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Autoimmune Pancreatitis or Pancreatic Cancer?☆

¿Pancreatitis autoinmune o cáncer de páncreas?

Autoimmune pancreatitis (AIP)[1] is a benign fibroinflammatory disease that frequently presents as obstructive jaundice, which may or may not be associated with a pancreatic mass. It shows characteristic histological changes, and there is excellent response to corticosteroid therapy, as published in the 2011 International Consensus on AIP.[2]

We report the case of a 59-year-old male with no prior history of interest who was transferred to the general surgery department due to symptoms compatible with post-ERCP acute cholecystitis after having been admitted to the Gastrointestinal Department because of obstructive jaundice secondary to a mass in the head of the pancreas. CT scan showed evidence of increased pancreatic gland size, related with acute pancreatitis vs a neoformation, as well as dilatation of the intra and extrahepatic bile duct (Fig. 1). Endoscopic ultrasound showed that the entire pancreatic gland was increased in size, with a neoformation in the head measuring 43×32 mm, which was in contact with the superior mesenteric vein by 12 mm. The Wirsung duct had a beaded appearance with a clear caliber throughout. The extrahepatic bile duct was dilated and had defined walls, with no interior content, but the distal section was displaced by the previously described mass. ERCP revealed irregular stenosis of the proximal intrapancreatic common bile duct and dilatation of the main bile duct. Lab analyses showed elevated total bilirubin, at the expense of direct bilirubin (3.2 mg/dL), normal tumor markers and slightly elevated immunoglobulin G4 169 mg/dL (adults: 9–104 mg/dL). The pancreatic biopsy taken during endoscopic ultrasound was not conclusive for malignancy.

Given the poor evolution of the acute clinical symptoms and the uncertain pancreatic diagnosis, we decided to

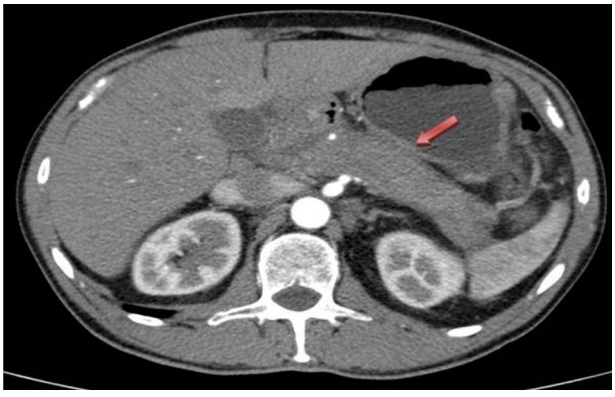

Figure 1 – CT image showing the pancreatic gland that is increased in size.

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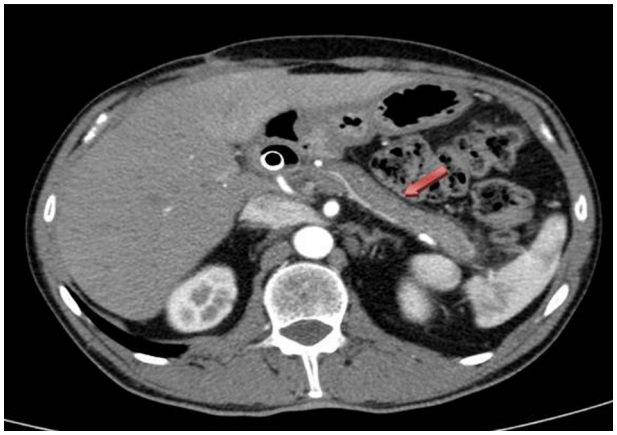

Figure 2 – CT image demonstrating the decrease in the inflammatory process after the initiation of corticosteroid therapy.

perform urgent surgery, and a cholecystectomy and pancreatic biopsies were performed. The postoperative period progressed favorably, and the patient was discharged on the 7th day post-op. The pathology results from the pancreatic biopsies were compatible with type 1 AIP because histologically there was extensive fibrosis with an isolated whirl pattern, lymphoplasmacytic aggregates, polyclonal plasma cells, many of which had IgG4 expression in some fields of up to >50 positive cells/high power field. Given these findings, treatment was initiated with corticosteroids, which achieved radiological improvement as seen on CT of the pancreatic inflammatory process (body and tail) one month after the start of treatment (Fig. 2).

AIP is an uncommon disease (prevalence: 2% of chronic pancreatitis) whose symptoms (obstructive jaundice) and radiology results (pancreatic mass or obstructive bile duct lesion)[3] are similar to those of pancreatic cancer. This leads to a high percentage of pancreatic surgical resections in a benign disease that otherwise responds well to treatment with corticosteroid therapy.

Histologically, AIP presents well-defined changes that are easily distinguishable from changes that occur in other types of pancreatitis (chronic alcoholic or obstructive), since the lymphoplasmacytic infiltrate is dense and more pronounced around medium and large-sized ducts, compressing the ductal lumen (horseshoe or star duct images are very characteristic of AIP), which differs from the ductal dilatation characteristic of chronic pancreatitis of other origins. Characteristic findings of AIP include lymphoplasmacytic sclerosing pancreatitis (LPSP) without granulocytic lesions; meanwhile, the pathognomonic findings of type 2 AIP are idiopathic duct-centric pancreatitis (IDCP) with granulocytic lesions.[4]

Currently, there are no specific serological markers for the diagnosis of AIP. Elevated serum levels of IgG4 is a characteristic finding of type 1 AIP (type 2 AIP never presents increased IgG4). Some studies accept the cut point of 135 mg/dL for IgG4 values suggestive of AIP versus pancreatic cancer with a sensitivity and specificity of 95 and 97%, respectively.[5]

In accordance with the International Consensus for the diagnosis of AIP,[2] the recommended treatment is prednisone at an initial dose of 35–40 mg/day[6] or 0.6–1 mg/kg/day for 4 weeks. Afterwards, if there is clinical and radiological response, the dose is gradually reduced over 3–4 weeks. Some groups recommend maintaining treatment with corticosteroids at low doses (2.5–5 mg/day) for 3 years in type 1 AIP, given its elevated rate of recurrence. The persistence of elevated IgG4 and proximal stenosis of the bile duct after treatment are factors for recurrence. The reintroduction of corticosteroids or the start of immunosuppressants are therapeutic alternatives used in these cases.[7]

Given the reported evidence of pancreatic adenocarcinoma or intraductal papillary mucinous neoplasm in patients with AIP,[8,9] close follow-up of these patients is necessary.

Thanks to the surgical experience accumulated in patients with acute pancreatitis (technical difficulties: vascular injury and hemorrhage[10]), we propose that in patients with uncertain diagnosis (atypical findings: atypical clinical evolution, involvement of other organs, peripheral halo around the mass, lack of pre-stenotic duct dilatation or the elevation of serum IgG4 to diagnostic levels), it is important to reconsider the situation and assess surgical biopsies prior to pancreaticoduodenectomy or to assess the response to corticosteroids, as long as pancreatic cancer has been ruled out with a high level of certainty (pancreatic masses with an atypical radiological image and negative cytology for malignant cells).

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Conflict of interests

The authors have no conflict of interests to declare.

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Abdominal Cocoon: A Rare Cause of Intestinal Obstruction☆

Síndrome de Cocoon: una rara causa de oclusión intestinal

Sclerosing encapsulating peritonitis (SEP) is a cause of intestinal obstruction of unknown etiology. Also known as abdominal cocoon syndrome, it is characterized by a fibrocollagenous membrane that either completely or partially encompasses the small bowel.[1] This causes patients to repeatedly seek medical care for symptoms of intestinal obstruction. Both symptoms as well as radiological images are non-specific, and diagnosis therefore requires elevated suspicion.[2,3]

We present the case report of a 54-year-old male patient with no prior history of interest, except for bilateral endoscopic hernioplasty that was completely extraperitoneal and recent elective surgery for cholelithiasis. It was during this latter surgery that sclerosing peritonitis was incidentally diagnosed when a whitish membrane that affected the small bowel and descending colon was found intraoperatively, blocking the supramesocolic compartment as well.

Afterwards, the patient came to the Emergency Department on repeated occasions due to colicky abdominal pain and subacute intestinal obstruction, so he was hospitalized once again. Physical examination detected abdominal distension associated with a palpable mass in the mesogastrium. A CT scan demonstrated medialized small intestinal loops, some with wall thickening, that were adhered amongst themselves and to the anterior abdominal wall; there was also free interloop fluid, which was probably related to adhesion-related syndrome. We decided to schedule surgery, and found a membrane covering the jejunum and a large part of the ileum (Fig. 1). We performed almost complete exeresis of the membrane, that could be separated from the serosa of the intestine. The integrity of the bowel loops was confirmed up to the ileocecal valve; no perforations were observed, and resection was not required. The patient's condition progressed favorably, and he was discharged 6 days after surgery with complete resolution of the symptoms.

Abdominal cocoon syndrome is a very uncommon disease of unknown etiology that is classified as either idiopathic or secondary. This latter form is more common, and there have been descriptions of cases of secondary SEP associated with peritoneal dialysis, tuberculosis, treatment with beta-blockers, familial Mediterranean fever, etc. The idiopathic form of SEP is relatively more frequent in tropical countries,

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