Hemolytic uremic Syndrome. Presentation of case.

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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 not 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 three main features; microangiopathic hemolytic anemia, thrombocytopenia and acute renal failure1. The most important abnormalities are damage on erythrocytes, platelets, capillary endothelial and glomerular cell membranes. HUS is caused by enterotoxins and bacterial polysaccharide 2. 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 3. 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 1-4. 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 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 2-12.

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 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 tachypnoic with a respiratory rate of 38/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 serum 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 400nl x kg x day, furosamide, 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 Shiga toxin (Stx), for this reason it is also named Shiga toxin Escherichia 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 3-5. 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 no complications, the disease usually resolves within a week. Other infectious causes include Shigella dysenteriae, Pseudomonas aeruginosa, salmonella typhi, campylobacter fetus jejuni, yersinia, bacteria, portillo virus, coxsackie, influenza, Epstein Barr, rotavirus, HIV, and microtobioitos.

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 capillaries 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.

Atypical pathophysiological mechanisms are associated with immunological processes like neuraminidase of pseudomonas aeruginosa which damages the sialic acid of erythrocytes. A Thomson-Friedreich (T-F) antigen that integrates with IgM has been shown to cause hemolysis, endothelial damages and thrombosis.

HUS has 4 phases on clinical manifestations:
1. Prodromal, fever, vomiting, diarrhea, abdominal pain, hemolytic anemia, thrombocytopenia, acute renal failure, pallor.
2. Acute, hemolysis, fever, hepatosplenomegaly, thrombocytopenia, acute renal failure.
3. Chronic, hemolytic anemia, thrombocytopenia, acute renal failure.
4. Sequelae, hemolytic anemia, thrombocytopenia, acute renal failure.

Management of HUS

Strict fluid management: Hydration and electrolyte balance and correction of hypovolemia 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.

References