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Case Report Open Access

Celiac Disease Presenting as Acute Pancreatitis: A Case Report and Review of the Literature

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Abstract

A 12-year-old girl presented with acute abdominal pain and vomiting. Raised serum amylase with the radiological findings supported a diagnosis of acute pancreatitis. Subsequent investigation confirmed celiac disease. Very few children with celiac disease present with an acute pancreatitis. The mechanism by which celiac disease is associated with pancreatitis is discussed here.

Keywords: Pancreatitis; Celiac disease; Abdominal pain

Introduction

Celiac disease is a chronic inflammatory disease of the small intestine caused by lifelong gluten sensitivity [1]. It occurs in genetically predisposed individuals and manifest as an immune-mediated Enteropathy.

Celiac disease has predilection for Western Hemisphere, affecting up to two percent of population. Symptoms are highly variable between patients.

Acute pancreatitis (AP) is an inflammatory condition of the pancreas that can be mild or life threatening. Genetic susceptibility, anatomical abnormalities, medications, trauma, infectious etiologies and metabolic causes are the primary risk factors for pediatric pancreatitis.

Pancreatic insufficiency has been reported in patients with celiac disease, [2] and an association has been noted between celiac disease and pancreatitis [3-5]. Very few pediatric case reports described the association between celiac disease and acute pancreatitis. Patients with celiac disease are at increased risk for development chronic pancreatitis than general population.

We report a 12-year female who presented with simultaneous celiac disease and acute pancreatitis

Case Presentation

A 12-year girl was referred to the ER with one day history of severe epigastric abdominal pain and vomiting. Her past medical history was significant for poor weight gain in the last year without abdominal pain or other gastrointestinal symptoms. She was afebrile, with moderate non-rebound tenderness over the epigastrium and normal vital signs. Her weight was 33 kg (3rd percentile) and the height was 152 cm at the mean.

She was diagnosed with acute pancreatitis based on high serum amylase level 470 U/L (normal 40-140 U/L) and abdominal ultrasound showing edematous bulky pancreas with normal common bile duct and gall bladder. The results of complete blood count, liver function tests, total bilirubin, triglyceride and electrolytes were normal, including serum Ca. Serum lipase test was not available at time of diagnosis.

She had no history of abdominal trauma or family history of pancreatitis, pancreatic cancer or cystic fibrosis.

She was admitted to hospital for pain management and intravenous fluid hydration. She had low serum albumin down to 2 g/dl (normal 3.5-5.5).

Vomiting has resolved and oral intake was resumed three days after admission. Her abdominal pain has significantly improved.

To investigate her hypoalbuminemia and poor weight gain, a tissue transglutaminase IgA (tTG) was obtained. Its titer was elevated at 55 (normal <12 mg/dl).

An upper endoscopy showed diffuse ulcerations, edema and scalloping of the duodenal mucosa with edema surrounding the ampulla of Vater. Duodenal biopsies showed severe villous atrophy compatible with marsh 3 classification confirming the diagnosis of celiac disease. Patient was started on gluten free diet. At 3-months follow up, she gained weight (10th percentile). She had normal albumin level, abdominal ultrasound and pancreatic enzymes level. She remained with no symptoms of abdominal pain or vomiting.

Discussion

Although celiac disease is considered a common disorder, an acute pancreatitis is less common with an estimated overall incidence across ages of 1 in 10,000 cases per year [6].

The association of celiac disease and acute pancreatitis therefore occurs most often by a chance. Epidemiological study by Ludvigsson et al. [5] has suggested that patients with celiac disease have a higher risk than the general population for the development of acute pancreatitis. A hazard ratio was 3.3 for pancreatitis in a Swedish registry of 14,239 patients with celiac disease. Of 9348 patients with celiac disease

younger than 16 years old, 8 were identified with pancreatitis. The youngest patient was 2 years old.

Halabi reported a 3-year-old boy with a history of intermittent abdominal pain and vomiting from the age of 1 year. He was diagnosed with acute pancreatitis. Subsequent investigation confirmed celiac disease. The limitation of that case report was that the patient had negative celiac serology but villous atrophy that improved after gluten free diet. Our patient had definite celiac disease based on both serology and histological changes [7].

Bultron et al. reported a 9 year boy with acute pancreatitis and coexisting celiac disease that was diagnosed at the age of 7 years [8]. The patient was on gluten free diet but endoscopy was not done at the time of pancreatitis to confirm the compliance for the gluten free diet.

Rodrigo et al. described the association of acute and acute recurrent pancreatitis with gluten enteropathy. From a total of 185 adult patients, 34 (18%) met the criteria for gluten enteropathy, but the limitation of this study that only 3 patients had positive celiac serology which questions the the real incidence of celiac disease [9].

Several factors can contribute to the association between celiac disease and pancreatitis. Malnutrition impairs pancreatic secretion leading to pancreatic acinar atrophy [10]. In addition, villous atrophy in general is associated with pancreatic insufficiency. However, there is no evidence to show improvement of pancreatitis, either by episode frequency or pain severity, after correction for malnutrition.

Celiac disease can also lead to papillary inflammation and stenosis that could sensitize the pancreas to develop an acute inflammatory response. Persistent inflammation and irreversible changes to the pancreatic structure and function might contribute to the long-term increased risk of pancreatitis in patients with celiac disease.

Patel et al. proposed that celiac disease might be associated with pancreatitis due to papillary stenosis resulting from localized duodenal inflammation [11]. From a cohort of 169 patients with suspected papillary stenosis and recurrent pancreatitis, they identified 12 (7%) with histologically proven celiac disease. Ten patients fulfilled strict manometric criteria for papillary stenosis and underwent sphincterotomy. None of the patients had malnutrition.

Our patient had severe ulcerations in the duodenum including the ampulla of Vater. This can be a possible mechanism in our patient, although manometric studies were not done to confirm this possibility.

Additionally, celiac disease and acute pancreatitis may share common immunological pathway that involves Th1-associated cytokine-mediated response. Polymorphisms in tumor necrosis factor-alpha, a proinflammatory TH1 cytokine, have been implicated as susceptibility markers for celiac disease [12]. Tumor necrosis factor also plays a role in the pathogenesis of pancreatitis. Thus, up regulated inflammatory cytokines in celiac disease may predispose susceptible individuals to pancreatitis; however, this postulated pathway requires further investigation.

In summary, we report on a young patient with concomitant acute pancreatitis and celiac disease. One must have high index of suspicion for presence of pancreatitis in setting of celiac disease, especially in when presenting with abdominal pain and vomiting.

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