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Pancreatic cystic lymphangioma: Report of a case

Pankreasın kistik lenfanjiyomu: Olgu sunumu

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We herein present a patient referred to our clinic with the complaints of flatulence and left upper quadrant abdominal pain who was diagnosed to have pancreatic cystic neoplasia radiologically. The septated cyst was defined to be 9x12 cm in diameter by abdominal computed tomography and by ultrasonography originated from the tail of the pancreas. Distal pancreatectomy procedure with complete resection of the cystic lesion was performed in this patient. Pathologic examination revealed pancreatic cystic lymphangioma (PCL). Although PCL is very rare in adult patients, it can cause confusion due to the presence of other cystic pathologies of the pancreas. Complete excision of the cyst is mandatory to prevent recurrences. In our case, no recurrence was detected after a two-year follow-up.

Key words: Lymphangioma, pancreas

Batın sol üst kadrandra ağrı ve hazımsızlık şikayetleri ile kliniğimize refere edilen ve mevcut radyolojik incelemelerinde pankreasın kistik neoplazisi tanısı alan bir olguyu sunmayı amaçladık. Batın bilgisayarlı tomografisinde ve ultrasonografide izlenen 9x12 cm boyutlarındaki septalı kistin pankreas kuyruğundan kaynaklandığı saptandı. Hasta ameliyata alınarak distal pankreatektomi işlemi yapıldı. Patolojik tanı pankreasın kistik lenfanjiomu olarak bildirildi. Erişkin hastalarda pankreasın kistik lenfanjiomları nadir görülse de, pankreasın diğer kistik patolojileri ile karışmaktadır. Kesin tanı kistin çıkarılması ve histopatolojik değerlendirme ile oluşturulmaktadır. Nükslerin önlenmesi için kistin tam rezeksiyonu gerekmektedir. Olgumuzda da 2 yıllık takip sonrası nüks saptanmamıştır.

Anahtar kelimeler: Lenfanjiyoma, pankreas

INTRODUCTION

Lymphangiomas are rare congenital benign tumors arising from the lymphatic system (1), and are mostly encountered in the neck and axillary regions of pediatric patients (95%) (2). Lymphangioma cases have also been reported in other organs, such as the liver, spleen, lungs, mediastinum, colon, retroperitoneum, pancreas, omentum, pericardium, pleura, kidneys, ureters, bone, scrotum, penis, and cervix, etc. (2). Intraabdominal lymphangiomas are extremely rare, with a reported incidence of less than 1 in 20,000 to 1 in 250,000 hospital admissions (3). Pancreatic cystic lymphangioma (PCL) is mostly seen in female adults and particularly represents an exceptional report (3, 4). Since its first description by Koch in 1913, a literature review revealed only 64 PCL cases (4).

Lymphangiomas are classified into three groups as cystic, capillary and cavernous (2). Although rare, the diagnosis of PCL is not easy (3) and should be taken into account with other cystic neoplasms of the pancreas.

CASE REPORT

A 50-year-old female patient was referred to our clinic with the complaints of flatulence and left upper quadrant abdominal pain. On physical examination, a palpable mass without clear borders was detected on the left subchondral region. All laboratory values, tumor markers and serologic tests were within normal limits. Hemagglutination test for hydatid disease was negative. Abdominal computed tomography (CT) scan revealed a lobulated hypodense cystic mass 9x12 cm in diameter, originating from the tail of the pancreas (Figure 1). In ultrasonography (USG), the mass was defined to be a septated fluid-containing cyst. Upper gastrointestinal endoscopy revealed nothing but the erosive gastritis and there was no external compression to the stomach. Fine needle aspiration biopsy

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(FNAB) of the pancreas was performed, and lymphoid cell components in a benign cystic lesion were reported in the histologic examination.

The patient was operated with the preoperative diagnosis of pancreatic cystic neoplasia. Following the left subcostal incision, abdominal exploration was performed and a lobulated cystic lesion 10 cm in diameter originating from the tail of the pancreas and extending into the transverse mesocolon was detected. There was no invasion of the cyst into other organs or vascular structures. Distal pancreatectomy including the cyst and the distal one-third of the pancreas was performed. A clear liquid was aspirated from the cyst. The pancreas was closed with U-suture. Macroscopic examination revealed a septated cystic lesion measuring 10x3x2 cm extending into the peripancreatic tissue; histopathology reported the cyst as peripancreatic lymphangioma with ectasia of the lymphatic vessels. Mature lymphocytes and rare macrophages were detected in the aspirated fluid (Figure 2). There was no postoperative complication.

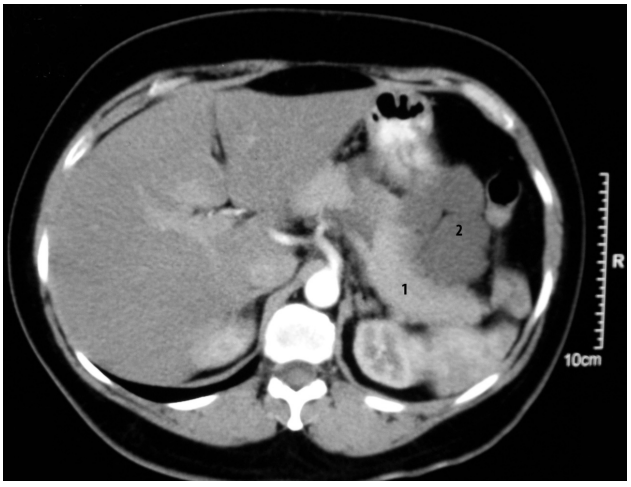

Figure 1. Abdominal CT scan revealed a lobulated hypodense cystic mass 9x12 cm in diameter originating from the tail of the pancreas. Pancreas (1) and cystic mass (2).

DISCUSSION

Lymphangiomas are endothelium-lined benign tumors that arise from the lymphatic system owing to congenital malformations. These malformations result in blockage of the lymphatic flow; thus, cystic dilatation of lymphatic channels occurs (5). Another theory is an inflammation leading to obstruction in lymphatic channels (3). Lymphangioma is mostly encountered in the pediatric age group. They are characterized by cystic and cavernous spaces and are mostly localized in the neck and axilla (95%), whereas abdominal involvement is seen in about 1% of all cases (3,4). PCLs, described for the first time by Koch in 1913, are extremely rare lesions (4), and fewer than 100 cases have been reviewed in the literature (6).

Macroscopically, cystic lymphangiomas are multiloculated soft cystic masses that contain either serous or serohemorrhagic or lymphatic fluid. Dilated lymphatic channels of varying size divided by thin septae are observed histologically. The cystic wall is lined by thin and flat endothelial cells. Islands of lymphocytes can be present in the lumen and/or neighboring tissue (4). In our case, the cyst was found to be septated and contained clear fluid macroscopically, and the diagnosis of PCL was made on histopathologic examination.

Abdominal lymphangiomas (ALs) account for less than 1% of all lymphangiomas and can be seen in any age group but are more frequent in female adults (1, 3). In a study by Igarashi et al. (2) that included 45 pancreatic lymphangioma cases, the female:male ratio was found to be 29:16. ALs generally present with vague abdominal symptoms such as chronic abdominal pain, nausea and vomiting, distension, and palpable abdominal mass. These symptoms may develop acutely in children, whereas in adults, months or even years may pass from the onset of the symptoms until the diagnosis. Furthermore, although rare, acute abdomen can occur due to the complications such as intestinal obstruction, rupture and/or hemorrhage (7).

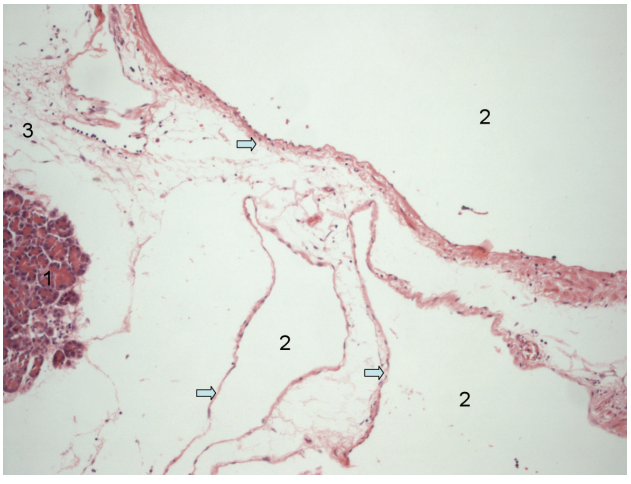

Figure 2. Histopathologic examination of the cystic pancreatic lymphangioma. Normal pancreatic tissue (1), ectatic lymphatic vessels (2) and peripancreatic tissue (3) are shown. Arrows indicate the single-layered squamous endothelial wall of the cystic cavities.

If the lymphangioma arises within the pancreatic tissue substance or if it is attached to the pancreas with a pedicle, it is termed as pancreatic lymphangioma (4). In our case, lymphangioma arose from the tail of the pancreas. In Igarashi's review study (2), it was observed that PCLs mostly derived from the body and tail of the pancreas.

Abdominal US, CT, magnetic resonance imaging (MRI), angiography and FNAB can be employed for the preoperative diagnosis of lymphangiomas. In USG, septated cysts can be visualized. Although CT provides additional information about the characteristics of the lesion, the diagnosis is often not direct. The capsule and the septations of the cyst and their thickness and the characteristics of the fluid within can be evaluated with CT. Preoperative FNAB is still controversial as it might cause hemorrhage, rupture or tumor implantation in malignant cases (4). MRI does not yield any further information (1). None of the examinations described above are pathognomonic. A preoperative diagnosis of PCL is difficult. Definitive diagnosis can only be made by histopathologic examination of the resected lesion (7), and the diagnosis is supported immunohistochemically by positive staining of factor VIII-R antigen, CD31, and CD34 markers (2).

Although clinical follow-up with periodic imaging of patients with PCL has been suggested (8), surgery is the current treatment of choice (3, 4). Laparotomy is generally preferred; however, laparoscopic resection can also be performed in suitable cases. Total resection of the lesion is mandatory. In our case, we needed to perform distal pancreatectomy for the total removal of the lesion, and no recurrence has been observed after a two-year follow-up. We noted that in a review study by Igarashi (2) including 45 PCLs, six distal pancreatectomies were performed, and distal pancreatectomy procedure was also preferred in two cases by Casadei (4).

In summary, PCLs are very rare tumors and carry a potential risk of local invasion. Definitive diagnosis can only be made by histopathologic examination of the excised lesion. Total resection of the tumor is mandatory in case of recurrence. Therefore, as in our case, partial pancreatectomy may be needed in some patients.

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