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Internal hernia and volvulus of the small bowel following liver transplantation

Received: 2 May 1996
Received after revision: 7 October 1996
Accepted: 28 October 1996

This paper was presented as a poster during the Joint Congress on Liver Transplantation in London in September 1995

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Abstract Internal herniation with volvulus of the small intestine is an uncommon, but potentially fatal, complication after liver transplantation. We present here four cases in which the herniation occurred around the Roux-en-Y loop used for the biliary reconstruction. One patient died due to intestinal and liver allograft necrosis; another lost almost the entire small intestine and has since undergone successful intestinal transplantation. Two patients survived following surgery that involved reduction of the hernia and closure of the mesenteric defect. Clinical diagnostic implications emphasize early diagnosis and appropriate operative intervention.

Key words Liver transplantation, internal hernia · Internal hernia, liver transplantation

Introduction

Liver transplantation has become accepted therapy for the treatment of patients with end-stage liver disease. Complications after liver transplantation may result from technical issues or problems related to immunosuppression, such as infection or rejection. These problems may manifest themselves in the gastrointestinal tract with signs and symptoms indistinguishable from other complications such as leakage from enteric and biliary anastomosis, intestinal obstruction and, rarely, internal herniation with volvulus of the small bowel. This occurs through the mesenteric defect created with

the formation of the Roux-en-Y loop performed for the choledochojejunostomy. The defect can occur in the small bowel mesentery or transverse mesocolon, if the loop is to be made retrocolic. Current standard surgical practice involves closing these defects to avoid internal herniation. We report four patients who presented with internal herniation and volvulus following liver transplantation.

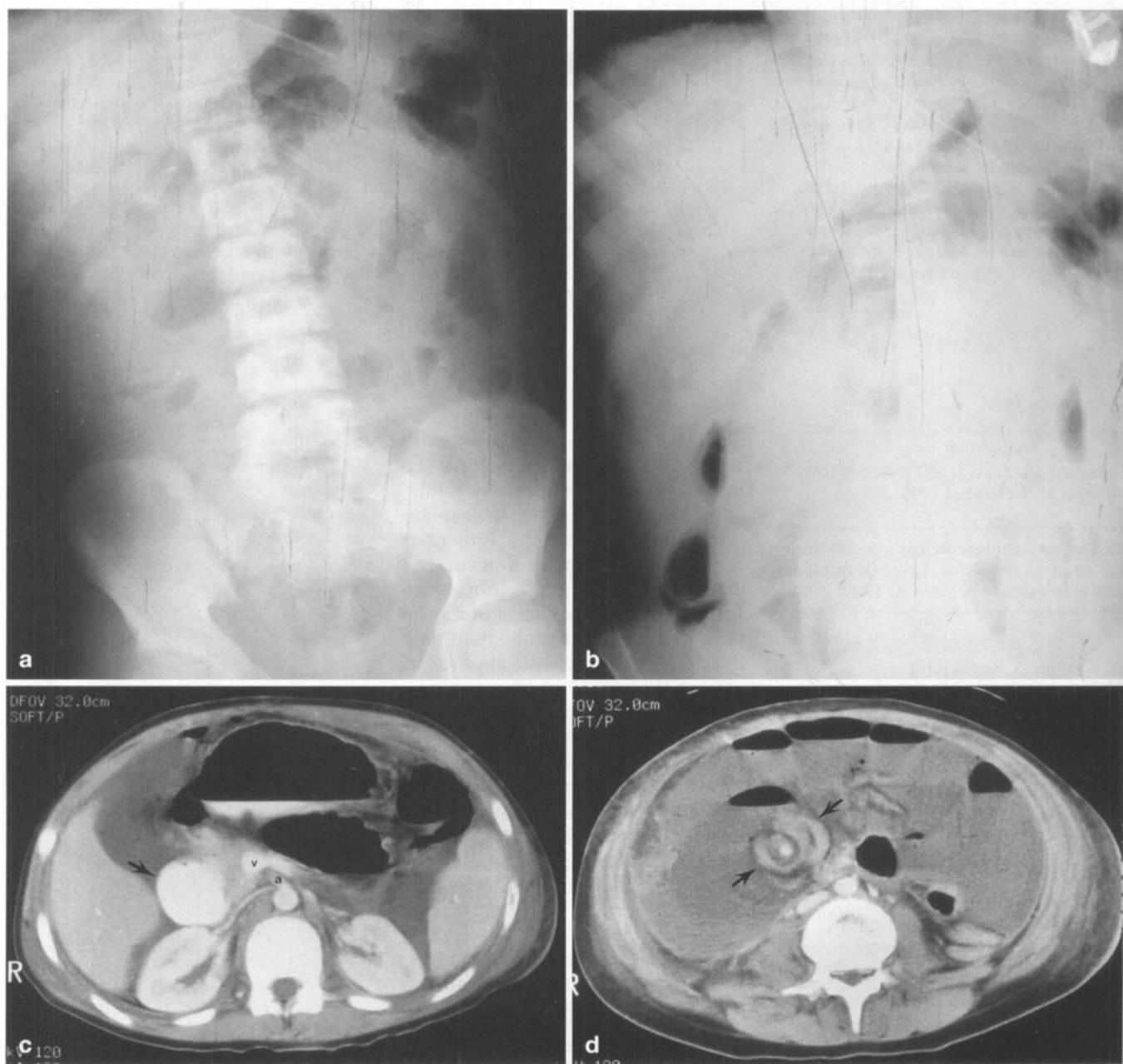


Fig. 1 **a** Film of abdomen taken in supine position. **b** Left lateral decubitus abdominal radiographs are nonspecific with diffuse, moderately dilated bowel loops and scattered air-fluid levels. **c** CT scan of the abdomen reveals moderate abdominal ascites. Note the dilated proximal duodenum (*arrow*) but the normal anatomic position of the superior mesenteric artery (*a*), to the left and posterior in relation to the superior mesenteric vein (*V*). **d** CT slice several centimeters inferior to **c**. Note the diffusely distributed, markedly dilated, fluid-filled small bowel loops with air-fluid levels and poor wall enhancement. There is a whorl appearance in the right posterior abdomen (*arrows*) consisting of twisted loops of bowel and mesentery around the superior mesenteric artery. This appearance is diagnostic of small bowel volvulus

Case reports

Patient 1

A 12-year-old girl presented to her local emergency room with a 6-day history of abdominal pain and vomiting. She had undergone liver transplantation for biliary atresia 10 years earlier and was under cyclosporin (CyA) immunosuppression. For several years she had experienced intermittent, self-limited episodes of abdominal pain that were not severe.

On admission, the patient was lethargic with features of hypovolemic shock requiring resuscitation. She had diffuse abdominal guarding and rigidity with absent bowel sounds. Her white blood

cell (WBC) count was elevated at 13.4 with 64% bands. Upper gastrointestinal endoscopy was obtained because of hematemesis and revealed esophagitis and stress fundic ulceration. As her general condition and abdominal signs continued to deteriorate, she was transferred to this hospital 24 h later. Plain films of the abdomen on arrival revealed mildly dilated bowel loops with non-specific, scattered, air-fluid levels (Fig. 1a, b). Abdominal ultrasound demonstrated moderate ascites, a normal liver allograft with patent vessels, and many dilated, fluid-filled, aperistaltic bowel loops.

Abdominal CT images revealed ascites and diffuse, markedly dilated, fluid-filled small bowel loops with poor enhancement of the bowel wall. A "whorl" appearance, extending around the axis of the superior mesenteric artery, was evident in the right mid-abdomen, inferior to a dilated, proximal duodenum (Fig. 1c, d). These findings suggested small bowel volvulus with probable gangrenous small bowel. The superior mesenteric artery and vein were normally located anatomically with no evidence of bowel malrotation, indicating an unusual etiology for volvulus (Fig. 1c). The entire colon appeared in the appropriate anatomic location, with normal wall enhancement.

At operation, the patient was found to have internal herniation of the entire small bowel through the rent in the mesentery around the Roux-en-Y limb of the choledochojejunostomy, with consequent volvulus and gangrene. Bowel resection was performed, leaving only 20 cm of healthy jejunum and 5 cm of terminal ileum. Tube choledochostomy with external drainage was performed due to necrosis of the Roux-en-Y limb, which was draining the common bile duct of the liver allograft. Forty-eight hours later, the patient was re-explored and a choledochoduodenostomy was performed. She recovered and remained on oral FK 506 immunosuppressive therapy and total parenteral nutrition (TPN) for 8 months, after which she underwent successful small bowel transplantation. She is presently well and off TPN.

Patient 2

A 14-year-old girl was admitted with symptoms of episodic nausea and vomiting. She had undergone orthotopic liver transplantation under tacrolimus (FK 506) immunosuppression for Alagille's syndrome 2 years previously. These symptoms precipitated repeated hospitalizations for work-up, which included barium upper gastrointestinal and small bowel series, a barium enema and contrasted abdominal CT scan, endoscopy, a percutaneous transhepatic cholangiogram, HIDA scan, and Doppler ultrasound scan of the abdomen, all of which were normal. She intermittently received medical therapy with reglan and cisapride; however, in view of the persisting clinical signs and symptoms, she underwent an exploratory laparotomy.

At operation the patient was found to have internal herniation and associated volvulus of the small bowel through the defect in the small bowel mesentery created by the Roux-en-Y limb of the choledochojejunostomy. The hernia and resulting volvulus were reduced, and the bowel was found to be viable. The mesenteric defect was closed with interrupted silk. Postoperatively, the patient made a good recovery and has remained asymptomatic.

Patient 3

A 12-year-old girl underwent orthotopic liver transplantation for alpha-1 antitrypsin deficiency under CyA immunosuppression. On the 13th postoperative day, she developed intermittent, central abdominal pain. Examination showed the abdomen to be dis-

tended and tender. An abdominal radiograph revealed diffusely dilated bowel loops. She had an elevated WBC count at 12000 cells/cm; electrolytes were normal.

Within a few hours the patient became hypotensive and passed blood per rectum. At laparotomy, she was noted to have herniation and volvulus of jejunum through a window between the Roux-en-Y loop and colon with borderline ischemia of approximately 60% of the small bowel. The hernia was reduced and the defect in the mesocolon was closed using dexon. The color of the bowel improved after 30 min; therefore, the abdomen was closed and re-exploration was scheduled for the next day. She continued to be hypotensive and was on dobutamine and epinephrine support.

Laparotomy the following day revealed ischemia of the liver, Roux-en-Y loop, and small bowel. Following bowel resection with end-to-end anastomosis, she continued to deteriorate and expired the same day.

Patient 4

A 38-year-old woman underwent orthotopic liver transplantation for cryptogenic cirrhosis in 1992 under tacrolimus immunosuppression. Nineteen months later she was admitted to a local hospital with complaints of abdominal pain and distension. Abdominal films showed features of small bowel obstruction. After initial resuscitation, she underwent exploratory laparotomy. At operation, significant small bowel distension was noted, with an obvious internal hernia of the small bowel through the mesenteric defect in the Roux-en-Y loop. The hernia was reduced and the mesenteric defect closed. She was discharged from the hospital ten days later.

Discussion

The clinical spectrum of abdominal pain following liver transplantation is both wide and diverse. Mechanical obstruction, adhesive or otherwise, as in these cases, can occur at any time. Infections of the gastrointestinal tract to which these immunocompromised patients are prone may present with similar complaints. Post-transplant lymphoproliferative disease can also present with intestinal obstruction or perforation. Gastrointestinal bleeding and abdominal tenderness may accompany these infections. Consequently, the approach to these patients, both regarding the symptom complex and the intensity of the work-up, must be individualized.

Internal herniation following liver transplantation is uncommon; a review of the literature produced one case report of a similar complication in a postpartum woman [3]. In a previously published series from this institution, this complication was noted only once among the 397 liver transplant recipients who were studied (patient 3 of this series [2]). Both the small bowel mesentery and mesocolon are potential sites for herniation. Although these defects are surgically closed, incomplete closure or breakdown of the surgical sutures may occur, leading to a potential site for internal herniation. The initial herniation probably involves a small segment of the intestine and is transient, causing intermittent abdominal discomfort or pain. This may lead to more and

more bowel entrapment with each episode, eventually resulting in acute abdomen, when the obstruction is complete, leading to ischemia.

The diagnosis is essentially based on a high index of suspicion. The presence of symptoms of acute intestinal obstruction in the absence of deterioration in liver function should raise suspicion for this diagnosis. Premonitory symptoms may include pain and vomiting, which probably represent subacute obstruction that may be episodic and suggest chronicity (patients 1 and 2). Patients may be on steroids for immunosuppression, which can mask abdominal signs. A "look and see", rather than wait and see, policy should be followed and early laparotomy performed to avoid serious complications.

Plain films of the abdomen were nonspecific (Fig. 1 a, b) and unhelpful diagnostically in our cases. CT scanning was the most useful imaging study for evaluating the abdomen, both to rule out other pathology and to suggest a diagnosis. In one child (patient 1), diagnosis of small bowel volvulus with ischemic bowel was suggested on CT (Fig. 1c, d). In the absence of volvulus, both CT and other imaging studies may not diagnose the internal hernia (as in patient 2). Hence, negative results on radiological examination should not influence the decision to operate if clinical suspicion exists.

Other reports have indicated the reliability of CT in diagnosing small bowel volvulus [1, 4]. The "hurricane" or "whorl" sign, representing bowel and mesentery twisting around the superior mesenteric artery, appears to be a specific imaging sign for small bowel volvulus, although this has not, to our knowledge, been reported previously in a patient with internal bowel herniation with volvulus.

Although the small bowel volvulus was diagnosed on CT, the internal hernia itself was not specifically suspected on the CT exam. Bowel loops were not focally dilated or confined to a localized area of the abdomen. However, the normal relationship of the superior mesenteric artery and vein, as well as the relatively inferior

and right-sided location of the volvulus and the normal location and appearance of the colon, suggested an unusual underlying cause for small bowel volvulus.

When the possibility of internal herniation and volvulus is entertained, prompt evaluation and exploration are particularly important in these immunocompromised patients in whom a minor septic focus can result in life-threatening infection. We feel that resection of obviously necrotic bowel and re-exploration 24 h later to assess portions of bowel that were of doubtful viability at the previous operation is a reasonable approach. The extent of resection may be more generous and immediate in the face of hemodynamic instability or liver allograft dysfunction.

The reconstruction and restoration of bowel continuity depends upon the condition of the patient, the extent of small bowel resected, the viability of remaining bowel, and whether the Roux-en-Y loop was involved in the ischemic process. In patients 2 and 4, the hernia was reduced with complete recovery. Patient 1, however, required resection of most of the jejunum and ileum, including the Roux loop. After resection of the Roux loop, external biliary drainage was instituted. Construction of the choledochoduodenostomy as a subsequent procedure was effective in allowing internal drainage of bile while awaiting small bowel transplantation. Patient 3 became unstable because of both small bowel and liver ischemia and did not survive subsequent resection of ischemic bowel. The necrotic and ischemic bowel likely produced significant damage to the liver allograft with rapid deterioration and death.

Intestinal transplantation has now emerged as a viable therapeutic modality for a selected group of patients with short gut syndrome. The results of this procedure are encouraging [5, 6] and justify aggressive surgical management in these very complex patients who may have lost a significant portion of their small bowel due to this disastrous complication.

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