

# Firing of an Implantable Cardiac Defibrillator: An Unusual Presentation of Celiac Crisis

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## ABSTRACT

Celiac crisis, an atypical presentation of celiac disease, is characterized by acute diarrhea and severe metabolic derangements. This diagnosis is often missed in the differential of acute diarrheal illness. Our patient is a 69-year-old man who presented with ICD firing and was found to have profound metabolic derangements. Further evaluation revealed undiagnosed celiac disease and his symptoms resolved with a gluten-free diet. Celiac crisis should be considered in all patients presenting with acute diarrhea, metabolic acidosis, and severe electrolyte abnormalities as management can be life-saving.

## INTRODUCTION

Celiac crisis is an unusual presentation of celiac disease (CD) characterized by severe diarrhea and acute metabolic derangements. Although rare, with an incidence of 1%, celiac crisis can lead to profound electrolyte abnormalities and death.<sup>1</sup> It is an underdiagnosed entity and often missed on the differential diagnosis for acute diarrheal illness. The following report describes a case of celiac crisis in a 69-year-old male who presented with implantable cardiac defibrillator (ICD) firing.

## CASE REPORT

A 69-year-old man with past medical history of ischemic cardiomyopathy (ejection fraction 20%-25%) presented to the emergency department after 2 consecutive episodes of ICD firing. He was in his previous state of health until 10 days prior when he started having intermittent watery diarrhea with diffuse abdominal discomfort. He denied any nausea, vomiting, melena, hematochezia, or weight loss. The patient's home medications included pantoprazole, furosemide, metoprolol, and lisinopril. Four months prior to presentation, the patient was admitted to an outside hospital with severe iron deficiency anemia (hemoglobin of 4.8 g/dL) and received 4 U of packed red blood cells with appropriate response in hemoglobin. Evaluation included an endoscopically normal esophagogastroduodenoscopy and colonoscopy. Duodenal bulb biopsies were reported as peptic duodenitis; gastric biopsies showed active chronic gastritis showing organisms morphologically consistent with *Helicobacter pylori*, and the patient received treatment with clarithromycin and metronidazole.

Upon his current presentation, vital signs were stable, and his abdominal exam was benign. An electrocardiogram showed a corrected QT interval prolongation at 538 milliseconds. Initial laboratory workup was notable for hypocalcemia (<5.0 mg/dL), hypomagnesaemia (0.8 mg/dL), hypokalemia (3.1 mmol/L), and hypoalbuminemia (2.2 g/dL). Hepatic function panel was within normal limits, but international normalized ratio was mildly elevated at 1.3. Complete blood count showed a hemoglobin of 12.2 g/dL. The patient had a transient troponin elevation that peaked at 2.44 ng/mL on hospital day 1. Stool studies were negative for infectious etiologies.

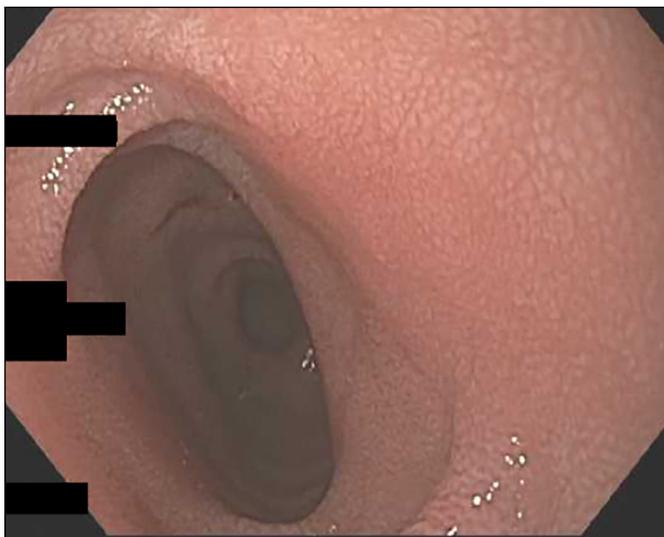
During the hospital stay, the patient's hypokalemia and hypomagnesaemia corrected with supplementation, but his hypocalcemia persisted. A 25-hydroxy vitamin D level was checked and was less than 4 ng/mL. The combination of

ACG Case Rep J 2016;3(4):e137. doi:10.14309/crj.2016.110. Published online: October 12, 2016.

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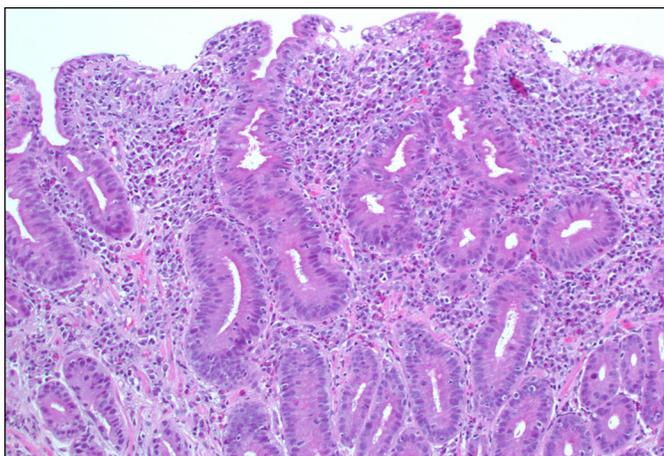
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**Figure 1.** Endoscopic photo of the patient's duodenum. The duodenal mucosa appears atrophic with loss of folds.

vitamin D deficiency, iron deficiency, hypocalcemia, and mild coagulopathy suggested a malabsorptive syndrome, and a tissue transglutaminase antibody was obtained on hospital day 2, returning strongly positive at 132 U (normal, 0-19 U).

Repeat esophagogastroduodenoscopy revealed decreased folds and flattening of folds in the second part of the duodenum (Figure 1). There were no complications of ulcerative jejunitis. Pathology from both the duodenal bulb and second portion of the duodenum showed total villous atrophy with intraepithelial lymphocytosis and epithelial repair (Marsh score 3b), confirming the diagnosis of CD (Figure 2). The gastric biopsy was negative for *H. pylori* organisms, verifying



**Figure 2.** Biopsy from the patient's second portion of duodenum showing almost total villous atrophy. The lamina propria is expanded by plasma cells and lymphocytes. The surface epithelium shows loss of its columnar shape and mucin depletion. A marked increase in the number of intraepithelial lymphocytes is present. Marsh score 3b.

eradication of his previous infection. He had no further episodes of arrhythmias during admission, and diarrhea improved on a gluten-free diet. He was discharged home in stable condition on oral calcium 1250 mg three times daily and vitamin D 50 000 IU once weekly.

## DISCUSSION

Our patient presented atypically after ICD firing with symptomatology consistent with celiac crisis. Although rare, when not recognized, celiac crises can lead to profound electrolyte derangements with potentially life-threatening complications. In our patient, severe electrolyte disturbances led to a prolonged QT interval, probable cardiac arrhythmias, and ICD firing. Furthermore, his underlying CD may have potentiated his cardiac history as it has been shown that individuals with CD are at an increased risk for ischemic heart disease.<sup>2</sup> This case also highlights the challenges in making the diagnosis of CD, as the patient's severe electrolyte abnormalities were initially attributed to what was thought to be a viral gastroenteritis. With concern focused on his heart and cardiac workup taking precedence, CD could have easily been missed if his very low vitamin D and calcium had not been taken into careful consideration. The presence of persistent hypocalcemia led to the diagnosis of vitamin D deficiency, which, in association with history of iron deficiency, hypoalbuminemia, and mild international normalized ratio elevation, suggested possible malabsorption. In a literature review of celiac crisis case reports, half of the cases presented similarly with marked hypocalcemia.<sup>1</sup>

Although our patient presented with celiac crisis, the vast majority of patients present with a more typical indolent course. It remains to be determined whether celiac crisis is the initial presentation of CD or an exacerbation of an underlying undiagnosed CD.<sup>1</sup> Our patient's duodenal bulb biopsy 4 months prior to this presentation was interpreted as consistent with peptic duodenitis; however, on future review did have some degree of partial villous atrophy. We believe that the unexplained severe iron deficiency anemia 4 months prior to presentation was caused by undiagnosed CD, and the subsequent crisis represents an exacerbation of underlying disease as opposed to an acute initial presentation of CD.

Interestingly, our patient's celiac crisis occurred 4 months after *H. pylori* treatment. This is of particular interest as studies investigating the relationship between *H. pylori* and CD have yielded conflicting results. Evidence suggests the presence of an inverse relationship between the declining prevalence of *H. pylori* in recent decades and the increasing prevalence of CD, even after correction for confounding variables such as age, gender, and socioeconomic status.<sup>3</sup> One postulated hypothesis to explain this relationship is that *H. pylori* confers protection against the

development of CD by modulating gluten immunogenicity.<sup>3</sup> Although additional studies are necessary to further evaluate this relationship, the eradication of *H. pylori* in our patient 4 months prior to his celiac crisis lends support to a possible protective role.

The patient's symptoms and electrolyte abnormalities responded appropriately to a gluten-free diet. This has been echoed in previous reported cases of celiac crisis, which all responded to a gluten-free diet.<sup>4</sup> Given the ease of managing a possibly life-threatening condition, the recognition and diagnosis of CD is crucial. Finally, CD should be considered in the differential of all patients presented with chronic and acute diarrhea, metabolic acidosis, and severe electrolyte disturbances, as prompt recognition and management can be lifesaving.<sup>5</sup>

## DISCLOSURES

Author contributions: C. Bou-Abboud collected the data and wrote the manuscript. J. Katz reviewed the manuscript and is

the article guarantor. W. Liu provided pathology slides and reviewed the manuscript.

Financial disclosure: None to report.

Informed consent was obtained for this case report.

Received October 8, 2015; Accepted April 6, 2016

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