A giant hemolymphangioma of the pancreas

A case report and literature review

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Abstract

Rationale: Hemolymphangioma of the pancreas is an extremely rare benign tumor; only 10 patients with this disease have been reported to date, the majority of whom were women.

Patient concerns: We describe a 28-year-old man who presented with abdominal pain and discomfort. Computed tomography and magnetic resonance imaging data showed a huge heterogeneous solid cystic mass at the retroperitoneal pancreatic head. The maximum cross-sectional lengths of the pancreatic lesion were approximately 12 × 8.5 × 12 cm³.

Diagnosis: Hemolymphangioma of the pancreas.

Interventions: The patient underwent a pylorus-preserving pancreatectoduodenectomy; surgical excision is the main treatment for this type of tumor.

Outcomes: The patient followed up regularly in the outpatient department for 6 months after surgery, and no sign of recurrence was found.

Lessons: Although it is uncommon, clinicians ought to consider the diagnosis of hemolymphangioma of the pancreas upon detection of a pancreatic cystic lesion exhibiting fat or calcification.

Abbreviations: CT = computed tomography, DWI = diffusion-weighted imaging, MRI = magnetic resonance imaging, T1WI = T1-weighted imaging, T2WI = T2-weighted imaging.

Keywords: hemolymphangioma of the pancreas, MRI, surgical resection, uncommon benign tumor

1. Introduction

Hemolymphangiomas are rare benign tumors arising from the mesenchyme that comprise malformed lymphatic vessels and venules. They mainly occur in the head and neck region and are less commonly found in the pancreas, spleen, small intestine, and vermiform appendix.[1,2] There have only been 10 reports of hemolymphangiomas arising in the pancreas to date, which attests to their rarity. The major symptoms of hemolymphangiomas are atypical and can thus be easily confused with other cystic-solid or cystic tumors of the pancreas; this greatly complicates the preoperative diagnosis of this benign tumor.[3]

In this study, we analyzed the magnetic resonance imaging (MRI) and computed tomography (CT) findings in a patient with hemolymphangioma of the pancreas, and also reviewed the relevant literature in an effort to improve the accuracy of diagnosing this disease using imaging.

2. Case report

The study was approved by the ethics committee of Taizhou People’s Hospital (Jiangsu, China) and informed consent was obtained from the patient. A 28-year-old man was hospitalized for right upper abdominal pain, he had experienced for approximately 2 days. Physical examination revealed a soft abdomen, no abdominal varicose veins, and a mass approximately 8.0 × 10.0 cm³ at the right upper quadrant that had an unclear border, poor activity, and tenderness (but no rebound tenderness). No abnormalities were found on laboratory testing. Plain CT showed a cystic-solid mass of mixed density with a size of approximately 12 × 8.5 × 12 cm³ in front of the right kidney and behind the pancreatic head. There was a small amount of adipose tissue and scattered spot-like calcified shadows with a CT value of 15 to 30 Hounsfield units. Enhanced CT showed that the solidified part of the lesion was slightly strengthened, and the lesion’s boundary with the surrounding adipose tissues was unclear as it partially wrapped around the duodenum (Fig. 1).

MRI showed a huge oval heterogeneous mass in the right rear of the pancreatic head, with slightly high intensity and scattered low intensity on T1-weighted imaging (T1WI) and high/low mixed intensity on T2-weighted imaging (T2WI). Dynamic enhanced MRI revealed mild enhancement of the mass with a slight delay to strengthen the signal; the head of the pancreas was deformed and pushed anteriorly with no expansion of the main pancreatic duct. The lesion was considered a retroperitoneal mature liposarcoma or ganglioneuroma because of the presence of adipose tissue (Fig. 2).

For surgery, a median incision was made at the upper abdomen after lumbar anesthesia. There was no effusion in the abdominal cavity, no implantable nodules in the pelvic peritoneum, and no
swollen lymph nodes in the hepatoduodenal ligament. Pylorus-preserving pancreatectoduodenectomy was performed to remove the tumor, which was approximately 12 cm in diameter with a complete capsule located at the pancreatic head.

As for pathological findings, expanded lymphatic vessels and vascular-like structures were visible on microscopy, especially in the former. Amorphous necrosis in the lesion was accompanied by multinucleated giant cell hyperplasia (Figs. 3 and 4). Immunohistochemistry staining results were as follows: cytokeratin (−), epithelial membrane antigen (−), CAM5.2 (−), S-100 (−), carcinoembryonic antigen (−), vimentin (+), Wilms’ tumor susceptibility gene 1 (−), CD34 (+), D2-40 (+), CD31 (−), and Ki-67 (+). The final pathological diagnosis of the lesion was a hemolymphangioma at the pancreatic head.

3. Discussion

Hemolymphangioma that arises from the mesodermal tissue is an extremely rare deformity involving lymphatic vessels and microvessels. The pathogenesis of this disease is not completely understood, and can be attributed to congenital vascular dysplasia, dysplastic lymphatic vessels, and venous lymphatic occlusion.\cite{1-5} Pathologically, a hemolymphangioma with veins and lymphatic vessels is composed of polycystic lymphatic and blood vessels; these cysts are partially connected to, and enveloped in, the endothelium. The tumors are often positive for D2-40, CD31, and CD34. D2-40 is specifically expressed in lymphatic endothelial cells, while CD31 and CD34 are specifically expressed in vascular endothelium; CD34 is stably expressed in tumor capillaries and capillary endothelial cells, and is a sensitive marker for these tissues.\cite{6-10} D2-40 and CD34 expressions were positive in our patient’s tumor, which supported the diagnosis of hemolymphangioma of the pancreas. Hemolymphangioma can occur in any part of the body. To date, limited cases of hemolymphangioma have been reported in the pancreas, limbs, spleen, and other organs. The first patient was reported by Couinaud et al\cite{7} in 1966, and was also located in the pancreas. A literature review using PubMed through January 31, 2018, using the keywords “pancreas and hemolymphangioma” revealed only 10 publications describing as many cases in total.\cite{1-5,7-12} As shown in Table 1, this tumor type is more common in women (n = 9) than in men (n = 2). Its major symptoms are atypical and include abdominal pain, distention, and discomfort.\cite{1,12} Some nonspecific symptoms are diarrhea and weight loss,\cite{9} as well as gastrointestinal bleeding.\cite{10} Generally, laboratory data reveal no abnormal findings.

Hemolymphangioma of the pancreas is a single, polycystic or cystic-solid mass with enlarged size; its cystic structures may be attributed to the continuous rupture and fusion of tumor microvessels and lymphatic vessels. Pathological findings show a large amount of lymphatic fluid as well as a small number of lymphocytes and erythrocytes in the cystic cavity. Lymphocyte infiltration is detectable between the vessel walls and septum, which are composed of endothelial cells.\cite{3} CT revealed the cystic area of the lesion without enhancement; the solid part, cystic walls, and septum were visible with no, mild,\cite{11,12} or uneven enhancement.\cite{11} This indicates that enhancement is related to the proportion of vessels and lymphatic vessels in the lesion, while nonobvious enhancement may be related to a slow blood flow within the vessels with reduced or atypical hyperplasia.\cite{3} A report by Figueroa et al\cite{11} detailed a patient misdiagnosed with hemangioma using enhanced CT. In our patient, the lesion was a cystic, noncompartmentalized entity, and the solid portion was visible on CT with mild enhancement. There was also a small amount of adipose tissue and spot-like calcified nodes, which have not been described in patients reported previously.

To date, only 2 cases of hemolymphangioma of the pancreas have been detected by MRI, with a low signal from the cystic component on T1WI and a high signal on T2WI.\cite{11,12} Low/high signals of the cystic component on MRI are related to the proportion of lymphatic vessels or vascular composition in the cystic fluid as well as hemorrhage. Diffusion-weighted imaging (DWI) offers a differential diagnosis of lesions; in our patient, the lesion contained a small amount of adipose tissue and calcified nodes. MRI showed mainly high signals on T2WI accompanied with some low and high/low mixed signals for fat suppression, homogeneous signals on T1WI, and mixed high signals on DWI. Dynamic enhanced MRI revealed mild enhancement of the mass with a slight delay to strengthen the signal. In this case, the
A hyperintensity region on T1WI after enhancement was considered to represent veins with slow-flowing blood.[3]

Clinical manifestations are nonspecific for a hemolymphangioma of the pancreas. Although imaging findings have certain features, it remains difficult to distinguish this type of tumor from a pancreatic cystadenoma, carcinoma, or solid pseudopapillary tumor. Cystadenoma is more common in older women, and often involves the tail of the pancreas and presents as a single cystic or polycystic low-density shadow(s). The solid component of the cystadenoma is obviously strengthened on contrast-enhanced CT. Solid pseudopapillary tumors of the pancreas, which exhibit borderline malignancy, often occur in young women, and mostly present as a cystic mass with polished edges, calcification, and uneven, delayed enhancement of the solid component on contrast-enhanced CT. Most patients with pancreatic pseudocysts have a history of pancreatitis and present with elevated amylase levels. A pancreatic pseudocyst is a single cystic mass with a thickened wall that can be strengthened on contrast-enhanced CT. A fuzzy peripancreatic fat gap and thickened prerenal fascia can also be visualized on CT. Hemolymphangioma of the pancreas, which is anatomically difficult to reach owing to its large size, should be distinguished from pancreatic cystadenoma.

Figure 2. Magnetic resonance imaging (MRI) revealed an oval-shaped tumor of mixed signal intensity behind the pancreatic head on both T1-weighted imaging (WI) and T2WI with visible mild dynamic enhancement with a delay to strengthen the signal. T2WI with fat suppression showed small areas of low signal. (A) Plain MRI; (B) arterial phase MRI; (C) venous phase MRI; (D) T2WI. Arrows indicate pancreatic cystic lesion.
cystadenoma and from retroperitoneal and other abdominal tumors. Our patient was initially misdiagnosed with retroperitoneal lesions following imaging examinations because the pancreas appeared to be pressured and a small amount of adipose tissue was present. As stated above, another case of hemolymphangioma of the pancreas was misdiagnosed as a hemangioma of the left hepatic lobe.[1]

Surgical resection is the main treatment for this disease, including tumor resection or partial pancreatectomy.[4] Pancreatoduodenectomy is routine for patients with no preoperatively identified diagnosis.[9,10] There was only 1 patient with hemolymphangioma in the spleen who was reported to have undergone laparoscopic surgery.[13] Laparoscopic surgery on hemolymphangioma of the pancreas has not been reported to date. All reported patients have good prognoses with no recurrence until now, but 1 case was reported to have duodenal invasion and gastrointestinal bleeding. Therefore, regular follow-up with imaging is necessary.[11] Although uncommon, clinicians should still consider hemolymphangioma of the pancreas during differential diagnosis upon the presence of adipose tissues or calcification in a pancreatic cystic lesion.

**Author contributions**

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Investigation: Jianguo Xia.
Methodology: Jianguo Xia.
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