The Budd-Chiari syndrome in pregnancy

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Summary
A case of Budd-Chiari syndrome in a young woman, which started probably in the last trimester of pregnancy, is described. The diagnosis was made clinically and was confirmed by inferior venacavography and on exploratory laparotomy. The possible connection of the syndrome with the pregnancy is discussed.

Introduction
The Budd-Chiari syndrome, due to occlusion of the hepatic veins by tumour or thrombus arising either locally or by extension from the inferior vena cava, is a rare condition. In his thorough review of the syndrome, Parker (1959) described 149 cases. In the majority of the reported cases the diagnosis was made at post-mortem (Parker, 1959, Westcott, 1973) and, in a few of them, only during the last 20 years, was the diagnosis made in life, owing to the development of new techniques such as liver biopsy, hepatic venography, selective hepatic arteriography, inferior venacavography and liver scintiscan.

The patient in the present case was a young, pregnant woman, and the diagnosis was made ante mortem. On this occasion, the possible connection of the syndrome with pregnancy is discussed.

Case report
A 25-year-old housewife was admitted to the medical department of Evangelismos hospital in February 1976, because of gross abdominal distension. The patient had had a normal delivery 12 days before admission. During the last trimester of pregnancy the abdomen had been so unusually large that the family obstetrician had suspected twin gestation. At the same time she complained of a moderately severe right hypochondrial pain. The enlargement of the abdomen persisted after the delivery. The presence of ascites was confirmed and the patient was admitted to hospital. On examination the patient was slightly pale and moderately jaundiced. The abdomen was grossly distended owing to the presence of ascites. The inferior epigastric veins were distended because of established collateral circulation. The liver was enlarged, hard, nodular and slightly tender. There was no hepato-jugular reflux.

Laboratory investigations
Haematocrit, 35%; WBC, 9.8 × 10^9/l (polymorphs 80%); platelets, 250–0 × 10^9/l; ESR, 76 mm; serum bilirubin, 214 μmol/l (conj., 10 μmol/l); SGOT, 65 i.u./l; SGPT, 65 i.u./l; alkaline phosphatase, 22 KAu; fasting blood sugar 4.7 mmol/l; blood urea, 6.3 mmol/l; serum sodium, 147 mmol/l; potassium 5.4 mmol/l; serum proteins, 70 g/l (albumin 36 g/l, globulin 34 g/l); immunoglobulins: IgG 12.5 g/l; IgA 2.86 g/l; IgM 2.00 g/l. Prothrombin time 11.3/13.5 sec.; α-feto-protein, negative; HBsAg, negative. Bromsulphthalein retention, 18% after 45 min.

Examination of the ascitic fluid
Protein 300 g/l, many white cells (polymorphs 65%), culture negative, examination for acid-fast bacilli negative, examination for malignant cells negative (three times). Chest X-ray showed elevation of the right diaphragm. ECG showed sinus tachycardia. The liver scan (Fig. 1) showed great...
Fig. 1. Liver scan anterior view: the hypertrophied caudate lobe is seen as an area of increased uptake superimposed on the central proportion of the liver.

Fig. 2. Liver biopsy: congested and haemorrhagic liver parenchyma with thrombotic hepatic veins. HE, × 10.
Fig. 3. Needle liver biopsy. Moderate portal fibrosis, some dilated central veins. HE, ×40.

Fig. 4. Hepatic vein filled with thrombus. HE, ×40.
Fig. 5. Needle liver biopsy. Moderate portal fibrosis, liver plates nearly normal. HE, ×40.

Fig. 6. Needle liver biopsy. Dilated central hepatic veins without signs of congestion. Some sinusoids show moderate dilatation. HE, ×40.
enlargement of the liver, especially of the left lobe, with a characteristic excess uptake of the isotope at the centre of the liver, without filling defects (Westcott, 1973). The spleen was moderately enlarged. Because abdominal distension had caused severe discomfort to the patient, paracentesis was done four times under simultaneous plasma administration. Diuretics had been tried initially with very poor results. However the ascitic fluid was quickly reproduced. The third day after her admission, the patient had a haematemesis and melaena. The haematocrit had fallen to 30%. The stools were normal 2 days later. During this time the patient had a blood transfusion. The presence of tender hepatomegaly, without significant deterioration of liver function, the early development of portal hypertension in a person who had no history or clinical signs of chronic liver disease, the rapid reproduction and the high protein content of the ascitic fluid, in connection with the finding of the liver scan led the authors from the start to a diagnosis of hepatic vein block. A liver biopsy (Fig. 2) and an inferior venacavography were performed in order to investigate the case further and to confirm the clinical diagnosis. The liver biopsy taken from a central area showed nothing characteristic apart from slight dilatation of sinusoids and a moderate fibrosis at the portal zones (Fig. 3). The inferior venacavography (Figs 2 and 3) showed a typical smooth narrowing of the hepatic segment of the hepatic vein (Westcott, 1973), this was probably due to the pressure of the hypertrophic caudate lobe. The main hepatic veins could not be visualized following Valsalva manoeuvre. An hepatic venography was arranged for the following day. Unfortunately, the patient had a severe haematemesis which was uncontrollable despite blood transfusion and the insertion of the Blakemore-Sengstaken tube and she was taken to the operating theatre as an emergency.

Operative findings
During operation, the liver was found to be enlarged and congested and the spleen twice the normal size. A gross collateral circulation, including oesophageal varices, was present and the portal vein pressure was found to be 54 cm of water. Because of severe and continuous blood loss from the oesophageal varices, a Tanner’s operation was performed. The patient died the next day. Post-mortem examination was refused.

A surgical liver biopsy showed an intense congestion with haemorragic infiltration and thrombosis of the hepatic vein radicles (Figs 4, 5 and 6).

Discussion
The clinical diagnosis of Budd–Chiari syndrome followed by confirmation of inferior venacavography and liver biopsy is well documented. The histological appearance of the liver at biopsy was different from that at surgery because the material taken at biopsy was from an area of minimal damage (caudate lobe) and not because of alteration of histology due to the progression of the disease. Unfortunately, the angiographic investigation was restricted to the inferior venacavography but, according to Clain et al. (1967), Deutsch et al. (1972), and Westcott (1973), this technique alone is usually sufficient to give the correct diagnosis. For this reason and also for the fact that it is a very simple technique, inferior venacavography has been widely used in the investigation of occlusion of the inferior vena cava and the hepatic veins. The smooth stenosis of the venous lumen in the present case and the failure of visualization of the hepatic veins, suggest occlusion of the veins by a thrombus rather than by a neoplasm. The liver scan appearance is also suggestive of the syndrome. The central uptake of the liver scan is due to hypertrophy of the caudate lobe, which is not unusual in the Budd–Chiari syndrome. This can be explained by the unique vascular supply and drainage of the caudate lobe. The caudate lobe receives blood from both right and left hepatic arteries and from two or three separate portal veins. Of greater importance in the Budd–Chiari syndrome is the caudate lobe venous drainage which is by two, three or, rarely four veins into the intrahepatic inferior vena cava (Westcott, 1973; Hales and Scatliff, 1966). This should make the caudate lobe much less affected than the right and left lobes, and probably accounts for the marked regenerative hypertrophy of the caudate lobe in the Budd–Chiari syndrome.

The occlusion of the hepatic veins probably began during the sixth month of pregnancy, making this case very significant, because no trace could be found of a similar case in the literature. There have been reported only nine cases which were observed close to time of delivery (Kahn and Spring, 1940; Deutsch et al., 1972; Chiari, 1899; Lange, 1886; Thran, 1899; Mann and Hall, 1904; Krass, 1957; Hannock 1968).

The aetiology of the Budd–Chiari syndrome remains unknown in 67% of cases. The increasing frequency of the syndrome in women taking oral contraceptives, and the increased incidence in pregnant women of venous thrombosis due to alteration of coagulation factors (factors VII, VIII, X and fibrinogen) (MRC Working Party Report, 1967) lead one to entertain the possibility of a connection between pregnancy and the Budd–Chiari syndrome in the present case.

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