Acinar Cystic Transformation of the Pancreas: A Diagnostic Problem

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Abstract- Acinar cystic transformation (ACT) or acinar cell cystadenoma is a rare benign tumor of the pancreas. Their clinical presentation is nonspecific, thus hampering their detection and frequently leading to misdiagnosis. In this report, we present a case of ACT of the pancreas in a 54-year-old man that was vague abdominal symptoms. A magnetic resonance imaging was shown a well-defined cystic mass located in the head of the pancreas, measuring 57×47×23 mm. Our patient was treated by pancreaticoduodenectomy. In the macroscopic examination of the mass, a multicystic lesion was found in cream-colored pancreatic tissue. Histological and immunohistochemical studies examination revealed a cystic mass containing multiple cysts at varying sizes, lined by epithelial cells without atypia and positive staining of CK7, CK8/18, and CK19. A high index of clinical suspicion is required to diagnose this tumor. The surgical approach is to ensure accurate diagnosis and to avoid complications.

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Keywords: Acinar cystic transformation; Pancreas; Cystic tumors

Introduction

Acinar cystic transformation (ACT) is a rare benign tumor involving the pancreas, which can be detected incidentally on imaging or surgery. ACT was described first in 2000 by Albores-Saavedra and has also been called acinar cell cystadenoma (1). Although it is generally detected incidentally, the cystic lesion can reach the size of the entire pancreas. These tumors may remain undetected unless local invasion to the surrounding tissue and vascular supply. The most common location of ACT is the head of the pancreas and less frequently entire pancreas and tail (2). They arise from the transformation of the acinar or ductal epithelium into the cystic differentiation. Although it is stated that it has genetic features, the epidemiology, and cause of the tumor are unknown (3). Here we present clinical, radiological, and pathological findings of such a rare case of ACT.

Case Report

A 54-year-old male patient was referred to our surgery clinic complaining of pain and tenderness at the right upper quadrant of the abdomen. A clinical examination did not detect any palpable mass. There was a history of diabetes mellitus, and no weight loss was reported. He had normal vital signs, and laboratory findings were normal except for increased tumor marker CEA2 (4.67 ng/ml .upper limit three ng/ml).

Magnetic Resonance Imaging (MRI) was shown a slightly hypodense, well-circumscribed, cystic mass located in the head and uncinate process of the pancreas, measuring 57×47×23 mm (Figure 1). In this lesson, solid areas were not observed, but multiple cysts were noted. This mass was no compressing peripancreatic vascular structures, with no obvious invasion. It was reported that as a cystic lesion, but there might be intraductal papillary mucinous neoplasms (IPMNs). We performed a pancreatoduodenectomy procedure.

In the macroscopic examination of the mass, a multicystic lesion containing serous fluid, measuring 43X26X15 mm., was found in cream-colored pancreatic tissue (Figure 2). There was not any communication between the lesion and pancreatic ducts. The distance of the lesion to the duodenum was 0.5 cm, and no invasion was found. Histological examination revealed a cystic mass containing multiple cysts at varying sizes, lined by epithelial cells without atypia (Figure 3). Moderate pancreatitis was observed in the pancreatic parenchyma around the lesion. Immunohistochemical studies showed positive staining of CK7, CK8/18, and CK19, and

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negative results for CK20, p53, chromogranin, and synaptophysin. In our case, musin markers (MUC1, MUC2, MUC5AC, and MUC6) and mismatch repair proteins (MLH1, MSH2, MSH6, PMS2) were found negative.

The postoperative period was normal, with no complication, and the patient was sent home on d 7.

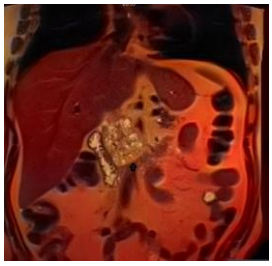

Figure1. MR image of the patient with a well-circumscribed lesion. Multiple cysts were located in the head and uncinate process of the pancreas. (arrow)

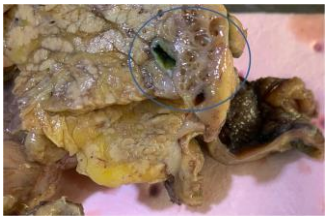

Figure 2. In the macroscopic examination of the mass, a multicystic lesion was found in pancreatic tissue

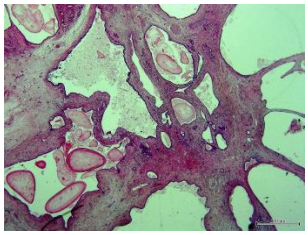

Figure 3. Histological examination revealed multiple cysts at varying sizes. (HEX4)

Discussion

ACTs are an uncommon cystic lesion of the pancreas. The mean age at diagnosis is 49 years, and the incidence is slightly higher in women than in men. The clinical presentation of ACT is variable (C). The symptoms do not generally differ from those of other cystic pancreas tumors, and diagnostic work-up is also similar to that for any other tumors. Often the tumor is "silent" until it reaches a large size, at which point it may cause nonspecific abdominal pain and discomfort.

Most ACT's originate at the head of the pancreas and, most commonly, have an exophytic grown pattern. CT and MRI findings are nonspecific, but the presence of multiple cysts and absence of connection with the pancreatic ducts are supportive for diagnosis. In the differential diagnosis, the neoplastic cystic tumors include mucinous cystic neoplasms, serous cystic neoplasms, and intraductal papillary mucinous neoplasm (IPMN) should be considered. The preoperative endosonography guided fine-needle aspiration biopsy is not recommended as a probable tumor can spread.

Histological findings are dissenting of acinar units to form a benign cystic structure (4). ACTs do not connect with the pancreatic ductal system, in which these finding rules out IPMNs, serous and mucinous tumors. In addition, mucinous differentiation, ovarian stroma, and papillary growth are not detected by HE staining (5). In our patient, the cystic lesion was surrounded by a single-layered cell, and no papillary protrusion and ovarian-like stroma were found with HE. It is known that histologic criteria do not provide predictive power, although greater tumor size is not associated with the mitotic activity. However, the diagnosis may be problematic because pathologists see only a few numbers of these tumors each year. The question arises as to whether classifying ACTs, defined as a transformation or a neoplasm. The answer to this question is that the applied immunohistochemical tests are the gold standard. ACTs are immunohistochemically positive for CK7, CK8, CK18, CK19, lipase, and trypsin, whereas they are negative for synaptophysin,α- amylase, and chromogranin. In our case, a definitive diagnosis was made owing to immunostaining.

Surgical resection has been the mainstay of therapy. Achieving negative pathologic margins of resection generally is not difficult because it does not diffusely infiltrate the peripancreatic tissue. In conclusion, a high degree of suspicion of the ACT should be raised in patients with cystic pancreatic lesions.

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