and led to patient admission, was a possible manifestation of the initial rupture into the lesser sac or splenic pedicle, which result in a temporary tamponade. The secondary rupture corresponded with the patient's collapse. SAA, being such a rare diagnosis, was not considered in this situation, and a possible opportunity to intervene was lost.

In symptomatic SAA rupture during pregnancy, the aim is immediate resuscitation via a caesarean laparotomy and splenectomy or splenopancreatectomy and ligation of the splenic artery. Urgent surgical treatment for haemorrhage control is the priority, together with the fluid replacement<sup>12,13</sup>. The surgical technique usually depends on the location of the SAA (80% located in the distal portion of the splenic artery)<sup>14</sup>. Since SAA rupture is a catastrophic event, the question is: "could we prevent the high rates of morbimortality associated with SAA in pregnancy?". McMahon DP et al performed a review to ascertain the possible benefit of screening pregnant women for SAA, in which the authors concluded that routine screening during pregnancy is not warranted due to the rarity of the condition, but that the identification of those at greater risk of harbouring an asymptomatic SAA, along with the early institution of treatment may prevent maternal and foetal mortality<sup>15</sup>. They continued to specify that further studies are needed to determine which high-risk population would benefit the most from screening and if demanding screening tools such as splenic artery duplex ultrasound would be sensitive enough to diagnose an asymptomatic SAA<sup>15</sup>. This patient is a very rare example of a potential cause of a persistent abdominal pain.

Maternal collapse is uncommon in pregnancy but requires a prompt and organized response from all health care professionals involved in patient management to achieve the best outcome possible<sup>16</sup>. Due to the rarity of these events in a Maternal Units, imple-

mentation of continuous training with simulations, other exercises and the formation of rapid-response teams is key. Knowledge regarding differential diagnosis to the most common causes of maternal collapse, such as rupture of SAA, is crucial as the collaboration from other specialities may be necessary and without clinical suspicion their inclusion on the case may be delayed<sup>17,18</sup>.

**Conflicts of interest:** none

**Hospital's ethics approval:** confirmed

**Patient Consent:** confirmed

## REFERENCES

- Lynch MJ, Woodford NWF. Rupture of a splenic artery aneurysm during pregnancy with maternal and foetal death: A case report. *Med Sci Law*. 2008;48(4):342-345.
- Sadat U, Dar O, Walsh S, Varty K. Splenic artery aneurysms in pregnancy - A systematic review. *Int J Surg*. 2008;6(3):261-265.
- Gourgiotis S, Alfaras P, Salemis NS. Spontaneous rupture of splenic artery aneurysm in pregnancy: a case report. *Adv Med Sci*. 2008;53(2):341-343.
- Tulsyan N, Kashyap VS, Greenberg RK, et al. The endovascular management of visceral artery aneurysms and pseudoaneurysms. *Journal of Vascular Surgery*. 2007;45(2):276-283.
- Al-Habbal Y, Christophi C, Muralidharan V. Aneurysms of the splenic artery - A review. *Surgeon*. 2010;8(4):223-231.
- Mattar SG, Lumsden AB. The management of splenic artery aneurysms: experience with 23 cases. *The American Journal of Surgery*. 1965; 169(6), 580-584.
- Pavlis T, Seretis C, Gourgiotis S, Aravosita P, Mystakelli C, Aloizos S. Spontaneous Rupture of Splenic Artery Aneurysm during the First Trimester of Pregnancy: Report of an Extremely Rare Case and Review of the Literature. *Case Rep Obstet Gynecol*. 2012; 2012:1-3.
- Arslan S, Karadeniz E, Colak A, Yapca OE. Spontaneous Rupture of Splenic Artery Aneurysm in Pregnancy. *J Emerg Med Case Reports*. 2016;7(4):70-73.
- Bedford PD, Lodge B. Aneurysm of the splenic artery. *Gut*. 1960; 1(4), 312-320.
- de Vries JE, Schattenkerk ME MR. Complications of splenic artery aneurysm other than intraperitoneal rupture. *Surgery*. 1982;91(2):200-204.
- He M, Zheng J, Zhang S, Liu W, Zhu M. Rupture of Splenic Artery Aneurysm in Pregnancy. *The American Journal of Forensic Medicine and Pathology*. 2010;31(1):92-94.
- Parangi S, Levine D, Henry A, Isakovitch N, Pories S. Surgical gastrointestinal disorders during pregnancy. *Am J Surg*. 2007;193(2):223-232.
- Abbas MA, Stone WM, Fowl RJ, et al. Splenic artery aneurysms: Two decades experience at Mayo Clinic. *Ann Vasc Surg*. 2002;16(4):442-449.
- Telfah MM. Splenic artery aneurysm : pre-rupture diagnosis is life saving. *BMJ Case Report* 2014; 2014: bcr2014205115
- McMahon DP, Ward WH, Harwood JL et al. An Institutional Review of Splenic Artery Aneurysm in Childbearing-Aged Females and Splenic Artery Aneurysm Rupture During Pregnancy . Is Screening Justified?, *Mil Med* 2012;177:96-98.
- Naidoo M, Med S. Best Practice & Research Clinical Obstetrics and Gynaecology Maternal collapse : Training in resuscitation. *Best Pract Res Clin Obstet Gynaecol*. 2015;:1-9.
- Ballout RA, Ghanem R, Nassar A, Hallal AH, Labib M. Case Report Splenic Artery Aneurysm (SAA) Rupture in Pregnancy : A Case Report of a Rare but Life-Threatening Obstetrical Complication. *Journal of Women's Health and Development* 2 (2019): 020-028.
- Schauberger CW, Chauhan SP. Emergency cesarean section and the 30-minute rule: definitions. *American Journal of Perinatology*. 2009; 26(03), 221-226.

## ENDEREÇO PARA CORRESPONDÊNCIA

Catarina Peixinho  
Hospital Pedro Hispano  
E-mail: catarina.peixinho1989@gmail.com

**RECEBIDO EM:** 25/11/2019

**ACEITE PARA PUBLICAÇÃO:** 01/05/2020