

Case Reports

Celiac Crisis with Severe Hypokalemia and Paraplegia as a First Presentation of Celiac Disease in a Child

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Abstract

Celiac disease is an autoimmune disorder, affecting the small intestine primarily, leading to severe villous loss. A celiac crisis is a rare complication of celiac disease, typically affecting children below two years of age, characterized by explosive watery diarrhea, impressive abdominal distension, dehydration, hypotension and lethargy. This severe picture is accompanied by profound electrolyte abnormalities including dangerously low potassium levels and severe acidosis. The awareness of the condition could lead to prompt diagnosis and prevent significant morbidity.

Keywords: Celiac crisis, paraplegia, hypokalemia, celiac disease.

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Introduction

Celiac disease (CD) is an autoimmune disorder, triggered by an auto-antigen, affecting the small intestine primarily, leading to severe villous loss. It is the most common malabsorption disorder in the western hemisphere, with an incidence of 0.3-1 %.^{1,2}

A celiac crisis is a rare complication of celiac disease,⁴ typically affecting children below two years of age,³⁻⁵ characterized by explosive watery diarrhea, marked abdominal distension, dehydration, hypotension and lethargy. This severe picture is accompanied by profound electrolyte abnormalities including dangerously low potassium levels and severe acidosis.

Our patient was a 9-year-old girl, admitted twice with watery diarrhea, severe dehydration and paraparesis with severe hypokalemia. The diagnosis was made on her second admission.

Case presentation

A 9-year-old girl presented to the emergency room with watery diarrhea for the last 4 days with severe weakness which progressed to her inability to walk. Her past history was irrelevant except for abdominal distension and loose motions. She had negative family history of a similar condition. Her physical examination showed a dehydrated, afebrile child with tachycardia (HR: 160/min) and hypotension (BP 80/65 mmHg). She was failing to thrive (both ht. and wt. < 3 %) with severe abdominal distension. Both lower limbs were weak with hyporeflexia. Edema (++) was found in her lower limbs. Her labs showed severe hypokalemia and acidosis (Table 1).

During her hospitalization, the patient developed cardiac arrhythmia, which required admission to the ICU. Her potassium level dropped to 1.4 mEq/L which required multiple boluses of KCl.

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After normalization of her potassium, she was discharged on oral potassium as a case of familial hypokalemic paralysis. Her poor socioeconomic status was blamed for her failure to thrive.

Table (1): Laboratory Results.

	<i>First Admission</i>	<i>Second Admission</i>
<i>Na (mEq/L)</i>	127	130
<i>K (mEq/L)</i>	1.4	2.0
<i>Urea (mmol/L)</i>	30	22
<i>Creatinine(Umol/L)</i>	0.7	0.4
<i>pH</i>	7.3	7.22
<i>HCO₃ (mmol/L)</i>	14.8	8.0
<i>Ca (mg/dl)</i>	7.2	8.8
<i>Hb. (g/dl)</i>	7.0*	10
<i>Albumin (g/L)</i>	2.9	2.5
<i>PT</i>	-----	47 sec / 14
<i>ALT (U/L)</i>	-----	31
<i>AST (U/L)</i>	-----	27
<i>Urine analysis</i>	No proteinuria	No proteinuria
<i>Urine collection for (Na, K, Crtn)</i>	Normal	-----
<i>Blood Film</i>	Dimorphic anemia, no acanthocytes	-----

* *Received PRBCs transfusion*

The patient presented again three weeks later with the same picture. Her serum K was 2.0 mEq/L. With severe metabolic acidosis, her pH was 7.22 and a serum HCO₃ of 8.0. Potassium renal losses were excluded. Pediatric gastroenterology service was consulted.

Three stool examinations for ova and parasites, specifically giardia were negative. The stool exam showed positive for fat droplets. She was anemic and hypoalbuminemic and her prothrombin time was prolonged (Table 1). Her wrist x-ray showed ricketic changes. Her liver enzymes were normal (Table 1). Unfortunately, celiac serology was not available.

After stabilization and correction of her coagulopathy, an EGD examination was done, which showed:

- Evidence of gastroparesis; a food bolus found in the stomach after more than 10 hrs of fasting
- Scalloping and grooving of the folds in the second and third part of the duodenum

Duodenal biopsies showed:

- Total villous atrophy, heavy lymphocytic infiltrate (Marsh stage 3), consistent with celiac disease (unfortunately slides not available).

Patient was started on gluten-free diet, iron and vitamins supplementation, in addition to Domperidone (for the gastroparesis). On the follow-up visit 2 weeks later, she had added weight but complained of carpopedal spasm due to hypocalcemia. She responded to calcium supplementation. Her lower limb weakness improved dramatically and the patient was discharged walking. Three months later, the patient gained 5 kg and her blood count, albumin, INR and electrolytes were normal.

Discussion

A celiac crisis is a rare entity which is potentially fatal. It can present as acute exacerbation in celiacs or as in our case to be the first presentation.^{5, 6, 8} Most of our understanding of the condition is coming from case reports or case series.^{3 - 10}

The exact incidence is not known, but it is dropping secondary to our better understanding of the disease and early recognition of disease clues. However, why patients develop a celiac crisis while others run a much more benign course is not known. The combination of severe mucosal inflammation, immune activation and disruption of normal patterns of motility might be the cause.⁸

Celiac crises can be fatal if untreated.⁴ Severe hypokalemia manifested with neurological signs is a well-known presentation.^{9, 10} Complete recovery is anticipated with a correction of electrolytes and strict gluten-free diet.^{9, 10} Though the development of more serious complications like cardiac arrhythmia is unusual.

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Serology nowadays plays a key role in the diagnosis of CD. This will establish an accurate and specific diagnosis and allow for prompt treatment. Unfortunately, a celiac antibody screen was not available at the time of presentation. However, the endoscopic findings, histology and the dramatic response to a gluten-free diet make celiac disease the most probable diagnosis.

The management is dependent on the correction of the fluids and electrolytes, followed by establishing the diagnosis and starting a gluten-free diet. Corticosteroids should be considered in the cases of celiac crisis when a gluten-free diet, in conjunction with fluid and electrolyte repletion, does not result in rapid improvement.^{6-8.}

The increased requirements of the nutritional substrate with a rapid increment of weight, which was not met by the patient's diet, led to the symptomatic hypocalcemia, which responded very well to calcium and vitamin D supplementation.

In summary, this patient had enough clues for diagnosis. The delay of this diagnosis was potentially fatal, so general pediatricians should be aware of the condition and have a high index of suspicion when dealing with diarrhea, severe electrolyte imbalance and hypoproteinemia.

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الأزمة الزلاقية المترافقة بهبوط حاد بالبوتاسيوم والشلل النصفي كمظهر أول للداء الزلاقي (اعتلال الأمعاء بالغلوتين)

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الملخص

الداء الزلاقي (اعتلال الأمعاء بالغلوتين) هو واحد من اضطرابات المناعة الذاتية، التي تؤثر في الأمعاء الدقيقة في المقام الأول مؤدياً إلى ضمور الزغبات المعوية. والأزمة الزلاقية هي اختلاط نادر من اختلاطات الداء الزلاقي (اعتلال الأمعاء بالغلوتين) تؤثر عادة في الأطفال دون سن السنتين. وتمتاز الأزمة الزلاقية بالإسهال الشديد، وانتفاخ البطن، والجفاف، وهبوط ضغط الدم. ويرافق ذلك اختلال في أملاح الدم (هبوط خطير في مستويات البوتاسيوم في الدم) وزيادة في حامضية الدم. إن نقص الوعي بهذه الحالة المرضية قد يؤدي إلى مضاعفات حادة بما فيها الوفاة.

الكلمات الدالة: الأزمة الزلاقية، الشلل النصفي، نقص البوتاسيوم في الدم، الداء الزلاقي.