Massive bleeding in pregnancy from ruptured oesophageal varices complicating portal hypertension: a cautionary tale

Alison Crocker MA MRCOG, **Joanna Girling** MRCP FRCOG and **Christina Cotzias** MRCOG

West Middlesex University Hospital – Obstetrics, Twickenham Road Isleworth, London TW7 6AF, UK

Summary: Portal vein thrombosis (PVT) is a rare complication of pancreatitis and can cause portal hypertension and oesophageal varices. Variceal rupture carries a high mortality. We report a case of successful pregnancy complicated by two episodes of massive variceal bleeding in a woman with PVT, and discuss how this might have been prevented.

Keywords: haemorrhage, high-risk pregnancy, hypertension, maternal–fetal medicine

INTRODUCTION

In portal hypertension (PHT) a collateral porto-systemic circulation with varices commonly develops¹ in the oesophagus, anterior abdominal wall² or extradural space.³ Outside pregnancy, massive bleeding from oesophageal varices carries a mortality rate of approximately 30%.⁴ Most data on PHT in pregnancy relate to women with cirrhosis,⁵ but case reports in pregnancy include ruptured varices in the splenic,⁶ intra-abdominal⁷ and abdominal wall vasculature.⁷ Consensus opinion on management of PHT in pregnancy includes a multidisciplinary approach to lower portal pressure, diagnose and manage oesophageal varices, prevent further thrombosis and bleeding and plan delivery.

We present the case of a woman with PHT secondary to portal vein thrombosis (PVT) after acute pancreatitis in a previous pregnancy, who had a successful pregnancy outcome despite recurrent massive haematemesis from oesophageal varices.

CASE REPORT

A 31-year-old Bangladeshi woman booked at seven weeks in her fifth pregnancy. She had had two uneventful vaginal deliveries, then developed gestational diabetes in her third pregnancy with term vaginal delivery, and subsequent type 2 diabetes. At 32 weeks in her fourth pregnancy she developed pancreatitis, acidosis, haemodynamic compromise, gross lipaemia (serum triglyceride 138 mmol/L) and fetal demise. The cause of the hyperlipidaemia was unclear but was deemed to have caused the pancreatitis which was otherwise of uncertain aetiology unrelated to cholelithiasis or alcohol abuse. Vaginal delivery followed induction of labour and her triglycerides were normalized. Five weeks postnatally, she was admitted with abdominal pain, diagnosed with PVT and anticoagulated. Thrombophilia screen was negative. On follow-up, her

haematological picture that included thrombocytopenia (platelet count $79 \times 10^9/L$) was felt to reflect possible myelofibrosis, but her platelet count normalized: in retrospect it is likely that this was due to hypersplenism. She was recommended long-acting reversible contraception and stated that she did not wish to conceive again.

Six months later, at booking in the index unplanned pregnancy, her platelet count was $93 \times 10^9/L$ and haemoglobin 9.2 g/dL. Despite extensive counselling about the risks of pregnancy, she declined termination. The multidisciplinary team treated her with insulin, propranolol 80 mg once daily, unfractionated heparin (since it has some lipoprotein lipase activity⁸) and aspirin. Abdominal ultrasound showed splenomegaly and an occluded portal vein. Her triglycerides remained normal and her resting pulse rate fell from 80 to 65 beats per minute after starting propranolol.

At 21+5 weeks, she became unwell, with a two-day history of haematemesis and melaena and haemoglobin of 5.7 g/dL with normal clotting. At endoscopy, ruptured oesophageal varices were diagnosed and banded. The risks of haemorrhage were felt to outweigh those of thrombosis so heparin was discontinued. The platelet count had fallen to $53 \times 10^9/L$ and the differential diagnosis again included myelofibrosis: four days later a bone marrow biopsy showed a cellular reactive marrow in keeping with hypersplenism. She had further elective endoscopies with variceal banding at 22 and 28 weeks: her platelet count was 69.

At 30+2 weeks, she presented with another massive haematemesis, was anaemic (haemoglobin 8.5 g/dL) and had endoscopic banding of bleeding varices. The same day she delivered a healthy 1.61 kg daughter by caesarean section. Large collateral varicosities in the visceral peritoneum were divided to gain access to the uterus: blood loss was 1500 mL.

She was transferred to King's College Hospital Liver Unit on day 1, and was managed with terlipressin, omeprazole, clotting support, further endoscopic variceal banding and six weeks of thromboprophylaxis. She was transferred back to our unit on day 4 and discharged home on day 15. Interval endoscopy

Correspondence to: Alison Crocker
Email: alisoncrocker@yahoo.co.uk

showed two quiescent columns of varices. She had a Mirena intrauterine system fitted six weeks later.

DISCUSSION

PVT is unusual in pregnancy,⁹ being most frequently seen in patients with other conditions causing hypercoagulable states. PHT is also uncommon in pregnancy, but irrespective of aetiology, lower oesophageal varices develop in up to 90% of cases. They may appear for the first time or become more numerous during pregnancy, because of the raised portal pressure caused by pregnancy-driven increased blood volume and increased azygous vein flow; this also increases the risk of bleeding.⁷ Endoscopic band ligation is the treatment of choice for bleeding varices in pregnancy. Beta blockers are used to reduce portal pressure to prevent variceal bleeding.

Preconceptual assessment of women with chronic disease is recommended by the Centre for Maternal and Child Enquiries (CMACE),¹¹ to facilitate discussion of the risks of pregnancy and optimization of medication. In this case, after the fourth pregnancy our patient was not planning further pregnancies. The fifth pregnancy was relatively soon so elective endoscopy had not taken place: this is usually carried out about a year after PVT when varices are commonly seen. While elective endoscopy is not routine in pregnancy, in retrospect there is the possibility that elective first trimester variceal banding could have prevented or delayed the variceal haemorrhages, and we would recommend this in future if it has not been carried out prior to conception. However, as with our patient, new varices may develop in pregnancy: so endoscopic surveillance is also recommended, although it may not prevent haemorrhage. Magnetic resonance imaging has been successful in diagnosing ectopic varices in sites which may compromise anaesthetic or surgical access at delivery, but is not sensitive enough to exclude oesophageal varices.

Pregnant women with oesophageal varices must be managed by a multidisciplinary team of obstetricians, gastroenterologists, anaesthetists and paediatricians. Beta blockers reduce portal pressure and minimize the chance of bleeding;¹⁰ treatment response is gauged by a drop in resting pulse rate, which appeared to be satisfactory in this case. However, beta blockers do not prevent all episodes of bleeding.

The rescue treatment of choice in pregnancy is endoscopic band ligation. If there is a severe variceal bleed then consideration should be given to delivery to reduce the risk to the mother, balanced against the fetal risks of iatrogenic

prematurity. Postnatally women should ensure effective contraception.

DECLARATIONS

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Guarantor: AC.

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