

Chapter 286

Surgical approach in young patient with autoimmune pancreatitis type I

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1 INTRODUCTION

Chronic pancreatitis is a benign inflammatory disease that results in fibrotic replacement of the pancreatic parenchyma, generating a syndrome characterized by changes in pancreatic and structural function.

The clinical picture may be asymptomatic. Whenntomatic the patient may present pain with several periods of recurrence, simulating reaggravation of pancreatitis or continuous pain. The pain may not be associated with the severity of the changes in the pancreas, being overlapping of the pancreatic changes associated with pressure, ischemia, or inflammation in the pancreas, in addition, the patient may develop allodynia and hyperalgesia. As for steatorrhea, there must be a loss of 90% of the function of the pancreas for it to occur. It usually appears after 5 to 10 years of disease progression.

As for laboratory changes, the patient may present with normal amylase and lipase due to the destruction of acinar cells. Bilirubin and alkaline phosphatase, when elevated, may indicate compression of the intrapancreatic choledochal tissue due to fibrosis and edema.

Regarding imaging exams, tomography and cholangioresonance imaging were more accurate, with sensitivity between 50-80% and specificity of 90% in some studies.

The tests used to evaluate pancreatic function can be direct as the test with secretin or indirect as quantification of fecal fat, chymotrypsin, or fecal elastase, among the indirect tests, the latter has greater sensitivity.

Chronic pancreatitis has several etiologies among them the most common are alcoholic, idiopathic, genetic, obstructive, and autoimmune.

The complications are exocrine insufficiency, with osteopenia in 40% and osteoporosis in 25% of cases, and endocrine characterized by type 3c diabetes mellitus, chronic pain, choledochal obstruction, slender obstruction, gastroparesis, pseudo arterial aneurysm, splenic vein thrombosis, ascites, pleural effusion, and adenocarcinoma.

This study aims to report a case of chronic autoimmune pancreatitis type II requiring surgical surgery.

2 CASE REPORT

RMBF, male, 20 years old, born in Jundiaí-SP, from Votuporanga-SP, stock replenishment. Patient with abdominal pain since the age of five. At the time, acute appendicitis was suspected, and was submitted to conventional appendectomy. For 15 years, the patient persisted with epigastric pain attacks with irradiation to the back that took him to the emergency room at least once a year.

At the age of 14, an investigation was initiated in the city of origin, through abdominal tomography, a pancreatic tumor was suspected, and a patient was then referred to the cancer reference center in Barretos-SP. He was submitted to biopsy by Endoscopy, which was negative for neoplasia, with negative IgG4, also presenting normal serum IgG4. The hypothesis of autoimmune pancreatitis type II was proposed.

In 2017 the patient began follow-up with the bile duct team, using pancreatin 60000 ui/day and corticosteroids in periods of exacerbation of the disease.

In April of 2022, he was hospitalized due to new abdominal pain, fever, nausea, vomiting, heartburn, and a weight loss of 16 kg in four months. At admission, the patient was in a regular general condition, slightly discolored and dehydrated, anicteric, tachyca, normotensive, with a flaccid abdomen, painful to deep palpation of the epigastrium, without signs of peritonitis. At the time, he presented positive serology for dengue, with the following laboratory tests: canalicular changes and transaminases (FAL 1008; GGT 1309; TGO 438; TGP 118) and mild elevation of bilirubin (BBT 1.48/ BBD 1.1), other tests within the normal range. An abdominal resonance was performed, which showed non-uniform dilation "in rosary" of the main pancreatic duct in its entire extension, measuring up to 16 mm, with dilation of the bile ducts, from the ampullary region to the hepatic periphery, measuring up to 17 mm, without filling failures in the ductal lumen. ERCP was requested, in which erosive esophagitis grade C was observed in Los Angeles, a large quantity of gastric stasis, and bulbar stenosis due to edema of the second duodenal portion preventing the progression of the apparatus. Thus, PPIs and a parenteral diet were initiated. Tomography of the abdomen showed a pancreas with reduced dimensions, multiple and sparse alkylifications in the parenchyma,

abrupt thinning of the distal choledochus, with dilation of the intra- and extrahepatic bile ducts upstream, and marked gastric distention.

The patient then presented with the complications of chronic pancreatitis: choledochal stenosis, duodenal stenosis culminating in intestinal subocclusion, and dilation of the main pancreatic duct with calcifications and intraductal stones. Thus, he remained hospitalized in the service for nutrition and preoperative preparation, requiring laboratory improvement, since he presented thrombocytopenia in the presence of dengue. On 05/30/2022 the patient underwent Frey surgery + distal pancreatectomy + splenectomy + cholecystectomy + choledocoplasty + gastroenteric anastomosis with Roux-en-Y reconstruction. In the intraoperative period, a large number of firm adhesions of the pancreas with adjacent viscera was observed, in addition to segmental portal hypertension with important gastric varices. A laminar tube drain was positioned in the left hypochondrion and a Kher drain was in a distal choledochus. The approximate surgical time was eight hours, with a transfusion of a packed red blood cell.

The patient was referred to the ICU for the postoperative period. In the first two days, there was a significant worsening of renal function (Cr 2.8/ Ur 154) and the need for vasoactive drugs in high doses, in addition to significant volume expansion, about 8L in the 1st PO.

A corticosteroid was maintained, already in use before surgery. Started meropenem, vancomycin, and octreotide, this remained for five days. On the third day, clinical improvement was observed with the weaning of vasoactive drugs, being switched off at the 5th PO and the patient extubated at the 6th PO. In the 7th postoperative period initiated enteral diet by Dobbhoff tube and good acceptance. On the 8th postoperative day, the patient was discharged from the ICU.

The biopsy of the specimen evidenced the pancreas with extensive fibrosis, periductal mixed inflammatory infiltrate, and chronic pancreatitis favoring the diagnosis of "autoimmune pancreatitis type 2". Gallbladder with polypoid cholesterosis. Spleen with fullness and vascular congestion. Neoplasm-free reactional lymph nodes.

The patient was discharged from the hospital on the 20th postoperative day, accepting an oral diet, with a Kher drain in low output, and a left laminar tube drain removed.

In the postoperative period, type 3c diabetes mellitus were observed with the need for insulin and exocrine insufficiency with return of pancreatin.

3 DISCUSSION

Defined by Yoshida et al. in 1995, IAP corresponds to an entity marked by the fibroinflammatory process rich in IgG4 cells, when of type I. It is a rare entity, more prevalent in males and over 50 years of age. Painless jaundice associated with obstruction of the common bile duct is the most typical form of presentation.

Type I AIP is related to IgG4. Usually, serum levels are elevated in pancreas biopsies as well. It may be associated with extra-pancreatic conditions in 60% of cases, such as biliary stenosis, hilar

lymphadenopathy, sclerosing sialoadenitis, retroperitoneal fibrosis, and interstitial nephritis. Type II AIP is limited to the pancreas and is not associated with infiltration of positive IgG4 plasma cells or elevations in serum levels. However, in 15-30% of cases, it may be associated with inflammatory bowel disease

AIP corresponds to an important clinical condition since early therapy can improve the patient's morbidity and mortality. Initially, the treatment is clinical, and the surgical treatment is reserved for refractory pain or complications.

The Mayo Clinic Diagnostic Criteria for autoimmune pancreatitis comprises the presence of one or more of the following items:

1. Histological Diagnosis.
2. Changes on imaging, with the diffusely enlarged pancreas, with expressionless edges and delayed enhancement with or without capsule-like edge.
3. High levels of IgG4 in serum.
4. Involvement of other organs (alluvial glands - Sjögren's syndrome), stenosis of the bile duct, pulmonary nodules, autoimmune thyroiditis, and kidney.
5. Response of pancreatic and extra-pancreatic manifestations to corticosteroid therapy.

Major radiological examinations include computed tomography, cholangio resonance, and endoscopic ultrasound.

Surgical intervention is reserved when the debilitating pain is persistent or due to complications of the disease. Surgical options include simple drainage procedures, gastroduodenal pancreatectomy (Whipple), cephalic pancreatectomy (Frey's technique), and total and distal pancreatectomy. The Frey technique aims to remove the obstructive mass and allow drainage of the pancreatic duct, consists of a limited resection of the pancreatic head with preservation of the duodenal wall, and is associated with low morbidity and mortality and better results in the patient's symptomatology.

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