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Chronic pancreatitis as presentation of Crohn's disease in a child

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Abstract

It is reported that a pancreatic disease may precede the diagnosis of inflammatory bowel disease (IBD) both in children and in adults. Idiopathic chronic pancreatitis, however, occasionally co-exists with the IBD, mainly at pediatric age. We report a case of a patient who progressively developed the features of a chronic pancreatitis, before the diagnosis of Crohn's Disease (CD). Ten months after the onset of the first episode of pancreatitis the patient developed bloody diarrhea, mucus stools and biochemical findings of inflammation. The colonoscopy revealed a diffuse colitis without involvement of the last loop and the gastroscopy showed inflammation of the iuxta-papillary area. The histological findings confirmed the diagnosis of CD that involved the colon and the duodenum. In conclusion, in children the idiopathic chronic pancreatitis may be an unusual presentation of CD. Thus, if other known cause of chronic pancreatitis are not found, a not invasive work up to exclude the IBD should be warranted. An early coincidental diagnosis of the IBD may delay the progression of the pancreatic disease.

Key words: Inflammatory bowel disease; Crohn's Disease; Pediatric age; Bloody diarrhea; Pancreatic disease

Core tip: We report a cases of chronic pancreatitis associated with Crohn's Disease (CD). We have not been able to find reports of this association in the pediatric medical literature. The present case suggests that in children the idiopathic chronic pancreatitis may be an unusual presentation of CD. Thus, if other known cause of chronic pancreatitis are not found, a not invasive work up to exclude the inflammatory bowel disease should be warranted. The early recognition of the CD, indeed, may help in delay the progression of the pancreatic disease.

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TO THE EDITOR

It is well known that pancreas can be involved in the course of inflammatory bowel diseases (IBD)[1]. The pancreatic disease can occur in cases of biliary lithiasis or of the administration of 5-aminosalicylates (5-ASA), corticosteroids, and azathioprine (AZA) or 6-mercaptopurine[1,2]. Most cases of pancreatitis are clinically silent

and the frequency of clinical pancreatitis is markedly lower than that of asymptomatic hyperamylasemia or of evidence of exocrine pancreas insufficiency[1]. The potential association of the IBD with the pancreatic diseases makes therefore a periodic assessment of the pancreatic function in all patients affected by IBD advisable. A pancreatic disease may also precede the onset of the IBD. It is shown that an acute pancreatitis may represent the picture of presentation of the IBD in children such as in adults[3]. As recently reported by Broide *et al*[3] the prevalence of acute pancreatitis as symptom of the onset of the IBD is 2.17% in children and 0.06 in adults[3]. Two previous reports found a prevalence of acute pancreatitis preceding IBD in 27%[4] and in about 5%[5] of the cases. Therefore it was suggested[3] that in children, after an episode of acute pancreatitis, specific attention should be paid to other IBD susceptibility factors, that could indicate investigations by colonoscopy and gastroscopy. Idiopathic chronic pancreatitis, on the other hand, was occasionally reported in association with the IBD, mainly at pediatric age[6-8]. We report a case of a patient who progressively developed the features of a chronic pancreatitis, before the diagnosis of Crohn's disease (CD). When she was 4 years old, she was admitted with abdominal pain, slightly raised C-reactive protein and of erithro-sedimentation rate and high serum level of amylase and lipase. No gallstones were found, but only edema and enlargement of the pancreatic gland were reported on abdominal ultrasound. She received the treatment of the acute pancreatitis (intra-venous fluids, bowel rest, antibiotic, protease and gastric acid inhibitors), with beneficial effects on the pain and of the biochemical indices. Thereafter, she developed numerous episodes of acute pancreatitis. Therefore we planned the imaging and blood examinations to exclude all the causes of the chronic and recurrent pancreatitis. Serological tests for Cytomegalovirus, Epstein-Barr virus, Echovirus, Rubella, Adenovirus, Coxsackie virus, *Legionella* and *Mycoplasma* resulted negative. Immunoglobulin G serum level was into the normal range, so to exclude an autoimmune pancreatic process. The sweet test was negative. Genetic analysis for the mutations of cystic fibrosis transmembrane conductor regulator gene, for the cationic trypsinogen (*PRSS1*) gene, and serine protease inhibitor Kazal type 1 (*SPINK1*) were all negative. The magnetic resonance cholangiopancreatography showed a picture of chronic pancreatitis. This result along with the persistence of severe abdominal pain led us to schedule an endoscopic retrograde cholangiopancreatography to have a clear pancreatogram and therefore to define the chronic process but also to perform the pancreatic sphinterectomy[9]. The sphinterectomy was followed by partial extraction of the pancreatic concretions and by the placement of a plastic stent into the main pancreatic duct for transpapillary drainage. The pancreatogram showed pancreatic calcifications and the distortion of the main pancreatic duct, both findings consistent with established chronic pancreatitis. The sphinterectomy and the placement of the stent reduced the abdominal pain but did not impede the course of the pancreatic failure that required the enzymes replacement. Ten months after the onset of the first episode of pancreatitis the patient developed bloody diarrhea, mucus stools and relevant increase of inflammation indices. The following not invasive work up to define the suspected IBD showed positive anti-saccaromyces cerevisiae antibodies and fecal calprotectin. Stool cultures and stool test for *Clostridium difficile* toxins A and B were all negative. Upper and lower endoscopies were therefore scheduled. Colonoscopy revealed a diffuse colitis without involvement of the last loop. The gastroscopy revealed a duodenal involvement, with inflammation of the iuxta-papillary area. Then the histological examination confirmed the diagnosis of CD localized in the colon and in the duodenum. Furthermore the nutritional treatment with an amino-acid based formula by naso-gastric tube induced a partial regression of the intestinal symptoms that relapsed when she restarted a diversified oral nutrition. The use of 5-ASA and of AZA determined an immediate increase of the pancreatic enzymes; thus the patient received corticoid alone, that improved the intestinal symptoms, without influencing the pancreatic function. The clinical phenotype of the CD in this patient was very severe, with several relapses requiring repeated courses of steroid treatment and therefore we began the biological drugs. We started with infliximab but, after the second administration that induced a severe anaphylactic reaction, we temporary suspended this treatment and we started a period of bowel rest by home parenteral nutrition. The following re-exacerbations were treated by short courses of steroids that also showed beneficial effects on the pancreatic exacerbations, with immediate regression of the pancreatic pain and of the inflammatory indices. When she was 13 years old she begun the adalimumab that determined a prolonged period of remission. The girl is now 16 years old, she reached the pubertal development and she is treated by adalimumab, without severe re-exacerbations of the intestinal disease. The pancreatic function is supported by the pancreatic enzymes. To our knowledge in literature are reported 16 cases of chronic pancreatitis associated with CD[5-8] and none of them occurred at pediatric age. Therefore we report the first pediatric case of chronic pancreatitis as picture of presentation CD. In our experience this is the first case of CD presenting as chronic pancreatitis and therefore we may consider this association very rare according with the literature data. It' s not clear if the pancreatic inflammation may be a metastatic presentation of CD or the complication of the duodenal involvement[1]. In our patients the CD-associated pancreatitis was due to the duodenal and to the iuxta-papillary area involvement, causing duodenal reflux and papilla obstruction. In this case the diagnosis of IBD was made only 10 mo from the onset of the pancreatic complaints, when the patient had already developed the intestinal signs suggestive of intestinal inflammation. When the CD was recognized the pancreatic disease had already progressed towards a chronic relapsing process

with intractable pain and exocrine pancreas insufficiency. We may speculate that in this case the earlier diagnosis of CD might reduce the severity of the pancreatic disease, delaying the course of the pancreatic failure. Both pancreatic pain and biochemical inflammation recovered indeed after short treatments with steroids, so confirming that the progression of the pancreatic disease might be influenced by a timely diagnosis of CD. In conclusion in children the idiopathic chronic pancreatitis may be an unusual presentation of CD. Thus, if other known cause of chronic pancreatitis are not found, a not invasive work up to exclude the IBD should be warranted. An early coincidental diagnosis of the IBD may delay the progression of the pancreatic disease.

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