Postpartum Veno-occlusive Disease Treated with Ascitic Fluid Reinfusion

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SUMMARY

Three patients presenting in the postpartum period with veno-occlusive disease not related to the ingestion of pyrrolizidine alkaloids are described. The patients were treated by intravenous reinfusion of the ascitic fluid. This form of therapy has, to our knowledge, not previously been reported in the management of this condition.


It is well known that veno-occlusive disease, or occlusion of the smallest tributaries of the hepatic venous system, occurs after ingestion of pyrrolizidine alkaloids found in plants of the genera Senecio, Crotalaria and Heliotropium. Previous outbreaks of veno-occlusive disease have been reported in South Africa after the consumption of bread made from contaminated wheat. Sporadic cases have also occurred in South Africa and Jamaica, mainly in children. In this article we describe 3 cases seen in the postpartum period. The diagnosis was confirmed on hepatic vein catheterization. The management included diuretic therapy and intravenous reinfusion of the ascitic fluid using the method described by Giraud.

CASE REPORTS

Case 1

A 22-year-old Black woman presented 1 month after delivery of her first child at an outlying clinic. The birth was normal and no complications were noted. One week after delivery she noted progressive swelling of her abdomen and feet. There was no history of previous liver disease, alcohol abuse or herbal ingestion. On examination she was thin, with marked ascites, and a 3-cm enlarged, firm, smooth liver was palpable. The rest of the examination was normal apart from mild pedal oedema. Laboratory investigations revealed a haemoglobin level of 9.9 g/100 ml, a white cell count of 7 500/μl, a blood urea level of 85 mg/100 ml and a prothrombin index of 62%. The serum albumin level was 2.9 g/100 ml, with globulins 3.4 g/100 ml, and bilirubin, transaminase and alkaline phosphatase levels were within normal limits. The ascitic fluid had a protein content of 1.6 g/100 ml. The chest radiograph was normal, but a barium swallow demonstrated oesophageal varices. Liver biopsy showed marked centrilobular congestion and necrosis. Catheterization of the hepatic vein showed patent main hepatic veins.

Treatment was started with diuretics, but the abdomen remained tense and required paracentesis on several occasions. When the peritoneal fluid was drained, it was collected in a sterile bag and reinfused intravenously. Two weeks after admission the patient became jaundiced and confused. Her condition deteriorated rapidly despite therapy for liver failure. Treatment with streptokinase was started because extension of the centrilobular venous thrombosis was feared.

The patient died suddenly after a massive haematemesis. A postmortem examination confirmed terminal bleeding from oesophageal varices. The liver was moderately enlarged, smooth and of ‘nutmeg’ appearance. The heart was normal and the inferior vena cava and main hepatic veins were patent. Histological examination of the liver showed massive centrilobular necrosis and congestion, with recent thrombus in some of the centrilobular veins.

Case 2

A 23-year-old Black woman was admitted with ascites which had started 1 week after the birth of her first child. Her baby was born in hospital and, apart from mild hypertension and a period of confusion after delivery, no complications were noted.

There was no previous history of liver disease. The patient had eaten ‘wild spinach’ early in her pregnancy, but had not taken herbal remedies. There was no family history of a similar illness. She was well nourished, but had gross ascites, a moderately enlarged, tender liver and mild pedal oedema. Her blood pressure was normal and the rest of the examination was non-contributory.

The haemoglobin level was 12.1 g/100 ml, white cell count 16 300/μl, platelet count 260 000/μl and erythrocyte sedimentation rate 20 mm/1st h. The blood urea level was initially 98 mg/100 ml but later returned to normal. The serum albumin level was 2.1 g/100 ml, globulin level 3.0 g/100 ml, and the prothrombin index 70%. The bilirubin was 1.2 mg/100 ml, the transaminase levels were slightly elevated and the alkaline phosphatase was normal. The chest radiograph was normal. No varices were seen on a barium swallow. On catheterization the hepatic veins were patent, the free hepatic venous pressure was 25 cm H₂O, the mean hepatic wedge pressure was 53 cm H₂O and a hepatogram showed a ‘mottled’ sinusoidal picture (Fig. 1).
The ascites was severe and re-accumulated rapidly after paracentesis, despite diuretic therapy. The fluid was tapped and reinfused intravenously on three occasions at weekly intervals. Total volumes of 10 litres, 7 litres and 5 litres were reinfused. The patient tolerated the reinfusion well and responded with a satisfactory diuresis. She was discharged with mild ascites and was clinically normal when seen 1 year later when all therapy had been stopped.

Case 3

A 21-year-old Black woman presented with a 3-week history of abdominal swelling and pain which had started 2 weeks after the birth of her first child in hospital. No complications had been noted and there was no history of herbal ingestion after the delivery. The main findings on physical examination were massive ascites with a 3-cm enlargement of the liver and minimal pedal oedema. She was well nourished and the rest of the examination was normal.

Laboratory investigations revealed a haemoglobin level of 11,6 g/100 ml, a white cell count of 7 000/μl, a platelet count of 129 000/μl, and an erythrocyte sedimentation rate of 22 mm/1st h. The serum albumin level was 2,8 g/100 ml, globulins 4,6 g/100 ml, and the prothrombin index was 65%. The blood urea, bilirubin, transaminase and alkaline phosphatase levels were within normal limits. The ascitic fluid had a protein content of 1,4 g/100 ml. A chest radiograph, barium swallow and liver scan were normal. Liver biopsy showed centrilobular congestion, haemorrhage and necrosis. Hepatic vein catheterization showed a free hepatic venous pressure of 14 cm H₂O and a wedged hepatic venous pressure of 50 cm saline; injection of contrast medium showed a sinusoidal picture.

Treatment was started with diuretics but little improvement was achieved. The abdominal distension necessitated paracentesis on several occasions, and the fluid was reinfused intravenously. The ascites gradually improved and had disappeared completely at follow-up.

DISCUSSION

The sudden onset of ascites with a tender, smooth, enlarged liver suggests obstruction of the hepatic venous outflow. In the absence of cardiac failure, constrictive pericarditis, inferior vena-caval thrombosis or malignant infiltration, the obstruction may be in the larger hepatic veins (Budd-Chiari syndrome) or the smaller venous radicles (veno-occlusive disease). The clinical presentation is similar and liver biopsy shows an identical picture in both conditions. The Budd-Chiari syndrome is suggested if isotopes are concentrated in the caudate lobe on a liver scan, but the scan is often difficult to interpret in the presence of gross ascites. Hepatic venography excludes obstruction of the large hepatic veins and shows a typical sinusoidal picture in the liver in veno-occlusive disease.

The distinction between veno-occlusive disease and hepatic vein thrombosis has relevance to the treatment. If a large vein is thrombosed then surgical removal may be attempted or fibrinolytic agents administered. Anticoagulants may prevent further thrombosis. There is experimental evidence that in veno-occlusive disease the venous thrombosis occurs secondary to the centrilobular necrosis. Anticoagulants are thus unlikely to alter the prognosis and may well be contraindicated, since these patients may have oesophageal varices or peptic ulceration.

The management of veno-occlusive disease is aimed at relieving the ascites and preventing liver failure. The ascites is usually resistant to salt restriction and diuretic therapy. Large doses of potent diuretics can precipitate liver failure from electrolyte disturbances and dehydration. Repeated paracentesis is usually necessary to relieve the gross abdominal distension as the fluid re-accumulates rapidly. Rapid removal of large volumes of ascitic fluid may also precipitate liver failure and repeated paracentesis will eventually cause severe protein and electrolyte depletion. Patients often become severely debilitated and succumb to intercurrent infections.

Reinfusion of ascitic fluid has been recommended in patients with intractable ascites due to cirrhosis of the liver. The use of this method of treatment in patients with veno-occlusive disease has, to our knowledge, not been reported previously. Provided renal function is adequate, reinfusion of ascitic fluid intravenously will relieve the ascites without loss of protein or gross electrolyte changes. If a closed system is used the risk of infection is small. Diuresis occurs without the use of diuretics, although these agents can be given concomitantly. Our patients were reinfused when the abdomen became tense. At the onset this reinfusion was required weekly, and then less frequently as the condition improved. No complications arose from the procedure and we feel this is a very useful form of therapy.

The occurrence of veno-occlusive disease in the post-partum period has, to our knowledge, not been reported previously. The majority of reported cases are in children. A history of herb ingestion, or of drinking medicinal 'bush tea' is often elicited. Most cases are sporadic, and accidental poisoning may occur, since the Senecio and Crotalaria species grow wild over large areas of South
Africa. More frequent occurrences of veno-occlusive disease involve populations who have eaten contaminated wheat or cereals. In 1920 Willmot and Robertson described an outbreak of the disease in families living in the George and Mossel Bay districts. A similar outbreak was reported by Selzer and Parker in 1951. More recently, epidemics have been reported from India and Afghanistan.

The condition has rarely been seen among adults in Johannesburg. The striking similarity of the 3 cases reported in this article suggests that a common cause was operative. Despite careful questioning we could elicit no history of herb ingestion in the intrapartum period. All 3 women were primiparous and none of the infants showed evidence of liver damage. Occlusion of the larger hepatic veins has been described in patients on oral contraceptives. Venous thrombosis of the legs and pelvic veins is not infrequent in the postpartum period. In our patients the larger hepatic veins were patent and it seems unlikely that venous thrombosis was the primary lesion. Protein deficiency has been shown experimentally to enhance the effects of the pyrrolizidine alkaloids, and many of the reported patients were malnourished. Our patients were all well nourished, but it is possible that the liver is more susceptible to injury by alkaloids in the postpartum period.

Complete recovery may be expected in half of the patients presenting with acute ascites. Stuart and Bras found a higher mortality in adults than in children. Jaundice is a poor prognostic sign and heralds death from acute liver failure. Persistent hepatomegaly and ascites suggest chronic liver damage and the development of cirrhosis and portal hypertension.

REFERENCES


Herspes Zoster of the Chest Wall and Gynaecomastia

A Case Report

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SUMMARY

Herpes zoster in a young males at puberty, associated with aggravation of gynaecomastia on the same side as the intercostal nerve involvement, is described.


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Gynaecomastia can occur under a wide variety of circumstances, including puberty, refeeding, endocrine and drug therapy. The exact mechanisms of production and the pathophysiology are still unknown. A disturbance of oestrogen metabolism appears to be the most favoured hypothesis. Injury to the intercostal nerves during thoracic surgery followed by gynaecomastia has also been described. However, herpes zoster affecting the intercostal nerves and aggravating or causing gynaecomastia must be extremely rare. The present article describes such a case.

CASE REPORT

A 19-year-old White male student presented to the endocrine service at Groote Schuur Hospital with the com-