

Laparoscopic Splenectomy for the Treatment of Refractory Immune Thrombocytopenia in Pregnancy

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Abstract

Background: Immune thrombocytopenic purpura (ITP) is a condition with potential hazard during pregnancy for both mother and fetus if platelet concentrations fall below a critical level. This report describes the use of laparoscopic splenectomy following unsuccessful medical management.

Case: A 35-year-old primigravid woman with systemic lupus erythematosus (SLE) developed ITP several years before becoming pregnant. She was treated early in pregnancy with high-dose oral prednisone and weekly intravenous immunoglobulin (IVIg) alternating with anti-D immune globulin, but laparoscopic splenectomy was indicated at 20 weeks' gestation because of thrombocytopenia. Following surgery, she continued prednisone and intermittent IVIg therapy until spontaneous delivery at 34 weeks' gestation. A small accessory spleen was identified postpartum by nuclear medicine scan. Satisfactory platelet concentrations were maintained postpartum using danazol and prednisone.

Conclusion: Laparoscopic splenectomy is a therapeutic option for women with ITP during pregnancy that fails to respond to medical management.

Résumé

Contexte : Le purpura thrombocytopénique idiopathique (PTI) est une pathologie comportant des dangers possibles au cours de la grossesse tant pour la mère que pour le fœtus si la numération des plaquettes chute en deçà d'un seuil critique. Ce rapport décrit le recours à une splénectomie laparoscopique à la suite de l'échec de la prise en charge médicale.

Cas : Une femme primigeste de 35 ans atteinte de lupus érythémateux disséminé (LED) a développé un PTI plusieurs années avant de devenir enceinte. Elle a été traitée, aux débuts de la grossesse, au moyen de fortes doses de prednisone orale, ainsi qu'au moyen d'immunoglobulines intraveineuses (IgIV)

hebdomadaires en alternance avec des immunoglobulines anti-D; toutefois la splénectomie laparoscopique s'est avérée indiquée à 20 semaines de gestation en raison d'une thrombocytopénie. À la suite de la chirurgie, on a continué à lui administrer de la prednisone et un traitement IgIV intermittent jusqu'à l'accouchement spontané à 34 semaines de gestation. À la suite de l'accouchement, on a détecté une petite rate accessoire par imagerie médicale nucléaire. Une numération des plaquettes suffisante a été maintenue au cours de la période post-partum au moyen du danazol et de la prednisone.

Conclusion : La splénectomie laparoscopique est un traitement potentiel pour les femmes présentant, au cours de la grossesse, un PTI sur lequel la prise en charge médicale n'a pas d'effets.

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INTRODUCTION

Immune thrombocytopenic purpura (ITP) affects between 1 and 3 women per 1000 pregnancies¹ and may lead to hemorrhagic complications in the mother and fetus.² ITP during pregnancy can usually be controlled with use of corticosteroids, intravenous immunoglobulin (IVIg), or anti-D immunoglobulin. Occasionally, however, splenectomy is required when thrombocytopenia is severe and refractory to medical therapy. We present a case of surgical management of ITP by laparoscopic splenectomy in the second trimester.

CASE

Jessica (pseudonym), a 35-year-old primigravida, presented to her obstetrician at 8 weeks' gestation with a 13-year history of systemic lupus erythematosus (SLE) and a 6-year history of ITP. The diagnosis of SLE was made in 1991 after she developed severe renal dysfunction and arthralgias. A renal biopsy confirmed lupus nephritis, and she was treated with corticosteroids and cyclophosphamide. She responded well and remained in remission until a second episode of

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lupus nephritis in 2000. This was confirmed again by renal biopsy and treated with mycophenolate mofetil, hydroxychloroquine sulphate, and corticosteroids, with good response. When she began attempts to conceive, azathioprine was substituted for mycophenolate mofetil and hydroxychloroquine sulphate, but she became severely thrombocytopenic, and treatment with azathioprine was withdrawn. After she had conceived, the corticosteroid treatment continued; despite this, she remained significantly thrombocytopenic, and at a platelet concentration of $20 \times 10^9/\text{L}$, she began to experience extensive bruising of her extremities. Her renal function, liver function, and blood pressure remained normal. Her levels of complement and double-strand anti-DNA were normal, but she was anti-SSA positive. Antiphospholipid antibodies were not identified.

Jessica's care between 8 and 16 weeks' gestation was provided by a multidisciplinary team that included an internist, a haematologist, a nephrologist, an obstetrician, and a perinatologist. Her oral prednisone therapy was increased to 50 mg daily, and she received weekly IVIG (50 g) to stabilize her platelet concentration. She experienced significant headaches with the weekly administration of IVIG, and as a result, her weekly treatment was changed to alternating IVIG and intravenous anti-D immunoglobulin (50 g/kg). Despite this treatment, her platelet concentration remained low, eventually falling to $8 \times 10^9/\text{L}$. At 16 weeks' gestation, Jessica was admitted to hospital with vaginal bleeding. Although her platelet concentration again responded to treatment with IVIG, her condition led to concern that she would be unable to continue her pregnancy to term and deliver safely. In anticipation of the possibility of her undergoing splenectomy and because of the potential risk of postsplenectomy sepsis, she was immunized against *Haemophilus influenza b*, *Meningococcus*, and *Streptococcus pneumoniae*. Jessica was referred to a general surgeon experienced in laparoscopic splenectomy for consideration of possible surgery.

She underwent a laparoscopic splenectomy at 20 weeks' gestation. Insertion of a Verres needle approach was not used for inducing pneumoperitoneum, to avoid potential injury to the uterus. Instead, a peritoneal cutdown was made under direct visualization to the left of the midline below the costal margin, and a 10 mm port was secured. Three 5 mm ports were placed radially in an arc around the spleen in the left upper quadrant, which is standard in laparoscopic splenectomy. The placement did not require modification because of the enlarged uterus. A small spleen was visualized, and no accessory splenic tissue was seen. Although space for intra-abdominal manipulation was initially restricted by the enlarged uterus (which reached the level of

the umbilicus), it was no longer restricted when peritoneal CO₂ insufflation was complete (15 mm Hg intraperitoneal pressure). The spleen was freed from its ligamentous attachments, and an endoscopic vascular stapler was used to divide the splenic vessels. The spleen was easily transferred into a 15 mm Endocatch II bag and removed.

During the case, standard maternal physiologic monitoring (electrocardiogram, pulse oximetry, arterial line, and end-tidal CO₂ monitoring) was provided, without intraoperative fetal monitoring. Five units of platelets were transfused during surgery. Estimated blood loss was 150 to 200 cc, and the patient tolerated the surgery well. She was discharged home 5 days after the surgery in stable condition.

Following the splenectomy, Jessica's platelet concentration increased for 2 weeks. However, when an attempt was made to discontinue the use of prednisone, she became acutely thrombocytopenic and required IVIG therapy (see Figure). Throughout the remainder of her pregnancy, she required IVIG therapy, less often than preoperatively, and was maintained on a lower dosage of prednisone (25 mg daily). Her gestational diabetic screen was positive, but subsequent oral glucose tolerance testing was normal. Her blood pressure remained normal, and renal function was stable. She had no recurrence of vaginal bleeding and did not require readmission to hospital. Fetal surveillance, with serial biophysical profiles and nonstress tests, remained reassuring; fetal growth was satisfactory, and fetal echocardiography showed no abnormality.

At 34 weeks' gestation, Jessica had spontaneous rupture of membranes and was admitted to hospital. She had received IVIG several days before admission, and her platelet concentration on admission was $81 \times 10^9/\text{L}$. Epidural analgesia was administered and induction of labour with oxytocin was begun. At a cervical dilatation of 4 cm, fetal heart rate monitoring showed repetitive variable decelerations and sustained bradycardia. Emergency Caesarean section was performed without complication, with delivery of a live male infant weighing 2220 g. Estimated blood loss was average ($< 1000 \text{ mL}$), and the baby's Apgar scores were 8 and 9, at 1 and 5 minutes, respectively. The baby was admitted to the level II nursery and underwent phototherapy on day 3 for mild jaundice. No arrhythmia was evident, and his platelet concentration was $149 \times 10^9/\text{L}$.

Jessica's platelet concentration fell to $39 \times 10^9/\text{L}$ at 6 hours after delivery. She again responded well to IVIG therapy and required IVIG several times over the subsequent 2 months. Eventually, her platelet concentration stabilized, and the prednisone dosage was successfully tapered to 10 mg every 2 days. She began treatment with danazol and hydroxychloroquine sulphate and no longer required IVIG.

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