CASE REPORT

An Unusual Cause of Acute Abdomen in Adults:
Giant Cystic Lymphangioma of the Pancreatic Head.
A Clinical Case and Literature Review

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ABSTRACT

Context Cystic lymphangioma of the pancreas presenting as acute abdomen in adults has not been reported before. Case report We report the case of a young man who presented with severe pain in the upper abdomen and abdominal swelling. On imaging, he was found to have a giant multiseptate cystic lesion occupying almost the entire abdomen anterior to the pancreas. On exploration, a cystic mass involving the head of the pancreas and densely adherent to the antrum, and the second and third parts of the duodenum was found and a classic Whipple pancreaticoduodenectomy was done. Histology revealed a diagnosis of cystic lymphangioma.

Conclusion Cystic lymphangioma is a rare benign tumor of the pancreas and this is the first reported case in an adult presenting with acute abdomen. Though rarely diagnosed preoperatively, this entity should also be considered in the differential diagnosis of cystic lesions of the pancreas. Complete surgical excision is curative.

INTRODUCTION

There are many causes of acute abdomen; however, cystic lymphangioma of the pancreas presenting as acute abdomen in adults has not been reported before. After a thorough search of PubMed (http://www.ncbi.nlm.nih.gov/pubmed/), we did not find any adult patient with this presentation although there are many case reports in the pediatric age group. We report a young man with a cystic lymphangioma of the head of the pancreas who presented with acute pain in the abdomen and underwent a Whipple pancreaticoduodenectomy.

CASE REPORT

A 20-year-old man with no previous history presented with the sudden onset of severe, upper abdominal pain radiating to the back of two days duration. The pain was relieved temporarily with analgesics and was associated with fever, chills and rigor. There was no history of alcoholism, jaundice, vomiting, obstipation or gastrointestinal bleeding. After careful questioning he admitted having noticed gradual fullness of the abdomen for 3 months prior to presentation.

Clinical examination revealed abdominal swelling with generalized tenderness occupying the whole abdomen and almost reaching up to the pelvis. Blood investigation revealed polymorphonuclear leukocytosis (total leukocyte count 12,600 mm⁻³, reference range: 4,000-11,000 mm⁻³; polymorphs 91%, reference range: 40-75%). The serum amylase level was 297 U/L (reference range: 20-85 U/L). Liver function tests and CA 19-9 levels were normal. An ultrasound carried out outside our hospital found a large septate cystic collection in front of the pancreas and reported as acute pancreatitis with fluid collection. A CT scan revealed a large multiseptate cystic lesion (23.2x11.7x26.2 cm) anterior to the pancreas reaching up to the pelvis and displacing the gut loops behind and downwards (Figures 1 and 2). We decided to perform a laparotomy with a presumptive diagnosis of cystic neoplasm of the pancreas.

At laparotomy the peritoneal cavity was found to be fully occupied by a multicystic, septate mass (approximately 30x50 cm) containing a clear yellowish fluid, arising from the head of the pancreas, adherent to
the transverse colon, mesocolon, duodenum and stomach (Figure 3). The mass was inseparable from the antrum of the stomach, the duodenum and the head of the pancreas. There were no ascites or peritoneal or liver nodules. The mesenteric and peripancreatic lymph nodes were enlarged. A classic Whipple pancreaticoduodenectomy was carried out (Figure 4).

Postoperatively, the patient had a bile leak in the drain which gradually decreased and the drain was removed on postoperative day 16. He was started on a jejunostomy feed from postoperative day 3 and a normal diet from day 8.
Histopathological examination showed large septate lymphatic channels in a background of connective tissue stroma. The tumor contained serous fluid. The stroma had multiple cholesterol crystals, lymphocytes, smooth muscles and congested thin walled vessels. The lymph nodes showed reactive changes. The final histological diagnosis was of cystic lymphangioma of the pancreas (Figures 5 and 6). The surrounding pancreas parenchyma was unremarkable. The patient is doing well 12 months postoperatively.

DISCUSSION

Cystic lymphangioma is a rare malformation described for the first time by Koch in 1913 [1]. It probably occurs as a result of a congenital malformation of the lymphatics resulting in obstruction of the local lymph flow and the development of lymphangiectasia [2]. Cystic lymphangiomas are usually benign but can be locally invasive [3]. They are found predominantly in the pediatric population [4] and commonly located in the neck and axillary regions [5, 6]. A variety of other sites have been described including the mediastinum, pleura, pericardium, groin, bones and abdomen. In the abdomen, the mesentery and retroperitoneum are the most common sites, and this tumor rarely develops in the pancreas (1% of all lymphangiomas) [4, 7]. Until 2008, there were only about 70 reported cases of pancreatic cystic lymphangiomas [8]. As a result, reports in literature are limited to case reports with very few case series. The pediatric age group is most commonly affected with females outnumbering males [4]. Clinical presentation varies according to age group with children having a more acute presentation ranging from three days to two months and ranging from two weeks to one year in adults [4]. Our patient presented with acute pain in the abdomen which had not been reported before in the literature in adults. The cause of this acute presentation is not clear; the patient had fever and chills with an increased total leukocyte count and neutrophil count. Upon cutting open the specimen after resection there was clear yellowish fluid inside the cyst, not suggestive of infection or recent hemorrhage. A cell count of this cystic fluid was not carried out. On histology there was no evidence of polymorphonuclear cell infiltration and the surrounding pancreas did not show any evidence of pancreatitis.

Figure 3. Operative photograph showing the tumor.

Figure 4. Post-resection operative site.

Figure 5. Cut section of the tumor showing multiseptate cystic spaces.

Figure 6. Hematoxylin and eosin (x100) staining shows dilated lymphatic channels of varying sizes, separated by thin septa (black arrows). The cystic spaces are lined with flattened or cuboidal endothelial cells.
Awareness of a lump and pain in the abdomen are the most common presenting features. It is seldom associated with pancreatitis, weight loss and laboratory abnormalities [9]. A female preponderance and the huge size of the swelling may lead to the mistaken impression of an ovarian cyst [8]. A case report of this rare condition associated with blue rubber bleb nevus syndrome has also been described [10]. Most reports in the literature describe the body and tail of the pancreas as the most common regions involved [11]. There have only been 16 previously reported cases of this tumor arising from the head of the pancreas [12, 13]. Tumor size may vary between 3 and 20 cm in diameter (average 12 cm) [14]. Our patient’s tumor size was 30x50 cm.

Ultrasound typically shows a polycystic tumor, and very rarely calcifications [15]. CT and MRI features are almost identical. The tumor appears to be well-circumscribed, encapsulated, water-isodense, polycystic tumor with thin septa, containing a radiologically evident fatty component. The cystic aspect of this lesion can complicate differentiation from other neoplastic and non-neoplastic cystic tumors of the pancreas [16, 17].

Differential diagnoses include pancreatic pseudocysts, mucinous and serous cystadenomas, intraductal papillary mucinous neoplasms, echinococcal cysts, other congenital cysts and pancreatic ductal carcinoma with cystic degeneration [1, 8, 18]. Histopathological examination plays a definite role in the diagnosis. Histologically, lymphangiomas are classified into three types: simple capillary, cavernous and cystic [1, 18]. Cystic lymphangiomas of the pancreas are composed of dilated cystic spaces lined by flattened endothelium containing abundant lymphoid tissue and smooth muscle in the wall of the cyst [4]. Immunohistochemical staining of factor VIII-R antigen, CD31 and CD34, which are markers for the identification of lymphatic and capillary endothelium, can be complementary to the diagnosis [11]. Measurement of the triglyceride level and the appearance of the aspirated fluid by EUS-FNA may be helpful in diagnosing small, asymptomatic lesions. A triglyceride level above 5,000 mg/dL and milky white fluid is diagnostic of a lymphangioma [19, 20]. In our case, the cystic fluid was clear yellowish, and preoperative EUS-FNA was not considered as the patient was symptomatic and we presumed it to be a case of cystic neoplasm of the pancreas.

A complete surgical excision is curative for lymphangioma, with incomplete excision being the only reason for recurrent disease [8]. Depending on the tumor location and size, complete excision may involve a simple excision of the mass or may require pancreatic resections, such as a Whipple procedure or distal pancreatectomy. In small asymptomatic cases, where a confident diagnosis of cystic lymphangioma can be made with EUS-FNA, non-surgical management with regular follow-up can be carried out.

CONCLUSION

Cystic lymphangioma is a rare benign tumor of the pancreas and this is the first reported case in an adult which presented with acute abdomen. Although rarely diagnosed preoperatively, this entity should also be considered in the differential diagnosis of cystic lesions of the pancreas. Complete surgical excision is curative.

Conflicts of interest

The authors have no potential conflicts of interest.

References


