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Polycystic Disease of the Pancreas: Rare Case Report

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Abstract | **Case Report**

Isolated polycystic disease of the pancreas is an extremely rare condition, usually found incidentally at imaging on asymptomatic patients. Cystic lesions of the pancreas are most oftenly seen in association with polycystic kidney disease and Von Hippel Lindau syndrome. Cross-sectional imaging and cystic fluid analysis play an important role in differentiating these congenital cysts from neoplastic cysts and pancreatic pseudocysts. The indications for surgery remain rare due to the very long asymptomatic evolution. The monitoring of these lesions generally shows the stability of most cysts. We report the case of a 33 years old patient who presented a polycystic disease of the pancreas incidentally discovered in imaging.

Keywords: Isolated polycystic, polycystic kidney disease, surgery.

INTRODUCTION

Cystic lesions of the pancreas include a wide variety of lesions, tumors or not, they are increasingly diagnosed due to recent advances in imaging and in particular cross-sectional imaging (CT scan and MRI). Their prevalence estimated at 20%, they are most often diagnosed by imaging in asymptomatic patients.

The diagnosis remains difficult, despite all the technological advances, to differentiate between cystic tumors with degenerative potential (or more rarely, in this context, degenerated) and lesions with benign potential (pseudo cysts, true cysts and cystic tumors with benign potential).

Therapeutic management must consider the patient's life expectancy without any intervention compared to the risks induced by surgical excision.

We report the case of a patient who presented with polycystic pancreatic disease discovered incidentally on imaging.

CASE REPORT

33-year-old patient with a history of treated Helicobacter pylori gastritis declared cured after a negative control, who has presented to our care for 6 months of intermittent biliary colic progressing in a context of apyrexia and conservation of the general condition.

Clinical examination was without abnormalities: the patient being in a good general condition, hemodynamically stable, anicteric, the abdomen was supple without any palpable mass or hepatosplenomegaly. The lymphatic node areas were free.

The abdominal MRI (Figure 1) revealed the presence of multiple cystic lesions, of different sizes, scattered throughout the pancreatic parenchyma, the largest measuring 41.5 mm. These lesions did not communicate with the secondary or main pancreatic ducts. Their signal is liquid in hypo signal T1, hyper signal T2, without partition, calcification or vegetation. Their wall is thin, without wall nodules or enhancement after contrast: Evoking pancreatic polycystosis without signs of degeneration. The gallbladder contained multiple gallstones without signs of inflammation or blockage.

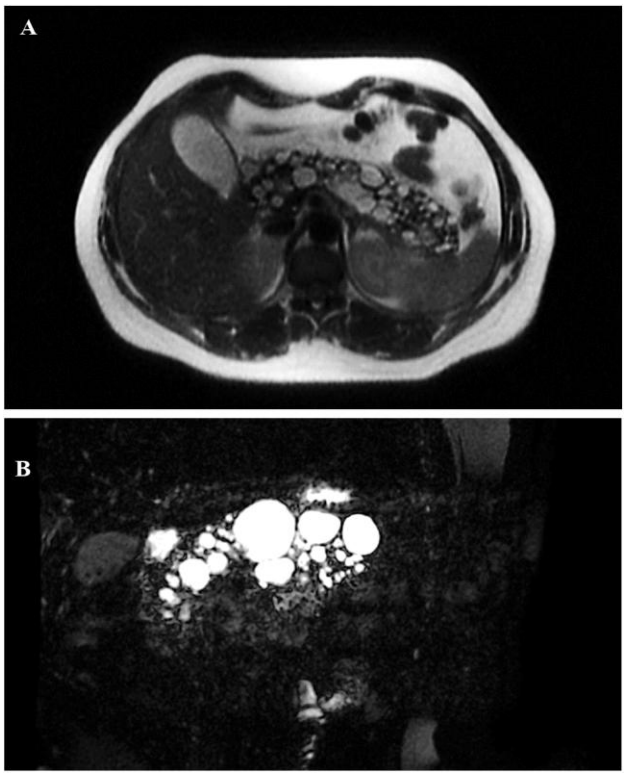

Fig-1: A + B: Abdominal MRI showing the presence of multiple cystic lesions of different sizes scattered throughout the pancreatic parenchyma

The decision of the multidisciplinary consultation meeting was surveillance for polycystic pancreatic disease and surgery for gallstone disease.

The patient was operated, having a laparoscopic cholecystectomy with simple postoperative consequences. The anatomopathological study of the surgical piece showed chronic nonspecific cholecystitis.

Postoperative recovery was uneventful and the patient was discharged from hospital the day after the operation. The evolution is favorable to date with complete disappearance of symptoms. A Pancreato-MRI is planned in a year as part of the surveillance.

DISCUSSION

Polycystosis of the pancreas is due to abnormal development of the pancreatic ductal system secondary to ciliopathy [1-3]. Recent studies have shown that the deficiency of hepatocyte nuclear factor VI and the gain in function of the protein containing Forkhead's box predispose to the development of pancreatic cysts [2, 3]. These cysts lack primary cilia similar to those seen in polycystic kidney disease [1].

Due to advances in cross-sectional imaging, cystic lesions of the pancreas are increasingly being diagnosed in asymptomatic patients. Their prevalence has been evaluated at 20% by imaging and autopsy [4, 5].

In polycystic disease of the pancreas (PDP), cysts constitute pseudo tumors without the potential for malignancy or risk of degeneration, unlike cystic tumors of the pancreas [6]. They can be localized in a part or spread to the whole of the pancreas.

It should be distinguished from multiple serous cystadenomas present in 15% of cases in asymptomatic patients with Von Hippel-Lindau syndrom [7], an autosomal dominant genetic disorder, which results in the development of a variety of tumors and cysts in the central nervous system (CNS) and visceral organs. Pancreatic lesions may be the only disease manifestation in 8% of cases [7].

Polycystic pancreatic disease can be associated with renal and hepatic cysts as part of the extra-renal manifestations of autosomal dominant polycystic kidney disease [8]. Pancreatic cysts are more common in women with polycystic hepato-renal disease and occur only in type 1 polycystic hepato-renal disease with a mutation on chromosome 16. Their frequency increases with age [8].

Polycystic pancreatic disease is in the majority of cases asymptomatic, but can very rarely be the cause of recurrent pancreatitis. Pancreatic cysts are found in 9% of patients by the age of 30 [8].

In polycystic pancreatic disease, the gradual increase in the size of the cysts results in parenchymal atrophy causing a feeling of heaviness which is often observed for any large benign pathology of the region such as polycystic liver disease. In our observation, the clinical signs were related to gallstone disease and were not specific to pancreatic involvement.

Finding PDP in an asymptomatic patient is always a difficult problem. Cross-sectional imaging (MRI, CT scan) or echo-endoscopy should orient the diagnosis towards a benign or malignant lesion [9, 10], but the first stage of interpretation always consists in ruling out the diagnosis of an associated pseudocyst of the pancreas usually with a history of pancreatitis [11].

Pancreatic MRI has excellent contrast resolution [12]. It makes it possible to clearly detect intracystic septa and adenoids and associated with Wirsungo-MRI sequences which are very T2-weighted, it detects elements containing stationary liquids very well, in particular microcysts and the Wirsung canal.

Endoscopic ultrasound also has good spatial resolution, which allows good detection of microcysts to confirm the diagnosis of serous cystadenoma and to look for endocystic abnormalities [13]. The major advantage of endoscopy is the possibility of performing directed punctures to measure intracystic markers and perform cytological analysis.

As with polycystic liver disease, evacuating puncture, under ultrasound or computed tomography, without intra-cystic injection, may be proposed to temporarily relieve symptomatic cysts [14]. Fine needle aspiration is often proposed in order to facilitate diagnosis by assaying tumor markers and histological analysis [14].

The rate of recurrence after hepatic cyst puncture is significant in the range of 23% to 57% in polycystic liver disease. The latter decreases significantly after fenestration (3 to 22%) [15]. The place of laparoscopic fenestration has been proven for hepatic cysts [16]. As with all abdominal surgery, laparoscopy has several advantages over open surgery such as the reduction in postoperative morbidity, the length of hospitalization, the cost, as well as the lower risk of postoperative adhesions and bronchopulmonary complications.

In polycystic pancreatic disease, the laparoscopic approach to the posterior omentum cavity is sometimes more difficult due to the size of the cysts and the inflammatory phenomena.

A case of laparoscopic fenestration for symptomatic polycystic pancreatic disease has been reported in the literature with simple postoperative consequences [17].

In pancreatic polycystosis, surgical indications remain rare due to the slow progression of the cysts, which for a long time remain little symptomatic [8].

Fenestration is offered in macrocystic forms and surgical resection is reserved for microcystic lesions and doubtful forms. After resection, recurrences are less frequent but the morbidity remains higher [16].

CONCLUSION

Polycystic pancreatic disease is a rare disease that should always be considered in the differential diagnosis when evaluating a patient with pancreatic cysts. It is most often associated with polycystic kidney disease or Von Hippel Lindau disease. Cross-sectional imaging and analysis of cystic fluid can differentiate these congenital cysts from a pseudocyst or pancreatic neoplasia.

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