CASE REPORT

DOI: 10.5336/caserep.2018-64110

Celiac Crisis: A Rare and Serious Condition Resulting in Death

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Received: 05 Dec 2018
Received in revised form: 22 Jan 2019
Accepted: 22 Jan 2019
Available online: 31 Jan 2019

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ABSTRACT Celiac disease is an autoimmune disease of the small intestine, resulted from villus damage and malabsorbtion due to ingestion of gluten; and production of antibodies to gliadin. The disease can manifest with various presentations. Celiac crisis was described as an acute, serious, life-threatening form of celiac disease about sixty years ago with a higher incidence among pediatric patients than adults. It is characterized by gastrointestinal symptoms like profuse diarrhea and severe metabolic distubances like hyponatremia and hypoproteinemia. Early suspicion and detection is important. This case report presents a 54-year-old female patient who died from celiac crisis and sepsis.

Keywords: Celiac disease; celiac crisis; water-electrolyte imbalance; sepsis

Celiac crisis is a serious, life threatening condition which has common manifestations including severe diarrhea, hypoproteinemia, metabolic and electrolyte imbalances. Signs and symptoms vary among patients.[1] Anderson and di Sant-Agnese first reported celiac crisis in 35 pediatric patients in 1953.[2] Most of the case reports present pediatric patients compared to adults.[3]

Treatment of cealiac crisis includes gluten-free diet, nutritional and fluid replacement and corticosteroid administration.[1] The present study aims to report a patient who died from celiac crisis and urinary tract infection.

CASE PRESENTATION

54-year-old female patient was admitted to emergency department with confusion, diarrhea, nausea and vomiting. She was somnolent, Glascow coma scale was 13. Blood pressure was 80/50 mmHg, heart rate was 110 bpm, body temperature was 38.1 °C. She looked malnourished and cachectic. Physical examination revealed abdominal distention and pretibial edema (3+) and the patient was transferred to Intensive Care Unit. She had Hashimoto thyroiditis and psoriasis as comorbid diseases. Celiac disease had been diagnosed fourty years ago. However the medical records at the time of diagnosis were unavailable. Marsh type 3C Celiac disease, villus atrophy and intraepithelial lymphocyte were detected by endoscopic biopsy and

pathologic examination ten years ago. The serologic markers for celiac disease were positive (anti endomysium IgA, tissue transglutaminase immunoglobulin IgA and IgG, anti-gliadin IgA and IgG) She had been using levothyroxin for thyroiditis. She was learned not to receive a gluten-free diet.

Initial laboratory tests showed mild anemia, increased cholestatic parameters, hyponatremia hypokalemia, hyperbilirubinemia, hypomagnesemia, prolonged prothrombin time and hypoalbuminemia. Artery blood gas analysis showed respiratory alkalosis. Serum TSH and cortisol levels were normal. Urinalysis detected pyuria. Hepatitis results and also blood and stool culture tests were negative. Laboratory values are shown below (Table 1). Hepatobiliary ultrasound detected hepatomegaly and massive free fluid accumulation in perihepatic, perisplenic and pelvic areas. Coagulation tests were abnormal, peritoneal fluid sampling was not performed as considered risky. Cerebral computerized tomography showed no acute pathology. Sequential organ failure assessment (SOFA) score was 7.

Fluid, electrolyte replacement and wide-spectrum antibiotic treatment were initiated. Hypotension persisted and inotrophic agents were added. Celiac crisis was considered and intravenous metilprednisolon was given daily. Urine culture test revealed *Klebsiella pneumaniae*. A briader spectrum of antibiotic was commenced according to antibiogram results. The patient was lost on the fifth day of hospitalization despite supportive treatment. The informed consent form was signed by the relatives of patient about case presentation.

DISCUSSION

Celiac disease is an autoimmune disease of the small intestine resulted from villus damage and malabsorbtion, triggered by gluten proteins. Cealiac crisis was described as acute, serious and life-threatening form of celiac disease.[1]

Nearly 20 adult celiac crisis patients were reported in literature. Female patients are predominant.[3] Definition criteria include the acute onset or rapid progression of gastrointestinal symptoms (severe diarrhea, vomiting), requiring hospital admission and/or nutrition with at least two of the following:

Signs of severe dehydration (hemodynamic instability and/or orthostatic changes); renal failure; neurologic dysfunction (peripheral neuropathy and tetany due to hypocalcemia); metabolic acidosis; hypoalbuminemia; electrolyte disturbances like hypokalemia, hyper/hyponatremia, hypocalcemia, hypomagnesemia; weight loss more than 4.5 kg.[4] Some patients also presented with prolonged prothrombin time. Our patient had the criteria mentioned above.

TABLE 1: Laboratory results and artery blood gas analysis results on admission.

	Result	Normal range		Result	Normal range		
Haemoglobin	9.9 g/dl	12-16	Serum magnesium	1.82 mg/dl	1.9-2.5	Arterial blood pH	7.48
Platelet count	169000/µl	150000-450000	Serum phosphorus	3.61 mg/dl	2.5-4.5	pCO_2	31
White blood cell count	13000/ µl	4000-11000	Serum calcium	8.29 mg/dl	8.5-10.5	pO_2	96
Serum Urea	43 mg/dl	17-43	Serum albumin	1.8 g/dl	3.5-5	HCO_3-	15.5
Serum Creatinine	0.8 mg/dl	0.5-0.9	Prothrombin time	26.3 sec	10-14		
ALT	20 U/L	5-40	INR	2.3			
Alkaline phosphatase	115 U/L	35-105	aPTT	50.2 sec	22-24		
Serum sodium	126 mmol/L	135-145	C reactive protein	138 mg/dl	0-5		
Serum potassium	3.3 mmol/L	3.5-5.1	Serum chloride	98	98-106		
Serum total bilirubin	2.07 mg/dl	0.1-1.1	Cortisole	20	6.2-19		
TSH	0.5 µU/ml	0.4-4					

The mechanism of celiac crisis is unclear. According to the combination of immun system hypothesis, an immune stimulus like gluten-included diet, infection and severe inflammation can start the crisis.[5] Our patient was not following a gluten-free diet and had urinary tract infection. Both reasons could promote severe disease.

Immediate diagnosis and treatment are of vital importance due to high morbidity. In all reported cases, hospitalization and correction of metabolic imbalance with intravenous fluids were required. Some part of the patients had to receive corticosteroids for treatment (with different drugs and dosages being reported).[4] In literature, successful treatment and results were reported. Most cases healed completely.[3,4] Our patient was lost despite proper treatment. Infection was severe, and impairment progressed rapidly.

CONCLUSION

We emphasize to be careful about gastrointestinal symptoms, metabolic disturbances for defining celiac crisis. Etiology should be investigated and the disease should be treated properly. Death risk should be considered.

Source of Finance

During this study, no financial or spiritual support was received neither from any pharmaceutical company that has a direct connection with the research subject, nor from a company that provides or produces medical instruments and materials which may negatively affect the evaluation process of this study.

Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Özlem Gül Utku;* ***Design:*** *İrfan Karahan;* ***Control/Supervision:*** *Aydın Çiftçi;* ***Data Collection and/or Processing:*** *Gökberk Gözükan;* ***Analysis and/or Interpretation:*** *Aydın Çiftçi;* ***Literature Review:*** *Gökber Gözükan;* ***Writing the Article:*** *İrfan Karahan;* ***Critical Review:*** *Aydın Çiftçi;* ***References and Fundings:*** *Özelm Gül Utku;* ***Materials:*** *İrfan Karahan.*

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