Hemolytic uremic Syndrome. Presentation of case.

[1]Dr Yoerquis Mejias Sanchez, [2] Dr Tsegereda Gebrehiwot, [3] Dr Orgel Duany Machado.
[1] Pediatric Hospital Eduardo Agramonte Piña, Camaguey, Cuba
[2] Orotta Pediatric Referral National Hospital, Eritrea
[3] Ministry of Health, Cuba
Corresponding author:
Dr Yoerquis Mejias Sanchez
Pediatric Hospital Eduardo Agramonte Pina
Camaguey, Cuba
Email yoeime@yahoo.com

Abstract

A 1 year child with a body weight is 8kg was brought to Orotta National Referral Pediatric Hospital because he had diarrhea and fever for one week. Initially the diarrhea was frequent watery yellow, and non bloody and during the last two days became mucous and greenish with bloody. Fever of about 38, vomiting coffee ground and failure to pass urine developed preceding death after two days.

Introduction

Hemolytic uremic syndrome (HUS) is a syndrome characterized by thee main features; microangiopatic hemolytic anemia, thrombocytopenia and acute renal failure[1-4]. The most important abnormalities are damage on erythrocytes, platelets, capillary endothelial and glomerular cell membranes. HUS is caused by enterotoxins and bacterial polysaccharide[3].

HUS was first described by V. Passer in Suiza, in 1955. Other cases were subsequently detected cases in Europe, North and South America, South Africa, Australia, India and Japan[1,2]. The incidence of HUS over the years has been increasing in children and has become an important cause of acute renal failure in this group of people[3,4]. It is the commonest cause of acute renal failure and the second cause of chronic renal failure and renal transplantation in children in Argentina[5,6]. The global incidence is about 2,1 cases for every 100.000 people per year with the maximum incidence in children less than 5 years of age where it is 6,1/100.000/year and the minimum of 0,5/100.000/year in people between 50 and 59 years[5,7]. Adults generally are rarely affected probably because they have acquired immunity. However, in children younger than 8 years the risk of developing HUS is high with between 7 to 10 percent who are affected can developing HUS. The etiology can be idiopathic, infectious and non infectious diseases and it can be typical or atypical[1,2].

Case presentation

A one year old with a body weight of 8kg was admitted to Orotta National Referral Pediatric Hospital because he had fever and diarrhea for one week. At first the diarrhea was frequent, watery with no blood. Later in the disease the diarrhea frequency increased with small amounts and blood started to appear in the excreta while the consistency changed to mucoid and greenish. Fever, vomiting coffee of ground and notable to pass urine developed during the last two days.

Physical exam:

The child was unconscious, pyrexial 38°C, pale, well hydrated with no peripheral edema. He was tachypnoeic with a respiratory rate of 34/min with normal vesicular breath sounds. Cardiovascular system was essentially normal with a tachycardia of 138/min and normal heart sounds.

Laboratory investigations showed leucocytosis of 16 with normal differential, Hb of 10.2g/dl, platelets 54 000. The blood film was normal. The results of urea, electrolytes and liver function tests were albumin 1,7, BUN 72, Creatinine 2,6, RBS 99, SGOT 153, SGPT 89, ALP 70, Na^+ 126, K^+ 7,8, and Chloride- 101.

Management:

The child was immediately transferred to ward C where he was given fluid 400ml x kg x day, furosemide, antibiotic, general supportive treatment. In spite of aggressive resuscitation the child unfortunately expired after 6 hours from the time of admission.

Summary of clinical problems in this case:

1- Acute Renal Failure (, HUS)
2- Gastroenteritis.
3- Underweight.

Discussion

The most important infectious cause of HUS is bacterial Enterohemorrhagic Escherichia coli (EHEC) serotype O157:H7. It can produce a very potent cytotoxin Shigatoxin (Stxs), for this reason it is also named Shigatoxin Echerichia Coli (STEC). The infection is frequently due to the consumption of contaminated meat and milk products, uncooked or without pasteurization and due to contaminated water consumption, fruits, vegetables or by means of the inter human contact. The hamburger which is not properly cooked is an important vector that causes more than half of the reported epidemic cases [2.5.7]. Shiga toxin E. Coli (STEC) Infection has an incubation period of about 2-4 days, the first manifestations are usually non-bloody diarrhea, abdominal cramps, at the 2nd to 3rd day of illness bloody diarrhea develops with low

fever, cramps are an important findings in infants and children. If there are not complications, the disease usually resolves within a week [9-11]. Other infectious causes include Shigella dysenteryae[1], pneumococcus, salmonella typhi, campylobacter fetus jejuni, yersinia, bacteroides, portillo virus, cocksackie, influenza, Epstein Barr, rotavirus, HIV, and microtatobiotos [1,2].

The typical pathophysiological mechanism involves release of Shigatoxin (Stx) which has a potent toxin and cytotoxin which damage the enterocytes and can produce bloody diarrhea. Circulating toxins bind to receptor (gb3) in small capillary in kidneys which damage vascular endothelia, liberating inflammatory mediators. Platelets are decreased, and erythrocytes suffer mechanical deformity causing hemolytic anemia. The lumen of renal capillaries decreases causing decreased oxygenation to the kidney. As a result glomerular filtration decreases leading to acute renal failure [1,2,9,11].

Atypical pathphysiological mechanisms are associated with immunological processes like neuraminidase of pneumococcus which damages the sialic acid of erythrocytes. A Thomson-Friedereich (T-F) antigen that integrates with IgM has been shown to cause hemolysis, endothelial damages and thrombosis [1,2,11].

HUS has 4 phases on clinical manifestations: Prodromal, free interval, acute phase and chronic phase or sequelae. 90% of the l manifestations occur in the prodroma phase are gastrointestinal like abdominal pain, fever, vomiting and diarrhea. The remaining 10% of the prodromal features can be respiratory, but there are rare and most of the time they are seen in older children. The acute phase usually occurs after 1-15 days and is characterized by hemolytic anemia, thrombocytopenia, acute renal failure, pallor weakness, fever, hepato-splenomegaly when hemolytic crisis is present. Hemorrhagic diathesis is a rare manifestation which is seen due to thrombocytopenia and can be manifested by gastrointestinal bleeding like hematemesis, melena, purpura or ecchymosis. The duration of thrombocytopenia can be about 7 days, when platelets start to be normal it is an indication of recovery. More than 50% of cases start with acute renal failure oliguria or oligo-anuria and can have arterial hypertension. The sequelae can be arterial hypertension and chronic renal failure [4,5,12].

Management of HUS

Strict fluid management: Hydration and electrolyte balance and correction of hyponatremia are very important. Anemia requires transfusion if Hb is less than 70 g/dl or hypovolemia. Furosemide is indicated if urine output is less than 1 ml/kg/h. Dopamine in renal doses can be administered to improve the perfusion. Hypertension is best managed using calcium blocker. Some indications for peritoneal dialysis include oliguria which is not responding to furosemide, over hydration, acidosis, refractory hypertension. In difficult cases of peritoneal dialysis hemodialysis and eventually renal transplantation can be considered following development of chronic renal failure [2,5,8,12].

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