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# A rare case of retroperitoneal hemolymphangioma

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ABSTRACT

*INTRODUCTION:* Hemolymphangioma, a rare vascular developmental condition, is characterized by malformed venous and lymphatic components in various proportions. Herein, we report a case of a retroperitoneal cystic tumor in an adult patient.
*PRESENTATION OF CASE:* A 68-year-old man presented to our hospital with complaints of abdominal pain and vomiting. His abdomen was distended with upper tenderness but without rebound tenderness. Computed tomography (CT) scanning demonstrated a retroperitoneal cystic tumor at the dorsal part of the pancreatic head. Thus, a diagnosis of liposarcoma or lymphoma was made. The patient was scheduled for surgery after his general condition became stable. Intraoperatively, the cystic tumor was found to have originated from the retroperitoneal space. The tumor was in contact with the pancreatic head, abdominal aorta, and inferior vena cava. There was no invasion into the surrounding tissue. The cystic tumor was resected completely. Histopathological examination revealed that the resected retroperitoneal cystic tumor was a hemolymphangioma. The patient had no recurrence during the 12-month follow-up.
*DISCUSSION:* Hemolymphangioma is a rare benign tumor, and its accurate diagnosis before surgery is still difficult. Disease presentation may vary from simple well-defined cystic lesions to aggressive ill-defined lesions, mimicking malignancy. Complete excision provides the best results with a low recurrence rate.
*CONCLUSION:* Further research is needed on the preoperative radiological diagnosis of such tumors and on how to determine tumor resectability in such cases.



## 1. Introduction

The present work has been reported in line with the SCARE-criteria [1]. Hemolymphangioma, a rare vascular developmental condition, is characterized by malformed venous and lymphatic components in various proportions. Nonsurgical treatments, including cryotherapy, laser therapy, radiotherapy, and local injection of sclerotic agents, do not show superiority to surgical treatments [2]. To the best of our knowledge, only a few cases of hemolymphangioma have been reported in the literature so far. Herein, we report a case of retroperitoneal hemolymphangioma, which was managed with surgical resection.

## 2. Presentation of case

A 68-year-old man was referred to our department from his primary care hospital for upper abdominal pain and vomiting with no other associated symptoms. The medical history of the patient included appendectomy and atherothrombotic brain infarction. On physical examination, his abdomen was distended with upper abdominal tenderness but without rebound tenderness. Family history was unremarkable. Laboratory analysis revealed that carcinoembryonic antigen (CEA), carbohydrate antigen 19-9 (CA-19-9), and interleukin-2 receptor were all within the normal ranges. Computed tomography (CT) scan with contrast medium revealed an almost low-density heterogeneous tumor with slight enhancement, which had both cystic and solid components. The tumor was oval, 66 × 47 mm in diameter, and located at the dorsal portion of the pancreatic head. There were no calcifications, but there was a possibility of invasion into the pancreatic head and duodenum (Fig. 1A, B). Positron emission tomography-computed tomography (PET-CT) demonstrated low fluorodeoxyglucose (FDG) uptake within the tumor (Fig. 1C). These radiological findings are suggestive of either a liposarcoma or a lymphoma.

*Abbreviations:* CT, computed tomography; CEA, carcinoembryonic antigen; CA 19-9, carbohydrate antigen 19-9; PET, positron emission tomography; FDG, fluorodeoxyglucose; US, ultrasound sonography; MRI, magnetic resonance imaging.

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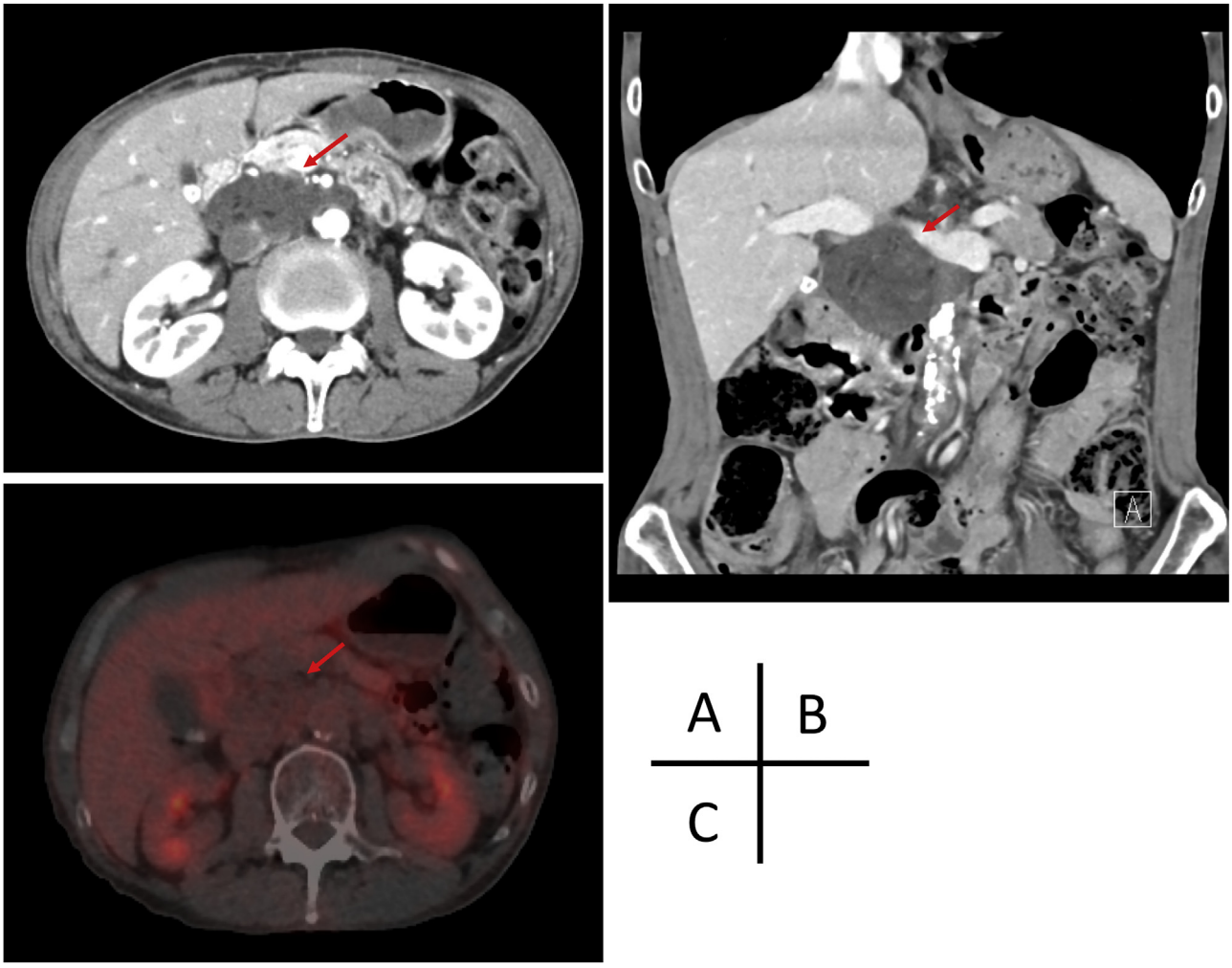


**Fig. 1.** Computed tomography (CT) shows a well-defined round, 66 × 47 mm cystic and solid mass with a rim of soft tissue in the retroperitoneal region and dorsal of the pancreatic head. Slight enhancement of the peripheral rim of soft tissue is seen after intravenous administration of the contrast medium (A: axial image, B: coronal image). This tumor has low fluorodeoxyglucose (FDG) uptake on positron emission tomography (PET) (C).

After carefully dissecting the pancreatic head, duodenum, inferior vena cava, and abdominal aorta, we removed the tumor en bloc through an open abdominal surgery. There was no invasion into the other organs. The operative time was 168 min, and the intraoperative blood loss was 70 ml. Macroscopic examination of the tumor revealed a capsulated mass, measuring 40 × 32 mm, consisting of cystic and solid components (Fig. 2). Hematoxylin-eosin staining showed that the tumor was composed of lymphatic and blood vessels with polycystic spaces (Fig. 3). Immunohistochemically, some endothelial cells were relatively positive for CD 31, and others were positive for D2-40 (Fig. 4). No atypical cells with hyperproliferation or mitotic division were seen. Considering the histological and immunohistochemical findings, a diagnosis of retroperitoneal hemolymphangioma was made. The postoperative course of the patient was uneventful. We confirmed that there were no abnormal findings detected during the postoperative CT reexamination, and the patient was subsequently discharged (Fig. 5). The patient was alive without recurrence at twelve months after the operation.

## 3. Discussion

Lymphangiomas can become evident at any age and may involve any part of the body; 50%–60% are seen at birth and 90% occur in children aged less than 2 years, and they commonly involve the head and neck. These lesions are rarely found in adult patients [3]. Intraabdominal lymphangiomas account for less than 5% of all lymphangiomas. The most common location is the mesentery, followed by the omentum, mesocolon, and retroperitoneum [4].

Hemolymphangiomas, on the other hand, are extremely rare and occur in various locations. Intraabdominal hemolymphangiomas account for 61.5% of all hemolymphangiomas. The most common location is the pancreas. Only one case of retroperitoneal hemolymphangioma has been reported [5].

Hemolymphangiomas are classified into two groups: congenital and acquired. Congenital hemolymphangiomas result from an obstruction of venolymphatic communication between systemic circulation and dysembryoplastic vascular tissue [6]. Acquired hemolymphangiomas occur owing to inadequate lymph drainage and damage to the lymphatic vessels resulting from surgery or trauma. Evidence suggests that imaging characteristics of hemolymphangiomas vary according to the location in the body, proportion of blood and lymphatic vasculature, and imaging modality [7].

These lesions can arise at any site, can be localized or more extensive, and can be superficial or deep [8]. Deep lesions without superficial involvement may remain unrecognized until the patient presents with clinical symptoms later in life [8]. The small intraabdominal cyst may not present any symptoms, unless it enlarges significantly and compresses adjacent organs. The larger cyst could cause acute abdomen or dull aching pain. Possible etiologies for

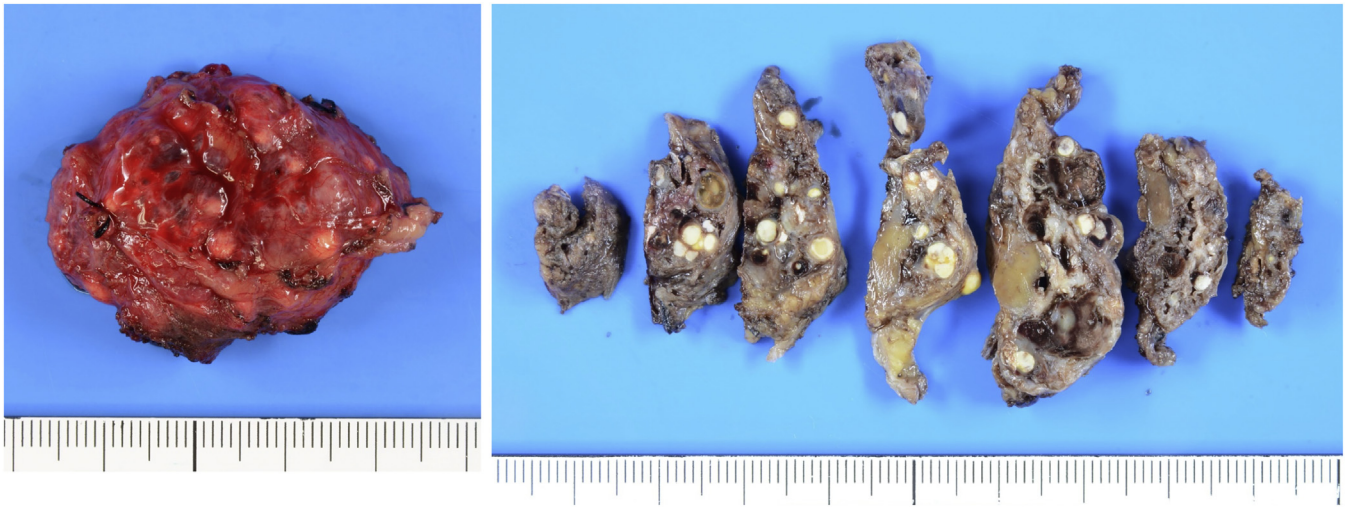

**Fig. 2.** The resected specimen reveals a capsulated mass, which measures 40 × 32 mm and consists of cystic and solid areas, macroscopically.

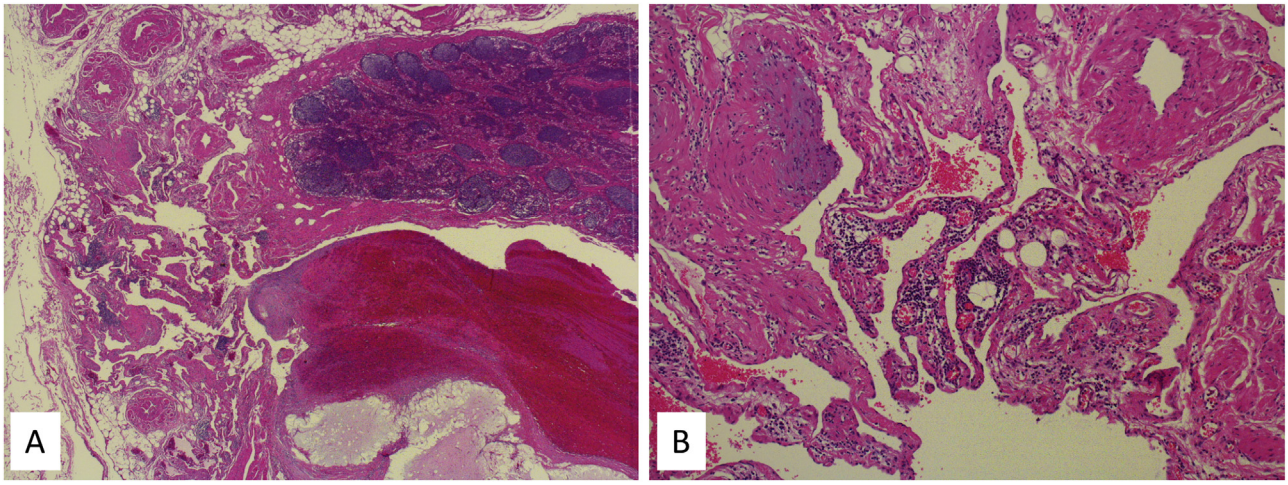


**Fig. 3.** Microscopic examination shows that the tumor is composed of lymphatic and blood vessels with polycystic spaces (hematoxylin and eosin stain). Magnifications: (A) ×2 and (B) ×10.

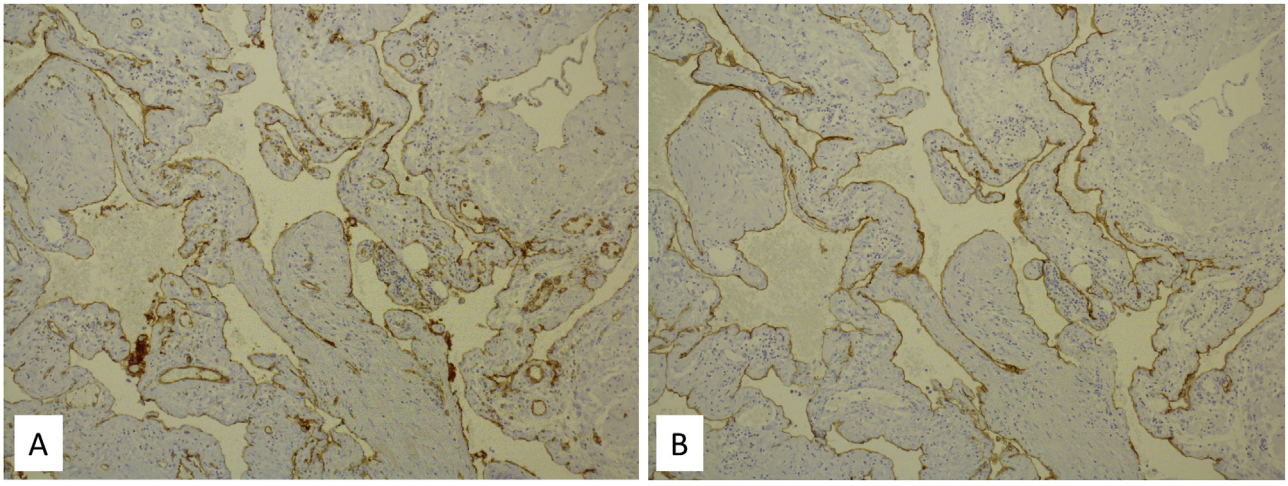


**Fig. 4.** Immunohistochemically, some endothelial cells are relatively positive for CD 31 (A), and other cells are positive for D2-40 (B).

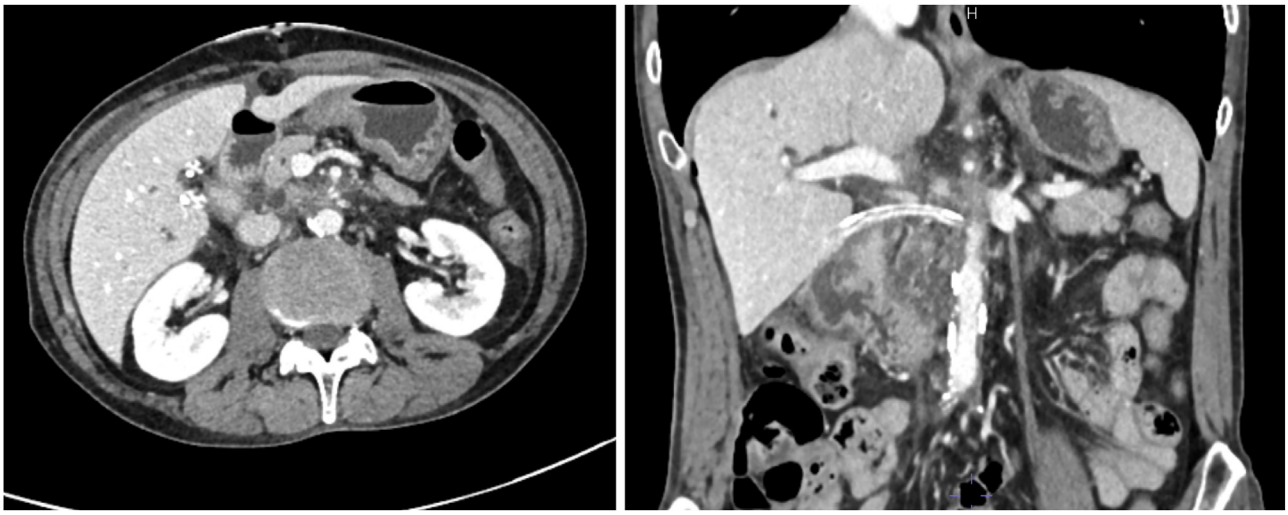

**Fig. 5.** Postoperative computed tomography (CT) shows no abnormal findings.

acute abdominal pain proposed in the literature include intestinal obstruction, torsion, infarction, traction of the mesentery as the cysts expand superimposed infection, and peritoneal irritation due to leakage of the cyst contents [4]. This tumor causes various symptoms depending on the site of occurrence. This patient presented with abdominal pain and vomiting, which could be due to the pressure of the huge lesion on the internal organs. Traumatic rupture, hemorrhage, and lymphorrhea followed by secondary infection are common complications of hemolymphangioma [2].

Hemolymphangiomas present as cystic or cavernous lesions that consist of dilated veins and lymphatic vessels interspersed with normal stromal tissue and vasculature; dilated vessels may contain thrombosis [9]. Immunohistochemical analysis demonstrates that hemolymphangiomas are positive for CD 31 and D2-40 [10]. Ultrasonography (US), computed tomography (CT), and magnetic resonance imaging (MRI) are useful for the diagnosis of hemolymphangiomas, with findings dependent on the amount of water-based substance and number of blood vessels in the lesion [7].

Complete excision provides the best results with a lower recurrence rate. However, careful follow-up is necessary. The recurrence rate varies depending on the complexity, anatomical location, and adequacy of the excision. It has been established in the literature that lesions that have been completely excised show 10%–27% recurrence, whereas 50%–100% of partly resected tumors may recur. Compared with surgical treatments, nonsurgical treatments, including cryotherapy, laser therapy, radiotherapy, and local injection of sclerotic agents, do not show superiority [2].

## 4. Conclusion

Hemolymphangioma is a rare benign tumor, and its accurate diagnosis before surgery is still difficult. Disease presentation may vary from simple well-defined cystic lesions to aggressive ill-defined lesions mimicking malignancy. Complete excision provides the best results with a lower recurrence rate. Further research is needed on the preoperative radiological diagnosis and on how to determine tumor resectability in such cases.

## Conflicts of interest

The authors declare no conflicts of interest.

## Funding

The authors declare that this study was not funded externally.

## Ethical approval

As a case report without Protected Health Information, no ethics approval was required for this project.

## Consent

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

## Authors' contributions

MO and TK drafted the manuscript. MO and TK contributed to patient care. MO, MK, JH, and MH performed the literature search. MK performed histopathological examination and diagnosis. MO, KT, JH, HM, and NH participated in the critical revision of the manuscript. All authors have read and approved the final manuscript.

## Registration of research studies

This is a case report.

## Guarantor

Toshihiko Kohashi.

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