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Case report

Acute abdomen with a parasitic smooth muscle tumor of uncertain malignant potential (STUMP) in pregnancy; a unique case report

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ABSTRACT

Introduction and importance: Diagnosis of a smooth muscle tumor of uncertain malignant potential (STUMP) during pregnancy is rare. Furthermore; the investigation of an intra-abdominal mass during pregnancy is clinically challenging due to anatomical changes and additional considerations of the developing fetus and radiation exposure. The unusual nature and diagnostic dilemma of such a case warrants a case report to serve as an educational prompt to clinicians who may encounter pregnant patients with undifferentiated intra-abdominal masses and/or suspecting of STUMP.

Case presentation: We report a rare case of a parasitic STUMP diagnosed during pregnancy. The patient presented with a new umbilical hernia and deranged liver function tests (LFT's) during her third trimester. MRI reported a large mass in the left mid flank with intra-abdominal varices extending into the umbilical hernia. She went on to develop an acute abdomen requiring laparotomy where a parasitic fibroid adherent to the omentum was excised and a preterm infant was delivered via caesarean section. Histology was difficult due to pregnancy related changes but ultimately confirmed a diagnosis of STUMP.

Clinical discussion: STUMP in pregnancy is rare and diagnosis is further complicated by histological challenges due to pregnancy related changes. Postoperative diagnosis is challenging due to lack of universally accepted diagnostic criteria and Uncertainty regarding prognostic factors makes management and follow-up of patients with STUMP challenging. Studies have shown that younger patients are more likely to demonstrate recurrence.

Conclusion: Investigation and management of intra-abdominal masses in pregnancy is challenging. It requires timely multi-disciplinary team (MDT) input. Additional complications and considerations relate to the preterm fetus. Knowledge and understanding of these difficulties will better equip clinicians working with such patients to formulate a structured and well informed approach to the pregnant patient with a new intra-abdominal mass. Diagnosis of STUMP during pregnancy may be challenging for the pathologist and require further exert opinion.

1. Introduction

This report describes an emergency preterm caesarean section (CS) due to an acute abdomen with known intra-abdominal mass. Histology of the mass reported a smooth muscle tumor of uncertain malignant potential (STUMP). STUMP during pregnancy is extremely rare with only two other cases described in the literature [1,2].

STUMP is an ill-defined subcategory of uterine smooth muscle tumors (SMTs). The WHO classification allocates STUMP as an intermediate tumor between a benign leiomyoma (LMs) and a malignant leiomyosarcoma (LMSs). The differentiation between a LM and a LMS is based on criteria around mitotic count activity, cytological atypia, and presence of tumor cell necrosis [3,4]. If these criteria are not met but the histopathological features are atypical, the cases are classified as STUMP [5].

There is no clinical difference between patients presenting with LMs, LMSs and STUMPs. Symptoms include abnormal uterine bleeding, pelvic pain or abdominal pressure. LM's, LMS's and STUMP are difficult to differentiate radiologically [3]. They are all usually seen within the myometrium and sometimes are pedunculated. Rarely, they are seen intra-abdominally. The STUMP in our case was intra-abdominal, separate from the uterus and parasitic. Postoperative diagnosis of STUMP is challenging due to the lack of universally accepted diagnostic criteria and may vary depending on the pathologists experience/expertise [5]

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which may lead to an over-diagnosis of this type of tumor [6].This makes estimates of STUMP incidence difficult [7,8].

The biological potential of STUMP differs to LMs or LMSs. STUMP has a superior prognosis when compared to LMSs but there seems to be more lymphatic and vascular dissemination associated with STUMP. Clinical behavior remains unclear as there are a limited number of studies related to STUMP [9–14]. Uncertainty regarding prognostic factors makes management and follow-up of patients with STUMP challenging. Studies have shown that the younger the patient at diagnosis the more likely that there is a recurrence of STUMP [3].

Our literature search found two case reports on STUMP diagnosed during pregnancy. The first case reports a pregnancy complicated by chorioamnionitis requiring caesarian section at 26 + 2 weeks [1]. Intraoperatively 2 subserosal myomas were resected and histopathology reported STUMP.

The second case describes a patient found to have a large intra-abdominal mass first seen on ultrasound at 14 weeks gestation [2]. Inflammatory markers and CA125 were both raised. A myomectomy was performed at 18 weeks' gestation due to worsening pain and dyspnea and a 4.2 kg tumor was removed. Histopathology reported a STUMP. Due to worsening pre-eclampsia a caesarean hysterectomy was performed at 34 weeks gestation and histology reported a leiomyoma without increased mitotic activity.

The aim of reporting this case is firstly to increase overall understanding and awareness of STUMP. Secondly the authors hope to highlight the added complexity that pregnancy and its related anatomical and physciological changes pose to investigation and management of any new intra-abdominal mass and the necessity for early MDT input.

2. Presentation of case

Our patient was Gravida 2 Para 1 having had a previous normal vaginal delivery at term. She had no other past medical or surgical history of note. She first presented to the emergency department (ED) at 19 weeks with abdominal pain and was discharged following a normal clinical exam and investigations. She represented to ED at 30 weeks with a cough. On examination she had an umbilical hernia which she reported had developed over the past 2 months. She was discharged with no follow up.

Obstetric USS at 32 weeks reported an incidental finding of a prominent maternal umbilical hernia with varicosities. Due to history of maternal itch bile salts and liver function tests were ordered. Results were deranged with ALT 459 U/L (*0–55 U/L*), AST 313 U/L (*0–55 U/L*), and bile salts of 45 umol/L (*<15 umol/L*). Obstetric referral was made at 32 + 3 to Waikato Tertiary Hospital.

The patient was seen at 32 + 6 weeks in antenatal clinic. She had a distended, reducible, non-tender umbilical hernia. History and investigations suggested a diagnosis of obstetric cholestasis for which ursodeoxycholic acid was commenced. An extended liver panel was negative. Concern remained around the nature of the umbilical hernia varicosities.

Follow up USS at 34 + 5 reported a large left hypochondrial mass separate from the spleen and kidneys, abutting the uterus, and with umbilical/para-umbilical varicosities. The mass was described as solid, heterogeneous, and fairly well demarcated. It was noted to be primarily hypoechoic and vascularized. There was no ascites and the ovaries were not visualized. Fibrinogen was elevated at 11.4 g/L (*1.5-5 g/L*) with normal INR and APTT. LFT's remained deranged but stable. CA-125 was raised at 103 (*36 U/mL*).

MRI was undertaken and reported a 17cm × 15 cm × 21 cm solid lobulated mass in the left mid flank/abdomen causing uterine displacement (Fig. 1). Extensive intra-abdominal varices including extension into an umbilical hernia were noted. No venous thrombosis or liver cirrhosis was seen.

The case was discussed at the Gynecology MDM and recommendation was for an upper GI endoscopy to rule out oesophageal varices. Concern regarding the stability of the vascularity surrounding the lesion lead to the recommendation of an elective Caesarean section at 37 weeks with both General and Vascular surgeons available. Regional gynecology oncology opinion was sought.

As the patient lived rurally and there was ongoing diagnostic uncertainty a planned admission at 35 + 5 was arranged. An upper GI endoscopy was performed and no oesophageal varices were seen. Oesophagitis was noted and omeprazole commenced.

At 36 + 3 the patient developed acute abdominal pain not relieved with opioid analgesia. There was distension in the left side of her abdomen with tenderness but no peritonism. The umbilical hernia and uterus were non tender. Bloods showed a normal, stable haemoglobin, platelet count and lactate. Pain was significant and not settling. MDT decision was for an acute exploratory laparotomy and LSCS as a joint case with an obstetric and a general surgeon.

A sub-umbilical midline incision was made and haemoperitoneum noted. An uncomplicated LSCS was performed where a fibroid was noted on the uterus. The abdominal mass was then located and noted to be mobile, solid and attached to the inferior aspect of the omentum with large omental varicosities (Fig. 2). No other attachment point was noted.

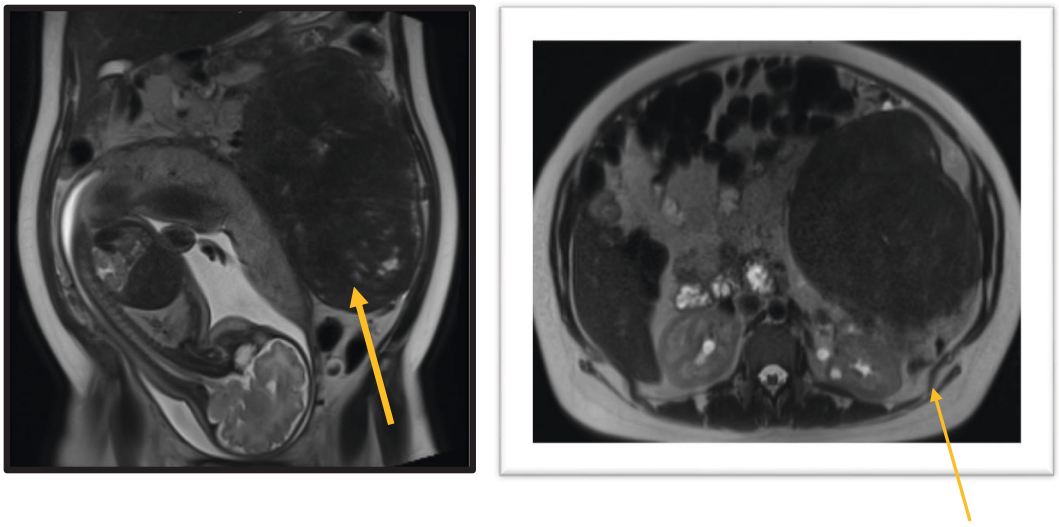

Fig. 1. T2 weighted coronal and axial MRI images demonstrating the intra-abdominal mass marked with yellow arrow. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

Fig. 2. Macroscopic view of STUMP and attached omentum with associated large varicosities.

An infra-colic omentectomy was performed with excision of the mass using Ligasure and large vessels secured with 1.0 Vicryl. The hernia was then identified; edges refreshed and then hernia repaired. Rectus sheath catheters inserted and closure of the sheath was using looped poly-dioxanone (PDS) with anchoring of the hernia sac to the sheath at closure. Estimated total blood loss was 1000 mL. There was planned overnight monitoring in the maternity high dependency unit (HDU).

The patient developed an ileus on day 3 requiring NG insertion with large aspirates. Her post-operative recovery beyond this was unremarkable and she was discharged on day 7. Outpatient follow-up occurred 4 weeks later consisting of clinical review and chest X-ray which was reassuringly clear of any evidence of metastasis. A week later the patient became acutely unwell with appendicitis; no mass recurrence was noted on imaging or intraoperatively. The patient will be followed up in the gynae-oncology outpatient clinic for 2 years total; every 6 months with clinical review and transvaginal (TV) pelvic ultrasound as per the MDM recommendation. TV USS 1 year postoperatively has shown only a 1.2 cm likely fibroid within the myometrium.

The excised mass measured 19 cm x150 cm x 120 cm and cut surface (Fig. 3) appeared to have areas of degeneration and small foci of possible calcification. Difficulty was encountered in histological assessment due to pregnancy related changes. Microscopically (Fig. 4) most of the neoplasm was arranged as loosely interlacing fascicles of smooth muscle fibre bundles. However, some areas demonstrated increased cellularity and areas of ischaemic necrosis. The mitotic activity of the neoplasm was low overall except for peri-necrotic areas.

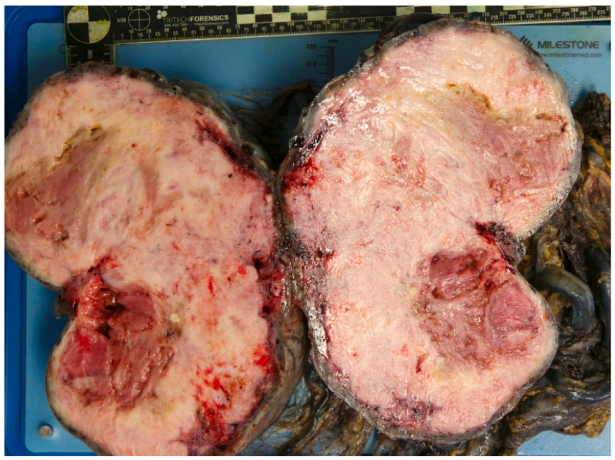

Fig. 3. Macroscopic cross-sectional view of the STUMP.

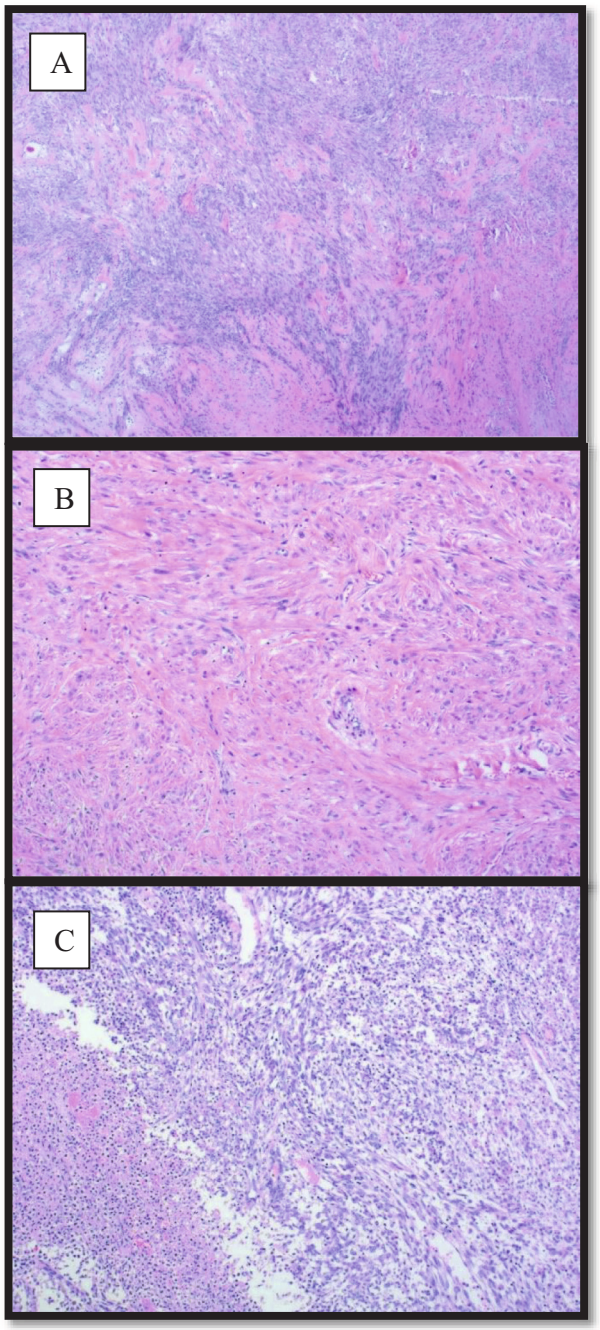

Fig. 4. Microscopic slides of STUMP A) 4xmagnification B) 10× magnification C) 10× magnification.

The neoplastic cells were positive for smooth muscle markers (SMA and Desmin) with low Ki67 proliferation index (up to 2 %). The neoplastic cells were positive for PR and ER but negative for p16, CD34, CD117, DOG1, Melan-A, and MDM2. The ectatic vessels within the omentum represented secondary changes. Based on these findings, an impression of an atypical smooth muscle tumor, best classified as STUMP; gynaecologic type, was reached. This was further confirmed by consultation with Auckland DHB specialists. Its location in the omentum with no connection to the uterus is compatible with so-called 'parasitic-type,' which represents a rare clinical presentation. The reported recurrence rate of STUMP when necrosis is present is up to 28 % in a limited number of cases [15].

This case has been reported in line with SCARE criteria [16].

3. Discussion

This is the third case reported in the literature of STUMP diagnosed during pregnancy and the only reported case of a parasitic STUMP during pregnancy. It is known that myomas can cause various pregnancy complications including preterm birth, antepartum and postpartum haemorrhage, pain due to "red degeneration"; with growth secondary to pregnancy hormones, and obstructed labour [2]. Myomas are not uncommon during pregnancy. A STUMP however is rarely seen during pregnancy but are potentially more problematic. As was reported in the second case study the significant growth and vascularity lead to worsening symptoms and warranted surgical intervention.

This case presented a diagnostic dilemma throughout its entirety. Assessment and management of new abdominal masses and/or hernias in pregnant patients are difficult due to anatomical changes. Radiological investigation is further limited during pregnancy and indeed interpretation is difficult due to anatomical changes related to pregnancy. The decision to operate and remove a highly vascular mass of unknown etiology was made secondary to acuity necessitating the late preterm delivery of the fetus.

Histological assessment of the mass was difficult due to pregnancy related change and the limitation of literature available on this topic meant evidence base was limited. The authors hope that this case report and overview of current literature on STUMP may assist clinicians with investigation, management and follow up for similar patients in the future.

4. Conclusion

Investigation and management of new intra-abdominal mass in pregnancy is challenging. Timely MDT input is required, particularly as complications may cause threat to both mother and fetus of which gestation is an additional concern. STUMP is rarely encountered in pregnancy and histological diagnosis may be delayed due to pregnancy related changes. More research is needed to guide follow up timing and duration for this patient group especially as literature demonstrates that younger patients with STUMP are more likely to experience recurrence [3].

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-chief of this journal on request.

Ethical approval

The study is exempt from ethical approval in our institution as it is a "Case report" and not a research study.

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Author contribution

Hannah Price = writing of original drafts.
Tanne Daniels = literature review and writing original drafts.
Tavaziva Mudzamiri = review of draft and supervision.
Archana Pandita = histology opinion and case contribution, review of draft.
Victoria Carlsen = review of draft, editing and supervision.

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Hannah Price.

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Declaration of competing interest

All authors declare they have no conflict of interest.

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