Clinical Case Report

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Splenic cystic lymphangiomatosis in association with omental varices and portal hypertension

A case report

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Abstract

Rational: Lymphangiomatosis is rare and benign, and slowly proliferating lymphatic vessels of unknown etiology and visceral lymphangiomatosis involving the spleen is rare. Since lymphangiomatosis may be asymptomatic or present as a sense of fullness, splenic cystic lymphangiomatosis is a disease of little concern.

Patient concerns: A 34-year-old woman suffering from progressive epigastric fullness after oral intake for two weeks.

Diagnoses: Physical examination showed a palpable mass which was more than 10 cm in size over the left hypochondrium. An abdominal computed tomography disclosed marked splenomegaly with multiple cystic lesions in the spleen, causing external compression with right-sided deviation of the adjacent organs and varices in the upper abdomen. Esophagogastroduodenoscopy revealed portal hypertensive gastropathy.

Interventions: Conventional total splenectomy was performed in this patient because of an enlarged spleen and unknown etiology, preoperatively. Upon surgery, splenomegaly with polycystic content and varicose vessels over the omentum were noted. Autologous spleen transplantation was not performed because of limited orthotopic and vascularized spleen.

Outcomes: The patient is doing well 18 months after splenectomy.

Lessons: This was a rare case presenting with splenic cystic lymphangiomatosis in association with omental varices and portal hypertension. Splenic cystic lymphangiomatosis should be considered in the differential diagnosis of patients with a palpable painless mass over the left hypochondrium.

Abbreviations: CT = computed tomography, GD = Gorham disease.

Keywords: omental varices, portal hypertension, splenic cystic lymphangiomatosis, splenomegaly

1. Introduction

Lymphangiomatosis is rare and benign, and slowly proliferating lymphatic vessels of unknown etiology and visceral lymphangiomatosis involving the spleen are rare.[1,2] It has been suggested that lymphangiomatosis is a congenital dysplasia of lymphatic tissue and is due to the abnormal development of the lymphatic vessels during fetal life.[3] Since lymphangiomatosis may be asymptomatic or present as a sense of fullness,[4] splenic cystic lymphangiomatosis is a diseases of little concern. Herein, we present the case of a woman with splenic cystic lymphangiomatosis. The splenic cystic lymphangiomatosis resulted in splenomegaly, external compression, omental varices, and portal hypertension. We have also reviewed the literature concerning splenic cystic lymphangiomatosis.

2. Case report

A 34-year-old woman had a medical history of chronic hepatitis B virus in the carrier state and splenic cysts without regular follow-ups. She suffered from intermittent epigastric fullness after oral intake for more than 5 years, and the discomfort progressed in the 2 weeks before visiting the clinic. She had no fever, nausea, vomiting, and weight loss. Upon physical examination, her vital signs were as follows: body temperature 36.5°C; respiratory rate 18 times per minute; heart rate 69 beats per minute; and blood pressure 143/84 mm Hg. Her abdomen was soft and mildly distended without tenderness, and hypoactive bowel sounds were noted. A palpable mass more than 10 cm over the epigastrium and the left upper quadrant were noted. Other physical examinations were unremarkable, including negative findings of lymph nodes over the neck, axillary, and inguinal areas. The patient had no pets. The laboratory data revealed anemia

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This article is a retrospective study and does not contain any studies with human subjects performed by any of the authors. So, the ethical approval was not necessary. Patient records or information was anonymized in this article. Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

The authors have no conflicts of interest to disclose.

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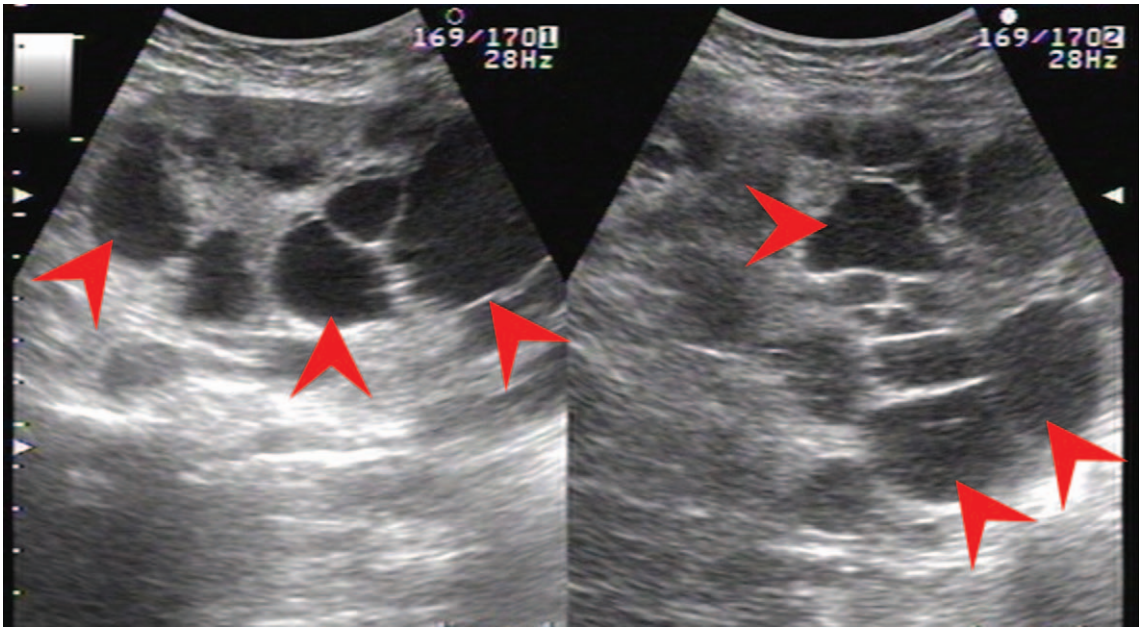

Figure 1. Abdominal sonography revealed multiple anechoic cystic lesions (arrow heads) in the spleen.

(hemoglobin 10.4 gm/dL, normal range 11.1–15.0 gm/dL) and thrombocytopenia (platelets 138×10^9/L, normal range 130–400×10^9/L), and normal aminotransferase levels. Tumor markers including α-fetoprotein (1.93 ng/mL, normal range <9.0 ng/mL), carcinoembryonic antigen (2.27 ng/mL, normal range <5.0 ng/mL), cancer antigen 19-9 (16.0 U/mL, normal range <35.0 U/mL), and cancer antigen-125 (41.5 U/mL, normal range <35 U/mL) were within normal limits.

Her previous abdominal sonography revealed multiple anechoic cystic lesions in the spleen without evidence of liver cirrhosis (Fig. 1). An abdominal computed tomography (CT) disclosed marked splenomegaly with multiple cystic lesions in the spleen, bilateral ovarian cysts, and a right renal simple cyst. The splenomegaly was causing external compression with right-sided deviation of adjacent organs and varices in the upper abdomen. No cystic lesions were noted in the lungs, liver, and bony structures (Fig. 2). Esophagogastroduodenoscopy for upper abdominal varices revealed a snakeskin appearance of the gastric body, favoring a diagnosis of portal hypertensive gastropathy, without esophageal or gastric varices (Fig. 3).

Splenectomy was performed with the diagnosis of upper abdominal varices and portal hypertension. The patient was placed in a supine position. We chose the conventional laparotomy method to remove the giant-sized spleen. Benz incision was created because the medial size of the spleen was 5 cm from right to the midline. While entering the abdominal cavity, the gastrosplenic ligament was first divided with HARMONIC FOCUS plus Long Shears (Ethicon, OH). After entering the lesser sac, the branches of splenic artery were individually ligated by silk tie for auto-transfusion. Splenomegaly with polycystic changes and yellowish fluid was noted during the operation. Varicose vessels were noticed

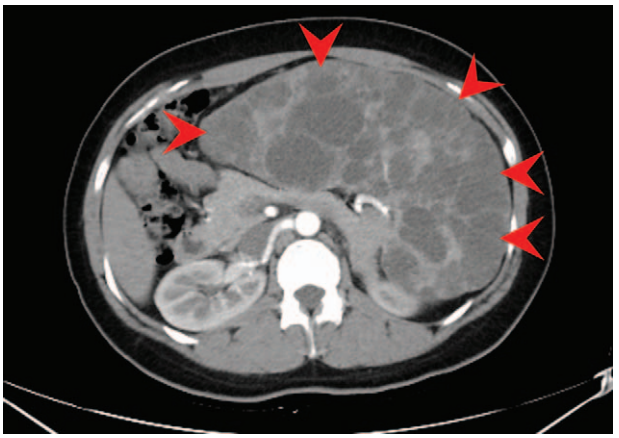

Figure 2. Abdominal computed tomography disclosed marked splenomegaly with multiple splenic cysts (arrow heads), causing external compression with right sided deviation of adjacent organs and varices in the upper abdomen.

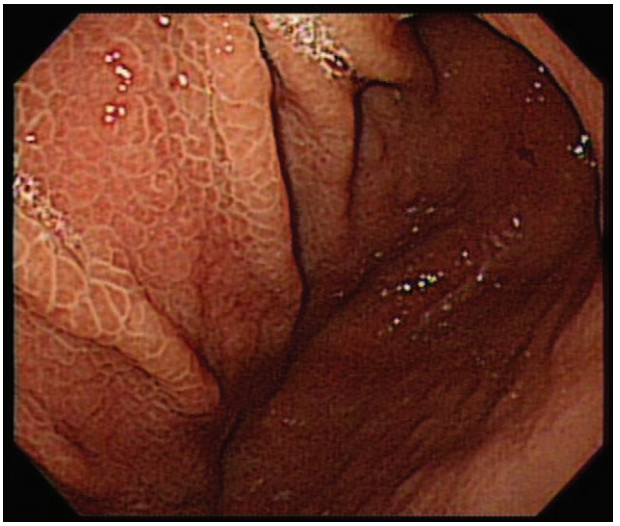

Figure 3. Esophagogastroduodenoscopy revealed portal hypertensive gastropathy.

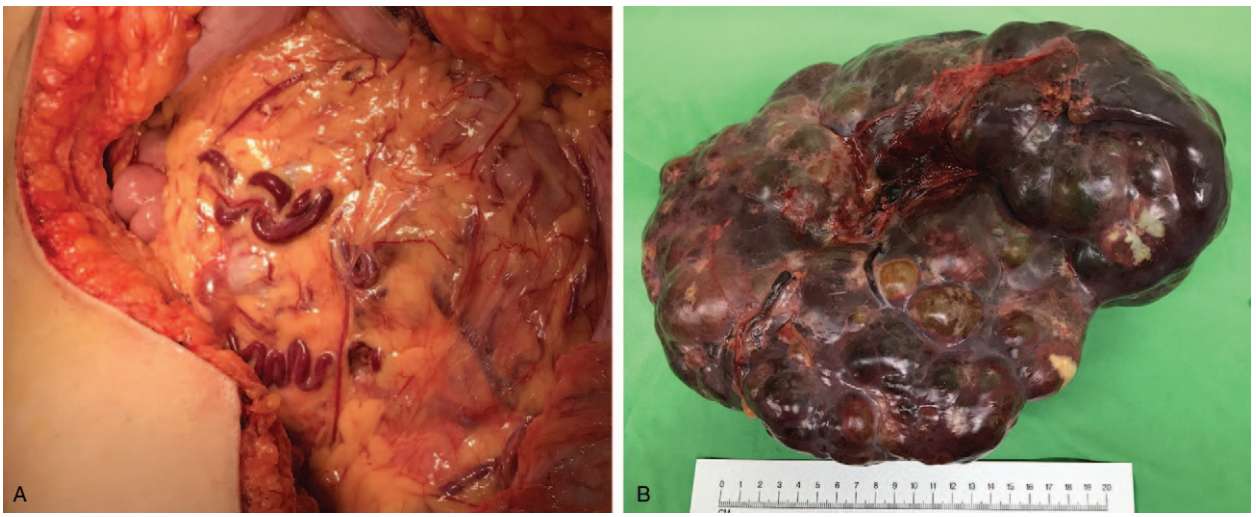

Figure 4. (A) Surgical finding of varices over the omentum. (B) The splenic specimen.

over the omentum (Fig. 4A). The splenic size was significantly reduced after auto-transfusion; thus, more space was available to easily finish conventional splenectomy. The total blood loss was 50 mL, and the total operation time was 158 minutes. The patient resumed oral intake since postoperative day 1, and was discharged on postoperative day 5 without any sequelae. The splenic specimen measured $28\,cm \times 21\,cm \times 9\,cm$ (Fig. 4B). Histologically, the splenic parenchyma had multiple, multifocal cystic spaces lined by flat endothelial cells (Fig. 5A). The lumen mostly contained proteinaceous fluid. Immunohistochemically, the endothelial cells were positive for CD31 (Fig. 5B) and negative for D2-40. The final diagnosis was splenic cystic lymphangiomatosis causing splenomegaly, omental varices, and portal hypertension from external compression. The patient is doing well 18 months after splenectomy.

3. Discussion

Lymphangiomatosis is a term used to describe a rare, slow-growing tumor characterized by thin-walled dilated vascular channels lined with endothelial cells filled with lymph.[2] Cystic lymphangiomatosis was first described by Rodenber in 1828.[5] The first case involving the spleen was reported by Frink in 1885.[6] It has been suggested that lymphangiomatosis is a congenital dysplasia of lymphatic tissue that results from abnormal development of the lymphatic vessels during fetal life.[3] Therefore, lymphangiomatosis is more likely considered a hamartomatous process rather than a neoplasm.[7]

Lymphangiomatosis affects somatic soft tissues in the neck, axilla, mediastinum, retroperitoneum, and extremities,[1,8,9] and it predominantly affects the neck (75%) and axilla (20%) in a case series involving children.[8] The lymphangiomatous process may diffusely involve multiple organ systems when the term "systemic" lymphangiomatosis is applied,[1,2,4] and it may be confined to a solitary organ, such as the liver,[9] spleen,[10] and kidney.[11] In the latter condition, lymphangiomatosis is called the "isolated" form.[4] It mainly occurs in infants and children and seldom affects patients older than 20 years.[12] The disease in children is often systemic, while it is mostly isolated in adults.[4] Visceral lymphangiomatosis involving the spleen is rare, and

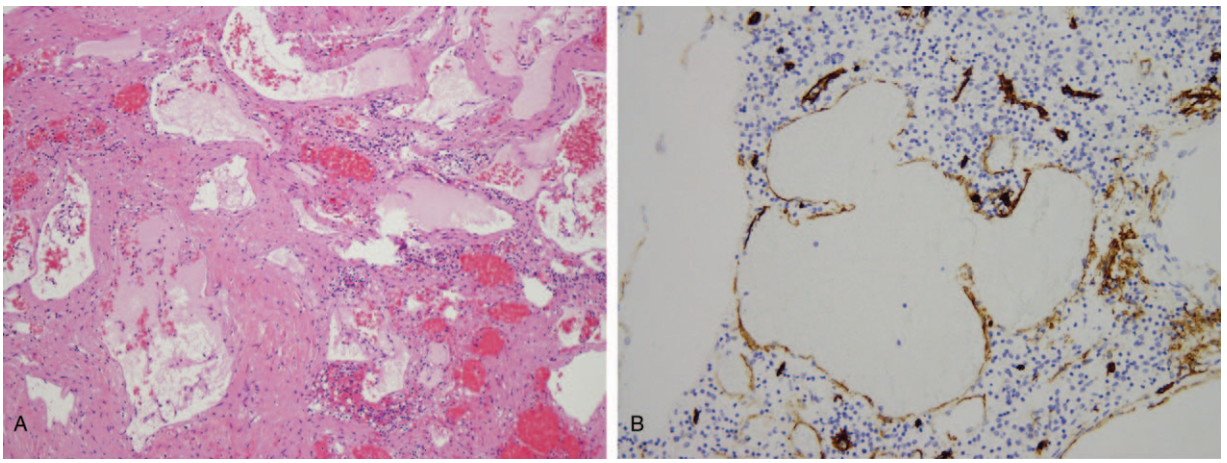

Figure 5. (A) Hematoxylin and eosin stain of multiple cystic spaces lined by flat endothelium histologically (×40). (B) Immunohistochemistry of CD31 (×100).

Bisceglia et al mentioned that <150 cases of both systemic and isolated lymphangiomatosis of the spleen have been reported until the end of 2013.[4]

Lymphangiomatosis may also involve bones, which was first described by Gorham in 1954 (named Gorham disease [GD]),[13] and this disease was classified as a distinct entity by Gorham and Stout in 1955.[14] Splenic cystic lymphangiomatosis could also be a manifestation of Klippel–Trenaunay–Weber syndrome, including a triad of port-wine stain of the skin, abnormalities of the venous and lymphatic systems, and hypertrophy of the soft tissue and bone involving an extremity.[15,16]

Histologically, lymphangiomas can present as solitary nodules, multiple nodules, or diffuse growth. It frequently consists of multiple cysts of varying sizes lined by flat endothelial cells, which contain eosinophilic serous fluid.[7] Based on the size of the dilated lymphatic channels, lymphangiomatosis can be classified as capillary (super-microcystic), cavernous (microcystic), or cystic (macrocystic).[7] The symptoms of lymphangiomatosis may be asymptomatic, left upper quadrant pain, sense of fullness, and abdominal distension. It may be complicate by coagulopathy, hypersplenism, bleeding or splenic rupture, and portal hypertension.[4] The relationship between hypersplenism and lymphangiomatosis is not well established, but Wadsworth et al showed the a child's hypersplenism-related symptoms, such as splenomegaly, thrombocytopenia, and progressive anemia, were resolved after splenic embolization.[3] An abdominal sonography usually reveals hypoechoic or anechoic cysts with possible internal debris depending on the size of the lymphatic spaces and protein content.[1,7] CT scan shows multifocal, well-demarcated, thin-walled, low-attenuating cysts with sharp margins.[2] The differential diagnosis includes true epidermoid cysts, parasitic cysts, mesothelial cysts, and hemangioma.[17] True splenic cysts will have definite epithelial lining, and the parasitic cysts of the spleen are due to hydatid disease cause by *Echinococcus granulosus*.[18] A serologic test for *Echinococcus* should be performed to exclude the diagnosis of hydatid cyst, but there is no available commercial kit in Taiwan. However, there was no hydatid sand in the splenic cystic lesions by the abdominal CT, and histology of the spleen did not show germinative layer, granulomatous palisading reaction, and scolices in this patient.[19] The splenic specimen showed typical findings of lymphangiomatosis, including multifocal cystic space lined by endothelial cells and proteinaceous content. Hemangiomas are also characterized by vessels lined by endothelium, but they are filled with red blood cells.[7] The endothelial lineage could be further demonstrated by immunohistochemistry of vascular markers, such as CD31.[4]

In our patient, the abdominal CT showed splenic cystic lesions with splenomegaly causing external compression and varices in the upper abdomen, and a preoperative diagnosis can be suspected. On the contrary, soft tissue lymphangiomatosis and GD may be confused with metastasis or hematologic malignancies in imaging studies.[20,21] Therefore, the definite diagnosis should be based on histology. Histology revealed that splenic parenchyma had multifocal, multiple cystic spaces lined by endothelium containing eosinophilic proteinaceous fluid in our patient, and these are the typical findings of splenic lymphangiomatosis.[7] Recently, a Kaposiform lymphangiomatosis was described, and it consistently affects the lung with the possible simultaneous involvement of the spleen.[22] However, Kaposiform lymphangiomatosis was reportedly composed of endothelial spindle cells which were different from the cystic lymphatic proliferations in the lymphangiomatosis in our patient. Lymphangiomatosis involving the spleen may be part of multi-visceral involvement.[5] In our patient, the CT revealed multiple splenic cysts without pulmonary, hepatic, and visible skeletal involvement, and only a right renal simple cyst was noted. Physical examination showed normal findings of lymph nodes over the neck, axillary, and inguinal areas. To our knowledge, this is the first case presenting with splenic cystic lymphangiomatosis in association with omental varices and portal hypertension.

Unlike smaller or localized lymphangiomas that can be treated by partial splenectomy, lymphangiomatosis warrants total splenectomy because of total splenic parenchymal involvement.[23] Leaving a splenic remnant in cases of total splenic involvement increases the risk of recurrence.[24] Splenic cystic lymphangiomatosis is a benign entity, but a malignant lymphangioma was also reported.[25] Therefore, we chose splenectomy for completely removing this giant-sized spleen with unknown etiology, preoperatively. Laparoscopic splenectomy has been a well-established procedure for patients with benign or malignant hematogenous disease, splenic cyst, and selected cases with splenomegaly.[26,27] In comparison with the open technique, laparoscopic splenectomy is associated with longer operative times, reduced blood loss, less wound pain, and shorter length of stay.[26] However, if the maximal diameter is larger than 15 cm, the intraabdominal space will be significantly restricted and the laparoscopic approach can be very challenging.[27] In addition, en block excision for a tumor lesion with unknown etiology is a basic surgical principle for arriving at the correct pathogenic result. The spleen plays an important role in hemostasis, immune response, and infection prevention,[28] and autologous spleen transplantation after total splenectomy is a possible method for preserving splenic function.[29] Unfortunately, definite diagnosis was unknown preoperatively, and the patient had limited orthotopic and vascularized spleen (<25%) required for autologous transplantation.[30] Therefore, we chose conventional splenectomy without autologous transplantation for this patient.

4. Conclusion

In summary, we reported a rare case of splenic cystic lymphangiomatosis with splenomegaly in association with omental varices and portal hypertension. The diagnosis is based on history, imaging findings, and pathology. Conventional total splenectomy was performed in this patient because of a large spleen and preoperative unknown etiology, and autologous spleen transplantation was not performed because of limited orthotopic and vascularized spleen. Splenic cystic lymphangiomatosis should be listed in the differential diagnosis in patients with a palpable painless mass over the left hypochondrium.

Author contributions

Conceptualization: Wei-Fan Hsu, Chun-Chieh Yeh.
Data curation: Wei-Fan Hsu.
Writing – original draft: Wei-Fan Hsu.
Writing – review & editing: Wei-Fan Hsu, Cheng-Ju Yu, Chun-Chieh Yeh, Wen-Hsin Huang, Cheng-Yuan Peng.

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