CASE REPORT

Successful maternal and perinatal outcomes in a term pregnancy with giant abdominopelvic leiomyomatosis

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Accepted 10 October 2017

SUMMARY

A 30-year-old second gravida with history of laparoscopic myomectomy and one previous caesarean section was admitted at 31 weeks and 2 days period of gestation (POG) with a diagnosis of diffuse abdominopelvic leiomyomatosis and moderate anaemia. After correction of anaemia with intravenous iron and erythropoietin, laparotomy was performed at 37 weeks POG. A healthy female baby weighing 2.9 kg was delivered by classical caesarean section followed by hysterectomy in view of multiple fibroid uterus with uncontrolled bleeding. Debulking surgery was performed, and multiple large intraperitoneal leiomyomata with encasing blood vessels were removed. There was another 15×15 cm leiomyoma arising from the diaphragm which was excised. She received 4 units of packed red blood cells and fresh frozen plasma intraoperatively. The postoperative course was uneventful, both mother and baby were healthy and discharged 7 days after surgery.

BACKGROUND

A considerable number of cases of diffuse peritoneal leiomyomatosis (DPL) following laparoscopic surgery and power morcellation during myomectomy or hysterectomy performed for uterine fibroids have been reported so far.^{1–4} Morcellation is required to permit extraction of large uterine myomas through small laparoscopic incisions.⁵ DPL is a rare condition where multiple smooth muscle-like nodules develop in any part of the peritoneal cavity. We present an unusual case of DPL associated with pregnancy with history of laparoscopic myomectomy managed successfully with caesarean hysterectomy and removal of all tumours.

CASE PRESENTATION

A 30-year-old second gravida was admitted in our antenatal ward at 31 weeks and 2 days period of gestation (POG) with abdominal distension. She had a caesarean delivery in 2008 for fibroid uterus with breech presentation, and a male baby weighing 2.7 kg was born who is alive and healthy. She underwent laparoscopic myomectomy in 2010 for a large (18×10 cm) uterine leiomyoma which was removed by power morcellation. She was hospitalised and medicated for a sudden episode of pain in the abdomen 5 years after myomectomy at some other hospital. Further evaluation revealed multiple intraperitoneal masses on CT

scan. Ultrasound-guided biopsy from tumour was suggestive of gastrointestinal stromal tumour (GIST). Based on the histopathology report, they started her on imatinib mesylate 400 mg per day for 6 months, but the patient did not respond to treatment. She then presented to our hospital for the first time in June 2016 when a repeat ultrasound-guided biopsy was performed which revealed benign mesenchymal lesion, possibly leiomyoma. However, she was lost to follow-up and reported in January 2017 with an amenorrhoea of 5 months and a positive urine pregnancy test. Her general condition was fine, and vitals were stable. Cardiovascular and chest examination was normal. Per abdominal examination revealed a grossly distended (figure 1) abdomen with a large 25×20 cm firm, irregular, lobulated and mobile mass extending from symphysis pubis to epigastrium. Fetal parts were palpable, and fetal heart rate was 140 beats per minute and regular. She had regular antenatal follow-up and received oral iron, calcium and vitamin supplementation. She was admitted at 31 weeks and 2 days POG for safe confinement.

INVESTIGATIONS

Ultrasound done at 19 weeks and 2 days POG suggested no gross fetal malformations. A large heterogeneous hypoechoic mass more than 20 cm in size was seen on the left side of the abdomen reaching up to the epigastrium. MRI done at 24 weeks POG showed multiple large heterogeneous masses with whorled configuration ranging in size from 5 to 20 cm in the abdomen and pelvis. The masses were also encasing the uterus, filling the lower abdomen and mid-abdomen and left subphrenic space compressing the left lobe of the liver and displacing the stomach inferiorly (figure 2A). A 6 cm midline lower abdominal defect was present with herniation of mass and mesentery through the defect (figure 2B). Mild ascites was present. Laboratory studies at admission demonstrated moderate iron deficiency anaemia with haemoglobin level of 7.4 g/dL. Screening for viral infections such as HIV and hepatitis B was negative, and blood sugars were normal. Liver function test, renal function tests and blood coagulation profile were within normal limits.

DIFFERENTIAL DIAGNOSIS

DPL is benign on histopathology but the diagnosis may be confused with peritoneal carcinomatosis or GIST preoperatively.¹

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To cite: Sharma JB, Kriplani A, Gupta M, et al. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-221954

Figure 1 Picture of the abdomen with huge distension with line drawing depicting the location of larger leiomyomas.

TREATMENT

She was given intravenous ferric carboxymaltose 2500mg in three divided doses 1 week apart along with four doses of injection of erythropoietin 4000 international units (IU) (total 16 000 IU) subcutaneously for treatment of anaemia, and her haemoglobin was built up to 10.7g/dL. She also received four doses of dexamethasone 6mg intramuscular 12 hours apart for fetal lung maturation. Fetal well-being was assessed with the help of obstetric ultrasound every 2 weeks and biophysical profile twice a week. Multidisciplinary team including obstetrician, anaesthetist, general surgeon and interventional radiologist were involved in her management. She was planned for caesarean delivery at 37 weeks POG in view of huge abdominal distension and increasing discomfort. Ten units of packed red blood cells (PRBCs) and fresh frozen plasma (FFP) were crossmatched and arranged. With the help of interventional radiologist, bilateral uterine artery balloon catheters were placed under fluoroscopy guidance, and balloons were inflated just before surgery. Combined general and epidural anaesthesia was given, and after establishing central venous access, the abdomen was opened by

Figure 2 (A) MRI of intrauterine pregnancy (arrow) with multiple abdominal leiomyomatosis. (B) MRI of herniating abdominal leiomyoma.

Figure 3 (A) Intraoperative picture of huge abdominal leiomyoma with large blood vessels. (B) Leiomyoma arising from the diaphragm (black arrow). (C) Specimen of the uterus (black arrow) with other leiomyomas.

a large midline incision from pubic symphysis to xiphisternum. Just beneath the peritoneum, there was a large 30×25 cm highly vascular mass (leiomyoma) arising from the right lower anterior abdominal and pelvic peritoneum, overlying the uterus (**figure 3A**). It was gently extruded outside the abdomen, after which the uterus could be visualised. A female baby weighing 2.9kg with an Apgar score of 9/10 was delivered by classical caesarean section as the lower uterine segment was obscured by large fibroid. There was another 25×20 cm mass arising from the left side of the abdomen and pelvic peritoneum. The peritoneal cavity and omentum were studded with multiple small leiomyomata. A large 15×15 cm tumour was also occupying the epigastric region over the liver with the pedicle arising from the diaphragm (**figure 3B**). Decision to perform hysterectomy was taken in view of multiple uterine fibroids and excessive blood loss. All the abdominopelvic tumours were removed after ligating their vascular pedicles and exposure of the ureters on both sides. The tumour in the epigastrium was also removed after clamping and cutting the pedicle arising from the diaphragm avoiding injury to the liver and stomach. Multiple other small tumours in the peritoneum were excised. The omentum was studded with small tumour deposits and hence, infracolic omentectomy was performed. As the appendix was adherent to one of the tumours, appendectomy was also done. The leiomyoma growing into the anterior abdominal wall hernia was removed. The picture of all masses removed is depicted (**figure 3C**). The hernial sac and the redundant abdominal skin were also excised. A 14 FG abdominal drain was inserted in the peritoneal cavity. The rectus sheath was identified and separated from the skin and closed with double (looped) monofilament nylon suture in continuous fashion. The skin was closed with interrupted 3-0 nylon suture. Uterine artery balloon catheters were removed after deflation.

Estimated blood loss was around 2.5 L, and 4 units each of PRBCs and FFP were transfused intraoperatively. The total duration of surgery was 4 hours. The specimen weighing 15 kg was sent for histopathology.

OUTCOME AND FOLLOW-UP

The patient was kept in intensive care unit for 24 hours and thereafter shifted to postoperative ward. The drain and catheter were removed on the third postoperative day. Both mother and baby were well and discharged on the seventh postoperative day.

Histopathological examination of specimen showed features of leiomyoma with focal areas of infarction and haemorrhage. There was no increase in mitosis and no features of necrosis or sarcomatoid changes. The tumour cells were immunopositive for desmin and smooth muscle antigen.

DISCUSSION

DPL is a condition characterised by benign smooth muscle cell proliferation in the peritoneal cavity. The multiple peritoneal nodules in DPL may stimulate peritoneal carcinomatosis.^{6–8} Although a rare entity, it may occur after the use of power morcellation to retrieve specimen during laparoscopic myomectomy or hysterectomy.^{2,5} Small pieces of myoma may fall in the peritoneal cavity during morcellation and may implant there leading to DPL. Even worse can be the spread of unsuspected cancer in leiomyosarcoma where morcellation is performed for presumed leiomyoma.^{2,9} Owing to the risk of dissemination of tumour cells in unsuspected leiomyosarcoma, the US Food and Drug Administration issued a warning against the use of power morcellation in 2014. In suspicious cases of leiomyosarcoma, it is preferable to perform either laparotomy (for hysterectomy or myomectomy) or mini laparotomy during laparoscopic procedures for retrieval of specimen instead of morcellation.¹⁰

The pathogenesis of DPL is unclear but recently some missense mutations in the mediator complex subunit 12 have been identified as the most frequent genetic aberration in uterine leiomyoma.¹¹ Concordant mutation in the uterine leiomyoma and peritoneal nodules might indicate peritoneal dissemination, whereas discordant mutation would indicate individual primary neoplasm.¹²

In the present case, diffuse and giant abdominopelvic leiomyomatosis followed laparoscopic myomectomy and power morcellation done few years back, leading to implantation and subsequent growth of myoma cells scattered in the peritoneal cavity. Our case is the first reported case of DPL in association with pregnancy. The myomas in our case were giant and at different locations in the anterior abdominal wall, pelvis, peritoneum, omentum and diaphragm. Similar finding of diffuse leiomyomatosis with deposits on the peritoneum, omentum and small and large bowel walls has been reported by other authors.^{13–15} Traditionally, myomectomy is avoided at the time of caesarean section owing to risk of excessive bleeding and hysterectomy. However, in the present case, caesarean hysterectomy along with removal of all the tumour masses, infracolic omentectomy (as there were small masses in the omentum) and appendicectomy (as it was adherent to myoma) were performed in the same sitting. Prophylactic balloon occlusion of anterior division of internal iliac arteries was done to reduce the blood loss during surgery. We propose that in a scenario such as this (giant leiomyoma with pregnancy), pregnancy can be continued until 36 to 37 weeks, and a planned laparotomy with removal of all visible leiomyomatosis can be performed after arranging adequate blood and blood products. A multidisciplinary team-work involving an experienced obstetrician, interventional radiologist, anaesthetist, general surgeon and neonatologist along with prior arrangement of adequate blood products can

result in successful maternal and perinatal outcomes as in the present case.

Learning points

- ▶ One should always think of diffuse peritoneal leiomyomatosis if diffuse masses are observed after a period of laparoscopic myomectomy for benign fibroids. The interval may vary from few months to few years.
- ▶ Carcinomatosis peritoneum and gastrointestinal stromal tumour are differential diagnosis. Ultrasound-guided biopsy is recommended to confirm diagnosis.
- ▶ Caesarean section followed by hysterectomy with removal of peritoneal and abdominal myomas can be safely performed in such cases.
- ▶ Adequate blood and plasma transfusion along with coordination between various team managers (obstetrician, anaesthetist, interventional radiologist, surgeon and paediatrician) is mandatory for successful maternal and fetal outcomes.

Contributors All authors contributed to the conception, design and acquisition of data and managed the patient. JBS and MG drafted the manuscript. AK and VS critically revised the manuscript. All authors gave final approval and agreed to all aspects of the work related to the accuracy and integrity of the case report.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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