Case Report

Total Pancreatectomy in Invasive IPMN: Case Report

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Introduction

Intraductal papillary mucinous neoplasm (IPMN) was first described in 1982¹ and has become an increasingly more common diagnosis among the pancreatic diseases.² It is characterized by an epithelium neoplastic producer of mucin with papillary growth pattern that substitutes the pancreatic ductal epithelium with varied degrees of invasion.³

The presentation varies from the absence of symptoms to states of recurring abdominal pain, repetitive pancreatitis, steatorrhea and diabetes mellitus.⁴

The decision for pancreatectomy is based on the presence of symptoms, the patient's clinical condition and the risk of lesion malignancy.⁵The entire pancreatic duct is under risk, with reports of multifocality and random lesions in up to 15% of the cases.²⁻³

A partial pancreatectomy, since a R0 resection is possible, is the accepted therapeutic option; however, in cases of multifocal disease, a total pancreatectomy can be necessary.⁶⁻⁷

We relate the case of a 45-year-old man with invasive IPMN involving the entire pancreatic duct, the employed treatment and patient evolution.

Case Report

A 45-year-old male patient sought the Department of Abdominal Surgery for evaluation of a discovered pancreatic nodule in routine exams. Two years prior, patient presented an episode of acute pancreatitis, at that time without etiological diagnosis. Patient denied abdominal pain, jaundice, fever or cholangitis, as well as alcoholism, dyslipidemia, diabetes mellitus and cholelithiasis. Patient had hypertension controlled with medication. The physical exam was without particularities.

Computerized tomography of the abdomen revealed a hypervascular nodule in the transition of the pancreatic head to body, 2.6 cm in the largest diameter, with dilation of the amount of the pancreatic duct and atrophy of the body and pancreatic tail (Figure 1). There was no dilation of the biliary vias or sign of involvement of the superior mesenteric vessels.

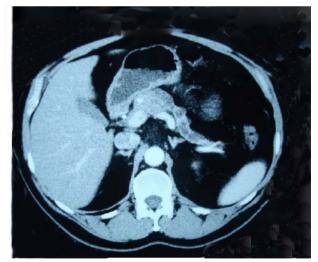


Figure-1 solid hypervascular nodule in the transition of the pancrehead to body, with dilation of the amount of the pancreatic duct and atrophy of the body

Correspondence Eduardo Zanella Cordeiro Rua Antônio Prudente, 211-Liberdade 01509 900 São Paulo Brazil Phone: 11 21895000 ramal 5119 E-mail eduardo.zanella@ig.com.br The fasting glucose level was of 111mg/dl. The tumorous markers (CEA, CA 19-9) were normal.

In view of a solid and hypervascular pancreatic nodule, surgical treatment was indicated, after staging exams discarded remote disease.

The surgical finding was a tumorous mass in the transition of the pancreatic head to body, with suggestive signs of chronic pancreatitis in the body and tail. Initially, due to the location of the lesion, a *duodenopancreatectomy* was opted for (Figure 2A); however, on sectioning the pancreatic parenchyma, in the height of the body, we observed vegetant lesion in the interior of the principal pancreatic duct, permeated by mucinous secretion (Figure 2B). Frozen section examination pointed to compromised margin for neoplasm. Enlarged resection through the extension of the pancreatectomy for approximately 3cm, and a new frozen section examination was made of the pancreatic margin, which was compromised. In face of the operative discovery of an invasive neoplasm with intraductal extension, not capable of R0 resection through duodenopancreatectomia, a total pancreatectomy was decided upon, making a body and tail pancreatectomy of the pancreatic remainder and splenectomy in monobloc (Figure 2C & 2D).

Reconstruction was made through lateral end hepaticojejunostomy anastomosis and entero-duodenal anastomosis in loop. The pancreatic bed and the biliodigestive anastomosis were drained with two Blake® suction drains. The elapsed surgical time was 7h30m. A concentrate of red-blood cells was transfused.

The control of glycemia was made with insulin in a continuous infusion pump during the first 48 hours. During the remainder of internment, NPH 30 UI



Figure-2 A) product of duodenopancreatectomy; B) pancreatic head opened by the Wirsung duct through the papilla, showing compromised margin; C) pancreatic body with necrotic and friable lesion extended along pancreatic duct; D) operating bed at end of resection

insulin was used in the morning and regular insulin (rescue therapy) in agreement with glycemia, with the glycemia values varying between 100 and 256mg/dl. The patient received ICU discharge on the second postoperative day. There was no fistula; the drains were removed and hospital discharge was given on the tenth postoperative day. Weightloss was 12 kg in the first month, with stabilization thereafter.

Histopathological tests revealed a papillary mucinous carcinoma of 5.6cm, presenting invasion of the peripancreatic tissues, with intraductal component affecting the entire extension of the Wirsung duct (Figures 3A & 3B). Absence of veined or lymphatic invasion, with one in sixteen lymph nodes compromised; free margins, staged as pT3N1M0.

After postoperative recovery, initiated adjuvance with gencitabine 1000mg/m² (D1, D8, D15 and D28), 6 cycles. The continuation is 6 months and there is no sign of disease in activity. The glycated hemoglobin is 7.5%.

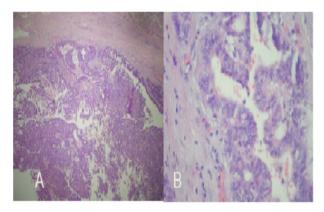


Figure-3 A) pancreatic duct filled by neoplasm with papillary differentiation; B) greater augmentation revealing the presence of mucin-filled vacuoles

Discussion

The World Health Organization classification divides IPMN into invasive and noninvasive. 3 Radiologicaly, the lesions can be divided in accord with the involved duct in IPMN as main duct, secondary duct and combined form 8 with the risk of malignancy being 64%, 18% and 40% respectively²

There is consensus that main duct and combined lesions are of surgical indication, as well as symptomatic lesions of the secondary duct for the risk of associated malignancy. The indication of surgery in lesions larger than 3cm or with solid mural component is a tendency.⁹

Secondary duct lesions of less than 3cm,

asymptomatic and without solid component can be accompanied with safety.¹⁰⁻¹¹ The analysis of CEA, CA 19-9 and CA 72-4 of the lesions, obtained through endoscopic ultrassound has negative predictive value of 100% for malignancy (when associated) and aids when conservative conduct is opted for.¹²

Patients with invasive IPMN have 5-year survival, when paired by staging, similar to patients with ductal adenocarcinoma ($31\% \times 24\%$), with overall recurrence rates of 58% after curative resection.³

The surgical margin is directly related to survival. In a series of 208 operated IPMNs, the average survival was 119 months for patients operated with free margin, 62 months when the margin was compromised by benign (noninvasive) neoplasm and 11 months when compromised by invasive neoplasm.3 Therefore, in the case of high degree dysplasia or invasive neoplasm in the margin, margin enlargement or re-resection is recommended⁵

Frozen section examination is an indispensable tool and some authors defend the intraoperative pancreatoscopy to aid in the extension of the resection⁵

When a R0 resection is not possible with a partial pancreatectomy, the total pancreatectomy becomes an option, and its morbidity is justified by a gain of the point of view of disease control, translated through the increase of survival in selected cases.6

In specialized centers, the total pancreatectomy is made with similar morbidity-mortality to a duodenopancreatectomy and with an acceptable quality of life; improvement in insulinic replacement and of the exocrine pancreatic secretion consequent.¹³

The postoperative evolution was very favorable with glycosylated hemoglobin of 7.5%, similar to that stated in the literature.¹³ The patient did not present signs of exocrine insufficiency, which was reflected in the stabilization of weight and in the absence of steatorrhea.

Even without evidence for adjuvance in invasive IPMN resection, the option in doing so in the present case was extrapolated from the results obtained with ductal adenocarcinoma.¹⁴⁻¹⁵

In short, the total pancreatectomy can be necessary in cases of invasive IPMN, multifocal or with involvement of the entire main duct, without eliminating to be offered the intention of providing a R0 resection. In spite of the limitations related to the total pancreatectomy, the same is a viable and safe option in selected cases.¹³

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