Coeliac crisis with severe hypokalaemia in an adult

Rosalie Magro, Edgar Pullicino

Abstract

Coeliac crisis is a rare life-threatening presentation of coeliac disease, in which acute dramatic metabolic derangements are present. It is observed mainly in children less than two years of age. In adults, coeliac disease usually has an indolent course and presents with mild gastrointestinal symptoms or may even be asymptomatic and present with long term complications including anaemia, osteoporosis and infertility. This case describes a 38 year old gentleman who presented with acute diarrhoea that led rapidly to severe metabolic disturbances including life threatening hypokalaemia. This case illustrates the heterogeneous clinical course of coeliac disease and the importance of considering it in the differential diagnosis of adult patients presenting with acute diarrhoea and metabolic disturbances.

Introduction

Coeliac disease usually has an indolent course in adults and usually presents with mild symptoms. Coeliac crisis is a rare presentation of coeliac disease in adults, in which acute diarrhoea is complicated by severe metabolic disturbances. This case report describes a 38 year old male who presented with coeliac crisis and improved following correction of the underlying metabolic and electrolyte disturbances. The presentation, management and aetiology of coeliac crisis and other similar case reports are discussed.

Case report

A 38 year old gentleman presented to the casualty department with a three week history of profuse non-bloody watery diarrhoea, severe lethargy and weight loss. Since three days the patient also developed bilateral lower limb weakness but no sensory symptoms. He also complained of mild abdominal pain, bloating and flatulence. His appetite was normal and there was no history of fever, vomiting or steatorrhoea. The patient gave a history of diarrhoea two years previously that lasted for seven weeks and resolved spontaneously. He was at the time investigated by means of a colonoscopy that was normal. On examination the patient was pale, dehydrated and appeared cachectic. His abdomen was soft and non-tender and he had no sensory abnormalities.

Initial investigations showed severe hypokalaemia (1.9mmol/L), hyponatraemia (131mmol/L) and normal anion gap metabolic acidosis (pH 7.34, HCO$_3^-$ 16.5mmol/L). Serum creatinine was elevated (140µmol/L). He also had hypocalcaemia (1.69mmol/L, albumin 25.7g/L, corrected calcium 2.04mmol/L) and hypophosphataemia (0.2mmol/L). His white cell count was elevated (17x10$^9$/L) with a raised neutrophil count (11.94x10$^9$/L). His liver function tests were normal. The patient’s ECG showed normal sinus rhythm with ST segment depression in leads V1 to V6 and had the presence of a U wave secondary to hypokalaemia.

The patient was rehydrated with an intravenous infusion and was given potassium supplementation via a central line. He was also administered intravenous calcium gluconate and intravenous vitamin K. He was admitted to ICU for intensive monitoring.

Keywords

Coeliac disease/complications, hypokalaemia/aetiology, diarrhoea/aetiology

Rosalie Magro* MD, MRCP(UK)
Department of Medicine, Mater Dei Hospital, Malta
Email: rosalienagro@gmail.com

Edgar Pullicino FRCP PhD
Department of Medicine, Mater Dei Hospital, Malta
Email: edgar.pullicino@gov.mt

*corresponding author
Further investigations included stool culture and sensitivity, stool examination for ova, cysts and parasites and *Clostridium difficile* toxin. These were negative for three consecutive times taken on alternate days. The thyroid function tests were normal. The patient was found to be hypoalbuminaemic (25.7g/L) and hypoproteinaemic (39.2g/L). Haematins revealed vitamin B₁₂ and folate deficiency (<111pmol/L and 2.3nmol/L respectively) despite a normal haemoglobin at presentation (15.2g/dL). The latter could have possibly been secondary to haemoconcentration since the haemoglobin dropped to 10.8g/dL with rehydration.

Once the dehydration and electrolyte depletion were corrected, the patient improved. The lower limb weakness resolved completely and he was transferred from ICU after two days. Coeliac disease was suspected as anti-endomysial antibody IgA was positive and tissue transglutaminase IgA and IgG were elevated (35.9U/ml and 20.6U/ml respectively). Thus a gastroscopy was performed nine days following his presentation, once the patient was stable. This confirmed coeliac disease, Marsh-Oberhuber type 3c as duodenal biopsies showed complete loss of duodenal villi and marked intraepithelial disease.

In our case the patient had a prolonged INR and hypocalcaemia, hypomagnesaemia, and hypoproteinaemia. Coeliac crisis is associated with a high morbidity rate, mandating immediate identification and treatment. To date, only twenty cases of coeliac crisis in adults have been reported in the literature and for this reason coeliac disease is rarely considered in adults presenting with acute severe diarrhoeal illness, even when infectious aetiologies have been excluded. Of the twenty cases, 15 were women and 5 were men, the mean age at diagnosis was 51.4 years overall; 73.4 years in men and 46.6 years in women. All patients described presented with severe diarrhoea. Other symptoms and signs included vomiting, dehydration, hypotension, peripheral neuropathy and tetany secondary to hypocalcaemia. A rare presentation is limb weakness secondary to severe hypokalaemia. This was present in the case that we have described but has also been reported in two other cases. Metabolic and electrolyte disturbances that have been reported include renal dysfunction presenting as increased creatinine level, metabolic acidosis, hypokalaemia, hypocalcaemia, hyponatraemia, hypomagnesaemia and hypoalbuminaemia. The case that we have described presented with one of the most severe reported hypokalaemia in coeliac crisis. Another rare presentation is bleeding diathesis secondary to prolonged prothrombin time. In our case the patient had a prolonged INR but there was no evidence of bleeding.

All reported patients required hospitalization, intravenous fluids with correction of metabolic and electrolyte disturbances and they were all started on a gluten-free diet. Few patients required parenteral nutrition and corticosteroids. In the majority of patients symptoms resolved with supportive care and a gluten-free diet alone.

The reason why some individuals present with coeliac crisis whereas the vast majority of patients with coeliac disease run a milder course is unclear. In some of the reported cases, coeliac crisis appears to be precipitated by a general immune stimulus such as pregnancy, surgery and infection. In most of the reported cases, including our case, no precipitating factor was present. It is unclear whether coeliac crisis in adults occurs at disease onset or whether coeliac disease is present but undiagnosed until a trigger leads to disease exacerbation. Two of the reported cases of coeliac crisis occurred in individuals who had already been diagnosed with coeliac disease but had not been following a gluten-free diet.

Thus, the cause of ‘coeliac crisis’ is unknown. In the reported adult cases, the patients presented with relatively acute onset watery diarrhoea, although chronic symptoms have also been present in some cases. This is similar to our case, in which the patient described a seven week history of diarrhoea that occurred two years prior to presentation with coeliac crisis.

The mainstays of treatment of coeliac crisis are initiation of a gluten-free diet, parenteral fluid replacement and nutritional support with correction of the underlying metabolic and electrolyte disturbances. Most of the reported cases including our case responded quickly to these interventions alone. For
individuals not responding promptly to gluten restriction, treatment involves the use of steroids, which are weaned off over a period of few months.1

Conclusion
This case demonstrates that although coeliac crisis is rare, it should be considered in the differential diagnosis of all patients presenting with an acute onset of severe diarrhoea with metabolic disturbances. Any patient found to have an increased tissue transglutaminase IgA in this setting should be placed on a gluten-free diet and have a small intestinal biopsy performed as soon as possible. Corticosteroids should be considered in cases of coeliac crisis when a gluten-free diet, in conjunction with fluid and electrolyte repletion, does not result in rapid improvement. Nutritional support often is required in the short term but most patients ultimately respond to gluten avoidance.

Acknowledgement
Dr D. Babic, Histopathologist, Department of Pathology, Mater Dei Hospital, Malta for the interpretation of the histology.

References

Interested in living and working in London for a year, or more?
Want to work in the exciting field of Emergency Medicine?
Keen on gaining the best training to pass the MCEM?

We are looking for doctors with at least 2 years postgraduate experience in acute specialties of which at least 6 months in Emergency Medicine, to work and train as a Senior Clinical Fellows in the Emergency Department of Queen Elizabeth Hospital, Woolwich in London. The hospital is located in the heart of historic South East London.

Successful candidates will join a dynamic emergency workforce treating a wide spectrum of emergency medicine presentations. This is a great opportunity to broaden your experience while working in a very friendly and supportive environment.

There is a strong training culture with protected four hours of training per week, incorporating lecture-room teaching and clinical supervision, attendance at relevant clinics and courses, and full immersion simulation sessions. Special focus is given towards passing the College of Emergency Medicine examinations.

There are also opportunities for flexible working and career progression to a more senior grade.

The post holders will work up to 48 hours per week (including night shifts and weekend duties). There are 3-4 night-shifts per month. Accommodation may be available.

For more details please contact Mr Ferdinod Ohanusi (Consultant in Emergency Department) on +44 208 836 4367/4364, fohanusi@nhs.net or Mr Hayder Hassan (Consultant in Emergency Medicine) on +44 2088364367/4364, hayder.hassan@nhs.net.

A more detailed description of these posts and information on how to apply is available by visiting our website http://jobs.slh.nhs.uk/job/v262216. Closing date: 30 September 2012.