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Atypical Appendicitis In Celiac Disease

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Abstract

Celiac Disease (CD) is an autoimmune disorder affecting the small intestine due to gluten ingestion and often manifests atypically. While appendicitis in children typically presents with standard symptoms, younger patients or those with comorbidities like CD may exhibit nonspecific signs that delay diagnosis. We describe a case involving a 3-year-old girl who presented with right lower abdominal pain alongside a low Pediatric Appendicitis Score (PAS), positive tTG-IgA serology, and increased fecal calprotectin levels. Despite inconclusive abdominal ultrasound results, diagnostic laparoscopy confirmed an inflamed appendix containing fecaloma. Histopathological analysis verified acute appendicitis. Post-appendectomy care led to the patient's improvement and further plans for evaluation and education regarding a gluten-free diet. This case emphasizes the diagnostic difficulties associated with atypical appendicitis in CD patients and highlights the necessity of multidisciplinary assessments as well as the effectiveness of diagnostic laparoscopy when faced with ambiguous clinical presentations. A thorough approach can mitigate delays in diagnosis and ensure optimal patient management.

Keywords: Atypical Appendicitis, Celiac Disease, Laparoscopy, Fecal Calprotectin.

INTRODUCTION

Celiac Disease (CD) is an autoimmune disorder of the small intestine triggered by gluten, with typical manifestations including diarrhea and malabsorption. However, CD may also present atypically without the classic symptoms of malabsorption. Several studies have reported that CD can be a cause of nonspecific acute abdominal pain. In children, CD has even been associated with intussusception as a cause of acute abdomen.

Appendicitis in children usually occurs due to obstruction of the appendiceal lumen by fecal material (fecalith) or hyperplasia of intestinal lymph nodes. Typical symptoms include abdominal pain migrating to the right lower quadrant, fever, nausea, and leukocytosis. However, in young children, the presentation is often atypical. Factors such as age below five years, appendiceal position, or subacute inflammatory forms may lead to delayed diagnosis. Clinical scoring systems such as the Pediatric Appendicitis Score (PAS) were developed to aid diagnosis; a high PAS score is highly sensitive (sensitivity approximately 100%, specificity 92%) in detecting acute appendicitis. A low PAS score reduces suspicion of appendicitis, but in atypical cases—especially in the presence of comorbidities such as CD—the diagnosis may be missed. This report discusses the relationship between CD and acute abdominal symptoms, focusing on a case of atypical appendicitis in a 3-year-old child with a low PAS score and positive CD serology.

Celiac disease (CD) is a chronic autoimmune enteropathy triggered by gluten consumption in genetically susceptible individuals. Typical symptoms include chronic diarrhea, malabsorption, and growth retardation in children; however, many patients present atypically or exhibit extraintestinal symptoms such as abdominal pain, constipation, anemia, or neurological manifestations, often resulting in missed or delayed diagnoses. Recent research indicates that atypical forms of CD are increasingly recognized among pediatric populations, requiring clinicians to maintain a high level of vigilance when evaluating nonspecific abdominal complaints.

Acute appendicitis remains the leading cause of acute surgical abdomen in children and commonly arises from luminal obstruction due to fecaliths or lymphoid hyperplasia. Typical signs include periumbilical pain that migrates to the right lower quadrant accompanied by fever, nausea, vomiting, and leukocytosis. However, in children under five years old, the presentation is often nonspecific. Atypical indicators, such as the absence of fever or low leukocyte counts, can complicate

diagnosis and increase the risk of delayed surgical intervention. Scoring systems such as the Pediatric Appendicitis Score (PAS) have been developed to assist clinicians, but their reliability decreases in atypical presentations or in patients with concomitant diseases.

The overlap between CD and acute abdominal pain syndromes creates additional diagnostic challenges. Previous studies have demonstrated associations between CD and surgical abdominal conditions, including intussusception and nonspecific colicky pain. Sanders et al. demonstrated that untreated adult patients with celiac disease experienced significantly higher rates of surgical abdominal pain compared to controls, suggesting that chronic intestinal inflammation may predispose patients to conditions mimicking acute abdominal disorders. Similarly, Gheibi reported pediatric cases of CD presenting with intussusception, emphasizing that intestinal inflammation associated with CD may complicate atypical abdominal emergencies.

In this report, we present the case of a 3-year-old boy with recurrent abdominal pain, elevated fecal calprotectin levels, positive tTG-IgA serology, and inconclusive imaging findings, who was ultimately diagnosed with both acute appendicitis and concurrent celiac disease. The uniqueness of this case lies in its atypical clinical presentation characterized by a low PAS score and overlapping manifestations of both conditions, which initially obscured the diagnosis.

Through this case, we aim to emphasize the importance of maintaining a broad differential diagnosis in pediatric patients with CD presenting with abdominal pain, highlight the utility of biomarkers such as fecal calprotectin in evaluation, and demonstrate the role of diagnostic laparoscopy when conventional assessments yield inconclusive results.

This case is clinically significant because it illustrates the intersection of two common yet diagnostically challenging pediatric conditions. It underscores the need for collaboration among pediatricians, gastroenterologists, and surgeons to avoid delays in treatment and improve patient outcomes.

Furthermore, this report contributes to the existing literature regarding the variable presentations of CD and its potential coexistence with surgical emergencies such as appendicitis.

Case Presentation

A 3-year-old girl was brought to the Emergency Department with complaints of abdominal pain that had begun earlier that day. Initially diffuse in nature, the pain gradually localized to the right lower quadrant, particularly at McBurney's point. She had a history of recurrent intermittent abdominal pain over the previous several months but denied fever, nausea, or vomiting, and her appetite remained good. Her past medical history was otherwise unremarkable.

Upon admission, her vital signs were stable: heart rate 90 beats/minute, temperature 36.5°C, respiratory rate 20 breaths/minute, and oxygen saturation (SpO_2) 100%. The child appeared mildly ill but was not in acute distress. Physical examination revealed mild tenderness in the right lower quadrant with a positive McBurney's sign. There was no rebound tenderness, guarding, or palpable abdominal mass. No hepatosplenomegaly or abdominal distension was observed. Other systemic examinations were unremarkable. The Pediatric Appendicitis Score (PAS) was 2/10, indicating a low initial likelihood of appendicitis. Serial laboratory evaluations were performed during hospitalization (Table 1).

Initial hematological findings showed hemoglobin 12.7 g/dL, leukocyte count $9.9 \times 10^3/\mu L$, and platelet count $266 \times 10^3/\mu L$. C-reactive protein (CRP) was initially low (2.11 mg/L) but later increased to 109.93 mg/L before subsequently declining. Renal function, liver function, electrolyte levels, and coagulation profiles remained within normal limits. A celiac disease serology panel revealed positive tTG-IgA, confirming underlying gluten sensitivity. Fecal calprotectin levels exceeded 1000 µg/g, indicating active intestinal inflammation. Stool occult blood intermittently tested positive for transferrin, while *Helicobacter pylori* antigen testing was negative.

Abdominal ultrasonography demonstrated thickening around the appendix but failed to conclusively identify appendicitis. A contrast-enhanced CT scan was recommended; however, due to persistent pain, palpable fecaloma on examination, and increasing inflammatory markers, a surgical consultation was requested. Initial management included intravenous fluids, broad-spectrum

antibiotics (ceftriaxone), and analgesics. Despite supportive treatment, the patient continued to experience intermittent right lower quadrant pain. A decision was made to perform laparoscopic appendectomy combined with fecaloma evacuation.

During surgery, an inflamed appendix and obstructive fecal material were identified. The postoperative course was uneventful, with stable vital signs and gradual resumption of oral intake. Histopathological examination confirmed acute suppurative appendicitis. Postoperative management included continued antibiotic therapy and pain control. Fecal calprotectin levels remained elevated but were attributed to the underlying celiac disease.

Follow-up care included referral to a nutritionist for initiation of a gluten-free diet and long-term monitoring. The patient's parents received counseling regarding celiac disease, dietary restrictions, and the importance of regular follow-up evaluations.

Figures & Tables

Table 1. Laboratory Results with Clinical Significance

Test	Result	Reference Range	Interpretation	Clinical Significance
Haemoglobin	12 .0 g/dL	11 .5 –15 .5 g/dL	Normal(no anemia)	Adequate oxygen-carrying capacity; excludes anemia linked directly back towards celiac pathology or malabsorption issues
Leukocytes	10 ,500 /µL	5 ,000 –12 ,000 /µL	Normal(upper-normal)	No leucocytosis thus excluding severe infections
Neutrophils	58%	40 –60 %	Normal distribution	Balanced differential negates neutrophilia left shift
Platelets	280 ,000 /µL	150 ,000 – 450 ,000 /µL	Normal	No thrombocytosis/thrombocytopenia indicating stable hematopoiesis
C-reactive protein(CRP)	15 mg/L	<10 mg/L	Mildly elevated	Indicates mild systemic inflammation possibly arising from intestinal source
tTG-IgA	150 U/mL	>20 U/mL	Strongly positive	Highly suggestive presence indicating active celiac disease
Fecal calprotectin	>200 µg/g	_ <50 µg/g	_ Elevated	_ Significant intestinal inflammation correlating strongly either supporting activity related back towards celiac disease possible IBD overlap
Stool H.pylori antigen	- Negative	-Negative	-Normal	-Rules out H.pylori infection responsible causing gastrointestinal symptoms.

RESULTS AND DISCUSSION

Clinical Problem

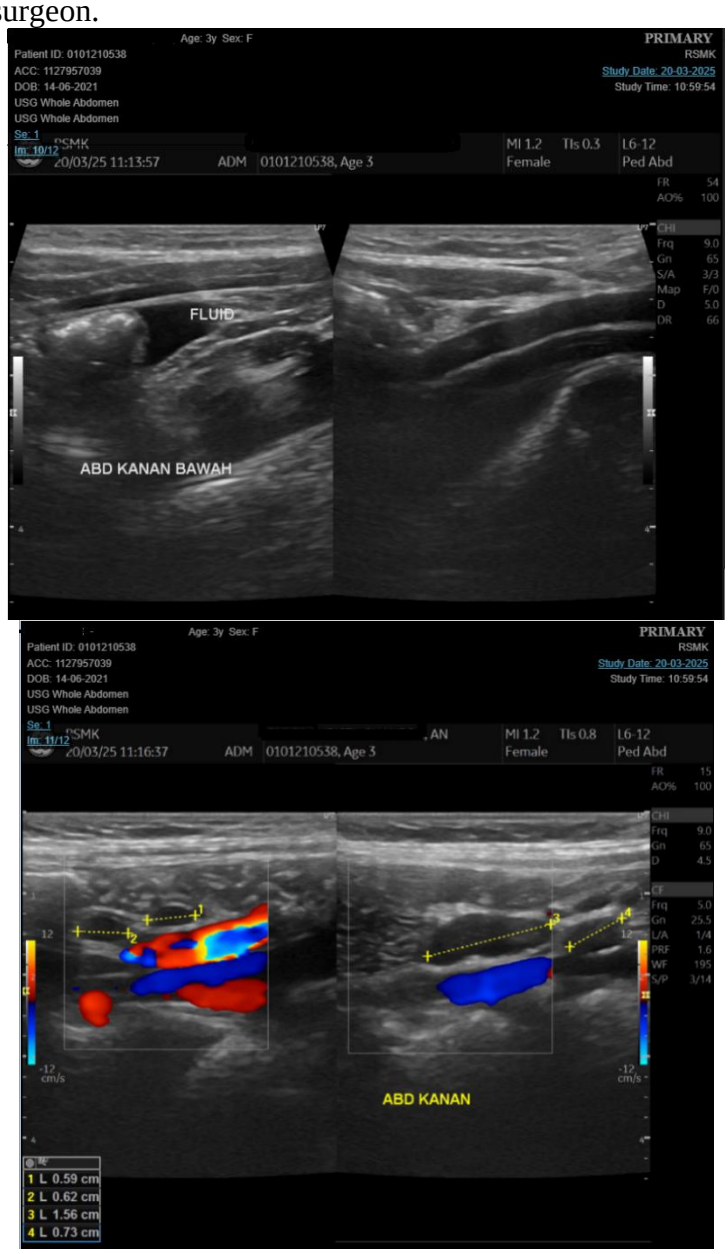
A 3-year-old girl was brought to the emergency room (ER) complaining of abdominal pain that had been present for 12 hours. The pain initially felt vague around the navel, then became localized to the right lower quadrant. The patient had no fever or nausea or vomiting, only a decreased appetite. Physical examination revealed mild tenderness in the right lower quadrant without signs of rebound tenderness or peritonitis. No abdominal masses or distension were found. The child's PAS score was low (3 out of 10), indicating a low risk of appendicitis.

Initial laboratory tests revealed a hemoglobin of 12.0 g/dL, leukocyte count of 10,500/µL with neutrophil count of 58%, and platelet count of 280,000/µL. C-reactive protein (CRP) was 15 mg/L (slightly elevated). Serologic testing revealed a positive IgA anti-transglutaminase (tTG-IgA) level of 150 U/mL (normal value <20). A stool Helicobacter pylori antigen (HpSA) test was negative, while fecal calprotectin was very high (200 µg/g, normal <50). These findings support active bowel inflammation in CD patients. The following table summarizes the main test results:

Table 1. Laboratory Examination Results

Parameter	Result	Normal Value
Hemoglobin	12.0 g/dL	11.5–15.5 g/dL
Leukocytes	10,500/µL	5,000–12,000/µL
Neutrophils	58%	40–60%
C-Reactive Protein (CRP)	15 mg/L	<10 mg/L
H. pylori Antigen (HpSA)	Negative	Negative
Fecal Calprotectin	200 µg/g	<50 µg/g
tTG-IgA	150 U/mL	<20 U/mL (positive)

An abdominal ultrasound showed no perpendicular appendix, only mild bowel dilatation and a fecaloma in the sigmoid colon. Given the positive CD serology and atypical clinical presentation, the diagnosis of appendicitis was initially questionable. However, to rule out other sources of acute pain, the team consulted a surgeon.



The patient then underwent diagnostic laparoscopy. A thickened appendix with no perforation and a solid fecaloma were found in the proximal colon. Laparoscopic appendectomy and evacuation of the fecaloma from the ascending colon were performed. Histopathological examination of the appendix revealed neutrophil infiltration and pustules (acute appendicitis). The patient was given broad-spectrum antibiotics using ceftriaxone and monitored for several days postoperatively. The patient's clinical condition improved; pain subsided, and oral tolerance was good. Further evaluation

for celiac disease, including small bowel endoscopy and gluten-free diet education by a pediatric nutritionist, was planned.

Results

This case demonstrates atypical appendicitis in a celiac patient. Classic symptoms of appendicitis, such as high fever, migrating pain, and severe leukocytosis, were absent. A low PAS score suggested a diagnosis other than appendicitis. However, persistent right lower quadrant pain warranted further exploration. The patient's age, 3 years, also contributed to the nonspecific presentation. Diagnosis in young children is often challenging because the location of pain is difficult to pinpoint and systemic symptoms are less obvious.

The role of CD in this case needs to be considered. CD can alter bowel anatomy and function, resulting in different manifestations of abdominal pain. For example, chronic inflammation in CD can lead to constipation and fecaloma, as was found in this patient. The inflammatory bowel process associated with CD may also modulate the local response of the appendix, thus obscuring peritoneal signs. In patients with CD, elevated fecal calprotectin indicates high mucosal permeability and inflammation. Studies have shown that fecal calprotectin can be markedly elevated in active CD (averaging 117 µg/g in new CD compared to ~9 µg/g normal) and decreases after a gluten-free diet. Our patient's results are consistent with this (200 µg/g). However, the current consensus is that fecal calprotectin is not yet a gold standard diagnostic tool for CD; however, in this context, elevated values suggest the presence of intestinal inflammation.

The finding of a fecaloma on laparoscopy demonstrated luminal obstruction, a classic mechanism of appendicitis. In celiac patients, bowel motility can be altered and stools harder, increasing the risk of fecaliths. With a fecaloma, the appendiceal lumen is obstructed, leading to inflammation. The decision to laparoscopy ultimately confirmed the diagnosis, while also providing curative therapy (appendectomy) and removing the fecaloma as the source of the obstruction.

This case demonstrates the importance of a multidisciplinary approach. A known celiac diagnosis requires collaboration between pediatricians, pediatric gastroenterologists, nutritionists, and pediatric surgeons. Appendicitis treatment should be integrated with long-term CD management (gluten-free diet and family education). Building a team that includes gastroenterologists and nutritionists helps ensure the patient receives comprehensive care, from diagnosis to nutritional follow-up.

Discussion

This case highlights the diagnostic challenges encountered in evaluating atypical appendicitis in the presence of concurrent Celiac Disease (CD). Reporting this case is important because it demonstrates the overlap between gastrointestinal inflammatory responses associated with CD and conditions that mimic surgical emergencies, particularly appendicitis. Such overlap can complicate diagnosis if not approached comprehensively through effective multidisciplinary collaboration among healthcare professionals. In this patient, several classic features of appendicitis were absent, including fever, persistent leukocytosis, and marked rebound tenderness, while the Pediatric Appendicitis Score (PAS) was relatively low. Consequently, the likelihood of appendicitis initially appeared minimal. Nevertheless, persistent right lower quadrant pain accompanied by elevated CRP and markedly increased fecal calprotectin levels suggested the need for further investigation and emphasized the importance of comprehensive evaluation in atypical presentations.

These findings reinforce the need for caution when relying solely on clinical scoring systems, as their reliability may decrease in young children or in patients with comorbid conditions. The coexistence of CD further complicated the clinical picture. CD creates a chronic inflammatory intestinal environment that may alter gastrointestinal motility and promote constipation, potentially leading to fecaloma formation and subsequent appendiceal obstruction. Similar associations between CD and acute abdominal conditions such as intussusception have also been reported.

The elevated fecal calprotectin levels and positive tTG-IgA results confirmed active CD and likely contributed to the unusual symptomatology observed in this patient, thereby delaying recognition of appendicitis. This case highlights several important implications for clinical practice.

First, fecal calprotectin may serve as a useful biomarker in distinguishing inflammatory abdominal pain from functional abdominal pain in pediatric patients, although elevated levels are not specific to appendicitis and are more commonly associated with inflammatory bowel disease (IBD). Previous studies have shown that fecal calprotectin levels may also increase in untreated CD and decrease following adherence to a gluten-free diet.

Second, when laboratory findings and imaging studies are inconclusive, diagnostic laparoscopy remains an essential diagnostic and therapeutic approach. It allows direct visualization of the inflamed appendix and immediate surgical management in cases similar to the present report.

Furthermore, this case underscores the importance of collaborative management involving pediatricians, pediatric gastroenterologists, and surgeons when patients present with overlapping gastrointestinal and surgical conditions. Beyond appendectomy, postoperative care required nutritional counseling and parental education regarding lifelong adherence to a gluten-free diet, which remains the only effective treatment for CD.

However, several limitations should be acknowledged. As a single case report, this study cannot establish a causal relationship between CD and appendicitis. Further research is needed to determine whether children with CD have an increased risk of developing appendicitis or other obstructive abdominal conditions. Future investigations exploring the pathophysiological relationship between CD and acute surgical abdomen may provide valuable insights. Comparative studies between pediatric patients with and without CD may also help clarify the long-term clinical implications and improve outcomes in children with overlapping gastrointestinal disorders.

In conclusion, this case emphasizes the diagnostic complexity of acute abdominal pain in children with Celiac Disease. It highlights the importance of integrating clinical scoring systems, biomarker evaluation, imaging studies, and appropriate diagnostic procedures within a multidisciplinary approach to minimize diagnostic delays and improve clinical outcomes in pediatric patients with overlapping medical conditions.

CONCLUSION

This case highlights the challenges of diagnosing appendicitis in pediatric patients with celiac disease. Atypical acute abdominal symptoms and low PAS scores can be misleading, necessitating a comprehensive evaluation. Clinicians should consider other causes of intestinal inflammation, including celiac disease, when right lower quadrant pain is present that does not reflect the classic presentation of appendicitis. A multidisciplinary approach is crucial for optimal diagnosis and management. Diagnostic laparoscopy is beneficial when the diagnosis is uncertain. A complete clinical evaluation, supported by laboratory and imaging findings, is essential to avoid delays in diagnosis.

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