Thoracoscopic repair of right-sided diaphragmatic hernia after liver transplantation for hepatoblastoma

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Abstract
Diaphragmatic hernia has been increasingly reported as a rare complication of liver transplantation. Here, we report a case of diagnosis and successful treatment of right-sided diaphragmatic herniation following transplantation of a left-lateral graft in a 14-month old patient treated for hepatoblastoma with neoadjuvant chemotherapy. We suspect a combination of factors contributed to this case of diaphragmatic herniation after liver transplantation, including the patient’s suboptimal nutritional status, prior chemotherapy, medial positioning of the graft, and intra-operative photocoagulation. This complication was repaired definitively and with low morbidity utilizing a minimally invasive approach.

Children with hepatoblastoma who are to undergo liver transplantation receive neoadjuvant chemotherapy. Preoperative optimization is difficult in these patients given the usual last-minute notice that a donor graft is available. Here, we report an association of neoadjuvant chemotherapy with diaphragmatic herniation following liver transplantation in an infant and the successful repair utilizing a minimally invasive technique.

1. Case report

A 14-month old boy with unresectable hepatoblastoma underwent orthotopic liver transplantation utilizing an in-situ split left lateral graft from an unrelated deceased donor. Pre-operative imaging revealed the mass to measure $7.2 \times 5.5 \times 7.4$ cm and be centered within the quadrate lobe abutting the bifurcation of the main portal vein. Prior to transplantation, the patient had undergone two rounds of chemotherapy consisting of cisplatin, 5-fluorouracil, vincristine, and doxorubicin, with his last round of chemotherapy ending five days prior to the date of transplant. In the immediate pre-transplantation period, nutritional optimization was attempted with total parenteral nutrition (TPN) in addition to nasogastric enteral feeds and ad libitum oral intake. At the time of transplant, the patient’s length and weight were 75 cm and 9.7 kg (12.7 and 12.5 percentile for age, respectively).

The transplant was completed in 3 h and 36 min with an estimated blood loss of 25 cc and 800 cc of crystalloid and 300 cc of packed red blood cells infused. The explant pathology revealed a 99% viable tumor that did not penetrate the capsular margin. The patient was extubated within hours after surgery and was saturating 100% on room air by post-operative day 3. On post-operative day 7, efforts to transition to PO intake were met with some coughing and choking. Over the next week, the patient experienced daily diarrhea and intermittent PO intolerance with low volume emesis. Stool cultures were found to be positive for adenovirus and a chest x-ray performed on post-operative day 16 revealed a right-sided infiltrate. Sputum cultures were obtained and empiric antibiotics were begun.
for the presumed pneumonia. Increased work of breathing was noted on post-operative day 19, although oxygen saturations remained 98% on room air. That same day, fluoroscopy for replacement of a feeding tube, a standard practice at our hospital, incidentally revealed right-sided diaphragmatic hernia with multiple loops of intestine within the chest. This was subsequently confirmed by chest x-ray (Fig. 1) and CT scanning. The liver graft remained well-positioned in the abdomen. The patient continued to have adequate oxygenation and ventilation on room air and intermittently tolerated enteral feeds with occasional low volume emesis. Thirty-two days after transplantation, after stool adenovirus cultures were confirmed negative, the patient was taken for thoracoscopic repair of the right-sided diaphragmatic hernia.

During the repair, viable colon and small bowel was noted to slide easily though a smooth rimmed $4 \times 6$ cm postero-lateral defect. All bowel was easily reduced into the peritoneal cavity and the diaphragm was closed primarily with non-absorbable interrupted sutures (Fig. 2). The operation was completed in 2 h and 30 min with minimal blood loss and successful extubation in the OR. A 16-french chest tube was left in place which was removed on post-operative day four. Both oral and post-pyloric feedings were resumed within 48 h and were tolerated well. The patient was discharged in stable condition 40 days after transplantation, and eight days after repair of the diaphragmatic hernia.

One year after discharge, the patient is without gastrointestinal or respiratory complaints. The liver graft function remains excellent and the patient has not required any further rounds of chemotherapy since transplant.

2. Discussion

Liver transplantation for terminal liver disease in infants dates to 1967, when Starzl and colleagues achieved survival to 13 months in a 1.5 year-old patient [1]. In 2000, unresectable hepatoblastoma, a malignancy that comprises more than 75% of pediatric primary liver tumors, was added to the list of diseases successfully treated with liver transplantation [2]. As with those patients who are eligible for liver-sparing resections, patients with unresectable hepatoblastoma who are to undergo primary transplantation are routinely treated with neoadjuvant chemotherapy [3].

A thorough review of diaphragmatic hernia after liver transplant by Shigeta and colleagues reported three cases and reviewed 20 reported cases in the literature. In four of these patients, the indication for transplant was neoplasia (2 hepatic hemangioendothelioma, 1 primary embryonal sarcoma, and 1 angiosarcoma) [4]. Further review of these case reports failed to reveal any mention of pre-operative chemotherapy [5–7]. Here, we report an association of neoadjuvant chemotherapy with diaphragmatic hernia.

Fig. 1. Chest x-ray revealing loops of bowel within right chest.

Fig. 2. A) Bowel encountered upon entering right chest with thoracoscope, B) Bowel being reduced into peritoneal cavity through diaphragmatic defect, C) Thoracoscopic closure of diaphragmatic defect, D) Diaphragmatic defect fully approximated with all bowel within peritoneal cavity.
herniation following liver transplantation in an infant and the successful repair utilizing a minimally invasive technique.

Both laparoscopic and thoracoscopic repairs of congenital diaphragmatic hernias are routinely performed in some centers, but acquired defects are even more rare [8]. In this case, thoracoscopy avoided many of the potential difficulties of a trans-abdominal approach after a major surgery that significantly alters the intra-abdominal anatomy, such as a split left-lateral liver graft.

Although transplantation for hepatoblastoma is becoming more common, experience with liver transplant after neo-adjuvant chemotherapy remains a formidable challenge. Unlike hepatic resection, timing of liver transplantation from a deceased donor is dependent on graft availability, making preoperative optimization logistically tenuous.

McCabe and colleagues have posited several factors that likely contribute to diaphragmatic herniation following liver transplant, with an emphasis on the interaction between patient factors and surgical technique [9]. In our patient, we believe the main factors contributing to this complication were the combination of argon beam diathermy (for achieving hemostasis of the large raw surface of the supra-hepatic diaphragm), the patient’s nutritional status (small for age), and the patient’s prior chemotherapy.

3. Conclusion

Right-sided diaphragmatic herniation remains a very rare complication of liver transplantation which requires prompt diagnosis and treatment. Transplant physicians and surgeons should heighten their index of suspicion for the possibility of this complication when a split graft is used and preoperative chemotherapy is involved.

Thoracoscopic repair is a viable option that provides definitive resolution with minimal morbidity. The video-assisted minimally invasive approach via the thoracic cavity allowed excellent visualization of and access to the defect while minimizing risk to the liver graft.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Conflict of interest

The authors have no relevant conflicts of interest to disclose.

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