Unexpected Gastrointestinal Stromal Tumor as Acute Abdomen in First Trimester of Pregnancy

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INTRODUCTION

A gastrointestinal stromal tumour (GIST) is a tumor of the gastrointestinal tract. We present a 36-year-old, gravida 2 para 1 lady presented with acute lower abdominal pain at 9 weeks period of amenorrhea (POA) associated with vomiting. She had a previous uncomplicated cesarean section 2 years ago. Abdominal examination revealed a tender and mobile 26-week-sized mass. A transabdominal scan revealed a right adnexal mass with a cystic component measuring 10 cm x 10 cm in size, as well as a viable singleton intrauterine fetus corresponding to her dates. She underwent emergency laparotomy in suspicion of a right twisted ovarian cyst. The inoperative finding revealed a large pedunculated and twisted gangrenous mass from the greater curvature of the stomach with normal uterus and ovaries. The histopathological examination of the mass reported as malignant Sarcomatoid epithelioid GIST (category 3b). The patient was managed by a multidisciplinary team approach involving the obstetrician & gynaecologist, surgeon and oncologist throughout her pregnancy course. The pregnancy progressed well, and she delivered via elective repeat cesarean section. Post partum, oesophago-gastric-duodeno-scopy and computed tomography assessment revealed no evidence of local disease or distant metastasis. She was advised for yearly follow-up with the surgical department.
Case presentation

A 36-year-old, Gravida 2 para 1 lady presented with acute lower abdominal pain at 9 weeks POA and vomiting at the emergency department. She had no known medical illness. Her previous cesarean section was done 2 years ago due to subclinical chorioamnionitis at 35 weeks with asymptomatic major placenta previa, and it was uncomplicated. This pregnancy was confirmed with a positive urine pregnancy test at 6 weeks POA. Unfortunately, there was no dating scan done. She did notice her lower abdomen was slightly increasing in size since the last delivery, but as it was never been painful. However, at 9 weeks POA, she developed a sudden onset of severe lower abdominal pain. It was at the umbilical area with a pain score of 10/10.

The pain was associated with vomiting, which were mainly food particles. There was no fever or any other bowel symptoms. There was no associated per vaginal bleeding or constitutional symptoms. Upon immediate assessment, she was clinically pink, tachycardic with a pulse of 110 beats per minute. Her blood pressure was normal. She had a low-grade fever (T=37.6°C). Perabdomen findings revealed a suprapubic transverse well-healed scar. Although the abdomen was soft, there was generalized tenderness with palpable mass felt at 26 weeks size of a gravid uterus. The mass has limited mobility, with a firm to hard inconsistency. Renal punch assessment was negative bilaterally. Other systemic findings were otherwise normal.

A transabdominal ultrasound scan revealed an IUGS with a viable fetus corresponding to her dates. There was a suspicious mass in the right adnexa measuring 10cm x 10cm in size, with some solid cystic appearance. The capsule was intact with no papillary projections. There was no free fluid seen, and both kidneys were normal. In view of her age and presented in early pregnancy, a right twisted ovarian cyst was the most likely provisional diagnosis; few other differential diagnosis such as a ruptured ectopic pregnancy and degenerating uterine fibroid in pregnancy were considered. Other non-obstetric causes such as an intestinal obstruction in view of her previous surgery and appendicitis were still being considered.

Management and outcome

She was counseled for an emergency laparotomy in suspicion of a twisted ovarian cyst in pregnancy with possible salphingo-opherectomy. Surprisingly, an intraoperative finding revealed an unremarkable uterus corresponded to her dates with normal ovaries and tubes. However, there was a large pedunculated circumscribed gangrenous mass situated at the pelvic area anteriorly to the uterus. It measured 11cm x 15cm x 9cm in size and has a thin 1cm elongated and narrowed stalk attached to the serosa of the greater curvature of the stomach Figure 1. The mass was twisted around its base, causing the mass to become gangrenous. The mass was excised by simple ligation at the stalk base and avulsed by the surgeon and weigh 535 gram. There was only minimal blood loss.

Figure 1: Intraoperative findings showed a large gangrenous mass with a twisted stalk attached to the greater curvature of the stomach.

The histopathological examination of the mass reported that the tumour cells were loosely cohesive, showing epithelioid morphology, having round to ovoid vesicular and hyperchromatic nuclei, inconspicuous nucleoli, and minimal to moderate amount of cytoplasm. There was also marked nuclear pleomorphism. Multinucleated forms were also seen. The mitotic figure was 1/50 hpf. These neoplastic cells were surrounded by myxoid stroma in large areas. The tissue slides were proceeded with immunohistochemistry studies and showed the tumour cells were positive for DOG1 Figure 2a, CD117 Figure 2b, CD34, SMA, Desmin, and negative for S100 and CKAE1/AE3. A diagnosis of sarcomatoid epithelioid GIST was concluded. Based on the pathological assessment and WHO Classification, the mass is categorized as 3b for prognosis stratification Figure 3.

Her postoperative recovery was uncomplicated. Initial intraoperative findings were explained to the patient and her husband. She was discharged without any complications. The patient was seen after 6 weeks to review her wound as well as the HPE report. Her pregnancy had progressed well since discharged. In view of its unexpected presentation in pregnancy, further discussion regarding the nature and clinical behavior of the GIST were made between obstetrician, surgeon and oncologist with the presence of the patient and her husband. Based on the pathological assessment and a large tumor size, the mass was best considered as possible high
Acute abdomen presented in pregnancy is usually associated with an ovarian incidence, which commonly requires surgical intervention. Yet ovarian cyst related pathologies such as twisted, ruptured, infected, and hemorrhagic cysts can be troublesome to the gynecologist as the decision for surgical intervention need to outweigh between the risk of fetal loss versus maternal morbidity and mortality. Patients usually present with severe and excruciating abdominal pain in the early pregnancy, sometimes associated with vomiting, pale and hypotensive episodes. The patient, in this case, did present with generalized abdominal pain, which was very severe. However, in ectopic pregnancy, the presentation is rarely associated with an abdominal mass. These patient’s symptoms were more suggestive of an ovarian cyst pathology, and the most common would be a twisted ovarian cyst.

Her symptoms and physical findings were suggestive of a twisted ovarian cyst and further supported with transabdominal ultrasonography of a huge solid-cystic mass in the pelvic area. Thus, she was counseled for suspicion of a twisted ovarian cyst in pregnancy and to proceed for an emergency laparotomy with possible salpingoopherectomy. The surgery was indicated, and it was done promptly. A large GIST presenting as an acute abdomen in pregnancy is a rare presentation. In this case, intraoperative findings revealed normal pelvic organs, and the origin of the tumor site was traced up following its stalk. An urgent referral to the surgical team was needed as it was found to be gastrointestinal related. It was also in the best interest to the patient. Gastrointestinal stromal tumors are mesenchymal tumors of the digestive tract and slow growing. This patient did notice that her abdomen was slightly distended since her previous cesarean section, yet it was asymptomatic.

Retrospectively, based on the size of the tumour itself, it could have been there for many years prior to this presentation. The stalk was very thin and elongated due to the large size, and perhaps the mass was fairly mobile with possible gravity influence to be on the lower part of the abdominal cavity for some time. Unfortunately for this patient, the tumor mass had twisted during her pregnancy and required an emergency resection. The diagnosis can be challenging as the signs and symptoms may not be specific and masked by the pregnancy itself.
Positive immunostaining for DOG1 and CD117 expressions confirmed the histopathological diagnosis (Kiśluk et al., 2016), which this patient's had. Based on the tumour size (>10 cm) and mitotic rate (less or equal to 5 per 50 HPFs), the prognostic group falls into group 3b, in which 12% of gastric GIST's patients show progressive disease during follow-up (Parab et al., 2018). Few studies mentioned that the positivity to the staining may give better prognostic value to longer survival, yet; should be clinically correlated with the presentation, size, and location (Kiśluk et al., 2016). While negative DOG-1 expression by staining showed to be an independent prognostic factor and lesser overall survival ($p = 0.002$) (Kang et al., 2010).

GIST is uncommon, with a reported incidence of 11 to 14.5 cases per million (Stubbs et al., 2011), and having a GIST with high suspicion of malignant histology in pregnancy is rare. Patient with GIST and suspicious for malignant histopathology is commonly treated with chemotherapy drugs such as Imatinib, which is a tyrosine kinase inhibitors for a certain duration as it is highly treatable (Stubbs et al., 2011; Scherjon et al., 2009; Charif et al., 2014). Cases of malignant GIST in pregnancy reviewed by G. Zarkavelis noted that resection was the main treatment followed by Imatinib chemotherapy to ensure good outcome (Zarkavelis et al., 2015). However, the risk of teratogenicity raised concern in the organogenesis phase, and the risk of fetal loss should be taken into concern during counseling. The review emphasized on multidisciplinary approach as it is a rare malignancy in pregnancy to ensure a good outcome.

Magnetic resonance imaging provides the best imaging to assess the extent of the disease in view of its soft tissue tumors characters (Charif et al., 2014). Yet, MRI in pregnancy itself predisposed to another risk of contrast exposure to the fetus, especially in the first trimester. Although, the electromagnetic waves in MRI is technically safe in pregnancy with no known biological effect to the fetus (Ray et al., 2016). The value of MRI might not be a benefit if there is no contrast used. These were among the important issues highlighted in discussion with this patient and were taken into consideration upon the decision opted regarding her pregnancy management, after reviewing her pathology report and incomplete staging of the disease antenatally against the slow-growing nature of her tumor.

The pregnancy was allowed to be kept, and she only underwent for MRI and OGDS later at 6 weeks postpartum. GIST in pregnancy are rare and there were limited evidences on the best option of treatment. Few studies showed several good pregnancy outcomes to mother and baby despite high-risk GIST requiring chemotherapy (Zarkavelis et al., 2015). Although this case is considered a rare and atypical presentation, yet it is a very important learning points to obstetricians and gynecologists on diagnosing acute abdomen in pregnancy, as delaying the appropriate management may jeopardize the mother and unborn fetus. It should be considered as one of the differential diagnoses for acute abdomen presented in pregnancy and once diagnosed. It should be managed with a multidisciplinary team effort to achieve optimal care to the patient and baby.

CONCLUSIONS

In conclusion, GIST presented with acute abdomen in pregnancies are not uncommon. It should be one of the differential diagnosis for acute abdomen presented in pregnancy, and once diagnosed, it should be managed with a multidisciplinary team effort to achieve the best outcome for the mother and unborn fetus.
fetus. Multidisciplinary team management and discussion with patients are crucial in deciding to proceed with further invasive investigation such as MRI and OGDS in pregnancy and the timing for treatment with chemotherapy with risk counseled.

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Conflict of Interest

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