Case Report: Multivisceral Transplantation for an Extensive Cystic Lymphangioma of the Mesenteric Root

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ABSTRACT

Background. We report the case of a 7-year-old girl with intestinal failure owing to a cystic lymphangioma compromising the root of the mesentery, not amenable to resection, leading to intestinal failure. Oncologic treatment was attempted to reduce tumor size with no response; therefore, she was listed for multivisceral transplantation.

Procedure. Resection of the tumor required resection of all abdominal organs with vascular inflow and outflow. A multivisceral graft (liver, stomach, duodenum–pancreas and spleen complex, small bowel, and right colon) was implanted. For vascular reconstruction, donor's superior vena cava was sutured to the recipient's suprahepatic veins in a common patch. For arterial inflow, an arterial conduit was placed directly to the infrarenal aorta, and sutured to an aortic patch of the graft. Cold ischemia time was 8:45 hours; warm ischemia time was 35 minutes. A double-layer gastrogastric anastomosis and piloroplasty was made; and the distal reconstruction was performed with ileocolic side-to-end anastomosis that allowed to perform of a Bishop-Koop ileostomy for endoscopic monitoring.

Outcome. The patient recovered well after the procedure and was discharged 36 days after transplantation with intestinal sufficiency. To the best of our knowledge, this is the first report describing cystic lymphangioma as an indication for multivisceral transplantation.

CYSTIC LYMPHANGIOMA is a benign condition derived from anarchic proliferation of lymphatic vessels; it is believed to be a congenital malformation rather than a true neoplastic lesion [1]. The most common sites of occurrence are head and neck, and it is a pathology usually found in pediatric patients. Mesenteric root localization constitutes about 1% of all cases; it presents with abdominal distension, palpable mass, or acute intestinal obstruction. The most common complication reported is volvulus. Intestinal failure owing to venous congestion, as in this case, is not reported in revised literature, to the best of our knowledge [2,3]. Surgical treatment, when the lymphangioma is symptomatic or complicated, is the best treatment. Nevertheless, when the tumor is located in the root of the mesentery and around the celiac trunk or the hilum of the liver, it makes the surgery difficult and challenging, and sometimes impossible to carry it out, outside off the transplantation setting. Multivisceral transplantation has been proved to be useful to treat benign or low-grade malignant tumors involving the root of the mesentery and/or celiac axis. We present a video of a pediatric patient with a cystic lymphangioma compromising the portomesenteric axis treated with multivisceral transplantation [4,5].

CASE PRESENTATION

A 7-year-old girl presented with an abdominal mass at the age of 4 years. The mass was localized in the root of the mesentery comprising the mesenteric vessels (Fig 1). Despite multiple medical

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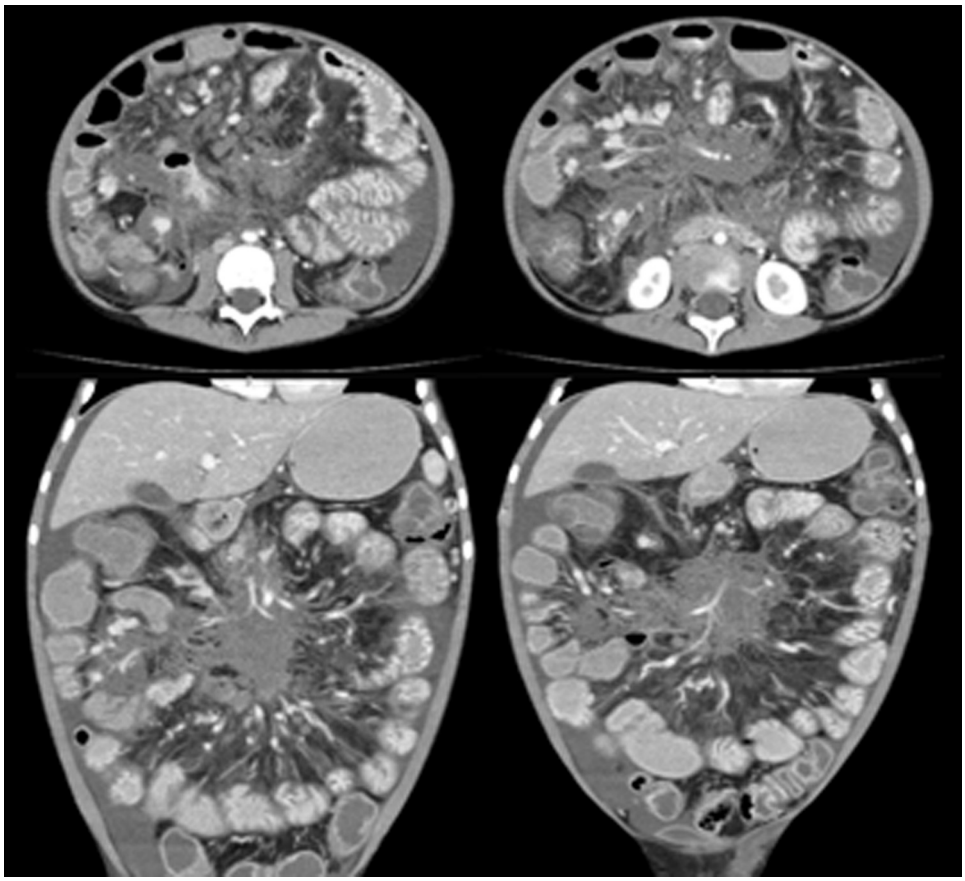

Fig 1. Computed tomography scan showing the tumor involving celiac and mesenteric axis, pancreas, and liver hilum.

therapeutic approaches, including chemotherapy (interferon, thalidomide, and bevacizumab) and surgical exploration, she evolved with a progressive weight loss, ascites, and diarrhea secondary to venous stasis. A surgical resection was attempted, but it was impossible owing to multivisceral involvement, and it would have required complete resection of all the abdominals organs (Fig 2). The case was referred for evaluation by our team, and a multivisceral transplant was proposed in the setting of a patient with frequent electrolyte disorders, worsening ascites, and weight loss owing to progressive intestinal failure.

Transplant Procedure

A xyphoid–pubic midline laparotomy was performed to access the abdominal cavity. Multivisceral resection was carried out, including all organs with inflow and outflow involvement. A 6-year-old trauma donor was offered and organs were procured, including liver, stomach, duodenum–pancreas and spleen complex, small bowel, and right colon. The engraftment started with venous vascular reconstruction, the donor's superior vena cava was sutured to the recipient's suprahepatic veins in a common patch. The arterial inflow was reconstructed with an arterial conduit placed in the infra-renal aorta of the recipient and sutured to the aortic patch of the graft. The graft was perfused with cold Ringer lactate solution to washout the University of Wisconsin solution before reperfusion, which occurred after 525 minutes of cold ischemia, and 35 minutes of warm ischemia. Upper gastrointestinal reconstruction was made with a hand-sewn double layer gastrogastric anastomosis, piloroplasty also performed. An end-to-side ileocolic anastomosis was made for distal gastrointestinal reconstruction, and a Bishop-Koop ileostomy for endoscopic surveillance. The patient received 2 U of red pack blood cells, and 4 U of fresh frozen plasma.

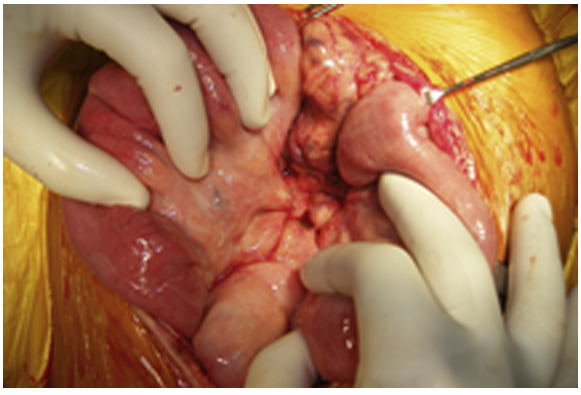

Fig 2. Intraoperative view of the tumor, retraction of the mesentery, as manifestation of mesenteric axis involvement can be seen. We also noted intestinal edema owing to superior mesenteric vein compression and involvement of the liver hilum.

Patient Follow-up

The patient remained in intensive care for 17 days, owing to failure of mechanical ventilation weaning caused by generalized edema. Oral intake was delayed owing to delayed gastric empting that was treated medically. After recovery, she was sent home at 36 days after the procedure with a fully functioning graft. During month 3 posttransplantation, the patient was readmitted with a severe Epstein–Barr virus primary infection, leading to develop a severe posttransplant lymphoproliferative disorder with central nervous system effects. She died owing to this complication with a fully functioning graft.

DISCUSSION

Cystic lymphangioma localized in the abdomen is a rare condition, reported only in 5% of the total cases of this disease. Only 1% affects the root of the mesentery. The condition is considered benign. Clinical manifestations are caused by mass effect of these lesions. The majority of clinical presentations reported include intestinal occlusion and complications owing to tumor compression; intestinal failure has not been reported as complication of the disease previously. In fact, this case not only has intestinal failure owing to mesenteric vein compression and stasis but also ascites and alterations owing to its location. Complete surgical resection was not feasible because it could require the resection of all abdominal organs; therefore, after careful evaluation multivisceral transplantation was considered as the best option. The early result supports the indication of the multivisceral transplant for this rare disease; the long-term results showed that multivisceral transplantation carried an increased risk for posttransplant lymphoproliferative disorder [5]. Although prevention and early diagnosis of the disease allowed better treatment, it remains as a serious and life-threatening complication [6].

REFERENCES

[1] Hardin WJ, Hardy JD. Mesenteric cysts. Am J Surg 1970;119:640–5.

[2] Chen CW, Hsu SD, Lin CH, et al. Cystic lymphangioma of the jejunal mesentery in an adult: a case report. World J Gastroenterol 2005;11:5084–6.

[3] Jang JH, Lee SL, Ku YM, et al. Small bowel volvulus induced by mesenteric lymphangioma in an adult: a case report. Korean J Radiol 2009;10:319–22.

[4] Nikeghbalian S, Aliakbarian M, Shamsaeefar A, et al. Multivisceral Transplantation for the treatment of intra-abdominal tumors. Transplant Proc 2013;45:3528–30.

[5] Singavi AK, Harrington AM, Fenske TS. Post-transplant lymphoproliferative disorders. Cancer Treat Res 2015;165:305–27.

[6] Abu-Elmagd KM, Mazariegos G, Costa G, et al. Lymphoproliferative disorders and de novo malignancies in intestinal and multivisceral recipients: improved outcomes with new outlooks. Transplantation 2009;88:926–34.

SUPPLEMENTARY DATA

Supplemental material for this article can be found at http://dx.doi.org/10.1016/j.transproceed.2015.12.067.