BUDD-CHIARI SYNDROME
AND PREGNANCY
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INTRODUCTION

Budd-Chiari syndrome (BCS) is a rare disorder, varied in aetiology, inconsistent in presentation, unpredictable in progression and challenging in therapy. We describe a rare case of successful pregnancy in a 16-year-old with a history of BCS.

CASE REPORT

A 16-year-old primigravida attended for antenatal care at 12 weeks gestational age. Three years previously, she took the combined oral contraceptive pill for six months. There was no significant medical history. There was a family history of thromboembolism. In her forties, the maternal grandmother was diagnosed with deep vein thrombosis. The booking bloods, dating scan and anomaly scan were normal. The pregnancy was uncomplicated. At 38 weeks gestational age, she went into spontaneous labour and had a normal vaginal delivery of a live male infant with birth weight of 3.13kg. There was no postpartum haemorrhage.

Four weeks postpartum, she developed right-sided pleuritic chest pain and shortness of breath. The physical findings were of jaundice, reduced air entry to the base of the right lung, ascites, tenderness to the left hypochondrium and pedal oedema.

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<th>Haematological and biochemical profile</th>
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<td>Haemoglobin</td>
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<td>White cell count</td>
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<td>Platelet count</td>
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<td>Red blood cell count</td>
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Kidney function tests were normal
Liver function tests were abnormal:
- alkaline phosphatase | 235 IU/L |
- alanine aminotransferase | 68 IU/L |
- gamma glutamic transpeptidase | 162 IU/L |
- albumin | 24 g/L |

The amount of protein in the urine over 24 hours was normal
Autoimmune antibody tests were normal

Bumetamide and spironolactone were commenced. The patient was anticoagulated using intravenous heparin which was changed later to oral warfarin.

Worsening jaundice, increasing ascites and pedal oedema, repeated abnormal liver function tests and the development of iron deficiency anaemia required a gastrointestinal specialist assessment. The patient did not improve greatly and an underlying hepatic cause for the pulmonary embolism and gastroenterology symptoms was suspected. She was then referred to a tertiary liver unit where the diagnosis of BCS was made and a transjugular intrahepatic portosystemic shunt (TIPSS) was performed with the use of a Viatorr stent. Three-monthly duplex surveillance liver scans were undertaken to ensure patency of the stent since occlusion of the hepatic or portal vein could lead to liver failure. In the following 18 months, she presented with two further episodes of pleuritic chest pain and was treated with anticoagulants.

Radiological findings

1. The chest radiograph was normal
2. Abdominal ultrasound:
   - normal liver and gallbladder
   - spleen enlarged
3. Lung ventilation perfusion and computed tomography scans revealed pulmonary embolism of the right lung

Two years after the first delivery, she presented with lower abdominal pain in her second pregnancy at five weeks gestation. Consultant-led care was provided with regular antenatal visits and twice-monthly blood tests for thrombocytopenia. Warfarin was changed to enoxaparin sodium at a therapeutic dose of 100mg daily. At 12 weeks gestation she developed pleuritic chest pain highly suggestive of pulmonary embolism but a lung scan was declined. The enoxaparin sodium dose was increased to 80mg twice daily. The booking bloods, dating scan and anomaly scan were normal. At 24 weeks gestational age she developed anaemia with a haemoglobin level of 10g/dl and was started on iron tablets. Threatened preterm labour was diagnosed at 28 weeks of pregnancy which resolved with oral nifedipine. The haemoglobin continued to fall to a level of 8g/dl. The kidney and liver function tests were normal. Throughout the pregnancy, serial growth scans and umbilical artery Doppler flow velocity waveforms were normal. She was reviewed regularly by the gastroenterologist and the haematologist.

At 34 weeks and five days gestational age, she was admitted in preterm labour at 9cm dilatation. A live male was delivered with a birth weight of 2265g with Apgar scores of 9 at 1 minute and 9 at 5 minutes. The third stage was managed actively with the use of syntocinon. There was an estimated blood loss of 450mls. The neonate was successfully treated in the special care baby unit for prematurity. Postpartum thromboprophylaxis was achieved with a ten-week course of enoxaparin sodium followed by longterm use of warfarin.
DISCUSSION

BCS is hepatic venous outflow obstruction. The aetiology cannot be determined in the majority of cases but may complicate thrombotic diathesis, including myeloproliferative disorders, such as polycythemia vera and paroxysmal nocturnal haemoglobinuria, tumours, chronic inflammatory diseases, clotting disorders, and infections.\(^5\) BCS rarely occurs in pregnancy or in the puerperium. Hypercoagulability and pelvic sepsis are possible mechanisms for its development.

The peak age of onset of BCS is in the third decade of life. Clinically, it is characterised by a triad of abdominal pain, hepatomegaly and ascites, and rapidly progressive hepatic failure. In the absence of surgical therapy, median survival is less than two years.

Duplex colour-flow Doppler ultrasound is the diagnostic procedure of choice. Magnetic resonance imaging (MRI) scanning helps in the assessment of hepatic venous blood flow. The utilisation of percutaneous liver biopsy and hepatic angiography had led to earlier diagnosis.\(^5\)

Selection of treatment is based on the degree of hepatic injury, liver biopsy results, potential for parenchymal recovery and pressure measurements. Hepatic vein stenoses have been treated successfully on a short-term basis with percutaneous dilatation. The concomitant use of stents has provided for extended patency rates, sometimes as high as 80% to 90%.\(^5\)

In cases of extensive thrombosis in which dilatation is unlikely to be effective, radiological management has been geared toward transjugular intrahepatic portosystemic shunt (TIPS) placement.\(^4\) TIPS is of use either as a primary or secondary method to relieve venous obstruction in BCS.

In the presence of fulminant forms of BCS, in cases of established cirrhosis or frank fibrosis, or for patients with defined hepatic metabolic defects (eg, protein C or protein S deficiency), liver transplantation is the treatment of choice.\(^5\) Liver transplantation in selected cases of BCS has reported a ten-year survival rate of approximately 75%.

CONCLUSION

BCS is a rare disease and difficult to diagnose. It needs to be suspected in all patients presenting acutely with hepatomegaly and ascites. A high index of suspicion may hasten diagnosis and management.

In pregnancy, a multidisciplinary approach is recommended for BCS, as it is mandatory to balance the health of a mother and an unborn child.

Most patients, irrespective of the primary treatment, will require anticoagulation therapy for life.

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REFERENCES