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Jejunal mesenteric lymphangioma treated by pancreaticoduodenectomy: A case report

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ABSTRACT

INTRODUCTION: A lymphangioma is a benign congenital malformation of the lymphatic system that generally appears in the head, neck, and axillary regions. Small bowel mesenteric lymphangiomas have been described in less than 1% of lymphangiomas.

PRESENTATION OF CASE: We report the case of a 20-year-old woman who presented with abdominal pain. Computed tomography revealed a large (22 cm in diameter) multi-cystic lesion inferior to the processus uncinatus of the pancreas. As the presumptive diagnosis was a lymphangioma of the jejunal mesentery, we decided to perform a laparotomy. Intraoperatively, the peritoneal cavity was found to be fully occupied by a multi-cystic lesion that arose from the root of the jejunal mesentery and the processus uncinatus of the pancreas. It was adherent to the duodenum and inseparable from the duodenum and the processus uncinatus. A subtotal stomach-preserving pancreaticoduodenectomy was performed. The tumor was diagnosed as a lymphangioma of the jejunal mesentery after histopathological analysis.

DISCUSSION: Although lymphangioma is benign, complete resection, including resection of the involved organs, is necessary. Incomplete resection has a 10% postoperative recurrence rate and may also be associated with complications. To the best of our knowledge, this is the first reported case of a mesenteric lymphangioma treated by pancreaticoduodenectomy.

CONCLUSION: Although the lymphangioma was pathologically benign, a pancreaticoduodenectomy was required to remove it completely. When a tumor's location and size cause impingement on surrounding structures, surgeons should consider performing a pancreaticoduodenectomy to treat similar cases.

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1. Introduction

Lymphangiomas are rare cystic tumors frequently located in the head, neck, and axilla of children [1]. Small bowel mesenteric lymphangiomas are extremely rare, especially in adults. These account for less than 1% of all lymphangiomas [2]. Lymphangiomas are benign tumors that appear to result from congenital malformations of lymphatic vessels [3]. The optimal treatment for mesenteric lymphangioma is complete resection [2]. Most mesenteric lymphangiomas are asymptomatic until they enlarge. Here,

we report a large mesenteric lymphangioma in a young adult treated with subtotal stomach-preserving pancreaticoduodenectomy. To the best of our knowledge, this is the first reported case of a mesenteric lymphangioma treated by pancreaticoduodenectomy. Because of the rarity of this condition, identified cases must be documented, reported, and elucidated. We are reporting in line with the statement: Updating Consensus Surgical Case Report Guidelines (SCARE) [3].

2. Presentation of case

A 20-year-old woman was referred to our hospital by a primary care physician due to abdominal pain and constipation for 7 days. Abdominal examination revealed distention. Most laboratory test results including carcinoembryonic antigen and carbohydrate antigen 19-9 levels revealed normal limits, except for a slightly elevated serum C-reactive protein level (CRP, 2.7 mg/dL). Abdominal contrast-enhanced computed tomography (CT) revealed a large, multi-cystic lesion (22 cm in diameter) inferior to the processus uncinatus of the pancreas. It extended up to the pelvis and

Abbreviations: CT, computed tomography; MRI, magnetic resonance imaging; CRP, C-reactive protein; EUS-FNA, endoscopic ultrasound-guided fine needle aspiration.

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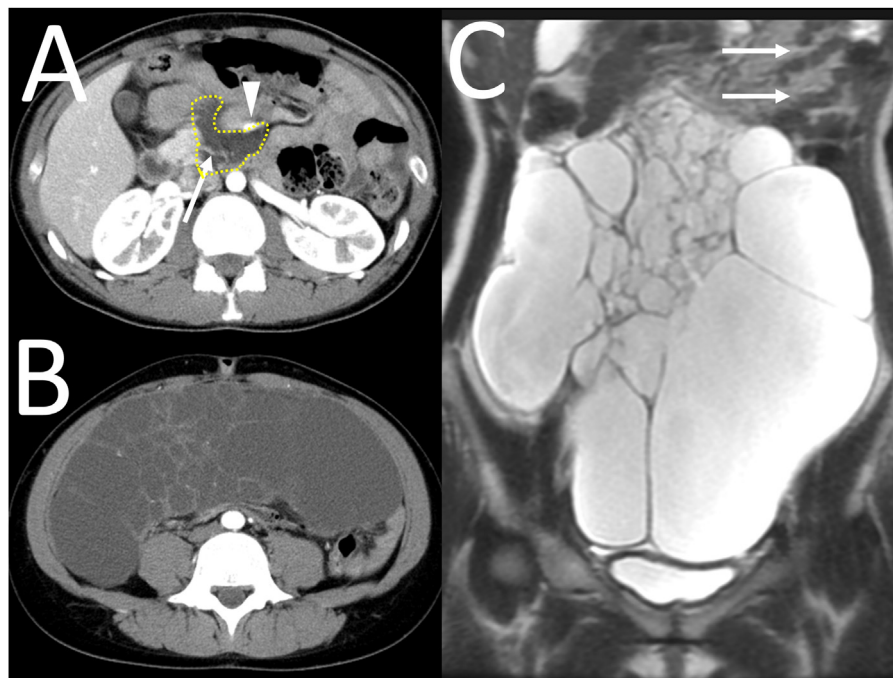


Fig. 1. Abdominal computed tomography (CT) and magnetic resonance imaging (MRI).

(A) Contrast-enhanced CT shows a cystic lesion arising from the processus uncinatus of the pancreas and the root of the superior mesenteric artery (arrowhead). The lesion is being supplied by the second branch (arrow) of the superior mesenteric artery.

(B) The cystic wall and septa are enhanced by the contrast medium.

(C) Coronal T2-weighted MRI shows hyperintense, huge, multiple, cystic lesions displacing the small intestine upwards (arrows).



Fig. 2. Intraoperative findings during the patient's laparotomy. The peritoneal cavity is fully occupied by a multi-cystic lesion arising from the root of the jejunal mesentery. It predominantly contains a clear yellowish fluid.

displaced the small intestine upwards (Fig. 1). The wall of the lesion and septa were enhanced by contrast medium. There was no nodule within the cystic lesion. It was supplied by the second branch of the superior mesenteric artery. Abdominal magnetic resonance imaging (MRI) showed hypointense T1- and hyperintense T2-weighted multiple cystic lesions (Fig. 1). Both ovaries were distinctly normal. The working impression was then a case of mesenteric lymphangioma. Total excision of the lesion was planned. During laparotomy, the peritoneal cavity was found to be fully occupied by a multi-cystic lesion that arose from the root of the jejunal mesentery and the processus uncinatus of the pancreas. It was adherent to the transverse mesocolon and duodenum (Fig. 2). It contained a clear yellowish fluid. A portion of the transverse mesocolon was resected with the cystic lesion; however, the branches of the middle colic vessels were preserved. The mass was inseparable from the duodenum and processus uncinatus. A

subtotal stomach-preserving pancreaticoduodenectomy was performed. Postoperatively, the patient's course was uneventful. She was discharged on the 18th postoperative day. Histopathologic evaluation of the lesion was conducted. Macroscopically, a huge, 22 × 21 × 8 cm, yellowish, multi-cystic lesion was observed (Fig. 3). The majority of the cysts contained a yellowish fluid; however, the small cysts near the root of the jejunal mesentery contained a milky white substance. Microscopic examination revealed that the multi-cystic lesion was composed of an irregularly dilated space separated by fibrous tissues and smooth muscle fascicles. The wall of the cyst was lined by a flat endothelial layer (Fig. 4). The stroma was composed of collagen, cholesterol crystals, and foreign body giant cells. The epithelium covering the cysts was positive for D2-40 and CD31 immunostaining. Histopathological examination was consistent with the diagnosis of a mesenteric lymphangioma. The processus uncinatus of the pancreas, duodenum, and two lymph nodes were involved with the lymphangioma. Eight months postoperatively, on follow-up, the patient was well and did not have any recurrence.

3. Discussion

Lymphangiomas are mass-forming lesions characterized by many thin-walled lymphatic spaces [4]. The etiology of mesenteric lymphangioma most likely favors a congenital abnormality of the lymphatic system that causes sequestration of the lymphatic tissues during embryologic development. However, other possible causes such as inflammation, abdominal trauma, abdominal surgery, radiation therapy, and lymphatic obstruction have also been suggested [4].

Mesenteric lymphangiomas are usually asymptomatic until they enlarge. Common symptoms include a palpable abdominal mass, abdominal pain, distension, vomiting, and constipation [5,6]. Unfortunately, these symptoms are common to many other diseases. Thus, it is challenging to make a specific diagnosis based on

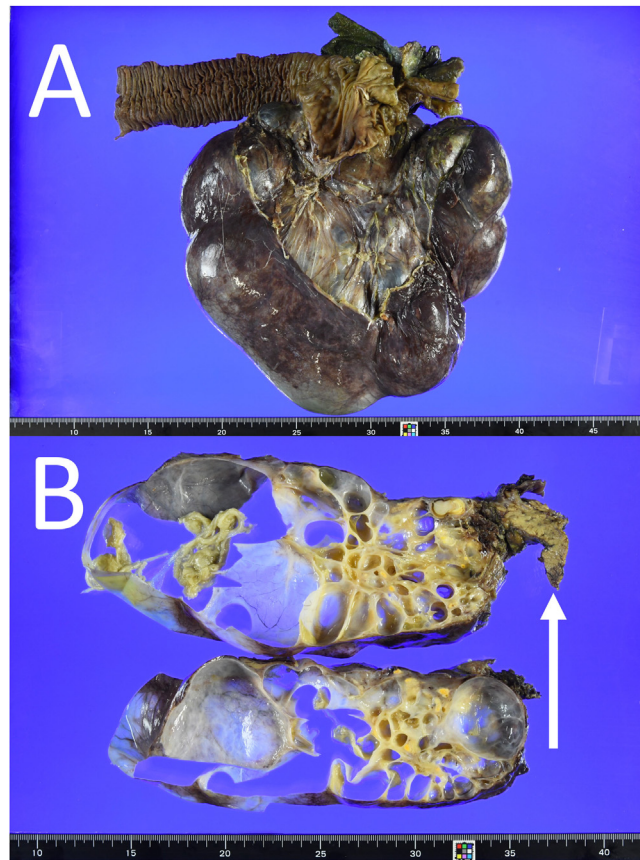


Fig. 3. Macroscopic examination of the multi-cystic lesion.
 (A) The formalin-fixed specimen shows a huge, 22 × 21 × 8 cm, polycystic lesion adherent to the processus uncinatus of the pancreas.
 (B) The cut section reveals that the tumor consists of multiple cysts which vary in size, from 1 mm to 10 cm. Arrow: processus uncinatus of the pancreas.

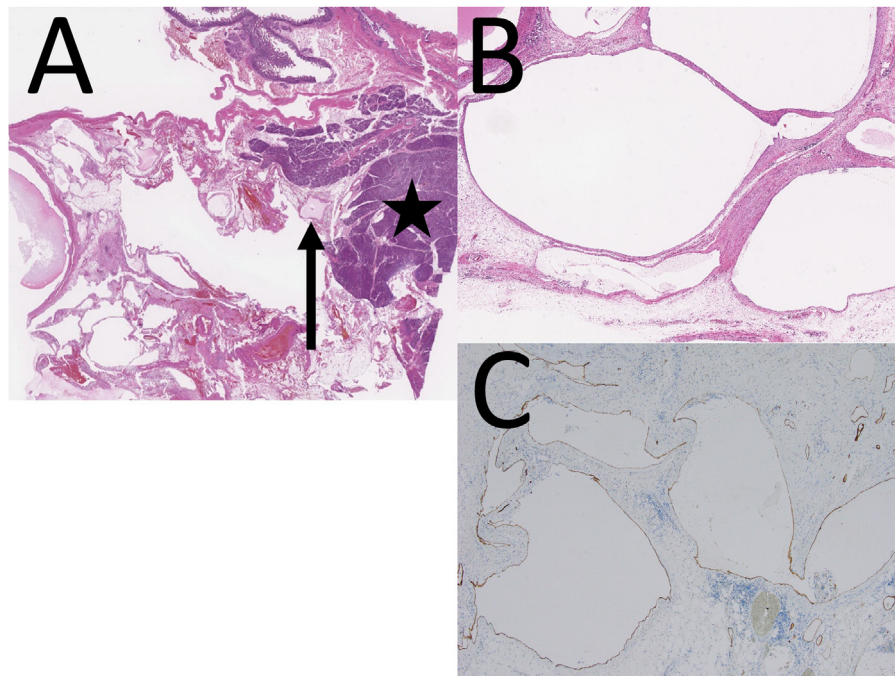


Fig. 4. Microscopic examination of the multi-cystic lesion.
 (A) The processus uncinatus of the pancreas is seen adjacent to a lymphangioma (Hematoxylin & Eosin, loupe). Arrow: a portion of the lymphangioma; Star: processus uncinatus of the pancreas.
 (B) The lesion is composed of multiple loculi; each locule has an irregularly dilated space separated by fibrous tissues and smooth muscle fascicles. The wall of the cyst is lined by a flat endothelial layer (Hematoxylin & Eosin, ×13).
 (C) Immunohistochemical staining shows that the epithelium covering the cysts is positive for D2-40 (×40).

these symptoms alone. Additionally, there are no blood tests to confirm the diagnosis. CT and MRI would usually show multilocular, well-circumscribed cystic masses whose wall and septa are enhanced by contrast media [2]. This finding was evident in our patient. Although these modalities aid in determining the diagnosis, they are insufficient in establishing an accurate preoperative diagnosis. Often, they are unable to determine the origin of the tumor. This is especially true if the size of the tumor is large. For the patient in the current study, CT revealed that the lesion was supplied by the second branch of the superior mesenteric artery. Therefore, we diagnosed its origin from the jejunal mesentery. The patient was appraised that pancreaticoduodenectomy would be needed if, intraoperatively, the tumor would be inseparable from the pancreas and duodenum. Even in asymptomatic cases, the primary treatment for this condition is radical excision of the mass because of its potential to grow enormously, invade adjacent structures, and develop multiple complications. Examples of such complications are intestinal obstruction, infection, rupture, and hemorrhage [1,5,7]. However, endoscopic ultrasonography guided fine needle aspiration (EUS-FNA) may be a safe and accurate method for the diagnosis and treatment of suitable cases [2]. In asymptomatic cases of small lymphangiomas diagnosed by EUS-FNA, non-surgical management with regular follow-up can be carried out [8]. In the current study, preoperative EUS-FNA was not applicable because the patient was symptomatic and the lesion was multi-cystic. Although the lesion has a benign nature, complete resection, including resection of the involved organs, is necessary [9,10]. Incomplete resection has a 10% postoperative recurrence rate [2]. Incomplete tumor removal may also be associated with complications such as infection, fistula, and hemorrhage [11]. As the intestines and mesenteric vessel branches may infiltrate, mesenteric lymphangiomas are usually resected with segments of the intestines [9]. In our case, we first performed partial pancreatectomy and duodenectomy because the lymphangioma infiltrated the duodenum and the processus uncinatus of the pancreas. However, the area in contact with the pancreas processus was unclear; thus, we performed a subtotal stomach-preserving pancreaticoduodenectomy to remove the tumor completely. Fahimi et al. reported that they performed a modified classic pancreaticoduodenectomy for a lymphangioma of the pancreas. Their technique involved resection of the head of the pancreas with preservation of the upper second duodenal portion and the ampulla of Vater [12]. This technique might be used in our case to preserve the ampulla of Vater. However, the safety of this unique technique has not yet been established.

We performed a PubMed® search on world literature between 1950 and April 2020 using the keywords “lymphangioma” and “pancreaticoduodenectomy.” There were eight articles about lymphangioma treated with pancreaticoduodenectomy [2,8,13–18]. All papers in English were included. Research papers on hemolymphangioma were excluded. The median age of the nine patients (including the case in this study) was 33 years (range, 20–53 years). There were four women and five men. The median tumor size was 22 cm (range, 3–27 cm). The origin of the lymphangioma was at the pancreatic head in six patients, the retroperitoneum in one patient, and the mesentery in the current case. One article did not mention it. To the best of our knowledge, this is the first reported case of a mesenteric lymphangioma treated by pancreaticoduodenectomy.

4. Conclusion

Here, we describe the case of a young woman who underwent pancreaticoduodenectomy for a large mesenteric lymphangioma. As seen in our case, when a tumor's location and size cause

impingement on surrounding structures, surgeons should consider performing a pancreaticoduodenectomy to treat similar cases.

Declaration of Competing Interest

Dr. Hirotaka Kitamura, Dr. Daisuke Yamamoto, Dr. Shinichi Kadoya, Dr. Hiroyuki Bando, Dr. Yurie Okayama and Dr. Hiroshi Minato have no conflict of interest of financial to disclose.

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Ethical approval

This study design was approved by the Ishikawa Prefectural Central Hospital Ethics Committee (approval no. 1599).

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.

Author contribution

Hirotaka Kitamura: a surgeon who operated on patient and contributed to writing the paper.

Yurie Okayama and Hiroshi Minato: pathologists who contributed on the pathological section of the paper.

Daisuke Yamamoto, Shinichi Kadoya and Hiroyuki Bando: provided assistance for medical practices and contributed to writing-reviewing the paper.

Registration of research studies

N/A.

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Availability of data and materials

The datasets used during the current study are available from the corresponding author on reasonable request.

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