Obstructive jaundice secondary to Morgagni's diaphragmatic hernia

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ABSTRACT
Obstructive jaundice is an uncommon cause of jaundice in children past the neonatal period. Operation on these patients carries some risks. We present a patient with Down's syndrome who developed jaundice, which resulted from recurrent Morgagni's diaphragmatic hernia causing displacement of the second part of the duodenum and obstruction of the common bile duct. We discuss the management and risks involved in these patients. We believe that obstructive jaundice due to Morgagni's diaphragmatic hernia has not been reported before.

Keywords: Obstructive jaundice, Morgagni's diaphragmatic hernia.

A 12-year old Down's syndrome girl presented with severe obstructive jaundice of more than 10 months' duration. The family resorted to traditional medicine before coming to the hospital. She had diaphragmatic hernia, Morgagni's type, operated upon 3 years prior to her admission. On examination, she was deeply jaundiced, skin and sclera, with scratch marks over her skin. The liver could be felt below the umbilicus with its inferior rounded edge along the mid line. The spleen was felt 2 cm below the costal margin. Chest x-ray showed bowel shadows in the chest on the right side raising the suspicion of recurrence of her diaphragmatic hernia. Laboratory investigations on admission showed total bilirubin to be 350 µmol/l, direct bilirubin 235 µmol/l, SGOT 224/100 ml, SGPT 240 IU/100 ml, prothrombin time 20 seconds (control 14) and partial thromboplastin time 55 seconds, (control 43 seconds).

Barium meal follow through study showed the bowel, starting from the end of the first part of the duodenum to be displaced in the chest cavity, confirming the diagnosis of recurrent diaphragmatic hernia (Fig. 1). Endoscopic retrograde cholangiopancreatography (ERCP) was carried out but it failed to cannulate the duodenal papilla. In view of the very high bilirubin, a percutaneous transhepatic cholangiography (PTC) was carried out to try to help in the diagnosis and to drain the biliary system in order to bring down the level of bilirubin before surgery. Percutaneous transhepatic cholangiography showed dilation of the biliary passages, kinking of the common bile duct and failure of passage of dye to the duodenum (Fig. 2). Percutaneous transhepatic cholangiography brought down the level of total bilirubin to 200 µmol/l and direct bilirubin to 150 µmol/l. Vitamin K daily injections failed to bring the prothrombin time down so a course of cryoprecipitate together with vitamin K was given. It was decided to operate on the patient when the PT was 16 seconds (control 14 seconds). The patient was well hydrated and 10 mg of frusemide was given just on anesthesia induction. The urine output was maintained during the operation, using fluid boluses added to her normal maintenance.

On exploration, there was no ascites. The liver was swollen, green with rounded edge and surface with fine nodularity. It was rotated to the right side with right lobe down and left lobe up. Part of the left lobe was prolapsed through the hernial defect into the chest. The spleen was enlarged. The duodenum, starting from the end of the first part, together with jejunum and parts of the ileum were prolapsed in the chest, on the right side. The cecum was in the normal place in the right iliac fossa. After repair of the hernia operative cholangiogram was carried out using the PTC catheter and this showed free drainage of the dye to the duodenum (Fig. 3). Liver biopsy was taken which showed bile duct proliferation and fibrosis in the portal tracts. It showed too some disturbance of hepatic architecture with bile pigment accumulation in the centrilobular areas.

The patient had a stormy post operative course. The urine output was 0.5 ml/kg/hour, on the first and second postoperative days. The patient was hydrated well. Her maintenance fluid was calculated according to her weight and height. This amount was added as an extra fluid given from the Department of Pediatric Surgery, King Fahad Hospital, Al-Baha, Saudi Arabia.

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intravenously per day. Frusemide 1 mg/kg was given i.v. twice on the first day post operatively. Mannitol 0.2 gm/kg was given on the second day to try to stimulate diuresis. The urine output started to improve from the third day onward. The patient developed collapse of the left lung which was resolved by intensive physiotherapy. A sympathetic right sided pleural effusion developed which took some days to resolve. Eventually the patient made a full recovery, with serum bilirubin, liver enzymes and the coagulation profile improving gradually.

The patient was discharged home 20 days postoperatively. Investigations on discharge were: total bilirubin 32 μmol/l, direct bilirubin 17 μmol/l, prothrombin time 14 seconds (control 13), partial thromboplastin time 36 seconds (control 34). The patient was lost to follow-up after the first post operative visit.

Discussion Morgagni’s diaphragmatic hernia is a very unusual cause for obstructive jaundice. Morgagni’s hernia is usually located anteromedially, on either side of the junction of the embryologic septum transversum and the thoracic wall. The hernia is usually discovered incidentally as a mass or air fluid level on chest x-rays obtained for other reasons. The diagnosis is confirmed by obtaining a lateral chest x-ray and/or by carrying out a barium follow through study. Jaundice, of the obstructive variety, is an
uncommon cause of jaundice in the pediatric age group, past the neonatal period. The most common causes of obstruction in these children are choledochal cysts. Obstructive jaundice due to Morgagni's diaphragmatic hernia has not been reported before.

A wide variety of other pathological processes can affect the wall of the extrahepatic bile ducts and result in obstruction. Ladd described 5 patients with jaundice, who, at laparotomy, were found to have mechanical obstruction caused by plugs of thickened bile. He suggested that the condition was secondary to distal common bile duct stenosis.1 Trauma, inflammatory processes or compression from outside, whether by a tumor, bands or pull on the biliary passages are other causes of common bile duct obstruction. Obstruction of the common bile duct has been reported in association with duodenal malformations,2 pancreatic hemangioendothelioma and hydronephrosis.3 Spitz et al, 1983, described obstructive jaundice due to chronic malrotation of the mid gut. The condition was relieved by a standard Ladd's procedure.4 In our case the cause of biliary obstruction was due, merely, to the pull and pinching of the junction of the common bile duct with the duodenum, resulting from the displacement of the duodenum into the chest. The displacement of the retroperitoneal fixed part of the duodenum, as well as the rotation of the liver, in our patient in this way could be explained by increased connective tissue elasticity, familiar to increased connective tissue elasticity, familiar to Down's syndrome patients. The preparation of the patient with obstructive jaundice for any surgical procedure is critical in order to avoid complications that include renal failure, which may occur in 5-10% of patients and sepsis.5 Several risk factors have been identified in patients with obstructive jaundice. Serum bilirubin >170 μmol/l, hematocrit 30 g/dl, alkaline phosphatase >100 units/l and creatinine >115 μmol/l are some of these factors.7 Coagulation defect is usually corrected with parenteral vitamin K, although to start with in our patient it was difficult to correct. Dehydration and hypotension, which could lead to acute tubular necrosis, are prevented by intravenous hydration, usually with 0.9% NaCl and close monitoring of fluid balance. Mannitol is given to protect renal function but patients must be well hydrated before its use, although a recent trial has questioned its benefit.6 Obstructive jaundice brings about some changes in the liver, the degree of which depends on the duration and the severity of obstruction. Fibrous tissue bands in the portal zones coalesce and the lobules are correspondingly reduced in size. Nodular regeneration of liver cells follows but a true cirrhosis rarely follows biliary obstruction.

Our patient had obstructive jaundice for over 10 months. This long period of obstruction produced a degree of liver damage. Following relief of obstruction, serum bilirubin goes back to normal slowly, as shown in our patient. This is partly due to reversal of the changes which happened in the liver, on the one hand and to slow dissociation of the bili-albumin which are covalently bound on the other hand. This improvement affects the coagulation profile as well, following relief of obstruction. The value of preoperative biliary drainage in patients whose bilirubin is very high, is controversial. Randomized controlled studies have not shown benefit.3

We believe that obstructive jaundice due to Morgagni's diaphragmatic hernia has not been reported before. There is an increased risk of operating on patients with obstructive jaundice. Urgent diagnosis and management of these cases is needed in order to avoid liver damage.

References