Unresponsive and refractory hypokalemia in celiac crisis- A predictor of high mortality

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Unresponsive and refractory hypokalemia in celiac crisis- A predictor of high mortality

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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 younger children, characterized by explosive watery diarrhea, abdominal distension, dehydration, hypotension and lethargy. This severe picture is accompanied by profound metabolic and electrolyte abnormalities including dangerously low potassium levels and severe acidosis. The awareness of this condition could lead to prompt diagnosis and prevent significant morbidity. Severe hypokalemia is refractory to treat and is predictor of high morbidity or mortality due cardiac arrhythmias and neurological abnormalities.

Keywords: Celiac crisis, Refractory hypokalemia, high mortality

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Introduction

Celiac disease or gluten enteropathy is an autoimmune mediated disorder elicited by the ingestion of gluten in genetically susceptible person and is characterized by chronic inflammation of small intestine associated with villous atrophy [1]. Celiac crisis is rare but potentially lethal complication of celiac disease and is associated with high morbidity. It is characterized by acute onset or rapid progression of gastrointestinal symptoms together with signs or symptoms of dehydration and metabolic disturbances which include hyponatremia, persistent hypokalemia, metabolic acidosis and shock. Celiac crisis usually has some underlying precipitating factor and is more common in patients with poor compliance with gluten free diet [2].

Case Report

Fourteen year old female child, second live issue of non consanguineous marriage of healthy parents and born at term following uncomplicated pregnancy presented with frequent episodes of diarrhea after six months of age, when she was weaned from breast milk. She was diagnosed at age of 3½ year of age as a case of celiac disease. Diagnosis was made by serology-anti tissue transglutaminase antibody and confirmed by GI endoscopy biopsy showing marsh stage 3 changes. She was started on gluten free diet. However, she had poor compliance and had frequent admissions for diarrhea with dehydration including one with encephalopathy. Last admission was one month back. Anti tissue transglutaminase antibody titers were high last time and GI biopsy also suggested of celiac...
This time she was admitted with acute onset of diarrhea and vomiting since two days. She had proportionate short stature (height 127 cm < 3rd percentile), weight 24 kg [<3rd percentile]. She had dehydration with hypotension and shock. She had pallor, grade 1 clubbing and there were multiple erythematous scaly and eczematous lesions with areas of denudation over both flexor and extensor surfaces. Complete blood count (CBC) revealed anemia with hemoglobin 9.2 g/dl, total leukocyte count 8400 (PMNLs 78%, leucocyte 19%, eosinophils 2%, monocyte 1%), thrombocytopenia with platelet count 15000/mm³, peripheral smear showing dimorphic anemia. Serum sodium was 136 mEq/L, hypokalemia with serum potassium level as 2.2 mEq/L, chloride 110 mEq/l, serum glucose was 108 mg/dl, magnesium 2 mg/dl, phosphorus 4 mg/dl, arterial blood gas analysis revealed metabolic acidosis (pH7.13, pO₂ 68 mmHg, pCO₂ 38 mmHg, HCO₃ 4 mEq/L), anti tissue transglutaminase antibody titres were significantly high, initial chest X ray was normal. Patient was admitted in Pediatric Intensive Care Unit and managed for shock with oxygen, IV fluid bolus with bicarbonate supplementation, and inotropic support. Potassium supplementation up to 80 mEq/L in intravenous fluids for persistent hypokalemia. Platelet transfusion has been given twice. Intravenous hydrocortisone were also given. Repeated CBC at the 24 hours of admission revealed as hemoglobin as 6 g/dl, White blood cell count as 6.800 mm³, thrombocytopenia with platelet count as 8000 mm³. Serum sodium was 131 mEq/L and serum potassium level was 1.9 mEq/L. Electrocardiography revealed as flat T wave due to hypokalemia. Blood cultures revealed no microorganisms. She has been received fresh whole blood transfusion and also three times platelet concentrates. Clinical condition worsened in spite of these interventions, shock and hypokalemia are unresponsive to treatment, and defined as multiorgan failure. She required mechanical ventilation. She died mainly due to massive gastrointestinal and pulmonary haemorrhage.

**Discussion**

Celiac crisis is rare but well known complication of celiac disease presenting with various systemic and metabolic manifestations like refractory hypokalemia, hyponatremia and shock. There is acute onset and rapid progression of gastrointestinal symptoms with signs and symptoms of dehydration and malnutrition. It may be associated with other autoimmune manifestations like in form of thrombocytopenia. Celiac disease is immune mediated disorder having a genetic basis with sensitivity to gluten in diet. It is a chronic inflammatory disorder associated with certain HLA subtypes DQA1 05 and DQB1 02. Genome wide association studies have shown risk variants in genes controlling immune response, some being shared with type 1 diabetes and other autoimmune disorders. The most evident expression of autoimmunity is presence of serum antibodies to transglutaminase. Several extra intestinal clinical manifestations of celiac disease [e.g. liver, heart, nervous system] are possibly related to antibodies. Symptoms usually appear, when weaning from breast milk is done and gluten is introduced in diet. Failure to thrive, chronic diarrhea, vomiting abdominal distension, muscle wasting and anorexia are present in most of cases [3]. As the age of presentation shift to later in childhood, extra intestinal manifestations become increasingly recognized, these includes iron deficiency anemia, short stature, arthritis and arthralgia, peripheral neuropathies, cardiomyopathy, endocrinopathy, encephalopathy, psychosis, sleep disturbances and other autoimmune manifestations [4,5]. Diagnosis is made by serology and confirmed by histology changes in biopsy specimens from endoscopy. The histological changes are graded according to Marsh criteria, grade ¼ changes are characteristics of celiac disease and grade 2 changes need positive serology [6]. Treatment is lifelong gluten free diet. After initiation of gluten free diet, there is a prompt response with rapid gain in weight and height within few months. An acute severe exacerbation of underlying mucosal inflammation leads to uncontrolled diarrhea, which in turn results in dehydration and loss of...
ions and bicarbonate in stool. This leads to metabolic acidosis and shock. Underlying malabsorptive state would predispose to hypoalbuminemia and hence a decreased effective arterial volume. These patients do not tolerate volume losses, and acid-base and electrolyte imbalance is more severe in them. There is also a compensatory rise in renin and aldosterone levels in response to a volume-contracted state, resulting in kaliuresis, thus worsening hypokalemia. This explains the high potassium requirement in celiac crisis. Despite urinary potassium and trans tubular gradient of potassium being high, urinary chloride are increased leading to negative urinary anion gap, indicating a primary extra-renal source of potassium loss. There are few such case report of celiac crisis presenting as severe metabolic disturbances and shock requiring as intensive care management and systemic steroids [7,8]. The constellation of clinical features reported and metabolic disturbances should prompt the clinician to investigate for the celiac crisis especially in poor compliant patient. Celiac crisis presenting as shock requires urgent intensive care management and correction of various metabolic disturbances and various bleeding diathesis. There is a role of systemic steroids .Delay in recognition of celiac crisis state and poor compliance in already known case of celiac disease leads to very high mortality especially when hypokalemia is present. Hypokalemia in celiac crisis is refractory to treat especially when present for a prolonged time and is predictor of high morbidity or mortality due cardiac arrhythmias and neurological complications.

References