Clinics in Surgery

Case Report
Published: 08 Feb, 2019

A Rare Case of Cystic Lymphangioma of the Pancreatic Head

Cavalcanti E[1], Lorusso D[2], Coletta S[1] and Armentano R[1]*

[1]*Department of Histopathology and Gastroenterology, Histopathology Unit of National Institute of Gastroenterology "S. de Bellis," Research Hospital, Italy*

[2]*Department of Surgery and Gastroenterology, Surgery Unit of National Institute of Gastroenterology "S. de Bellis", Research Hospital, Italy*

Abstract

The cystic lymphangiomas are taxonomically framed as benign tumors that originate from congenital malformations of the lymphatic vessels with consequent interruptions of the flow and dilatation of the lumen. These lesions considered non-neoplastic, but hamartomatous. To date, there are few reported cases of abdominal lymphangiomas and in particular pancreatic involvement is rare and represents a diagnostic and therapeutic challenge for the clinician. We herein report of a 68-year-old patient had undergone surgical resection with the diagnosis of cystic neoplasm of the pancreatic head. This case underlines that, in the pre-operative differential diagnosis, cystic lymphangiomas are difficult to be diagnosed from mucinous cystic tumors and the origin of the tumor is also hard to be detected before operation. Consequently this case leads us to reflect that we should combine an accurate diagnostic images and pathological examination to better clarify a preoperative diagnosis so as to avoid a complete pancreaticoduodenectomy for a benign tumor such as lymphangioma.

Keywords: Cystic lymphangioma; Pancreaticoduodenectomy; Neoplasia

Introduction

The cystic lymphangiomas are taxonomically framed as benign tumors [1,2] that originate from congenital malformations of the lymphatic vessels with consequent interruptions of the flow and dilatation of the lumen. The most frequent localization, mostly in pediatric age, [3] are the neck, the axillary cavity and the mediastinum. Several cases of lymphangioma in abdominal organs were reported, however, the pancre as is one of the rarest origins and lack any specific signs or symptoms that could allow a clinica l diagnosis. These lesions can be clinically interpreted as no-malignant neoplasms and the differential diagnosis should be considered in patients presenting with an abdominal mass. Mass resection is the treatment of choice [4]. We described the case of a 68 -year-old patient had undergone surgical resection with the diagnosis of cystic neoplasm of the pancreatic head.

Case Presentation

A 68-year-old male was admitted to our hospital with diagnosis of cystic neoplasia of the pancreatic head, performed by another institution. Although the instrumental examinations had excluded the invasion of the adjacent organs, the suspicion of a possible malignant cystic neoplasia was advanced, due to the presence of hypervascularized zones at a contrast-enhanced Computed Tomography (CT). Nothing in the patient's anamnestic history of the illness, except a persistent vague dyspeptic symptoms and a non-significant weight loss. The results of laboratory tests and the tumor markers performed, including Ca 19-9, are normal. Laparotomy revealed a pancreatic mass, with integrity of the adjacent organs and the peritoneum. Thus, duodenopancreatectomy was chosen using a pylorus preserving Whipple procedure. Supposing a diagnosis of malignant neoplasm, intraoperative histologic assessment of surgical margins was required. Macroscopically, the mass had a lobulated cystic appearance with a diameter of 6 cm (Figure 1); Cystic lymphangiomas appear as soft, multiloculated cystic masses that contain serous fluid. The microscopic features of the lesion are shown in Figure 2: dilated lymphatic channels of varying size are seen, separated by thin septa. The cystic spaces are lined with flattened or cuboidal endothelial cells. Diagnostic confirmation of pancreatic head cystic lymphangioma was defined by immunohistochemical positivity to Podoplanin (Figure 2). The patient's post- operative course was satisfactory and free of complications.

OPEN ACCESS

Correspondence:
Dott Elisabetta Cavalcanti, Department of Histopathology and Gastroenterology, National Institute of Gastroenterology "S. de Bellis," Research Hospital ,Via Turi 27, Castellana Grotte (Ba), Italy, Tel.: +39-0804994307;
E-mail: elisabetta.cavalcanti@irccsdebellis.it

Received Date: *16 Jan 2019*
Accepted Date: *05 Feb 2019*
Published Date: *08 Feb 2019*

Citation:
Cavalcanti E, Lorusso D, Coletta S, Armentano R. A Rare Case of Cystic Lymphangioma of the Pancreatic Head. Clin Surg. 2019; 4: 2328.

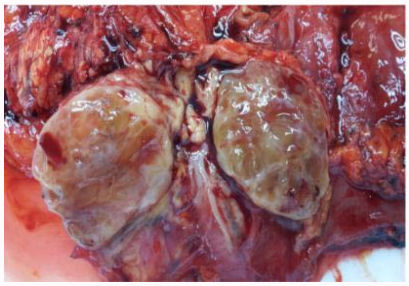

Figure 1: Surgical specimen of the pancreatic head and body by Whipple procedure, including the cystic lesion of the pancreas head. On the cutting surface it is evident the cystic concereration with serous fluid and the solid component.

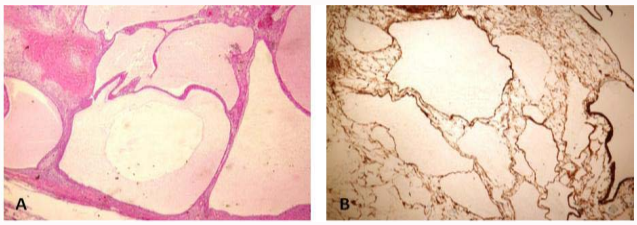

Figure 2: A: Dilated lymphatic vessels immersed in mesodermal tissue. H & E staining. Low power magnification: x2; **B:** IHC staining for podoplanin. Lymphatic channels lined by endothelium immunoreactive to antibody podoplanin. Low power magnification: x 4.

Discussion

Lymphangioma is an uncommon, benign malformation of the lymphatic vessels, considered non-neoplastic, but hamartomatous lesion. Based on the clinical and histopathological characteristics, different entities are recognized. A commonly used classification, based (depending) on the depth and the size of these abnormal lymph vessels, classifies these lesions into capillary, cavernous and cystic lymphangioma (Landing BH, Farber S. Tumors of the cardiovascular system. Atlas of tumor pathology. Washington, DC: Armed Forces Institute of Pathology 1956). The first two are predominantly skin, while cystic lymphangioma is predominantly intra- abdominal and retroperitoneal. Histologically capillary lymphangioma consists of a proliferation of capillary vessels that are distinguished from those of hemangioma due to the absence of endoluminal cells. Cystic lymphangioma (or cystic hygroma): is a vascular lesion of probable malformative nature typical of the neonatal age. It presents itself as a painless mass, mostly of large dimensions, located more frequently in the neck and limbs. Histologically, cystic lymphangioma is supported by a proliferation of widely dilated lymphatic vessels. Lymphangioma with pancreatic localization is extremely rare and clinically, may mimic pancreatic neoplasms [Khandelwal M, et al, 1995]. A few cases are described in the literature [5] and only the cystic and cavernous types have been reported in the pancreas [Paal E, et al, 1998]. These tumors appear to originate from the extra-lobular connective tissue peripancreatic, from lymphatic malformation of the dorsal duodenum. The clinical presentation can be asymptomatic or present with vague symptoms. CT imagings are essential diagnostic to define these abnormalities although it is difficult to differentiate abdominal cystic lymphangiomas from other pancreatic lesions such as in our case occurred. At times it becomes difficult to define a precise pre-operative diagnosis of the nature of the lesions. We should combine image and pathological examination to clarify a diagnosis. Although lymphangiomas are benign tumours, they can encroach on adjacent organs, in rare cases begin with signs of obstructive jaundice and that, resection of these invaded organs may be required for a complete excision. Differential diagnosis must always be established with cystic malignant lesions originating from pancreatic ducts and pseudocysts [6]. In our case, the instrumental examinations had excluded the invasion of the adjacent organs but suggesting a probable neoplastic lesion due to the presence of hypervascularized. In this case it may be necessary to perform a diagnostic laparoscopy to evaluate the lesion, but frequently the aspirated material by the cyst was not enough and useful for cytologically or histologically diagnosis; a possible indirect useful sign of the diagnosis could be the partial collapse due to a decrease in the intracystic pressure during the aspiration of the liquid. Therefore, the role of diagnostic laparoscopy must be considered before the laparotomic approach, in patients with a mass of doubtful meaning and in this case the choice intervention must be decided, after careful examination of all the parameters that can be evaluated. Actually surgical resection still remains the best treatment for lymphangiomas and it is usually curative, while incomplete excision is likely to lead to a local relapse. Partial pancreatectomy may be necessary in some cases. However, the definitive diagnosis of pancreatic lymphangioma can be made only by excision and histopathologic examination. In abdominal cystic lynphangioma cases the classic pancreaticduodenectomy may be cause significant derangement of the normal anatomy and, to some degree, the normal physiology of the upper gastrointestinal tract, therefore, it is desirable to perform a tumor resection block while preserving the upper portion of the second part of the duodenum and the Vater ampoule. In the literature, there are cases of tumor resection with preservation of the ampulla of Vater [7]. In our case, it was not possible to minimize the damage to the normal anatomy and physiology of the upper gastro-intestinal tract due to technical reasons and the size of the cyst. Therefore, in the pre-operative differential diagnosis, cystic lymphangiomas are difficult to be diagnosed from mucinous cystic tumors and the origin of the tumor is hard to be detected before operation. Consequently this case leads us to reflect that we should combine an accurate diagnostic images and pathological examination to better clarify a preoperative diagnosis so as to avoid a complete pancreaticoduodenectomy for a benign tumor such as lymphangioma.

Compliance with Ethical Standards

All procedures performed were in accordance with the ethical standards of the institutional research committee accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2000 [5]. Informed consent was obtained from patients for being included in the study.

References

1. Colovic RB, Grubor NM, Micev MT, Rankovic VI, Jagodic MM. Cystic lymphangioma of the pancreas. World J Gastroenterol. 2008;14:6873-5.
2. Koening TR, Loyer EM, Whitman GJ, Raymond AK, Charnsangavej C. Cystic lymphangioma of the pancreas. AJR Am J Roentgenol. 2001;177(5):1090.
3. Gray G, Friend K, Iraci J. Cystic lymphangioma of the pancreas. CT and pathologic findings. Abdom Imaging. 1998;23(1):78-80.
4. Lyngdoh TS, Konsam R, Th B, Marak B. Giant cystic lymphangioma of pancreas. ANZ J Surg. 2008;78(8):673-4.
5. Igarashi A, Maruo Y, Ito T, Ohsawa K, Serizawa A, Yabe M, et al. Huge

cystic lymphangioma of the pancreas: Report of a case. Surg Today. 2001;31(8):743-6.

6. Kullendorff CM, Malmgren N. Cystic abdominal lymphangioma in children. Case Report. Eur J Surg. 1993;159(9):499-501.

7. Fahimi H, Faridi M, Khorsandi M, Gholamin S, Molanaee S. Cystic lymphangioma of the pancreas: Diagnostic and therapeutic challenges. JOP. 2010;11(6):617-9.