

Acute Abdomen in Pregnancy... Expect the Unexpected

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Abstract

Acute abdomen during pregnancy it refers to any serious intra-abdominal condition related or not related to pregnancy that is accompanied by a sudden severe abdominal pain, muscular rigidity, tenderness, nausea, vomiting and other signs and symptoms that can help to guide in the prompt diagnosis. It complicates 1/500 - 635 pregnancies (0.2 - 2.2%) and about 0.5% - 2% of the patients will require surgery for non-obstetric causes. Represents a major challenge for first contact physician due to the anatomical and physiological changes happening during pregnancy. The wide range of causes and varied spectrum of clinical presentations besides the anatomical and physiological changes in "normal pregnancy" pose a formidable diagnostic and therapeutic challenge. It is of paramount importance a detailed clinical history and physical examination besides blood samples for laboratory testing and specific investigations. If the patient is hemodynamically unstable it might require urgent laparotomy. In this case a multidisciplinary approach and prompt diagnosis and definite treatment can make the difference between life and death.

We present a clinical case of a 30 year old gravida 2, abortion 1, 19 weeks of pregnancy who arrived to our emergency department with severe acute abdominal pain, palpable mass in left flank, nausea, vomiting and imminent signs and symptoms of hypovolemic shock. Her final diagnosis, outcome of the product and evolution showed us the importance of a detailed clinical history, physical examination and radiological evaluation. A review of literature in the topic was performed and we found that the cause of acute abdomen in our patient was not considered in the diagnostic approach of abdominal pain. The reason might be that the incidence of this condition is very rare. We present a review of abdominal pain in pregnancy and pinpoint the importance of making a complete clinical history, examination of the patient, diagnostic approach and treatment, besides we make a review of the cause of abdominal pain in our patient.

Keywords: *Acute Abdomen; Pregnancy; Abdominal Pain; Abdominal Mass; Hypovolemic Shock; Wunderlich Syndrome; Beck Triad; Renal Angiomyolipoma*

Introduction

The definition of acute abdomen in pregnancy is the same as in the general population. it refers to any intra-abdominal condition that produces severe, sudden pain, although sometimes depending on the cause it can be intermittent, it is accompanied by nausea, vomiting,

abdominal distension and other signs and symptoms that may help in the diagnosis. On physical examination there may be muscular stiffness, sensitivity, absence of peristaltic sounds, and specific signs that guide to find the definite diagnosis [1]. It has an incidence of 1 in 500 - 635 pregnancies, about 0.5% - 2% of the patients will require surgery for non-obstetric causes [2]. The physiological and anatomical changes associated with pregnancy make difficult to have a prompt diagnosis. After 12 weeks the uterus becomes an extrapelvic organ and as it continues to grow it displaces the intra-abdominal structures, its weight can reach 750 to 1000 grams this can make atypical the clinical presentation of abdominal pain often resulting in diagnostic uncertainty and therapeutic delay with increased risks of maternal and fetal morbidity [3]. When we face with a pregnant woman with abdominal pain we have to consider first the typical obstetrical causes by trimester (Figure 1).

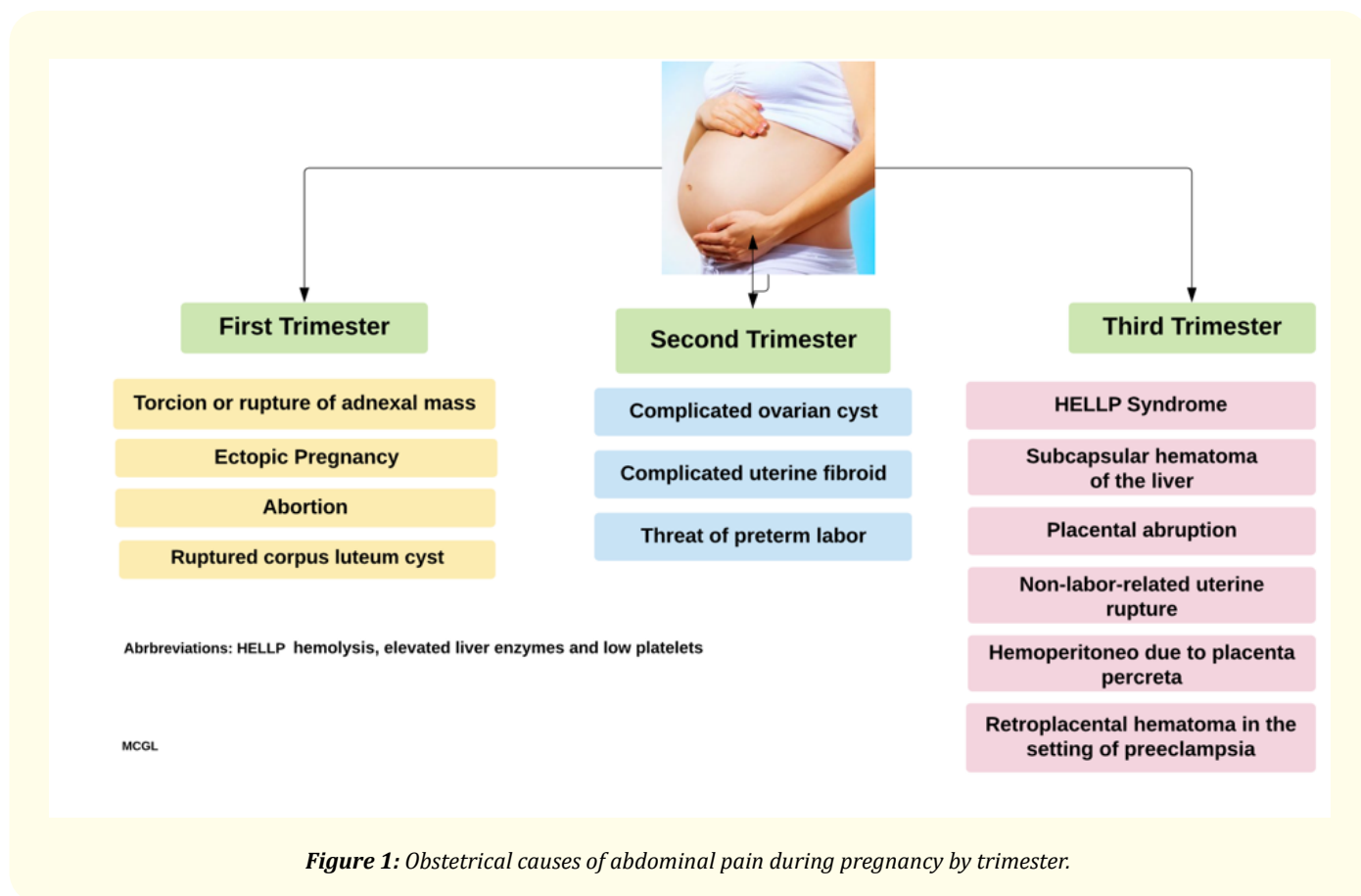


Figure 1: Obstetrical causes of abdominal pain during pregnancy by trimester.

There are non-obstetrical causes of abdominal pain during pregnancy, it is very important to make a complete clinical history, the semiology of abdominal pain must be complete.

The history of abdominal pain should include:

- Site (right upper quadrant, left upper quadrant, right lower pain, left lower pain, epigastric, mesogastric, diffuse) including radiation to other sites; (shoulder, left arm, right arm, upper back, lower back etc).
- Type of pain (colic, dull, burning, throbbing).

- Duration and timing of the pain (What was the patient doing when the pain started? The pain is intermittent or continuous? the pain last seconds, minutes, hours or has several days, months or years?).
- Exacerbating factors (what makes the pain to increase? The change of position? Deep breath?)
- Relieving factors: Medication, not moving, sleeping, or nothing relieves the pain?
- Associated symptoms (nausea, vomiting, fever, chills, diaphoresis, cough, dyspnea, diarrhea, constipation, blurred vision, cefalea, etc).

The first surgical cause of acute abdominal pain in pregnancy is appendicitis, the incidence is 0.04% and 0.2% [4,5]. This is followed by cholecystitis [6], pancreatitis [7] and bowel obstruction [1,8,9]. It is not our objective to review every cause of abdominal pain in pregnancy, we want to make a review of the causes of abdominal pain in pregnancy according to the location (Figure 2) and when we face a patient like ours, with left upper quadrant pain, palpable mass and hypovolemic shock (Beck triad), we must to suspect in rupture of a renal tumor (Wunderlich syndrome). This syndrome is a rare and life-threatening condition considered as an urologic emergency which is characterized by non-traumatic renal haemorrhage into the subscapular and perinephric space and was described by Wunderlich in 1856 [10]. The information we have of this syndrome in pregnancy we obtained it from reports of clinical cases. There are various etiologies which include benign and malignant renal neoplasms, vascular disease (vasculitis, renal artery arteriosclerosis, and renal artery aneurysm rupture), nephritis, infections, undiagnosed hematological disorders, and anatomical lesions.

We added the diagnosis of renal angiomyolipoma in figure 2 because we have to suspect it when we face with a pregnant patient with sudden severe abdominal pain, palpable mass in any flank and clinical signs and symptoms of hypovolemic shock. The commonest cause of Wunderlich syndrome in most series is renal angiomyolipoma (AML) which is a benign clonal neoplasm consisting of varying amounts of mature adipose tissue, smooth muscle, and thick-walled vessels within renal parenchyma. It was first referenced as an entity in 1900 and Fischer first described the histopathology in 1911 as including the three components of dysmorphic blood vessels, smooth muscle and mature adipose tissue from which the tumor derives its name.

It is seen in more than 50% of patients with tuberous sclerosis syndrome (TS) [11,12]. AML in TS is more likely bilateral and has a tendency toward accelerated growth. Clinical presentation depends on the degree and the duration of the bleeding. The classic presentation includes flank or abdominal pain, a palpable tender mass, and gross hematuria (Lenk's triad) [13,14]. Massive retroperitoneal hemorrhage represents the most significant and feared complication, an abdominal ultrasound should be performed, it may help in the diagnosis although sometimes that won't be possible due to the clinical condition of the patient [15-17].

Clinical Case

Written Informed consent was obtained from the patient for publication of her case.

A 30-year-old gravida 2 abortion 1, collapsed at home at 17 weeks of gestation with a singleton intrauterine pregnancy following the sudden onset of severe left-sided flank and left hypochondrium pain irradiated to left hemithorax. This pain began six hours before became very intense. She had an uneventful pregnancy until this presentation. She was brought to our hospital by ambulance. She gave a history of intermittent left sided abdominal pain that became very intense before she collapsed. The pain was accompanied by nausea and vomiting six times. She had no history of any trauma. Her previous medical history included several treatments for infertility, smoking and marijuana use before pregnancy and also had gestational diabetes treated with diet. She had a family history of hypertension and diabetes. On examination, she was pale and diaphoretic, blood pressure of 80/60 mm Hg, heart rate of 140 bpm, temperature 35 grades and a respiratory rate of 12. Her abdomen was soft, with appropriate distension for her gravid state and she had marked left-sided flank tenderness, pain and a palpable mass of approximately 20 cm was felt in the left hemiabdomen, painful, indurated, with frank evidence of peritoneal irritation, absent peristalsis. Obstetrically with fetal movements present, FCF 154 per minute. Due to hemodynamic instability, it was not possible to perform an imaging study. Management began with crystalloid and blood products and assessment is requested



Figure 2: Differential diagnosis of abdominal pain in pregnancy according to location.

by general surgery for acute abdomen and grade IV hypovolemic shock, shock index 1.7. Laboratory findings included blood cell count which was abnormal due to anemia, hemoglobin level was 8.6 g/dL (12.7 - 16.5), hematocrit 24% (41.3 - 43.7), leucocytes $22.87 \times 10^3/\mu\text{l}$ (4.8 - 10.2), and platelets $155.4 \times 10^3/\mu\text{l}$ (142 - 424). Her renal function reported urea 21 mg/dL (10.0 - 50.0), Creatinine 0.6 mg/dL (0.55 - 1.0), uric acid of 9.8 mg/dL (2.4 - 7.0), hepatic profile was not taken, electrolytes potassium 5.2, chloro 113, corrected calcium 9.6, sodium 136 mmol, phosphorus 3.0. Patient underwent urgent laparotomy. Her initial diagnosis was acute abdomen and hypovolemic shock. During surgery she was hypotensive, her blood pressure dropped to 40/25, hearth rate 100 per minute. A left kidney tumor 20 x 15 cm was found, lobed, very vascularized (Figure 3 and 5), attached to the left lateral face of vena cava that infiltrates the anterior wall

and body and tail of the pancreas, displaces descending colon and sigmoids without invading them, with profuse active bleeding and retroperitoneal hematoma of approximately 3000 ml, total bleeding 11 liters (Figure 4), patient underwent total nephrectomy and drainage of the retroperitoneal hematoma. Two compresses were left in the abdominal cavity. During the trans-operative period she had a diuresis of 750 ml. 5.2 liters of crystalloid solution, 2 liters of colloids, 16 globular packs, 6 units of fresh frozen plasma, 10 platelet concentrates and 10 cryoprecipitates were administered.

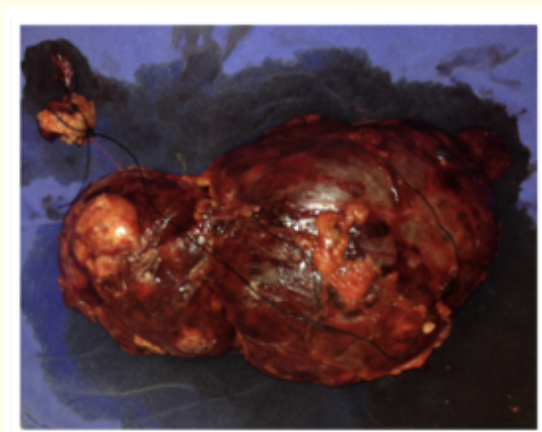


Figure 3: Specimen of left nephrectomy and renal angiomyolipoma.



Figure 4: Total bleeding (11 liters) of renal angiomyolipoma and retroperitoneal hematoma.

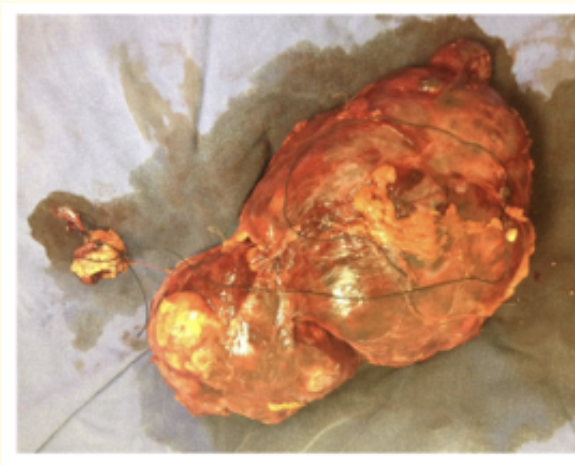


Figure 5: Macro of left nephrectomy and renal angiomyolipoma.

Trans-operative histopathological report was angiofibroma. Definite histopathological report showed a mesenchymal tumor consistent with angiomyolipoma, with ischemic degeneration and necrosis, residual renal parenchyma with acute inflammation, permeable renal hilum, no atypia or mitosis were identified (Figure 6-8).

After surgery patient was transferred to intensive care unit, under sedation and mechanical ventilation. An obstetric ultrasound was performed observing absence of fetal cardiac activity and a few hours later presents spontaneous abortion, meriting uterine curettage, without complications, apparent and estimating bleeding was 50 ml. Patient continued with inotropic support and mechanical ventila-

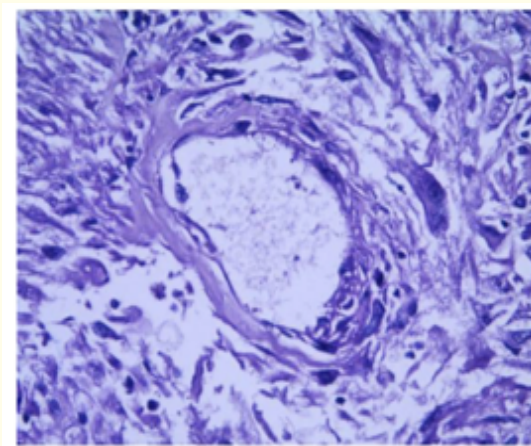


Figure 6: A medium caliber central blood vessel surrounded by muscle fibers with elongated nuclei without atypia in the nucleus or mitosis is observed at large magnification. H&E staining.

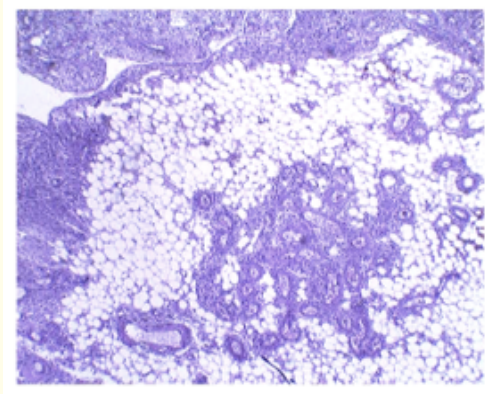


Figure 7: Panoramic view of an adipose component is observed, which is surrounded by vascular structures of various sizes.

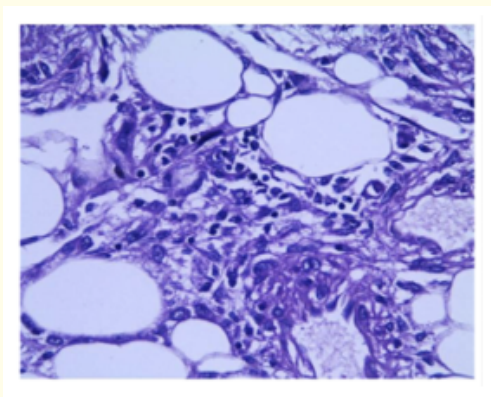


Figure 8: Fat cells (adipocytes) which are adjacent to muscle cells, fusiform and elongated as well as two vascular structures.

tion, on day 2 she underwent again surgery to remove the abdominal packaging, there was no bleeding at all. On day fourth she was successfully extubated. Three days later she was transferred to another hospital for urologic follow up. Patient was discharged home three days after. Patient was lost to follow up until three years later, she said she continued with abdominal pain, a CT scan of the abdomen was performed 6 months after the initial event and reported hepatic angiomyolipoma/predominance of the fatty component, angiomyolipoma in the right kidney and lesions and in iliac bones suggestive of benignity, she also had left limb deep vein thrombosis and post thrombotic syndrome. We do not have images of the CT scan, we performed an abdominal ultrasound 3 years after the initial event and found the hepatic angiomyolipoma and right renal angiomyolipoma (Figure 9 and 10).

Discussion

Abdominal pain is a common complaint in pregnant women. It should not be underestimated and requires assessment to rule out surgical obstetric and non-obstetric surgical causes. Due to the physiological and anatomic changes during pregnancy it may present with atypical or attenuated clinical symptoms; morphological assessment and imaging must be performed promptly to rule out causes that

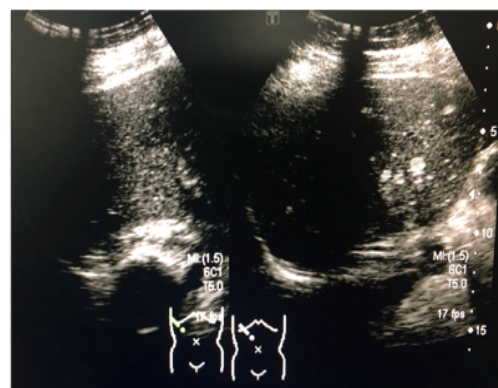


Figure 9: Liver with multiple echogenic nodular images of well-defined edges of 5 - 12 mm suggestive of hemangiomas.

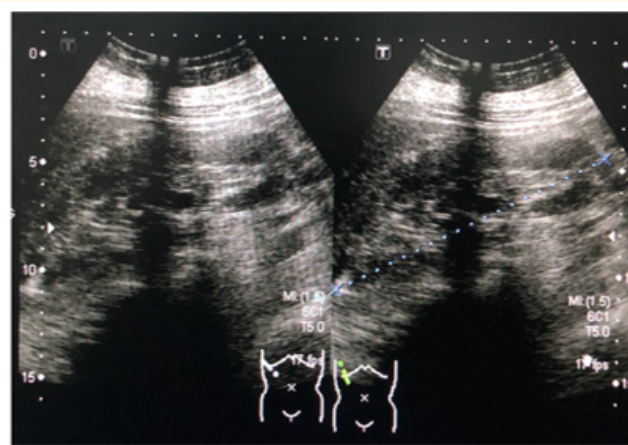


Figure 10: Right kidney with areas with loss of corticomedullar relationship associated with multiple well-defined echogenic nodules of less than 15 mm.

might require surgical intervention although sometimes that won't be possible specially in the case when the patient arrives to hospital with acute abdomen and hypovolemic shock as it happened with our patient. Multidisciplinary approach is mandatory in order to preserve life in the patient and the unborn child. In our case we didn't think in renal angiomyolipoma because this is the first case we have in our hospital, although we are a tertiary gynecology and obstetrics hospital with more than 15,000 births every year this is the first case we document. Angiomyolipoma are benign mesenchymal tumours. The major risk factors for bleeding are tumour size (> 4 cm) and the presence of tuberous sclerosis which is an autosomal dominant disease with a prevalence of 1 in 12,000. Our patient had a renal angiomyolipoma 20 x 15 cms and has lesions in liver, right kidney and in the skin that make very possible the diagnosis of tuberous sclerosis. The indications for intervention are pain, bleeding or suspected malignancy. Julien Raft., *et al.* performed a review of the cases

of angiomyolipoma in pregnancy from 1952 to 2004 and found seventy-two cases of association of renal angiomyolipoma and pregnancy, out of which 58 presented with haemorrhage as it did our patient. In only 26% of the cases, renal angiomyolipoma had been documented prior to pregnancy. In our case was an incidentaloma. Clinical presentation was similar to cases occurring among non-pregnant women: abdominal pain (88%), hypotension or shock (33%) and hematuria (24%), our patient did not have hematuria. Average size of the rupture was 11,7 cm. Our patient had a 20 x 15 cm size renal angiomyolipoma. Average gestation age upon occurrence of haemorrhage was 27 weeks with a minimum at ten weeks, our patient had 17 weeks Therapeutic strategies at the time of shock or hypotension were total nephrectomy in 79% of the cases, (as it happened in our case) 7% polar nephrectomy, 7% embolisation followed by nephrectomy and 7% abstention [16,18].

The exact cause of rupture of angiomyolipoma in pregnancy is not well understood. It is thought that abdominal mechanical pressure, hormonal or histological changes) could increase the risk of renal angiomyolipoma rupture. These patients should have a medical follow-up period at closer intervals during their pregnancy and the postpartum phase, this is in the context when you already know the diagnosis of angiomyolipoma in the patient. Anticonception should be performed due to another pregnancy specially in a patient with sclerosis tuberosus may endanger her life and the life of the offspring.

Conclusion

Abdominal pain in pregnant women can be a common presentation or a catastrophic emergency, especially when the patient arrives to emergency room with acute abdomen and signs and symptoms of hypovolemic shock, a thorough clinical history, examination, and laboratory tests should be done in the structured assessment. A Multidisciplinary approach is mandatory and should take into account the needs of the mother and the unborn baby.

Conflict of Interest

There is no financial interest or any conflict of interest.

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