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Type 2 Autoimmune Pancreatitis and Ulcerative Colitis an Uncommon Association: About One Case

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Abstract **Case Report**

Autoimmune pancreatitis is one of the less recognised associations and remains poorly understood. The overall reported prevalence of autoimmune pancreatitis in inflammatory bowel disease patients is 0.4% [5, 6] considering the only two studies in the literature specifically examining this issue, and that concern Asian populations. Autoimmune pancreatitis in the setting of inflammatory bowel disease is challenging to diagnose but a clinically important entity to recognize and treat. Here we report a case of a 43-year-old women followed for ulcerative colitis, presented with epigastric pain and dyspepsia symptoms associeted to steatorrhea and 12 kg weight loss in 2 months. Laboratory tests found fasting plasma glucose at 1.27 g/l. HbA1c was 8%, IgG4 Levels were normal and the fecal elastase level was low. Magnetic resonance imaging of abdomen revealed a swollen pancreas especially in the uncus. The Bilio-pancreatic endoscopic ultrasound showed heterogeneous hypoechoic lesion of the pancreas head. And the biopsy of the pancreas revealed signs compatible with autoimmune pancreatitis type 2. A final diagnosis of ulceratice colitis associated autoimmune pancreatitis type 2 was made and the patient was treated with corticosteroids, with impressive improvement during his follow-up. In the context of inflammatory bowel disease, autoimmune pancreatitis is a rare and challenging to identify condition. The creation of clear guidelines and diagnostic standards for autoimmune pancreatitis is anticipated to improve awareness of the condition and result in a rise in the number of confirmed cases during the ensuing years.

Keywords: Autoimmune pancreatitis, inflammatory bowel disease, fasting plasma glucose.

INTRODUCTION

The inflammatory bowel diseases (IBD), Crohn's disease (CD) and ulcerative colitis (UC), are multisystem diseases that cause chronic relapsing inflammation within the gastrointestinal tract, and are associated with extraintestinal manifestations in up to 50% of patients [1]. Autoimmune pancreatitis (AIP) is one of the less recognised associations and remains poorly understood.

AIP is a benign fibroinflammatory disease of the pancreas first reported in 1961. It acquired the name, "autoimmune", later in 1995, because it was associated with hypergammaglobulinemia, was accompanied by autoantibodies and responded dramatically to corticosteroids (CS) [2]. On imaging, the pancreatic inflammatory process is classically seen as a sausage-shaped pancreas with an irregular narrowing of the pancreatic duct [3].

Two types of AIP were subsequently recognized according to clinical profile, histopathological pattern and natural history; namely, type 1 AIP or lymphoplasmacytic sclerosing pancreatitis (LPSP) and type 2 AIP or idiopathic duct-centric pancreatitis (IDCP) [2]. Box 1 summarizes the typical features of each type of AIP [4].

Box 1: Comparison between the clinical profiles of type 1 and type 2 autoimmune pancreatitis.

	Type 1 AIP (lymphoplasmacytic sclerosing pancreatitis)	Type 2 AIP (idiopathic duct-centric pancreatitis)
Demographics	7th decade of life 3:1 male predominance	5th decade of life Equal gender distribution
Clinical presentation	Painless jaundice	AP and abdominal pain Painless jaundice
Association with IBD	Rare	May be present in up to 1/3 of patients
Pancreatic imaging	Diffuse pancreatic parenchymal enlargement with delayed enhancement Long or multiple strictures of the main pancreatic duct	
IgG4 elevation	Increased in 2/3 of patients	Usually normal
Pancreatic histology	Periductal lymphoplasmacytic infiltration Obliterative phlebitis Storiform fibrosis Increased IgG4 positive cells (>10 per HPF)	Granuylocytic infiltration of duct wall Absent or scant IgG4 positive cells (0–10 per HPF)
Involvement of other organs	Proximal biliary tree, salivary glands, kidney and retroperitoneal fibrosis	No
Response to steroids	Excellent	
Recurrence	Common	Rare

AP, acute pancreatitis; HPF, high power field.

Box 1: Comparison between the clinical profiles of type1 and type 2 autoimmune pancreatitis [4].

Therefore, we report a case of type 2 AIP associated with ulcerative colitis.

CASE REPORT

A 43-year-old female presented with epigastric pain and dyspepsia symptoms associeted to steatorrhea and 12 kg weight loss in 2 months. Her past medical history included UC diagnosed 5 years earlier was treated with 5-ASA with no follow-up. Laboratory tests found fasting plasma glucose at 1.27 g/l. HbA1c was 8%, IgG4 Levels were normal and Low levels of fecal elastase. Magnetic resonance imaging of abdomen revealed an enlarged pancreas (figure1).

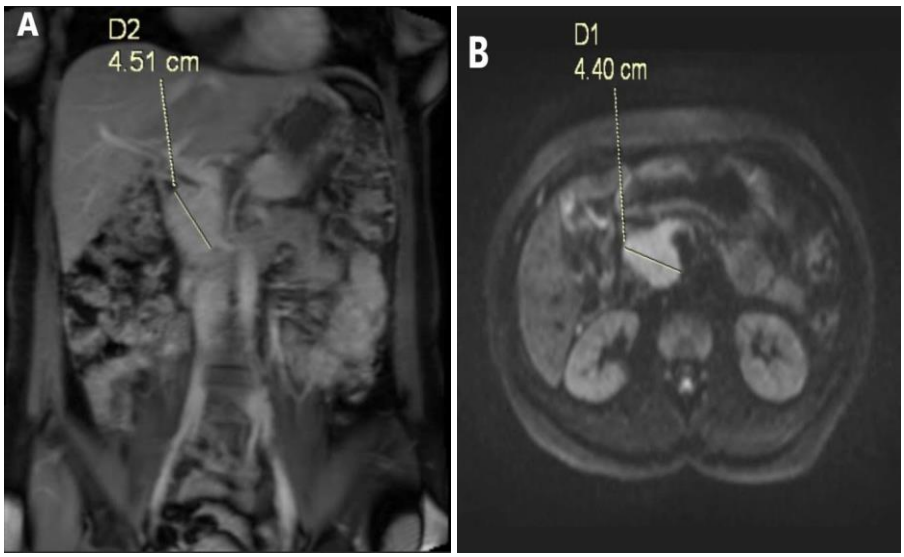

Figure 1: MRI: A- coronal section: globular appearance of the pancreatic gland with localized hypertrophy of the incus. B- Axial section

The Bilio-pancreatic endoscopic ultrasound (EUS) showed heterogeneous hypoechoic lesion of the pancreas head without vascular repercussions and the histopathological analyses (figure2) showed pancreatic ductules dissected by fibrosis, around which we find an inflammatory infiltrate rich in plasma cells. These findings were felt to support the diagnosis of AIP. The immunohistochemical study revealed anti IgG4 antibody negative but anti CD138 antibody positive in mature plasma cells representing 30% of the inflammatory infiltrate.

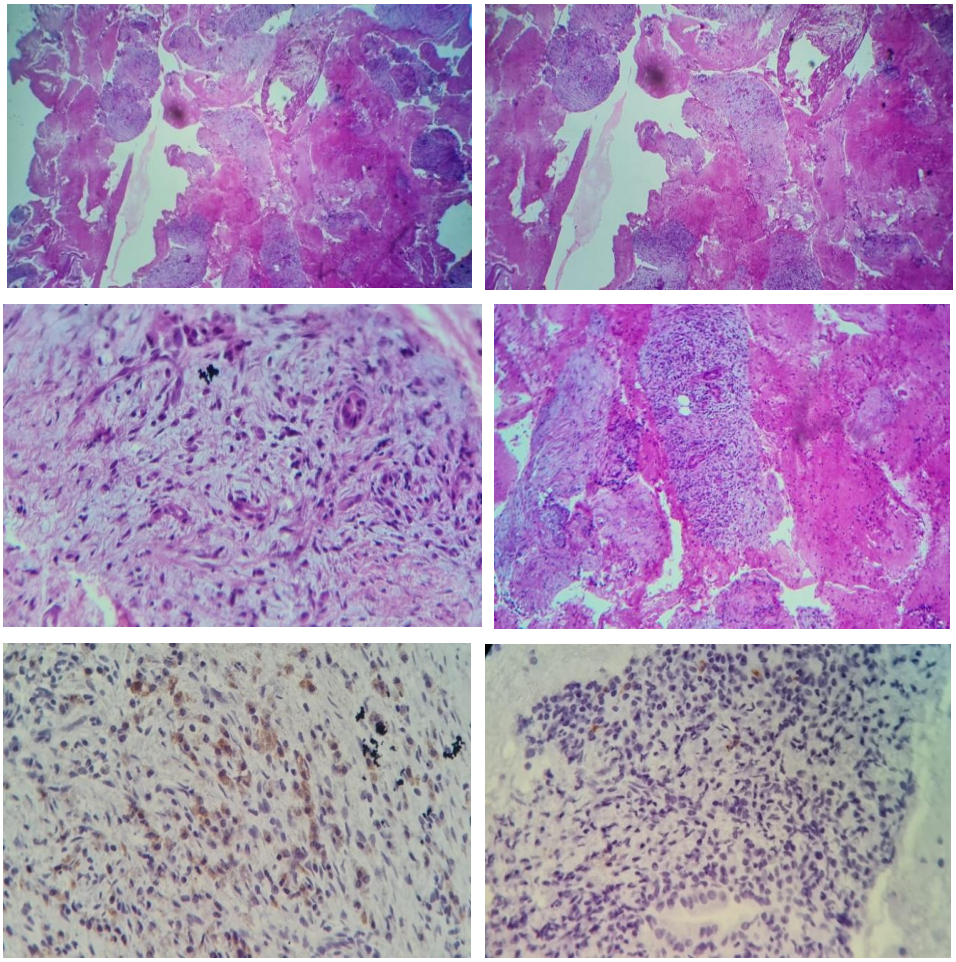

Figure 2: Anatomopathological examination shows pancreatic ductules dissected by fibrosis around which an inflammatory infiltrate rich in plasma cells is found; a fibrous tissue rich in inflammatory cells is also noted.
The immunohistochemical study carried out shows a positive anti-CD 138 antibody in mature plasma cells representing 30% of the inflammatory infiltrate, and a negative anti-CD 4 antibody.

Based on these results, the patient was diagnosed as having type 2 autoimmune pancreatitis associated with ulcerative colitis. Treatment with 40 mg per day of prednisolone was started with diabetes monitoring by an endocrinologist.

During their follow-up, the evolution was marked on the clinical level by a clear improvement with the disappearance of pain and diarrhea and a gain in weight. Biologically, there was an improvement of the blood sugar level and of the fecal elastase.

A progressive regression of corticosteroids was considered without relapse.

DISCUSSION

The overall reported prevalence of AIP in IBD patients is 0.4% [5, 6] considering the only two studies in the literature specifically examining this issue, and that concern Asian populations. This 0.4% figure is 100–400 times greater than the prevalence estimated for the general population [7]. Prospective multicenter studies are needed to clarify the true prevalence of AIP in IBD patients.

Shared antigenic molecules between the colon and the pancreas cause an immune response in both organs, which is a potential pathophysiological explanation for the link between IBD and AIP that has been found [8]. The histological similarities between type 2 AIP and UC are also intriguing [6]. Colonic crypt epithelium (cryptitis) and lumen (crypt abscess)

are affected by neutrophils in UC [9]. Similar to type 1 AIP, type 2 AIP also has neutrophils in the lumen and epithelium of the small and medium-sized ducts and pancreatic acini [10]. In turn, smoking (>10 packs per year) has been linked to increased prevalence of related diabetes and more frequent pancreatic injury on imaging [11].

Most IBD-AIP patients appear to be male (64%), young, and UC (80%) positive. Nearly 50% of UC patients have severe or pancolonic disease, while all CD patients have involvement of the colon. The progression of the AIP and IBD do not appear to be affected by one another. Overall, type 2 or Idiopathic Duct-centric Chronic Pancreatitis (IDCP) AIP was more prevalent than type 1 in IBD-AIP patients [4].

Autoimmune pancreatitis in the setting of IBD is challenging to diagnose but a clinically important entity to recognize and treat accordingly The case presented the association of AIP and UC. Our case met criteria for definitive type 2 AIP based on supportive imaging findings level 2 histology with clinical IBD and response to steroids.

Abdominal pain and acute pancratitis (AP) were the most frequent presentations of AIP, and diagnoses of the condition were made more commonly in the third and fourth decades of life. AIP should be taken into account in the differential diagnosis of AP in IBD patients as a result. Obstructive jaundice, back discomfort, and newly diagnosed diabetes were less common symptoms. Patients with and without IBD did, in fact, present with identical clinical characteristics [5, 12]. An abdominal computed tomography or magnetic resonance imaging (MRI) are valid initial tests to identify features consistent with AIP (diffuse pancreatic enlargement with loss of normal lobulated contour, or "sausage-pancreas"); or pancreatic cancer [3]. AIP diagnosis requires a high index of suspicion. The benefit of using MRI to perform cholangio-pancreatography sequences is that it can provide information about the pancreatic duct, which can be useful in IgG4 serum levels should also be measured, and extra-pancreatic symptoms should be looked for. To rule out cancer, EUS with fine-needle aspiration ***(***FNA) is required [13]. In the situation being discussed, EUS with pancreatic biopsy were considered in the diagnostic analysis. Pancreatic histology is the gold standard for the diagnosis of AIP and necessary for a conclusive diagnosis of type 2 AIP, in which the disease is limited to the pancreas and IgG4 levels are typically normal. This is in addition to ruling out malignancy.

Consequently, in routine clinical practice, the choice of whether to get a histology sample should be determined on a case-by-case basis and as part of a multidisciplinary approach, including the radiologist, the operator of the EUS, and the gastroenterologist. Recurrence rates were higher in type 1 (75–100%) than type 2 (15–20%) AIP in IBD-AIP patients [5, 6, 12, 14].

CONCLUSION

In the context of IBD, autoimmune pancreatitis is a rare and challenging to identify condition. The creation of clear guidelines and diagnostic standards for AIP is anticipated to improve awareness of the condition and result in a rise in the number of confirmed cases during the ensuing years.

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