

Pancreatic Lymphangioma: A Case Report and Review of Literature

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June 22, 2023

TITLE:

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Type: Case Report

Short Title: Pancreatic Lymphangioma

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Abstract:

Lymphangiomas are cystic lesions that arise due to malformations (congenital or acquired) in the lymphatic system and are composed of thin-walled septa lined by endothelial cells that enclose blocked lymph fluid. Lymphangiomas can occur in any part of the body, and at any age but generally, 90% occur in children < 2 years of age and 90% times in the upper part of the body like head, neck, and axillae. Abdominal lymphangiomas are rare (only 1% of all lymphangiomas) and pancreatic are even rarer. When occurring in the pancreas, they mostly present at later ages with recurrent abdominal pain +/- palpable abdominal mass and mostly in 'women', although most commonly they are an incidental finding. Here we present a case of a young 'male' with the complaint of epigastric pain that responded to acetaminophen, USG and CT raised the suspicion of cystic lymphangioma and EUS FNA showing pathognomic lab findings in the aspirated fluid. A definitive diagnosis can be made on histology. In asymptomatic patients, conservative treatment is preferred while in patients with symptoms and compression of adjoining structures due to size, En-bloc resection is curative with an excellent prognosis.

Key-words:

Lymphangioma, Abdominal Lymphangioma, Pancreatic Lymphangioma, Pancreatic cystic lesions.

Introduction:

Lymphangioma benign and slow-growing tumors of vascular origin that can be congenital or acquired (1). When congenital these can occur in isolation or can be a part of various syndromes (Turner, Noonan, Downs). When acquired, they are mostly a result of infection, inflammation, or cancer that can block the normal

lymphatic flow (2). Lymphangiomas are rare, representing 4% of vascular neoplasms in general and approx. 25% of all nonmalignant tumors of vascular etiology in the pediatric age group. They almost always exist at birth, but overt presentation depends on the rate of growth in which fluid accumulates and on the site of its origin. 90% of these tumors occur in children, and 90% of children are <2 years of age at the time of presentation. Abdominal lymphangiomas are very rarely seen and pancreatic lymphangiomas are rarest. Pancreatic lymphangiomas compose <1% of all pancreatic neoplasms. These are mostly found in women. Here we discuss a case where a young man presented to us with epigastric pain and workup led us to the diagnosis of pancreatic lymphangioma.

Case Presentation:

A young man came to our hospital with presenting complaint of epigastric pain for few days. Urgent ultrasound abdomen was performed which reported it a pancreatic cyst. The pain was relieved by pain killers. The radiology team was taken on board and CT pancreas dynamic was advised. CT pancreas dynamic was performed with images obtained in noncontract, arterial, venous, and delayed venous phases. The rest of the abdomen and pelvis were reviewed in the venous phase. There was a multilocular and multiseptated homogeneously hypoattenuating/nonenhancing fluid density omento-mesenteric mass predominantly occupying the lesser sac. The lesion was insinuating within the pancreatic body and tail parenchyma as well as along the small bowel mesentery with mild mass effect on the stomach which is anteriorly compressed. It was abutting the left adrenal gland, which was however normal. The pancreatic head and uncinate process appeared normal. It was causing displacement of the small bowel loops and surrounding the transverse colon and splenic flexure. Suspicion of lymphangioma was given on it.

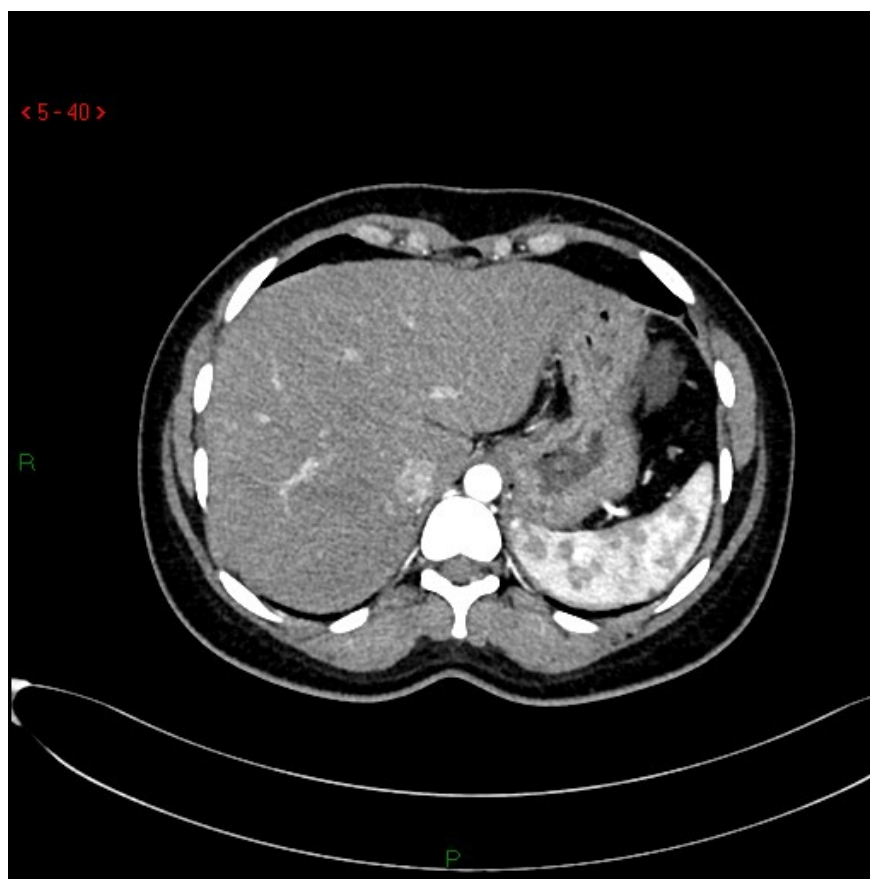


FIGURE 1: Axial Contrast enhanced CT in arterial phase showing omentomesenteric mass.



FIGURE 2: Axial Contrast enhanced CT in arterial phase showing lesion insinuating within the pancreatic body and tail parenchyma.



FIGURE 3: Axial Contrast enhanced CT in venous phase showing mass abutting the left adrenal gland.

Further evaluation by the ultrasound-guided aspiration to check for LDH and Triglycerides level in the aspirated fluid was advised. Informed consent was obtained. Sterile prep and drape were done. The patient was placed in the supine position. Local anesthesia with 2% Xylocaine was given. Under direct ultrasound guidance, an 18 G LP needle was inserted into the lesion in the epigastrium. 30 cc of straw-colored fluid aspirated. No complications were seen. This aspirated fluid was sent to the pathology department for further workup. LDH levels and Triglycerides levels were significantly raised in this body fluid and a diagnosis of pancreatic lymphangioma was made.

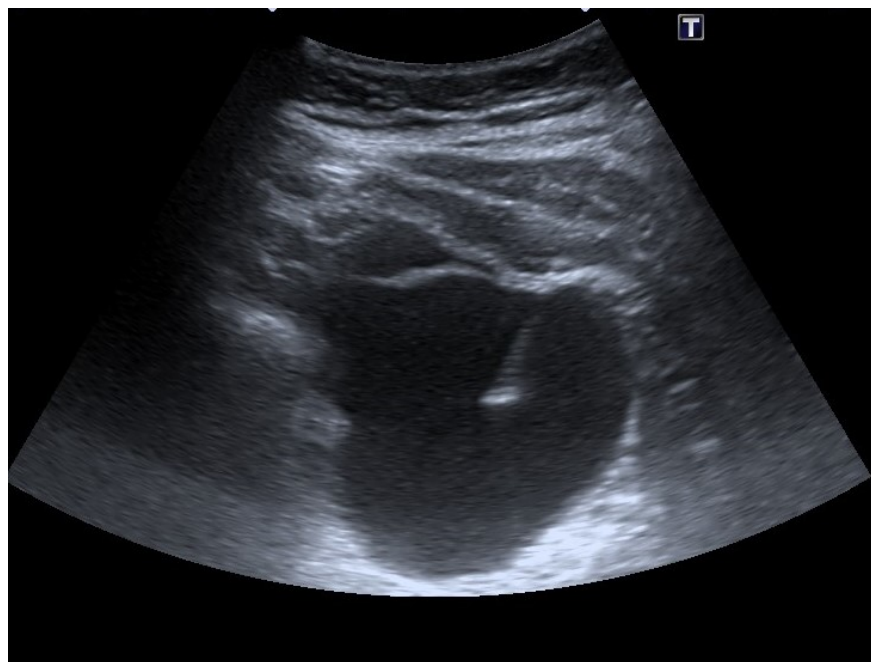


FIGURE 4: Spot image of ultrasound guided aspiration.

Discussion:

Lymphangiomas are the cystic dilatation, due to malformation (congenital) or fibrosis (acquired), of lymphatic channels. These are benign slow-growing cystic masses that cause lymphangiectasia due to lymphatic flow blockage. Gui et al described these segregated lymphatic channels as defective channels formed during the embryonic period rather than a true neoplasm [1]. These are generally reported in young children and are linked with congenital disruption syndromes such as Turner syndrome, Down syndrome, and Noonan syndrome [2]. During the development of an embryo, the disconnection of lymphatic vessels results in lymph obstruction that is enveloped by endothelial cells [3]. As a result of this multiloculated lymphangiomas are formed. Whereas acquired lymphangioma developed due to fibrosis, inflammation, and lymphatic vessel degeneration [4]. Frequently these lesions are found in the upper parts of the body mainly the neck and head (75%), axilla (15%) [5, 6, 7], unusual sites are the pericardium, pleura, liver, spleen, colon, groin, genital organs, and bones. Lymphangiomas developing in the adult pancreas comprise less than 1% of all Lymphangiomas [8].

Until now, only a few cases of lymphangiomas have been reported. The first case was described by Koch K, in 1913 a benign cyst formed due to blocked regional lymphatic ducts [9]. In medical literature, to our best knowledge, less than 85 to 90 cases have been described [10].

Clinical manifestation of pancreatic lymphangiomas is nonspecific and varies based on the site and size of the cyst[11]. Usually, they are symptomless and found as incidental findings [8]. when symptomatic are present, they are in the form of digestive discomfort, pain in the upper abdomen, or palpable masses. Although benign may invade the adjacent structure and when involve bordering structures like adrenal gland patient presents with specific symptoms like acute renal injury with the breakdown of skeletal muscle fibers.[12]

Hence, the combination of clinical presentation and laboratory evaluation may help identify the origin of the tumor and make appropriate diagnoses and differentials. In our case, the patient was symptomatic, he was a male patient who came with complaints of epigastric pain for few days. The average size of pancreatic lymphangiomas is around 12 cm in the greatest dimension [13], but in our case, the lesion was relatively larger. For evaluation, the CT pancreas dynamic was performed which revealed a multilocular

and multiseptated homogeneously hypoattenuating/non-enhancing fluid density omento mesenteric mass predominantly occupying the lesser sac. The lesion was insinuating within the pancreatic body and tail parenchyma as well as along the small bowel mesentery with mild mass effect on the stomach which is anteriorly compressed. It was abutting the left adrenal gland, which was however normal. The pancreatic head and uncinate process appeared normal. It was causing displacement of the small bowel loops and surrounding the transverse colon and splenic flexure.

Generally, pancreatic cystic lesions can be differentiated from lymphangiomas include pancreatic simple cysts pseudocysts, mucinous cystic neoplasms, serous cystadenomas, intraductal papillary mucinous neoplasms, and cystic pancreatic carcinomas. Imaging results may assist in preoperative diagnosis of upper abdominal lymphangiomas, even though none of the findings are confirmatory [13]. On Ultrasound scan, lymphangioma gives the impression as a complex cystic mass with intra-cystic septation or internal echoes, with or without internal calcifications. CT scan will reveal a low-density, thin-walled, uni, or multilobulated cystic mass with thin enhancing endo-cystic septation [8]. pancreatic cystadenomas share the same imaging findings and may present in similar ways suspension, can be ruled out on histology. Pseudocyst in the pancreas usually develops due to chronic or acute pancreatitis, USG or CT imaging will reflect the finding. These changes were not present in our case and therefore the possibility of pseudocyst was low.

Diagnosing pancreatic lymphangioma is highly suspect with the help of clinical findings and imaging investigations like CT, and EUS combined with FNA. In our case after clinical evaluation, we did CT which narrow down our differentials and leads us to ultrasound-guided Fine Needle Aspiration (FNA). Aspirated fluid was sent for cytology and biochemistry yielding very high LDH and triglyceride levels, making pancreatic lymphangioma highly likely and our working diagnosis. Simple cysts, serous cysts, or lymphangiomas usually yield clear thin, straw-colored fluids, composed of lymphomononuclear cells. Fluid from mucinous cysts will produce mucin, which can be detected grossly and microscopically. Moreover, the aspirated fluid from mucinous cyst will show mucin-producing columnar epithelial cells in varying amounts, arranged in papillae, clusters, or singles, these cells may or may not show cytological atypia. pancreatic Cystic carcinomas characteristically exhibit cytologically and morphologically malignant cells arranged in clusters, acinar or papillary patterns [8,13,14]

A definitive diagnosis is made on histopathological examination. Grossly cystic lymphangiomas are typically soft multilobulated masses containing serosanguinous or serous fluid. The cyst walls are usually single-layered cells without any solid areas. Under microscope dilated lymphatic channels of large and small sizes are seen, which are separated by thin septa and are intermixed with the pancreatic parenchyma [13]. Cysts are lined by flattened endothelial cells. Lymphocytes aggregates may be noticed inside the cysts or adjoining stromal tissues in Lymphangiomas [8,13]. Histologically lymphangiomas are classified into cystic, cavernous, and capillary [4, 7 15]. This classification was defined by Wegner in 1877 [16]. 2018 International Society for the Study Vascular Anomalies, subclassify the cystic lymphangioma as macro-cystic lymphatic malformation if cyst diameter is more than 1 cm, while cavernous lymphangioma is described as microcystic lymphatic malformation as the cyst diameter is less than 1 cm. Thus, lymphangiomas are classified into microcystic, macrocytic, and mixed lymphangiomas [17]. Capillary lymphangiomas are formed by tiny, poorly defined lymphatic vessels comprising of a thick cellular stroma [7]. Microcystic lymphangiomas are common in areas with dense connective tissue so decreased muscle expansion, such as the tongue and lip. Macro-cystic lymphangiomas are more common in areas with loose connective tissue, like the axilla, neck, and abdomen, so endothelial-lined channels can easily expand. [18]. Retroperitoneal macro-cystic lymphangiomas are predominantly multilocular [7]. In the pancreas, cystic and cavernous and a mixture of these two have been reported.

The treatment of lymphangioma is based on classification. In macrocytic or mixed lymphangiomas spontaneous regression usually occurs while it is rare in microcystic lymphangioma. Old age is more likely associated with spontaneous [19]. Pathologically, mixed lymphangiomas macrocystic are comparable but vary from microcystic lymphangioma in the balance of in and outflow of lymph fluid [19]. In microcytic lymphangioma, surgery is a reasonable approach because of the lack of spontaneous regression. While if the

patient remains asymptomatic and the lesion is not increasing, the patient can be managed conservatively with regular surveillance imaging studies. Surgical resection is the definitive treatment of lymphangiomas.

Different surgical choices may be considered, like simple cyst excision, total pancreatic resection, (Whipple procedure or distal pancreatectomy), which depends on the size and morphology of the cysts [14]. En-bloc resection of the pancreatic lymphangioma is the treatment of choice, which was done in the present case. Partial removal of cyst or aspiration of the content of cyst is not recommended because it may cause recurrences. Long term prognosis as complete resection of the pancreatic lymphangioma is excellent [13,20].

Conclusions:

Pancreatic lymphangioma is an unusual clinical finding of the pancreas which should be included and evaluated in the differential diagnosis of pancreatic cystic tumors. It is often asymptomatic and diagnosed on imaging. Definitive diagnosis is made with the combination of clinical evaluation, imaging studies and FNA analysis leads to suspicion which can be confirmed on histopathology. Lymphangioma is a benign lesion, if the patient is symptom-free and the lesion is not growing, a patient can be managed conservatively with regular surveillance and imaging is the best strategy. Surgical management is reserved in symptomatic patients, with typical morphology (microcystic) that does not favor spontaneous regression, or cyst expansion endangering adjacent structures and not required and our case reveals the typical imaging and pathognomic ES FNA results of a lymphangioma in an atypical organ.

Conflict of Interest: All authors declare no conflict of interest.

Funding Role: None to declare.

Author Contributions: All authors contributed equally to the drafting of study.

Ethical Statement: The patient signed an informed consent form, as per the ethical guidelines of hospital board.

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