

Atypical Initial Presentation of Crohn's Disease: Inflammatory Ampullary Pseudotumor Causing Obstruction of the Common Bile Duct - A Case Report and Review of the Literature of Pseudotumors Linked to Crohn's Disease

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MRI: Magnetic resonance imaging; CBD: Common bile duct; ERCP: Endoscopic retrograde cholangiopancreatography; IBD: Inflammatory bowel disease; ASA: Aminosalicic acid; EUS: Endoscopic ultrasound

1. Introduction

Crohn's disease extraintestinal manifestations have been well described in the literature and in many cases may precede the diagnosis of the disease. However, the findings of an inflammatory pseudotumor, at different organs of the body, is rare especially those presenting in the peripancreatic (such as ampulla of Vater) and pancreatic area, and obstructing the biliary tract. We report a rare case of Crohn's disease manifesting by a peripancreatic pseudotumor, at the level of the ampulla of Vater, causing an initial clinical presentation of an obstructive jaundice, in an elderly man, in whom a Whipple procedure was performed due to the high suspicion of a malignant peripancreatic tumor.

2. Case Report

A 68 years old man, smoker of 40 pack year, presented to our clinic complaining of one-year history of unintentional weight loss (around 5 Kg) with decreased appetite and intermittent mild abdominal discomfort. Physical examination revealed minimal diffuse tenderness mainly in upper quadrants but no other abnormalities. Blood test drawn and showed microcytic anemia with a hemoglobin of 10,7 so

a screening colonoscopy was done in April 2020 and showed two sessile polyps in the left colon, less than 5mm in size with non-complicated diverticulosis. Polyps were removed and sent to the pathology unit that showed an inflammatory nature with no malignant features. At that time, only first few millimeters of the terminal ileum had been assisted due to a difficult cannulation into the ileocecal valve. Despite those results, the patient persisted to have weight loss with increasing episode of abdominal pain and recently jaundice with and elevated direct bilirubinemia (1,8mg/dl), so an abdominal computed tomographic scan was done and revealed a mild intrahepatic duct dilatation and a dilated CBD reaching 1,5cm with thickening noted at the ampulla of Vater suggesting an ampulloma. A biliary plastic stent was inserted during endoscopic retrograde cholangiopancreatography (ERCP) after revealing an external obstruction of the CBD, where an erythematous ulcerated tumor of the ampullar was identified. Next, a Whipple procedure was performed because all the work up studies were in favor of an obstructive carcinoma. Macroscopical examination of the fragments removed from the stomach, duodenum, pancreatic head and the CBD showed an ulcerated ampulla, measuring 1,3x1,2x0,3 cm extending to the CBD. In addition, an ileal

fragment measuring 8x4x2 cm showed a stenotic lesion measuring 6x4 cm located at 1cm from the nearest pancreatic border. Histologic examination of the resected duodenum and pancreatic tissue and some peripancreatic lymph nodes showed, at the level of the ampulla, an ulcerative lesion composed of an acute leucocytic-fibrinous contents with a granulation tissue of polymorphonuclear cells and, at the level of pancreatic parenchyma, presence of lesions indicative of a chronic pancreatitis with suppurative cystosteatonecrosis. The rest of the histologic examination of the stomach part, CBD, duodenum, wirsung duct and all other lymph nodes was normal with no malignant cells identified. At the level of the resected ileal segment, a diffuse erosion with some crypt atrophy, crypt abscesses and a dense chronic inflammatory infiltrate, moderately active rich in eosinophils associated with hyperplastic reactive lymph node without any signs of malignancy or pathogenic agent, were identified, all of which were compatible with a Crohn's disease. Based on these findings, an inflammatory pseudotumor of the ampulla was linked to the diagnosis of Crohn's disease in this previously healthy patient. The patient started on biological treatment for maintenance therapy for Crohn's disease and after 9 month of the surgery, and to allow maturation of the ileo-ileal anastomosis, a full ileo-colonoscopy was done that showed scattered patchy erosions in the right colon with a periappendicular orifice inflammation and erosive anastomosis ileo-ileal compatible with Crohn's disease, but with clinical improvement of the symptoms.

3. Discussion

On the basis of the pathological and colonoscopic findings, in addition to the clinical follow-up, we can assume that this patient had a definitive diagnosis of a peripancreatic - ampullary pseudotumor linked to Crohn's disease, with a rare initial clinical presentation of weight loss and jaundice due to CBD obstruction.

The extraintestinal manifestation of Crohn's disease are numerous including joints, skin, bones, eyes, kidneys and liver. Ampullary involvement is rare, occurs in 0.5-4% of patient with Crohn's disease, usually associated with upper gastrointestinal Crohn's disease (duodenal and jejunal crohn's) and may present as erosions, ulcerative lesions and pseudotumors. In 2015, an isolated ampullary Crohn's disease was described in an elderly lady, with well controlled disease on infliximab, diagnosed by endoscopic ultrasound (EUS) and ERCP. It revealed a non-obstructing erythematous ampulla with ulcerations, compatible with ampullary Crohn's disease, treated by escalating the treatment to vedolizumab with the resolution of her symptoms. In contrast to our case, where an obstructing ampulloma linked to Crohn's disease was diagnosed on pathological examination after a Whipple procedure. In both cases, no granuloma was seen on pathology [1] (Figure 1).

Another case published in 2009, of Crohn's disease affecting the ampulla and the CBD described in an elderly man, similar to our case in term of the obstructive clinical presentation and the absence of con-

comitant duodenal involvement, but it occurred in a man with already a history of Crohn's disease maintained on 5 ASA [2] (Figure 1).

In the both cases mentioned above, patients had a history of inflammatory bowel disease (IBD) after initial presentation of bloody diarrhea, and during the course of the disease, the ampulla of Vater was affected. What makes our case special, is that the initial presentation of Crohn's disease was an obstructing jaundice due to this ampullary pseudotumor and not the classic presentation of Crohn's disease.



Figure 1: Dilated CBD on scan tomography of abdomen.



Figure 2: Patchy thickened ileum on scan tomography of abdomen.

Different manifestations of pancreatic disease have been also associated with IBD, most of them seem to be drug induced or related to biliary stones, with acute pancreatitis being the most common manifestation in this area with the highest morbidity rate. The first association was identified in 1950 on postmortem pathological examination, when Ball et al found a ductal pancreatic lesion in around 25%

of patients with ulcerative colitis and 38% of patients with Crohn's disease [3, 4]. Various clinical presentation of IBD-associated pancreatitis was reported, from asymptomatic disturbance in pancreatic enzyme, to mild symptoms of acute and/or chronic pancreatitis, to pancreatic insufficiency and very rarely to a pseudotumorous pancreatic lesion causing obstruction of the CBD. In fact, 2 cases were reported in the literature of IBD presenting as pancreatic pseudotumor. The first one described in 2001 in an elderly patient known to have ulcerative colitis on aminosalicylic acid (ASA), diagnosed with a pseudotumorous chronic pancreatitis associated with ulcerative colitis, based on ERCP and cytological analysis of stenosis brushings after being mistaken for a pancreatic ductal adenocarcinoma that led to a surgical resection of the tumor [3]. The second case reported also in 2001, in a patient presenting with jaundice that had undergone Whipple procedure for a suspicious pancreatic tumor, found to be after pathological examination an inflammatory pseudotumor linked to Crohn's disease that was diagnosed 6 months later when he started to have bloody diarrhea [5]. This latter case is somehow similar to our case with difference in that Crohn's disease, in our case, was diagnosed pathologically at the time of Whipple procedure due to the histological findings of the resected ileal stenotic lesion found during surgery. In addition, in our case, the pancreatic tumor was at the level of the ampulla of Vater, in contrast to the both written cases where the tumor was located in the head of the pancreas. In all three cases, Whipple procedure was performed due the high clinical suspicion of a pancreatic cancer. However, Whipple procedure carries a high morbidity and mortality rate approaching the 50% mark despite meticulous surgical technique and standard critical care postoperatively and necessitate a longer follow up with a frequent post operation complication such as pancreatic fistula, wound infection and enteric leakage [6].

Regarding ampullary tumor, these are rare tumors, that can be clinically indistinguishable from tumors of pancreatic head and they are usually histologically divided into a pancreaticobiliary and intestinal tumors [7]. Concerning pancreatic cancer, lesions were divided into a neuroendocrine pancreatic cancer and an exocrine pancreatic cancer with adenocarcinoma type responsible of around 90% of all pancreatic cancer type. In another side, benign pancreatic tumor exists and may manifest as pancreatic cysts (with different types, from serous cystic to mucinous cystic to intraductal papillary mucinous neoplasm), solid pseudopapillary neoplasms and rarely inflammatory pseudotumor of the pancreas [8]. Pancreatic inflammatory pseudotumors are rare benign lesions of the pancreas, that can present as a solid single mass in some case or a cystic mass in other cases, with predominant location in the head of the pancreas. Diagnosis is difficult due to the similarity in clinical presentation and imaging studies with pancreatic cancer with only way of diagnosis based on histopathological examination of the specimen [9]. Endoscopic ultrasound (EUS) with fine needle aspiration may play an important role in diagnosing pancreatic and peripancreatic cancer with a very high negative predictive and

a diagnostic yield higher than computed tomography especially for lesions less than 2-3 cm in size [10]. However, EUS was not done in our case due to financial problems.

As for treatment, regression of the inflammatory pseudotumors associated with IBD are by the use of the maintenance therapy of Crohn's or ulcerative colitis disease. To note that concerning the literature about IBD's associated pseudotumors, three cases were reported in the liver (two published in 2004 in two elderly man diagnosed to have inflammatory pseudotumor on pathology which had been linked later on to Crohn's disease that improved with maintenance therapy of Crohn's disease; and one published in 2009 in young lady diagnosed simultaneously with severe Crohn's colitis and liver inflammatory pseudotumor that regressed with maintenance therapy of Crohn's disease). One case was reported in the cerebellum (published in 2012 in a young man on immunosuppressive therapy for Crohn's disease) and one in the kidney (published in 2016 in a young lady on medical therapy for Crohn's disease that developed, several years after, a kidney mass found to be an inflammatory pseudotumor consistent with Crohn's disease, based on clinical, radiological and pathological exam that showed a non caseating inflammatory granuloma) [11-14].

4. Conclusion

Our case report is one of the rarest initial clinical presentation of Crohn's disease. The importance of this reported case reside in the necessity to include Crohn's disease and/or IBD inducing inflammatory pseudotumor in the differential diagnosis of common bile duct strictures before undergoing such a high complicated surgical procedure, by the meaning of a medical management using the maintenance treatment for Crohn's and/or inflammatory bowel disease instead of surgery, which decrease the morbidity, the mortality related and the length of the hospitalization stay.

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