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Spontaneous regression of a cystic peripancreatic tumor in a 12-year-old boy: A case report☆

Satoshi Yokoyama*, Akinori Sekioka, Hirofumi Utsunomiya

Department of Pediatric Surgery, Japanese Red Cross Society, Wakayama Medical Center, 4-20 Komatsubara-dori, Wakayama 640-8558, Japan

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ABSTRACT

We present a case of spontaneous regression of a cystic peripancreatic tumor in a 12-year-old boy. The cystic tumor was initially suspected to be a pancreatic pseudocyst associated with traumatic pancreatic damage. However, the differential diagnosis included the possibility of lymphatic malformation in view of the clinical and image findings. In anticipation of spontaneous regression, the patient has been followed without treatment. Elective drainage is available but fortunately the cyst has shown no sign of expansion. Close surveillance may be a treatment option for a cystic peripancreatic tumor.

Retroperitoneal cystic tumors are rare. Cystic lymphatic malformation is one of the more common intra-abdominal cystic tumors; the differential diagnosis includes cystic tumors of mesothelial, enteric or urogenital origin; dermoid cysts or teratomas; and pseudocysts from trauma or infectious origins. Outcomes following complete resection of retroperitoneal lymphatic malformation are generally good, however, patients with retroperitoneal lymphatic malformation are more likely to have incomplete excision of the cysts, and therefore have a higher rate of recurrence. We present a case of spontaneous regression of a cystic peripancreatic tumor in a 12-year-old boy.

1. Case report

A 12-year-old boy presented to the emergency room of our hospital because of two days of high fever with worsening abdominal pain associated with nausea and vomiting. Two days prior to admission he had sustained minor trauma to his upper abdomen during a quarrel with his brothers. He developed a high fever and moderate abdominal pain following the incident. At presentation, he complained of worsening abdominal pain. His temperature was 38.0 °C. Bowel sounds were faint. Palpation of his left upper abdomen produced pain without guarding or rigidity. Laboratory data showed hemoglobin 13.2 mg/dL, leukocytosis (18,600/mm3) and a remarkably increased C-reactive protein level (16.8 mg/dL). Serum and urine amylase were both in the normal range. An abdominal X-ray film showed caudal displacement of the transverse colon (Fig. 1). Contrast-enhanced computed tomography (CT) of the abdomen revealed a large cystic mass without calcifications compressing the lesser sac in the gastrosplenic space, and disruption of the continuity of the pancreatic body, suspicious for a pancreatic laceration (Fig. 2a–d). The patient was diagnosed with a traumatic pancreatic pseudocyst due to blunt trauma to the abdomen. He was treated conservatively with the administration of a broad-spectrum intravenous antibiotic (meropenem at 1500 mg/day) and total parental nutrition. The patient began to improve clinically with conservative management.

On the third day after admission, an ultrasound (US) scan revealed a complex cystic mass in the region of the gastrosplenic space (Fig. 3). The patient underwent magnetic resonance imaging (MRI) of the upper abdomen, as well as magnetic resonance cholangiopancreatography (MRCP). MRI showed low signal intensity in T1-weighted sequences and on T2-weighted MRI, a high signal intensity cystic mass with septations was seen occupying the left upper quadrant of the retroperitoneum (Fig. 4a–c).

MRCP revealed an intact pancreatic body and tail with adjacent fluid collections, but no clear evidence of main pancreatic duct transection (Fig. 5). The patient had an uneventful recovery and after seven days in the hospital he resumed p.o. intake. The elevated infection/inflammation laboratory parameters normalized. A repeat

* Corresponding author. Tel.: +81 73 422 4171; fax: +81 73 426 1168.
E-mail address: yokoyama.st@gmail.com (S. Yokoyama).

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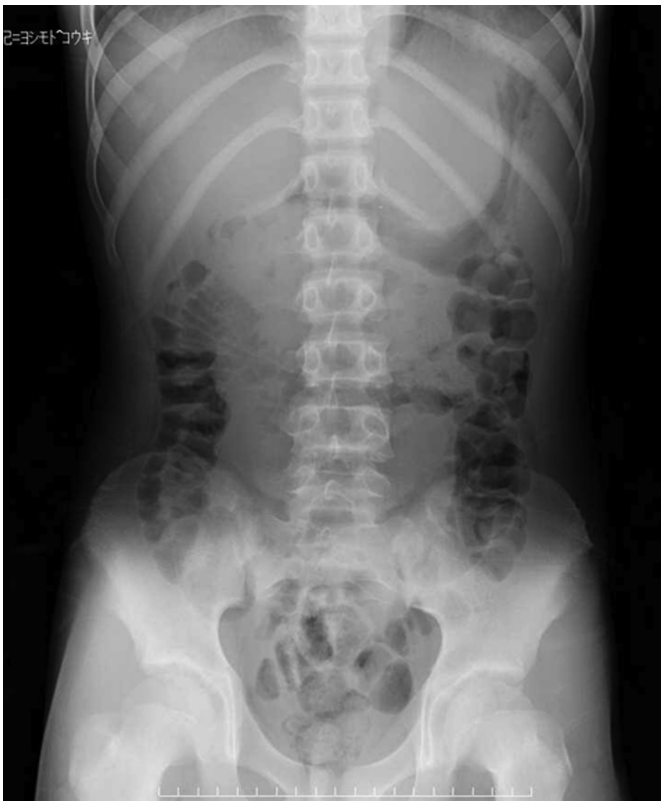

Fig. 1. An abdominal X-ray film showed caudal displacement of the transverse colon.

CT scan on the 12th day after admission showed the cystic mass was slightly reduced in size (Fig. 6). The patient was discharged from the hospital 14 days after admission.

A follow-up MRI scan four months later revealed that the cystic mass was no longer visible. The inside of the cyst did not show signals of blood (usually high T1 signal intensity) but rather of water (usually low T1 and high T2 signal intensity) (Fig. 7). This lesion regressed spontaneously without surgical treatment. Although the diagnosis was not histologically confirmed, a tentative diagnosis of a retroperitoneal cystic lymphatic malformation was finally made based on the imaging findings. On follow-up at 3 years, the patient was well and did not have abdominal complaints.

2. Discussion

Retroperitoneal cystic tumors are rare. Cystic lymphatic malformation is one of the more common intra-abdominal cystic tumors; the differential diagnosis includes cystic tumors of mesothelial, enteric or urogenital origin; dermoid cysts or teratomas; and pseudocysts from trauma or infectious origins.

Lack of rational nomenclature and confusing terminology of vascular lesions has often led to inappropriate diagnosis and treatment. In 1982, Mulliken and Glowacki proposed a classification system of vascular lesions referring to the endothelial characteristics and clinical behavior. This classification divides vascular anomalies into 2 categories: tumors and vascular malformations [1]. Regarded as an abnormality of morphogenesis rather than as a neoplasm, lymphangiomas are now referred to as *lymphatic malformations*.

A lymphatic malformation is a benign congenital anomaly resulting from anomalous lymphatic tissue with the potential to invest surrounding structures [2]. These lymphatic malformations are found in the head, neck, and axilla in 95% of cases, but approximately 5% are diagnosed within the intra-abdominal cavity with the retroperitoneum as the most common location [2–4]. Trauma, infections, surgery, or radiation therapy are generally considered responsible for their pathogenesis in acquired type of lymphatic malformations due to obstruction of lymphatic system [5].

Lymphatic malformations are usually asymptomatic, however the acute presentation of lymphatic malformations can cause abdominal pain, tenderness, distension, fever, leukocytosis, peritonitis, dysuria, and garding. Lymphatic malformations of the retroperitoneum are usually diagnosed in older children or adults [5–8].

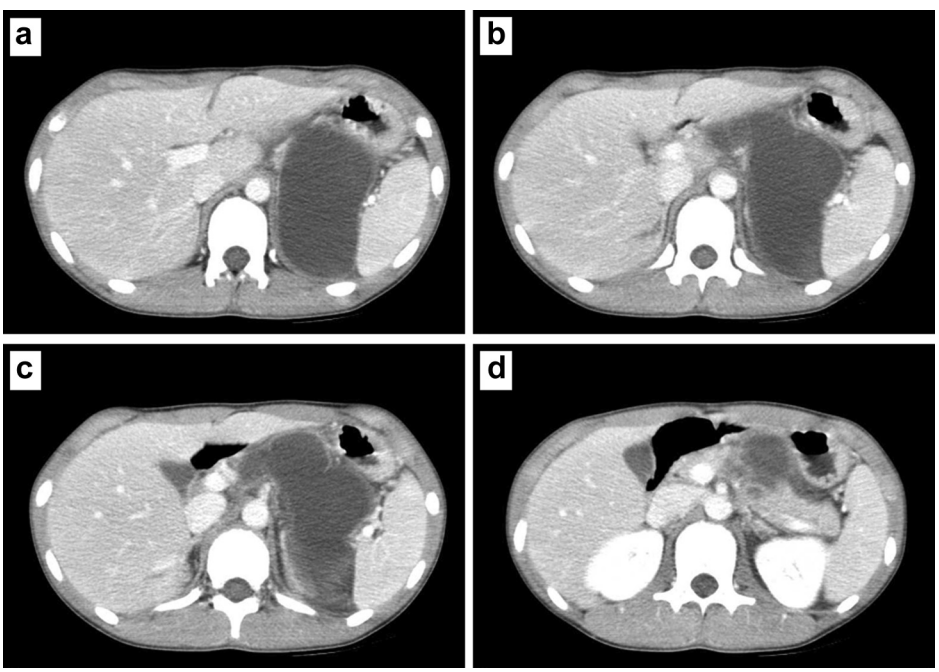

Fig. 2. (a–d): Contrast-enhanced CT of the abdomen revealed a large cystic mass without calcifications compressing the lesser sac in the gastrosplenic space, and disruption of the continuity of the pancreatic body, suspicious for a pancreatic laceration.

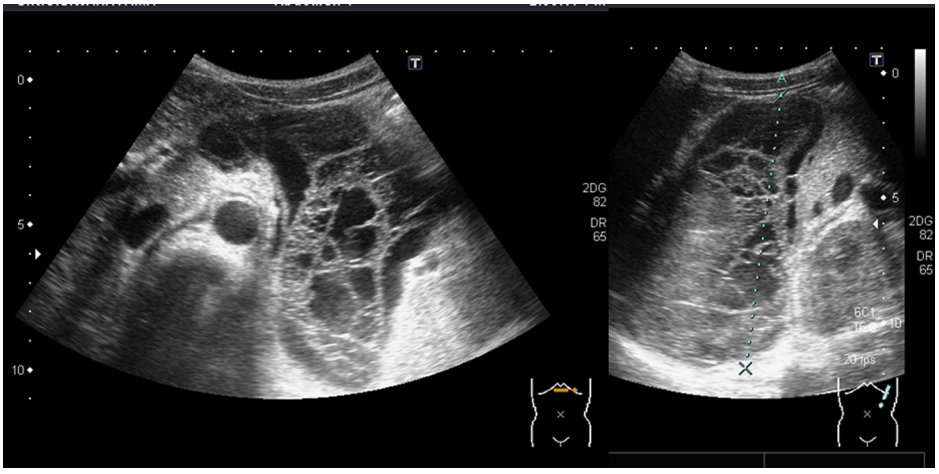

Fig. 3. An US scan revealed a complex cystic mass in the region of the gastrosplenic space.

The specific diagnosis of retroperitoneal lymphatic malformation is challenging. Assessment by diagnostic imaging, including US, CT, and/or MRI, is very important for the diagnosis of a lymphatic malformation. Diagnostic important point for cystic lymphatic malformation is an elongated shape and acrossing from one retroperitoneal compartment to an adjacent one. Also at CT, cystic lymphatic malformation typically appears as a large, thin-walled, multiseptate cystic mass [6,7]. Evaluation with US demonstrates thin walled, multi-septated structures without peristalsis, and Doppler US demonstrates no blood flow within the cystic walls [3]. MRI may better define the relationship between bowel, vasculature, other soft tissues, and the cystic walls of a lymphatic malformation. The mass typically surround and compress normal structures. Low signal intensity is seen on T1-weighted [3,8].

There are two main treatment options for lymphatic malformations; surgical excision and sclerotherapy. Surgical excision has historically been the treatment of choice, however, sclerotherapy affords a minimally invasive therapeutic option or adjunct for these malformations when vital structures may be at risk with complete resection [9]. When possible, the treatment of choice is complete surgical resection. It is recommended as soon as possible after the diagnosis is established, because complications such as infection, hemorrhage, and bowel obstruction can occur. As time passes, growth of the cyst might prevent complete removal [6,7,10]. Other treatment modalities such as drainage, and irradiation give poor results [11–13]. Sclerotherapy is recommended for unresectable lymphangiomas. Doxycycline is a sclerosing agent recently used in treating lymphatic malformations [14]. Generally, patients with retroperitoneal cysts are more likely to have incomplete excision of the cysts, and therefore have a higher rate of recurrence. When considering surgical treatment of lymphatic malformations, it must be kept in mind that these are benign malformations and that injury to important adjacent structures should be avoided. Abdominal lymphatic malformation usually involves adjacent organs, thereby making it difficult to resect the cysts completely without a combined resection. The risk of short bowel syndrome and possibility of damage to vital organs must be balanced with the benefit of aggressive excision.

In our case, the lack of a histologic diagnosis is a limitation. The cystic tumor was initially suspected to be a pancreatic pseudocyst, largely on the basis of reported incidence. However,

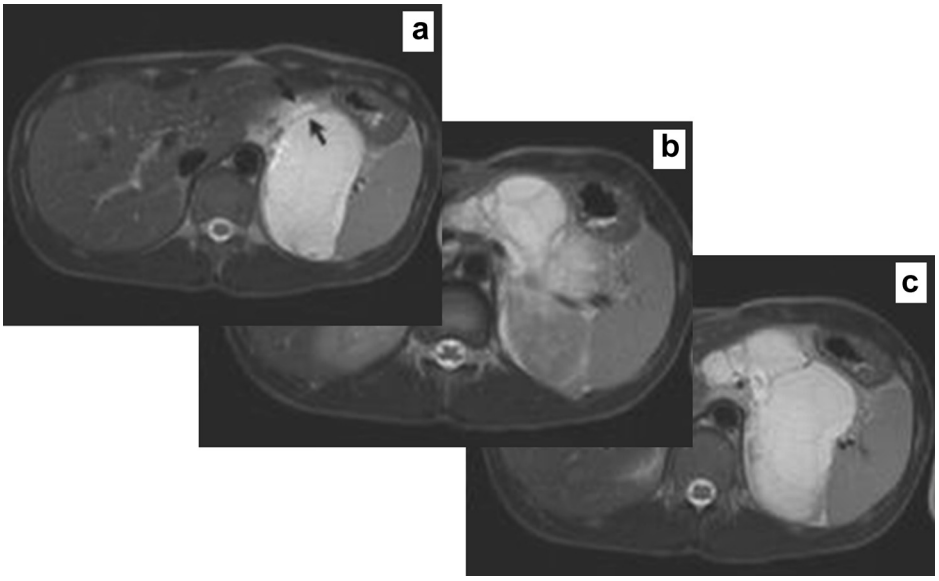

Fig. 4. (a–c): On Axial T2-weighted MRI, a high signal intensity cystic mass with septations was seen occupying the left upper quadrant of the retroperitoneum. Arrow: left gastric artery and vein.

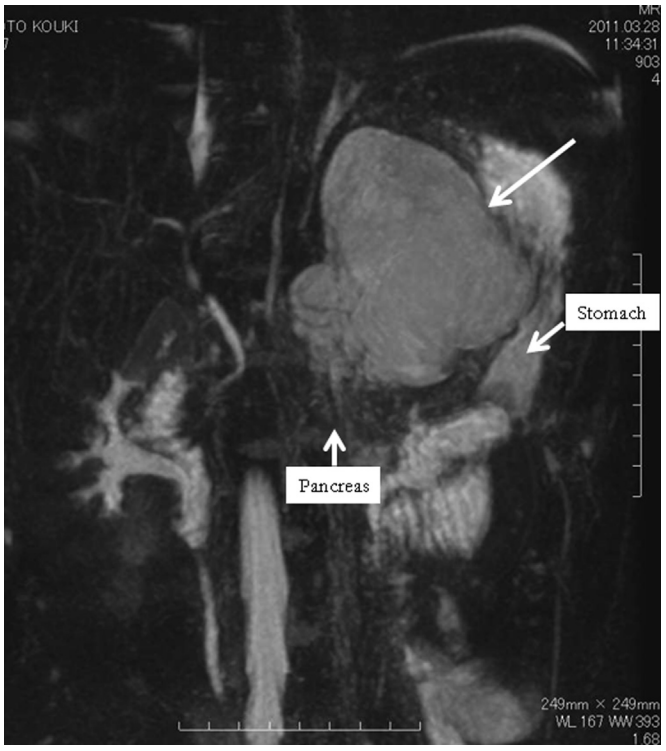

Fig. 5. MRCP revealed an intact pancreatic body and tail with adjacent fluid collections, but no clear evidence of main pancreatic duct transaction. Arrow: cystic mass.

the differential diagnosis included the possibility of lymphatic malformation in view of the following clinical and image findings [1]: CT scans on admission suggested a traumatic pancreatic pseudocyst; however, US, MRI and MRCP performed 3 days later revealed the presence of an irregular multiloculated mass at the pancreatic head. Although the mass was in direct contact with the pancreas, the pancreatic parenchyma remained intact and the mass did not exhibit significant changes of clinical concern despite its size. [2] On the third day after admission, MRI provided the inside of the cyst did not show signals of blood (usually high T1 signal intensity) but rather of water (usually low T1 and high T2 signal intensity).

[3] Serum amylase remained normal throughout patient monitoring. Although the cause of the cystic lymphatic malformation is unknown, we suggest two possibilities. It may have been the result of lymphatic obstruction due to previous abdominal trauma, or, more likely, a previously undiagnosed lymphatic malformation was revealed by acute trauma and secondarily infection.

Lymphatic malformations rarely undergo malignant change or spontaneous regression. Although lymphatic malformations are benign lesions, spontaneous regression is rarely seen and only 12.5–15.0% respectively [15]. Spontaneous regression of nuchal or mediastinal lymphangioma has been reported previously, but regression of an intra-abdominal lymphatic malformation, like our case, is very uncommon. Two reports of a total of three cases of spontaneous regression of a intra-abdominal cystic lymphangioma have been described before [16,17]. What all three cases had in common is that these were all postpartum females. In our case, there is no known theory regarding the spontaneous regression of cystic retroperitoneal lymphatic malformations. Although the diagnosis was not histologically confirmed, our case suggests that close surveillance may be a treatment option for a cystic peripancreatic tumor.

3. Conclusion

The cystic tumor in this patient was considered to have been caused by lymphatic malformation, which has a very low prevalence. We did not clarify whether the lymphatic malformation was congenital or acquired due to lymphatic obstruction resulting from the trauma or inflammation. In anticipation of spontaneous regression, the patient has been followed without treatment. Elective drainage is available but fortunately the cyst has shown no sign of expansion. Therefore, observation monitoring may be appropriate in some cases.

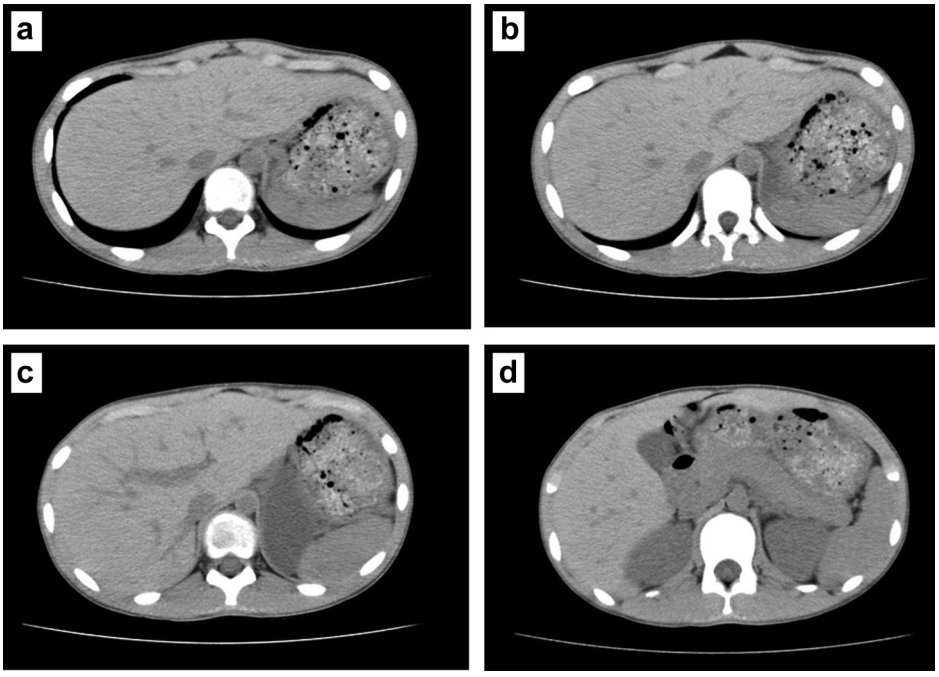

Fig. 6. A repeat CT scan after admission showed the cystic mass was slightly reduced in size.

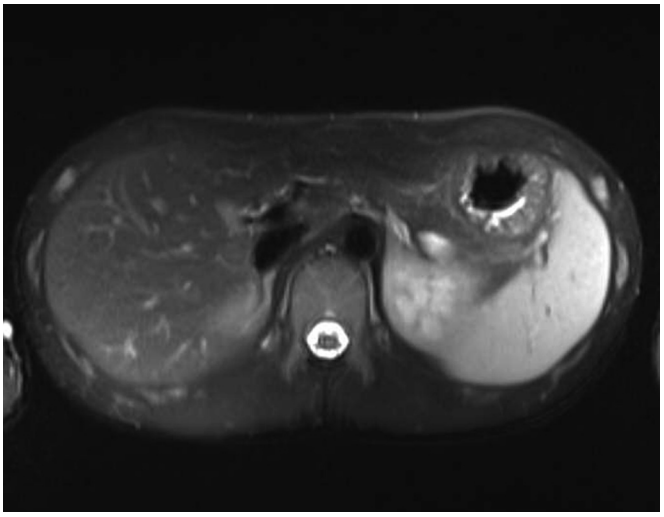

Fig. 7. A follow-up CT scan revealed that the cystic mass was no longer visible.

Acknowledgments

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