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Acinar Cystic Transformation (ACT) of the Pancreas: Focus on Radiological Aspect. Presentation of Two Cases and Review of Literature

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1. Abstract

1.1. Background: Acinar cystic transformation of the pancreas (ACT) are rare, benign cystic lesions of the pancreas often discovered in the context of non-specific abdominal pain or incidentally during imaging for another reason. Diagnosis is difficult by imaging only because of the lack of typical radiological criteria and most diagnoses are made by histopathological examination after surgical resection. Because no risk of malignant transformation is described in the literature, a conservative approach is the treatment of choice for ACT once the diagnosis has been made.

1.2. Case Description: We report two cases of ACT diagnosed by Magnetic Resonance Imaging (MRI) with typical radiological features: multiple cysts (>5), clustered pattern, presence of calcifications, no connection to the main pancreatic duct (MPD) including side branches, no MPD dilatation. Due to the typical imaging features and lack of malignant characteristics, conservative treatment with radiological follow-up was opted for, therefore avoiding surgical resection.

1.3. Conclusion: Pre-operative diagnosis of ACT is of great benefit as it can lead to a more conservative treatment by radiological follow-up instead of extensive surgical resection with its associated mortality and morbidity, especially for asymptomatic patients. Unfortunately, the limited diagnosed cases and the short-term follow-up remain an obstacle to establishing guidelines for diagnosis, management and treatment of ACT.

2. Introduction

Acinar cystic transformation of the pancreas, also called acinar cell cystadenoma, corresponds to rare cystic lesions of the pancreas first described in 2002 as incidental autopsy finding [1]. Since then, fewer than 100 cases have been described in the literature, mostly as single case reports, case series or retrospective studies [12]. ACT was classified as a non-neoplastic, benign, pancreatic cystic lesion by the 5th World Health Organization (WHO) classification of gastro-intestinal tumors in 2010 [2].

Although the aetiology of ACT remains unknown, a few hypotheses exist such as neoplastic origin, congenital malformation, genetic predisposition and obstructive or inflammatory aetiology with a possible heterogeneous nature of these lesions [3,13]. ACT are predominantly observed in women with a female to male ratio of approximately 2:1 and age range reported of 40-50 years [4]. Two thirds of patients present with non-specific abdominal pain while the others are usually asymptomatic [5]. On laboratory examination, serum lipase and amylase can be slightly elevated and fluid analysis of the cysts shows elevated lipase and amylase in 80% of the cases [7]. CEA and CA 19-9 levels are usually normal [5]. While the serum and cyst aspiration analysis usually remains non-contributive, the radiological aspects of ACT may permit a non-invasive diagnosis. Differential diagnosis of ACT include branch-duct intraductal papillary mucinous neoplasm (BD-IPMN), serous cystadenoma, mucinous cystadenoma or cystadenocarcinoma, post-obstructive pancreatic cysts, pseudocysts and pancreatic ductal adenocarcinoma with cystic degeneration [6].

Here we attempt to provide a review of this rare under-recognized entity with the emphasis on the radiological features. We hope that it could lead to a better diagnostic approach and avoid unnecessary biopsy or unnecessary surgical resections in patients with ACT.

3. Case Presentation

3.1. Case 1

A 65 year-old female patient with a medical history of idiopathic hypertension, tobacco consumption and lower limb venous insufficiency, presented to emergency department with non-specific right flank and abdominal pain. The physical examination and laboratory analysis, including serum amylase and lipase, revealed no abnormality. An abdominal ultrasound (US) demonstrated multiple hepatic lesions compatible with hemangiomas. An abdominal Magnetic Resonance Imaging (MRI) was performed, confirmed the presence of hepatic hemangiomas and revealed the presence of multiple small cystic lesions of the pancreas, distributed in a clustered manner throughout the entire pancreatic parenchyma, the largest one being at the body-tail junction and measuring 17mm. All lesions demonstrated a high signal intensity on T2-weighted images (T2WI), hypo-intensity on T1-weighted images (T1WI) with no contrast enhancement on dynamic sequences corresponding to multiple pancreatic cystic lesions (Figure 1). On magnetic resonance cholangiopancreatography (MRCP), the main pancreatic duct (MPD) was not dilated and no communication between the MPD and the cystic lesions was detected.

The patient underwent endoscopic ultrasound (EUS) which confirmed the presence of multiple cysts throughout the pancreatic parenchyma and demonstrating small calcifications in the wall of the lesions without communication to the MPD. Based on typical radiological and endosonographic findings and, after multidisciplinary discussion, the diagnosis of ACT was retained and conservative treatment with annual follow-up by imaging was proposed. Two years follow-up with MRI displayed no morphological changes of the cystic lesions, particularly no signs of malignant degeneration.

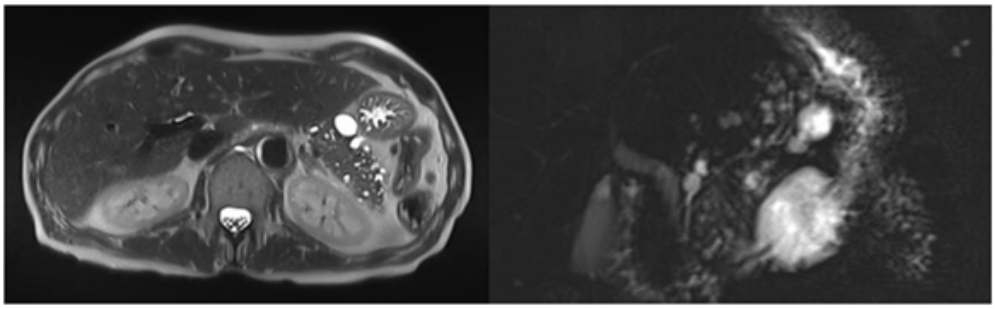

Figure 1: MRI findings of case 1

3.2. Case 2

A 74 year-old male patient was referred by his general practitioner to our hospital for further investigation of diarrhea and weight loss. His past medical history included regular alcohol consumption and sick sinus syndrome with a pacemaker implantation in 2021 and surgical history included bilateral inguinal hernia and umbilical hernia repair in 2021 and endoscopic resection of benign prostatic hyperplasia in 2015. Physical examination and laboratory testing were normal apart from a decreased faecal elastase level of 61mg/kg (normal range >200mg/kg) suggesting an exocrine pancreatic insufficiency. As per routine protocol, oesogastroduodenoscopy and colonoscopy were performed showing left colonic diverticulosis and excluding tumoral lesions. To evaluate the pancreatic parenchyma, an abdominal MRI was performed which revealed hepatic polycystosis and diffuse moderate atrophy of the pancreatic parenchyma surrounding multiple cystic lesions (Figure 2). The majority of the lesions were less than one centimeter while the largest one measured 12mm and was located in the tail. The lesions were distributed uniformly throughout the pancreatic parenchyma without obvious communication to the MPD, the latter was not dilated. Due to characteristic radiological findings and in the absence of malignant features, the diagnosis of ACT was made and a regular MRI follow-up was proposed which was unremarkable during the first year.

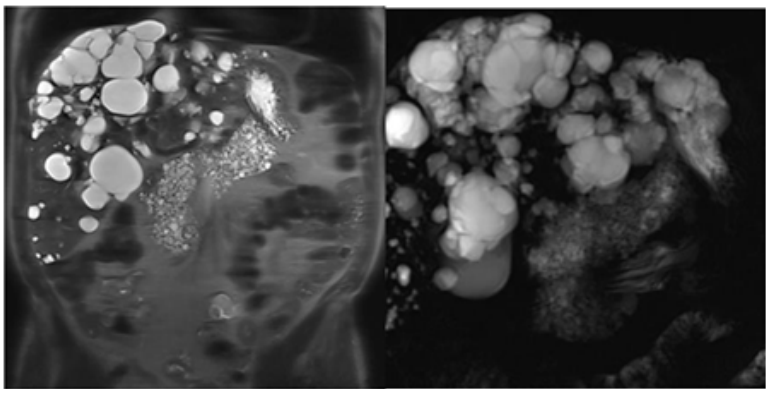

Figure 2: MRI findings of case 2

4. Discussion

Due to the rarity of ACT and lack of familiarity with this entity, the pre-operative diagnosis based on radiological findings remains difficult, renders recourse to EUS with fine needle aspiration (FNA) mostly mandatory. While the EUS aspect of ACT could be characteristic for an experienced gastroenterologist, FNA has a limited role in the diagnosis due to a high false negative rate of 25% related to low cellularity, loss of histologic architecture and non-specific liquid markers including amylase, lipase, CEA or CA 19-9. It is usually not sufficient to distinguish ACT from other potentially malignant pancreatic cystic lesions [4,7]. A new tool, the Moray microforceps biopsy device®, seems to provide a better sampling according to one case report from 2017, but diagnosis remains difficult [8].

Faced with an uncertain diagnosis, a total or a partial pancreatectomy (for example Whipple procedure) is usually performed and the final diagnosis is made on surgical specimen6. The cysts are mostly located in the head of the pancreas (in 50% of all cases) but can also be diffusely distributed throughout the pancreatic parenchyma as in present cases [4,14]. On gross examination, the cysts are usually multiloculated, clustered and separated by thin septa and vary in size between a few millimeters and several centimeters [9]. Histologically, the cysts are lined by an unstratified, flat to cuboidal epithelium mostly composed of acinar cells. Ductal cell metaplasia can also be present as well as squamous cells as described in a few cases [5,13]. The acinar cells were PAS-positive (Periodic Acid Schiff) on histological examination and they contain a clear cytoplasm with basal nuclei and apical eosinophilic zymogen granules4. The content of the cysts is serous and calcifications are typically present10. The adjacent pancreatic parenchyma is usually normal without signs of fibrosis nor chronic inflammation and there are no ovarian-like stromal changes seen in comparison to mucinous cystic neoplasm or IPMN1. No malignant features like nuclear atypia, necrosis or mitotic figures are described10. On immunochemistry, the cells are positive for trypsin and chemotrypsin consistent with acinar cell differentiation as well as for cytokeratine 7 (CK7) typical for ductal differentiation10. Ki-67 index is low (<1%) [10].

Diagnosis of ACT by radiological imaging remains difficult because of the lack studies reported in the literature. There is only one retrospective study dating from 2014 that compares radiological characteristics of ACT and IPMN which represents the main differential diagnosis [11]. Four criteria associated with the diagnosis of ACT seem to be statistically significant: presence of 5 or more cysts, clustered aspect of the cysts, presence of calcifications, no connection to the MPD or side branches [11]. The size of the cysts was not significantly different between ACT and IPMN. With these results, Delavaud et al. found out that if two out of four criteria were present, the sensitivity was by 100% and the specificity by 85% and in the presence of three out of four criteria 80% and 100% respectively for the diagnosis of ACT11. However, the study was limited by the low number of included patients and, although calcifications are one of the main diagnostic criteria, they are often not visible on MRI due to technical limitations in detection of calcifications and their small size.

In our study, we present two cases of ACT diagnosed by MRI with typical radiological features. The patients presented the following typical criteria on imaging: multiple cysts (>5), clustered pattern, presence of calcifications, no connection to the pancreatic ducts including side branches, no MPD dilatation (Table 1) [11,12,15]. The cysts were usually small (<2cm) and the cyst walls were thiny and non-enhancing on MRI and no intramural nodules were detected in the presented cases. In one patient, the diagnosis was confirmed on EUS without FNA, based on typical findings.

According to the aforementioned criteria, the diagnosis of ACT was reached based on the radiological features in our cases. Because there are no malignant transformations of ACT described in the literature, conservative treatment was proposed in these patients avoiding invasive biopsy and surgical resection with its associated morbidity and mortality. However, the lack of long-term follow-up in the literature renders regular MRI surveillance mandatory [16].

The main limitation of our study is the small number of cases for the establishment of precise diagnostic criteria. Furthermore, we have no pathological samples of the lesions and, even though our cases meet the typical radiological criteria mentioned above, we cannot confirm the diagnosis with certainty. Larger samples should be collected to appraise the validity of these criteria and to determine the sensitivity and specificity for the diagnosis of ACT. Additionaly, the follow-up in our study is short and long-term results of the evolution of these lesions are, as yet, unknown.

Table 1: Typical radiological features of ACT on MRI

Typical radiological features of ACT on MRI
• Presence of 5 or more cysts
• Clustered aspect of the cysts
• Presence of calcifications
• No connection to the MPD or side branches, no MPD dilatation

5. Conclusion

In conclusion, ACT are rare, benign lesions that should be considered in the differential diagnosis of pancreatic cystic lesions. Up to now, there are no cases of malignant transformation or metastatic spread described in the literature4. A better understanding of the aetiology of these lesions and establishment of precise radiological imaging criteria allowing for pre-operative diagnosis would lead to better management and also avoid unnecessary biopsy and surgical resection particularly in asymptomatic patients. Due to the rarity of ACT, more data needs to be collected for the development of robust guidelines.

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