

A case of splenic artery rupture secondary to nodular regenerative hyperplasia in pregnancy

This case report describes the outcome of a high-risk pregnancy of a 38-year-old woman of Tigrinyan origin, which was complicated by non-cirrhotic portal hypertension secondary to Nodular Regenerative Hyperplasia.

Dr James Goadsby, Miss Eleanor Parker, Miss Kenga Sivarajah, Miss Lisa Long

Nodular Regenerative Hyperplasia is a rare liver condition, first described in 1959, characterised by a widespread benign transformation of the hepatic parenchyma into small regenerative nodules⁽¹⁾. It is estimated to be the cause of 27% of cases of non-cirrhotic portal hypertension worldwide and is a common cause of portal hypertension in women of fertile age⁽²⁾.

Pursuing pregnancy is a high-risk endeavor for women with portal hypertension, with perinatal mortality rates of up to 18%. There is an increased risk of variceal bleeding as the physiological adaptations of the maternal circulation to pregnancy increases flow to collateral circulations and increases portal venous pressures⁽³⁾.

The third trimester and the second stage of labour are the most high-risk periods with up to seventy-five percent of women with varices experiencing at least one bleed during pregnancy. In cases of oesophageal varices banding can be utilised prophylactically to treat acute bleeding in conjunction with terlipressin⁽⁴⁾.

Booked in 1st trimester
P1- previous SVD 2014
PMHx: Non-cirrhotic portal hypertension caused by Nodular Regenerative Hyperplasia. Awaiting liver transplant.

- Classification: Severe *-peri-splenic, para-umbilical and short gastric varices alongside concurrent massive splenomegaly.*
- Evidence of an impairment of hepatic synthetic function at booking with Platelets of 30, an INR of 2.3 and Albumin of 30.

The patient had one admission at 26 weeks with reduced fetal movements however CTGs were normal and she was discharged the following morning.

During the first trimester, she was counselled by the obstetric team extensively about the increased risk of a variceal bleed during the pregnancy. This was quoted at 35- 40% and due to the high risk of serious morbidity and mortality, she was offered a termination of pregnancy.

- Antenatal cardiovascular investigation showed a hyper dynamic circulation with biventricular and bi-atrial enlargement, moderate tricuspid regurgitation and ejection fraction of 61.7%.
- OGD revealed no oesophageal or gastric varices however an MRI at 32 weeks reported concerning features of; significant liver atrophy and massive splenomegaly, large volume short gastric varices with a large peri-splenic variceal bed.

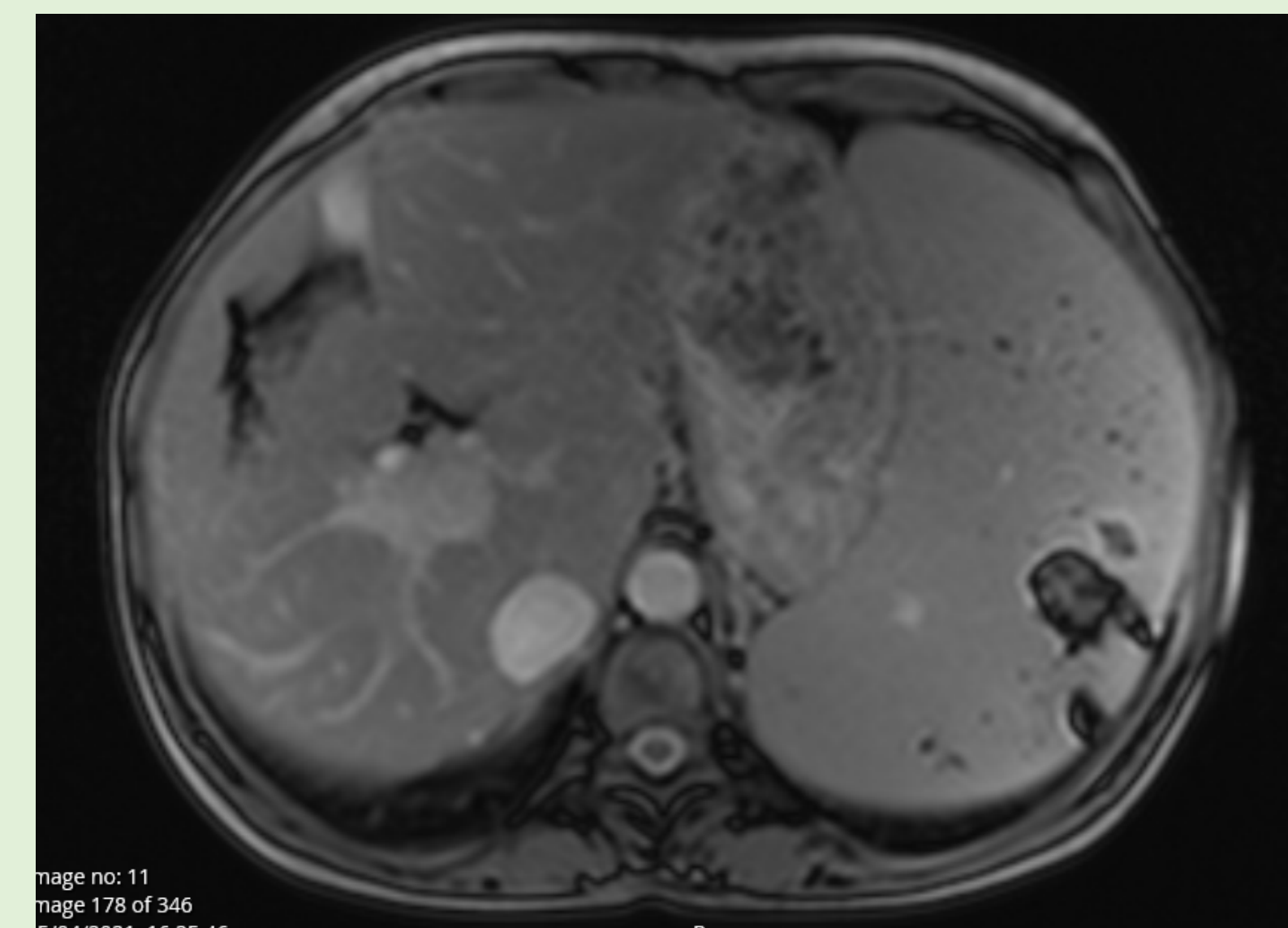
At 33+1/40 the patient was found collapsed at home and transferred by London Ambulance Service. She was intubated on arrival in A&E and successfully resuscitated after multiple episodes of loss of cardiac output. Perimortem C-Section was performed in resus with significant volumes of free intra-peritoneal bleeding noted. The estimated blood loss was 2000ml. The patient's abdomen was packed and she was transferred to trauma theatres with ongoing blood products transfusion.

Once transferred to theatre she underwent a laparotomy with successful ligation of bleeding varix in her splenic hilum.

After a 15-day intensive care stay and a 3-week rehabilitation as an inpatient the patient was discharged with outpatient input from the psychology team and bereavement midwifery team. She remains on the liver transplant waiting list.

She was counselled extensively in the post-natal period regarding contraceptive options and the risks of any future pregnancy. The patient had a subsequent first trimester pregnancy terminated and was advised to avoid pregnancy until she is one year post transplant.

This case highlights a rare cause of non-cirrhotic portal hypertension in a pregnancy women and the significant risks to mother and fetus of embarking on a pregnancy with poor synthetic liver function and known varices. Despite extensive counselling, close observation and timely emergency care this case sadly ended with neonatal demise and a near terminal event for the patient. This case emphasises the importance of multi-disciplinary management and contingency planning for high-risk events such as splenic artery aneurysm rupture.



MRI 32+1/40: significant liver atrophy and massive splenomegaly, large volume short gastric varices

References:

1. Hartleb M, Gutkowski K, Milkiewicz P. Nodular regenerative hyperplasia: evolving concepts on underdiagnosed cause of portal hypertension. World J Gastroenterol. 2011;17(11):1400-9.
2. Reshamwala PA, Kleiner DE, Heller T. Nodular regenerative hyperplasia: Not all nodules are created equal. Hepatology. 2006;44(1):7-14.
3. Aggarwal N, Negi N, Aggarwal A, Bodh V, Dhiman RK. Pregnancy with portal hypertension. J Clin Exp Hepatol. 2014;4(2):163-71.
4. López-Méndez E, Ávila-Escobedo L. Pregnancy and portal hypertension a pathology view of physiologic changes. Annals of hepatology. 2006;5(3):219-23.
5. Morton A, Laurie J, Hill J. Portal hypertension in pregnancy – Concealed perils. Obstetric Medicine. 2020;13(3):142-4.