Atypical Presentation of Main-Duct Intraductal Papillary Mucinous Neoplasm

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ABSTRACT

Context Intraductal papillary mucinous neoplasm is a ductal epithelial tumor characterized by dilation of the pancreatic duct due to mucus production and is a pre-neoplastic disease. The most used diagnostic studies are computed tomography scan and magnetic resonance cholangio-pancreatography. **Case Report** A Sixty-three-year-old female presented with fever and a painful palpable abdominal mass in the left flank. Ultrasonography revealed a supra-aponeurotic fluid collection that was punctured, obtaining purulent material. Computed tomography scan showed a supra aponeurotic collection communicated with a cystic image that extended behind the stomach and involved the pancreas, which also showed dilatation of the Wirsung duct. Magnetic resonance cholangio-pancreatography showed dilatation of the main pancreatic duct from the pancreatic head up to the tail of the pancreas and was also communicated with a large cystic cavity behind the gastric antrum. The intraoperative finding evidenced a communication of the supra-aponeurotic collection with a cystic tumor that involved the pancreas and the posterior surface of the stomach. Left pancreatectomy with splenectomy and antrectomy was performed, sectioning the pancreas at the neck. Frozen section of the pancreatic duct surface revealed severe dysplasia at the main duct. Resection was completed by performing pancreatoduodenectomy. The pathologic analysis revealed cystic papillary mucinous neoplasia with foci of high-grade dysplasia and invasive colloid-type carcinoma. **Conclusions** This is an atypical presentation of type 1 intraductal papillary mucinous neoplasm, but the diagnosis was suspected in the preoperative period with the computed tomography scan, the magnetic resonance cholangio-pancreatography and the upper GI endoscopy. It is important to know all the possible differential diagnosis to decide the best surgical procedure. In cases of invasive type 1 intraductal papillary mucinous neoplasm, involving other organs, an aggressive surgical resection is the best choice for the patient.

INTRODUCTION

Intraductal papillary mucinous neoplasia (IPMN) is a ductal epithelial tumor characterized by dilation of the pancreatic duct due to mucus production. It is a pre-neoplastic disease that is usually manifested by acute pancreatitis, abdominal pain, endocrine or exocrine pancreatic insufficiency or by complications of tumors associated with this disease such as jaundice or vomiting due to duodenal obstruction.

The diagnosis of IPMN increased as the quality of the imaging studies improved, being nowadays computed tomography scan (CT scan) and magnetic resonance cholangio-pancreatography (MRCP) the most used diagnostic studies [1]. The aim of this paper is to show an atypical presentation form of IPMN type1.

CASE REPORT

A Sixty-three-year-old female presented with fever and a painful palpable abdominal mass in the left flank. The patient had history of type 2 diabetes recently diagnosed and asymptomatic diverticula. Ultrasonography revealed a supra-aponeurotic fluid collection that was punctured, obtaining purulent material which was sent to culture **(Figure 1)**.

Diagnosis was completed with CT scan and MRCP. CT scan showed a supra aponeurotic collection communicated with a cystic image that extended behind the stomach and involved the pancreas, which also showed dilatation of the Wirsung duct. The uncinate process was preserved with a smaller dilation of the pancreatic duct **(Figure 2)**. MRCP showed dilatation of the main pancreatic duct from the pancreatic head up to the tail of the pancreas and was also communicated with a large cystic cavity behind the gastric antrum **(Figure 3)**. Upper gastrointestinal (GI) endoscopy was requested and a fistulous tract was identified at the level of the posterior surface of the antrum through which mucinous material was exited **(Figure 4)**.

Exploratory laparotomy was performed. The intraoperative finding **(Figure 5)** evidenced a communication of the supra-aponeurotic collection with a cystic tumor that involved the pancreas and the posterior surface of the stomach. The cystic tumor was communicated with the gastric antrum as was reported by the upper GI endoscopy. Intraoperative ultrasound confirmed the dilation of the main pancreatic duct in the pancreatic head without evidence of pancreatic tumor

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Abbreviations IPMN intraductal papillary mucinous neoplasm; MRCP magnetic resonance cholangio-pancreatography
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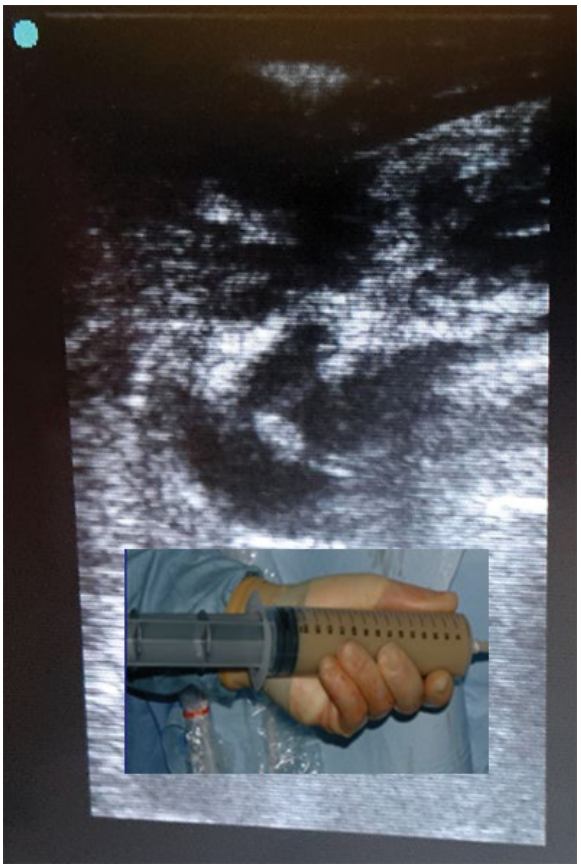

Figure 1. Ultrasound showing fluid supra-aponeurotic collection. Purulent material obtained by puncture.

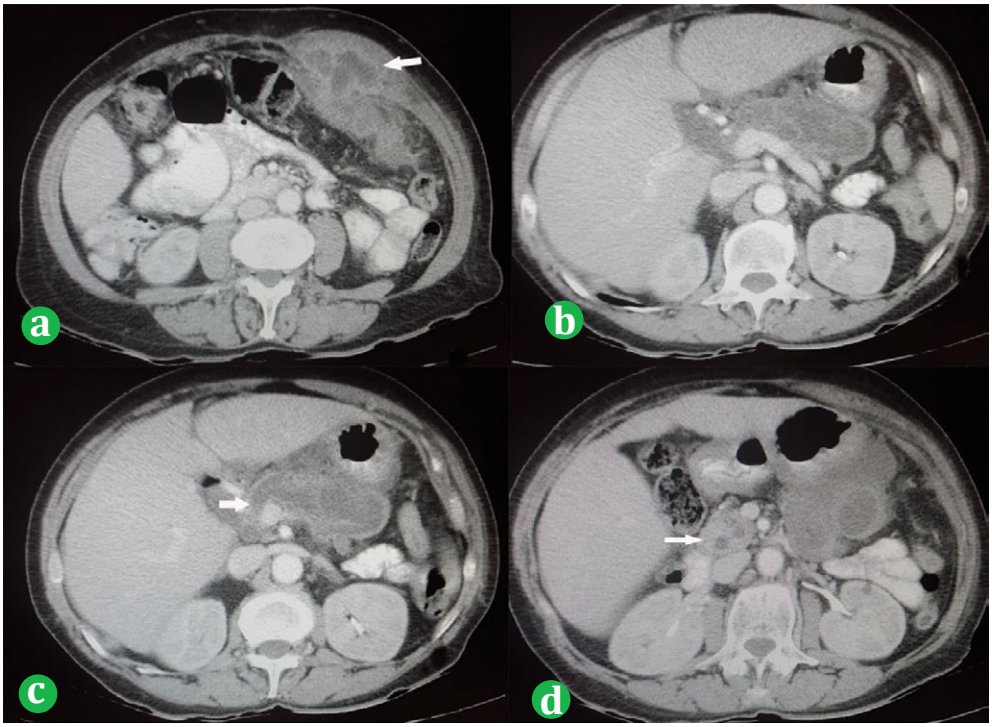

Figure 2. **(a).** CT scan, white arrows points supra-aponeurotic collection. **(b).** CT scan with cystic mass behind the stomach which involves the pancreas body. **(c).** White arrow shows Wirsung duct dilation. **(d).** White arrow points uncinate process with pancreatic duct dilation.

or lithiasis inside the duct. Left pancreatectomy with splenectomy and antrectomy was performed, sectioning the pancreas at the neck. Frozen section of the pancreatic duct surface revealed severe dysplasia with foci of carcinoma at the main duct. Resection was completed by performing pancreatoduodenectomy **(Figure 6)**. The pathologic analysis revealed intestinal-type cystic papillary mucinous neoplasia with foci of high-grade dysplasia and invasive colloid-type carcinoma. Stomach did not showed tumor invasion and the fistulous track showed mucinous material.

The patient was discharged 10 days after surgery without complications.

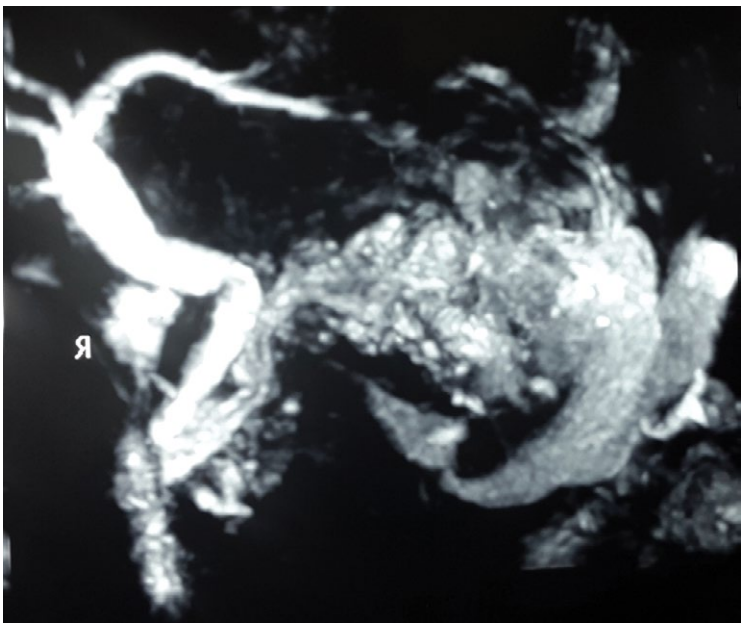

Figure 3. MRCP showing main pancreatic duct dilation and associated cystic lesion.

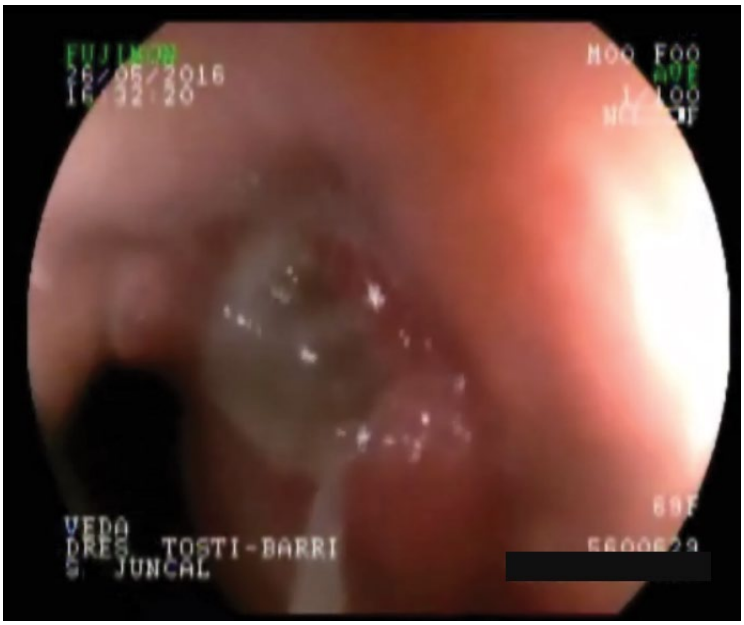

Figure 4. Upper GI endoscopy. Fistulous tract with mucinous material.

DISCUSSION

IPMN were first classified into a unified diagnosis by the World Health Organization in 1996 [2]. There are two types of presentation of this disease. The Type I involves the main pancreatic duct and has high incidence of malignant degeneration and Type II affects peripheral pancreatic ducts, having an incidence of malignant degeneration much lower than type I [3, 4].

The type I IPMN is characterized by a diffuse or segmental dilation of the Wirsung duct over 0.6 centimeters [5]. However, when analyzing the imaging studies, it must be considered that there are other pathologies that may cause dilatation of the pancreatic duct. Usually the IPMN dilatation is uniform and regular, unlike the dilation in the chronic pancreatitis which is characterized by an irregular dilation of the pancreatic duct. In some situations dilatations of IPMN can be confused with obstructive dilatations produced by small head tumors. The best diagnostic method to assess the pancreatic duct is the MRCP. The CT scan has special utility to detect the presence of an associated tumor and the related complications. On the other hand, the CT scan has the utility to rule out other pancreatic pathologies that can lead to dilatation of the pancreatic ducts. For a correct differential diagnosis it is essential to assess the clinical history of the patient.

In the analysis of the present case, there are three differential diagnoses. The first one is complications of pancreatic necrosis. The retrogastric fluid collection on CT scan and the wall abscess are images compatible with walled off pancreatic necrosis. However, this diagnosis was ruled out because the patient did not report history of any episode of acute pancreatitis. The second differential diagnosis is the rupture of a pseudocyst secondary to chronic pancreatitis. The cystic image on the CT scan and

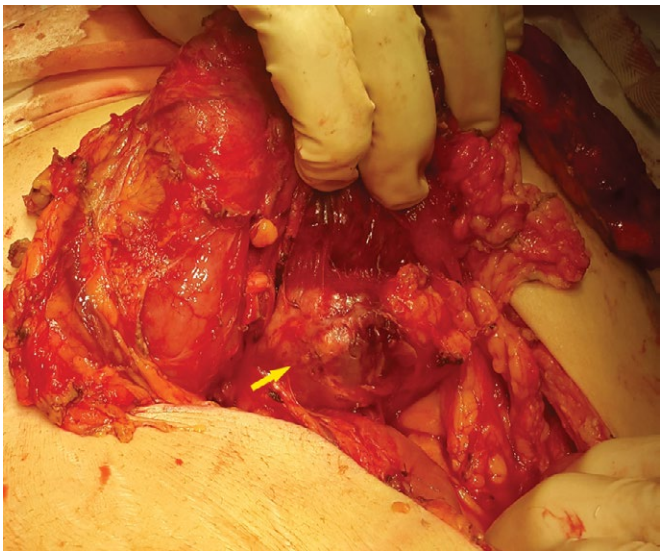

Figure 5. Intraoperative image. Yellow arrow points cystic tumor behind the stomach.

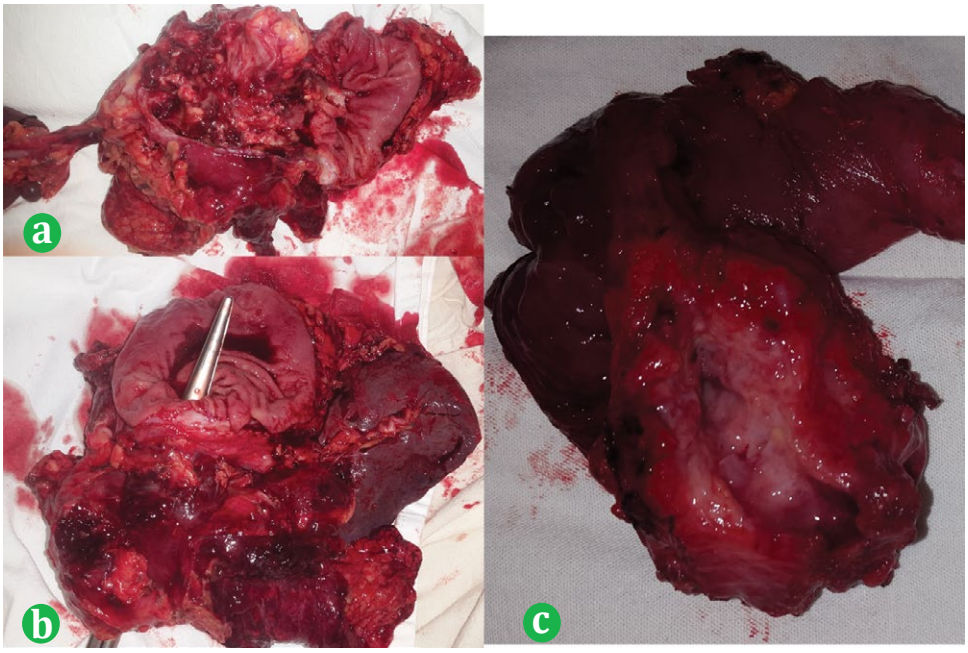

Figure 6. Specimen **(a).** Spleen, pancreas, stomach and open cystic tumor. **(b).** Scissors through the fistulous tract between stomach and the pancreatic tumor. **(c).** Pancreatoduodenectomy specimen.

MRCP is compatible with a chronic perforated and infected pseudocyst. But the patient did not have history of alcohol consumption, and there were lack of the characteristic images of chronic pancreatitis, such as intraductal or parenchymal calcifications or irregular Wirsung dilatation. Another differential diagnosis to consider is that cystic image is a retentional cyst caused by a pancreatic head tumor. This type of dilation is not frequent but has been described [6] and should always be consider in the differential diagnosis. There are situations where the tumor is very small and is not detected on the CT scan or MRCP. However, in this case, the presence of a head tumor was ruled out when performing intraoperative ultrasound. Echoendoscopy would have been useful in the preoperative evaluation to rule out the presence of a tumor in the pancreatic head but would not have changed the surgical behavior.

Typical presentation symptoms of IPMN are acute pancreatitis, abdominal pain, diabetes, exocrine insufficiency, or jaundice. The presentation in this patient with fever and wall abscess, makes it a totally atypical presentation which made us doubting about the diagnosis of IPMN.

The other issue to be discussed in this case is the extent of surgical resection. Studies have suggested that IPMNs grow according to the adenoma-carcinoma sequence, and therefore various histopathologies such as adenoma, Carcinoma in situ (CIS), and invasive carcinoma can be seen in the same patient. When deciding on a therapeutic strategy, it is very important to choose a suitable treatment method for each patient [7, 8, 9, 10]. In this patient a left pancreatectomy with splenectomy and antrectomy was performed and the pancreas was sectioned in a place where the caliber of the Wirsung duct was smaller than the rest of the pancreas. Frozen section revealed severe dysplasia with foci of carcinoma, forcing the total pancreatectomy and duodenectomy. Frozen section with mild or even

moderate dysplasia is not associated with a higher rate of recurrence than in negative cases [8], allowing the preservation of pancreatic parenchyma and avoiding the complications of total pancreatectomy. However, these patients should be closely monitored with MRCP or CT scan. If there is local invasion of other organs, an extended surgical resection is indicated.

CONCLUSION

In conclusion, this is an atypical presentation of type 1 IPMN, but the diagnosis was suspected in the preoperative period with the CT scan, the MRCP and the upper GI endoscopy. It is important to know all the possible differential diagnosis to decide the best surgical procedure. In cases of invasive type 1 IPMN involving other organs (like the stomach in this case), an aggressive surgical resection is the best choice for the patient.

Conflict of Interest

The authors declare that there is no conflict of interests regarding the publication of this paper.

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