CASE REPORT

Portal vein thrombosis during pregnancy

Papa Dasari, Sathyalakshmy Balusamy

Department of Obstetrics and Gynaecology, JIPMER, Puducherry, India

Correspondence to
Professor Papa Dasari,
dasaripapa@gmail.com

SUMMARY

A 22-year-old primigravida was diagnosed to have portal vein thrombosis during 20th week of gestation by ultrasound examination which was carried out to rule out congenital fetal anomalies. She had splenomegaly and thrombocytopenia. Investigations did not reveal any prothrombotic disorder. She was managed with anticoagulants which were started at 31 weeks of pregnancy. Labour was induced at 40 weeks of gestation and she delivered a healthy neonate without any complications. Anticoagulants were restarted after delivery and continued through the postpartum period and up to 6 months thereafter.

BACKGROUND

Cavernous transformation of the portal vein occurs as a sequela of portal vein thrombosis (PVT) and is a rare occurrence in pregnancy. The aetiology is diverse and there are no definite guidelines for management of these women during pregnancy due to the paucity of cases reported and therefore the treatment is on individualised basis. The present case adds to the existing literature and emphasises the treatment issues for an optimal outcome.

CASE PRESENTATION

A young woman aged 22 years attended our obstetric services at 30 weeks of gestation as portal vein cavernoma complicating pregnancy. In an ultrasound performed at 20 weeks to rule out fetal anomalies she was diagnosed to have chronic PVT with partial recanalisation elsewhere and she was advised to attend a higher institution for further care. An earlier report of ultrasound performed at 15 weeks of pregnancy did not have findings of PVT.

She had no history of haematemesis or blood transfusion. There was no significant medical or surgical illness in the past. There was no pallor, pulse rate was 84/min, blood pressure 110/70 mm Hg, no pedal oedema, no icterus and her cardiovascular and respiratory systems were within normal.

Abdominal examination revealed mild splenomegaly, uterus enlarged to 30 weeks, relaxed and fetal heart was 140/min. She was evaluated for thrombophilia as a possible cause of PVT.

INVESTIGATIONS**Ultrasound abdomen (done elsewhere)**

31 May 2011

- ▶ Single live intrauterine pregnancy.
- ▶ Gestational age 15 weeks.
- ▶ Placenta left anterolateral wall.

28 July 2011

- ▶ Single fetus 22 weeks gestational age without any congenital anomalies.
- ▶ Placenta anterior, Liquor average.
- ▶ Mild splenomegaly; PVT present.
- ▶ Pancreas, liver and both kidneys were normal.

29 July 2011

Hb 9 g%, white cell count 6900 cells/mm³, platelets 73 000/mm³, prothrombin time 18 s.

Liver function tests

- ▶ Serum bilirubin 0.9 mg/dl
- ▶ Total protein 6.1 g%
- ▶ Albumin 2.8 g%

Renal function tests

- ▶ Blood urea 14 mg/dl
- ▶ Serum creatine 0.5 mg/dl
- ▶ VDRL—non-reactive; HIV—non-reactive; HbS Ag—negative

Thrombophilia profile

- ▶ Antithrombin III, Factor VII and Factor VIII, Protein C and S—within normal range
- ▶ Antiphospholipid antibodies—negative
- ▶ Lupus anticoagulant—negative

23 November 2011

- ▶ Hb 9.1 g%; platelets 166 000/mm³; peripheral smear—normocytic normochromic RBCs

Ultrasonography

- ▶ Cephalic presentation; bipareital diameter (BPD)=35 weeks; abdominal circumference (AC)=34 weeks; femur length (FL)=34 weeks
- ▶ Estimated fetal weight 2670 g liquor average
- ▶ Portal vein showed increased periportal echogenicity and multiple tubular hypoechoic to anechoic structures noted in portal lumen and portal vein not seen separately suggestive of portal vein cavernoma. Spleen enlarged to 13 cm and no ascites.

29 November 2011

- ▶ Platelet count 350 000/mm³
- ▶ Prothrombin time 21.2; International Nationalised Ratio (INR) 1.3
- ▶ APTT control 28.5" test 29.8"
- ▶ Doppler findings suggestive of portal vein cavernoma

3 December 2011 (just after delivery)

- ▶ Hb 8.5 g%; platelet count 120 000/mm³
- ▶ 9 December 2011 (sixth postnatal day)
- ▶ Platelets 145 000/mm³ INR 1

To cite: Dasari P, Balusamy S. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2012-008325

12 December 2011

- ▶ INR 1.53 (on 6 mg of Warfarin)
- ▶ Ultrasonography—before discharge
- ▶ Portal vein cavernoma persisting

DIFFERENTIAL DIAGNOSIS

Prothrombotic disorder

TREATMENT

Medical and surgical gastroenterology consultation was obtained regarding management of PVT soon after admission. They advised conservative management and differed in anticoagulation. But because of the experience with a previous case of maternal death due to splenic vein thrombosis, it was decided to start on anticoagulation under close supervision by the obstetrician in-charge.

She was given injection unfractionated heparin 5000 units subcutaneously twice daily. Platelet count was repeated weekly. It gradually rose and at 38 weeks it was 350 000/mm³.

She was admitted at 38 weeks for safe confinement. With fetal status reassuring induction of labour was undertaken at 40 weeks when her amniotic fluid index was 8. Heparin injection was stopped 12 h prior to induction of labour. Cervical ripening was achieved by an intracervical foley catheter and cerviprime gel after 24 h. Labour was augmented with oxytocin and she delivered a live baby weighing 2.9 kg with APGAR of 8 at 1 min and 9 at 5 min, by vacuum extraction because of fetal distress.

OUTCOME AND FOLLOW-UP

She was started on heparin injection 6 h after delivery and tablet Warfarin 4 mg after 3 days. Repeat ultrasound performed after 10 days revealed the same findings of cavernoma. She was discharged on day 10 on Tab. Warfarin 6 mg once a day and advised to continue for 6 months. She came for postnatal check-up at 6 weeks and was found to be normal.

DISCUSSION

PVT complicating pregnancy is a rare occurrence. The aetiology of PVT is highly diverse. The common causes include cirrhosis, neoplasia, myeloproliferative disorders, inflammatory disorders like pancreatitis and thrombophilias. The hypercoagulable state of pregnancy itself can predispose to thrombosis of portal veins.^{1 2} Acute PVT presents with fever, abdominal pain and new onset ascites whereas chronic PVT presents with haemetemesis, splenomegaly and ascites. The complications of PVT are portal hypertension, thrombocytopenia, splenomegaly and the most feared being variceal bleeding. Pregnant women are especially at high risk of variceal bleeding which can lead to maternal morbidity and mortality. In a study of 26 pregnancies in 14 women with extrahepatic portal vein obstruction, pregnancy outcome was reported to be successful provided the disease is adequately controlled.³ Management of complications and long-term anticoagulation are the key issues in the treatment of PVT.

Cavernomatous transformation of portal vein is the development of collateral venous circulation which occurs as a sequela of PVT. It can occur as early as 6–20 days after PVT.⁴ Pregnancy in patients with portal vein cavernoma has been reported in seven cases so far. Most of the patients reported earlier had an underlying aetiology for portal vein obstruction like thalassaemia intermedia,⁵ congenital hepatic fibrosis,⁶ omphalitis,⁷ hepatic haemangiomas⁸ and protein S deficiency.⁹ The present case did not have any underlying disorder and she

was negative for thrombophilic disorders. All the patients reported earlier were diagnosed with portal vein cavernoma prior to pregnancy except in the patient with thalassaemia intermedia where diagnosis was established when imaging was carried out for abdominal pain. In the present case diagnosis was made while a routine imaging was performed to rule out fetal anomalies.

Thrombocytopenia is a well-known complication of portal vein cavernoma. Therefore, weekly determination of platelet count is essential to prevent haemorrhagic complications. A minimum platelet count of 50 000 is essential when caesarean section is to be undertaken. Bleeding from oesophageal varices is a dreaded problem in patients with portal vein cavernoma. In patients with history of variceal bleed prior to pregnancy, it is necessary to ensure that the varices are treated adequately in the prepregnancy period. None of the patients reported earlier had haemetemesis during pregnancy. Wax *et al*⁷ have described wire-less video oesophageal capsule endoscopy and MRI as safe and effective non-invasive techniques for evaluation of oesophageal and gastric varices in the pregnant state. Gastrointestinal (GI) endoscopy in pregnant women carries the risk of fetal hypoxia due to maternal over sedation and vena caval compression due to maternal positioning. American Society for Gastrointestinal Endoscopy has stated that upper GI endoscopy should be reserved only for women who have significant variceal bleed during pregnancy.¹⁰

Anticoagulation should be initiated in all patients to prevent extension of thrombus and recurrence of new thrombosis. It has been observed that partial and complete recanalisation of the portal vein is higher in patients treated with anticoagulants.¹¹ Pregnancy itself is a hypercoagulable state and therefore can predispose to thrombosis. Anticoagulation therapy was not given in the case reported by Bayratkar *et al* probably because of the presence of oesophageal varices. The associated thrombocytopenia and varices in patients with portal vein cavernoma are the limiting factors in the initiation of anticoagulation therapy. Heparin should be stopped at least 12 h prior to induction of labour and restarted after 6 h in vaginal delivery and 12 h after caesarean section. Anticoagulation should be continued for at least 6 months¹² and depending upon the underlying aetiology.

Of the seven cases reported earlier caesarean section was performed for five patients, three were performed for obstetric indications. In the case reported by Ducarme *et al*,⁹ the patient had jejunal varices and caesarean section was performed because of the expected higher risk of variceal bleed during vaginal delivery. Since variceal bleed is the most common complication which accounts for increased morbidity and mortality, elective caesarean section appears to be safe in these patients. Concurrent splenectomy with caesarean section was performed in one patient who had splenomegaly.⁶ The safety of such a procedure is not yet established. Two patients who were planned for vaginal delivery delivered without any intrapartum or postpartum complications.^{7 9} Neonatal outcome in the cases reported including the present one are good except in the patient with congenital hepatic fibrosis who had twin pregnancy with single fetal demise.

Based on the cases reported so far, a multidisciplinary approach including obstetricians, gastroenterologists and anaesthetists is essential to optimise the outcome. Preconception counselling is vital. Oesophageal and gastric varices should be treated in the prepregnant state. Anticoagulant therapy plays an important role and should be advocated in all patients.

Learning points

- ▶ Portal vein thrombosis (PVT) with pregnancy should be treated as a high risk case.
- ▶ Haematological monitoring is essential as thrombocytopenia is common.
- ▶ Anticoagulation should be initiated under careful monitoring to prevent extension of thrombus and recurrence of new thrombus.
- ▶ Splenic vein/PVT should be considered as a form of deep vein thrombosis.
- ▶ Anticoagulation in this condition prevents consumption of platelets and thus prevents consumptive coagulopathy and mortality associated with it.
- ▶ Anticoagulation benefits the patient and prevents morbidity and mortality associated with the condition especially pulmonary thromboembolism.

Contributors SB carrying out the instructions given by PD, consultant obstetrician in-charge of the patient. Both authors contributed to the manuscript and review of the literature for this case as well as the previous case report.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Goodrich MA, James EM, Baldus WP, *et al.* Portal vein thrombosis associated with pregnancy. A case report. *J Reprod Med* 1993;38:969–72.
- 2 Jouini S, Fakunle YM, Azazy A. Portal vein thrombosis in pregnancy: case report and review of the literature. *Ann Saudi Med* 2002;22:227–9.
- 3 Aggarwal N, Chopra S, Raveendran A, *et al.* Extra hepatic portal vein obstruction and pregnancy outcome: largest reported experience. *J Obstet Gynaecol Res* 2011;37:575–80.
- 4 De Gaetano AM, Lafortune M, Patriquin H, *et al.* Cavernous transformation of the portal vein: patterns of intrahepatic and splanchnic collateral circulation detected with Doppler sonography. *AJR Am J Roentgenol* 1995;165:1151–5.
- 5 Hackmon-Ram R, Holcberg G, Bashiri A, *et al.* Thalassemia intermedia and cavernous transformation of portal vein thrombosis in pregnancy. *Eur J Obstet Gynecol Reprod Biol* 2003;107:101–4.
- 6 Bayraktar Y, Tuncer ZS, Kabukcu A, *et al.* Pregnancy complicated by congenital hepatic fibrosis with cavernous transformation of the portal vein: a case report. *Am J Obstet Gynecol* 1997;177:459–61.
- 7 Wax JR, Pinette MG, Cartin A, *et al.* Cavernous transformation of the portal vein complicating pregnancy. *J Obstet Gynecol* 2006;108(3 Pt 2):782–4.
- 8 Tuncer I, Arslan H, Harman M. Two giant cavernous hemangioma caused cavernous transformation of the portal vein in a pregnant woman. *Turk J Gastroenterol* 2002;13:229–31.
- 9 Ducarme G, Plessier A, Thuillier C, *et al.* Pregnancy and delivery in patients with portal vein cavernoma. *Gynecol Obstet Invest* 2009;68:196–8.
- 10 Qureshi WA, Rajan E, Adler DG, *et al.* ASGE guideline: guidelines for endoscopy in pregnant and lactating women. American Society for Gastrointestinal Endoscopy. *Gastrointest Endosc* 2005;61:357–62.
- 11 Sogaard KK, Astrup LB, Vilstrup H, *et al.* Portal vein thrombosis; riskfactors, clinical presentation and treatment. *BMC Gastroenterol* 2007;7:34.
- 12 Kocher G, Himmelmann A. Portal vein thrombosis (PVT): a study of 20 non-cirrhotic cases. *Swiss Med Wkly* 2005;135:372–6.

Copyright 2013 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow