Hemolytic Uremic Syndrome

By

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Introduction

• The hemolytic uremic syndrome is the most common cause of acute renal failure in young children.

• It is classically characterized by the triad of microangiopathic hemolytic anemia.

• HUS has features common to thrombotic thrombocytopenic purpura, except than the latter tends to occur in young adult women as a relapsing illness with fever, serious CNS involvement and thrombocytopenia.
Etiology

- An acute enteritis with diarrhea caused by shiga like toxin producing E coli precedes 80% of more of HUS cases in developed countries. The reservoir of this organism is the intestinal tract of domestic animals.
- It is usually transmitted by undercooked meat or unpasteurized milk.
- Outbreaks also occur with diarrheal epidemics after swimming in contaminated pond, lakes or pools as well as eating contaminated milk, cheese from, child care centres, dairy or petting forms, and by person to person contact.
Clinical manifestations

• HUS is the most common in children younger then 4 yrs of age.
• Onset is usually preceded by gastroenteritis characterized by fever, vomiting, abdominal pain, and diarrhea that is initially watery, but then becomes bloody.
• Less commonly pts may present after an upper respiratory tract infection.
Cont....

- Sudden onset of pallor, irritability, weakness, lethargy and oliguria usually occurs 5-10 days after the initial gastrointestinal or respiratory illness.

- Physical examination may reveal dehydration, edema, petechiae, hepatosplenomegally and marked irritability.
Diagnosis

• The diagnosis is supported by the findings of a microangiopathic hemolytic anemia, thrombocytopenia and Ac. renal failure.

• Anemia....(acute onset) with microangiopathic changes (i.e. shistocytes, bur cells, or helmet cells) on peripheral blood smear.
• Renal injury (acute onset) evidenced by either hematuria, proteinuria, or elevated creatinine level (more than or equal to 1.0 mg/dl in a child younger than 13 yr, or more than or equal to 1.5 mg/dl in a person 13 yr or older).
Case classification

• **Probable**… an acute illness diagnosed as HUS that meets the laboratory criteria in a patient who does not have a clear history of acute or bloody diarrhea in the preceding 3 week. or

• An acute illness diagnosed as HUS that has onset within 3 wks after onset of an acute or bloody diarrhea and meets the laboratory criteria except that microangiopathic changes are not confirmed.
Cont....

- **Confirmed** .... an acute illness diagnosed as HUS that meets the laboratory criteria and begin with in three week after onset of an episode of acute or bloody diarrhea.
Complications

Complications include

- Anemia
- Acidosis
- Hyperkalemia
- Fluid over load
- Heart failure
- Hypertension
- And uremia
• External manifestations of CNS, GIT, heart and skeletal muscle may be life threatening.

• Other complications such as skin necrosis, parotitis, adrenal dysfunction and rhabdomyolysis have been reported.
Treatment

• Supportive care….fluids, electrolytes, nutrition & control of hypertension.
• Plasmaphoresis & FFPs transfusion.
• Peritoneal dialysis….it controls fluids and electrolyte abnormality and maintains normal intravascular volume. It may contributes to dissolution of vascular thrombi.
Prognosis

• More than 90% pts survive with better management of ARF.
• Death or end stage renal disease effects 12% of Pts.
• HTN, proteinuria, low GFR affects 25% Pts.
• Overall prognosis is associated with negative long term renal outcomes, when CNS symptoms are present during acute illness or dialysis is required.
Follow up

• Pts require long term follow up, because complications such as HTN CRF & proteinuria may not be apparent up to 20 yrs.

• Renal transplantation in pts with HUS can be successful. Although there may be disease recurrence.
THANK YOU