EM CASE OF THE WEEK.
BROWARD HEALTH MEDICAL CENTER
DEPARTMENT OF EMERGENCY MEDICINE

BROWARD HEALTH
Care Warriors

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Hemolytic Uremic Syndrome

A 55-year-old man comes to the ED with an 8-day history of diarrhea, fatigue, vomiting, jaundice, and dark urine. He denies recent travel. He recalls the symptoms beginning after eating at a salad bar at a local restaurant with the family. Laboratory evaluation reveals low hemoglobin of 9 g/dL, low platelets, low haptoglobin, high LDH, high creatinine, high indirect bilirubin. Liver function tests were normal. Peripheral blood smear is displayed to the right. What is the best initial treatment for this patient? [1]

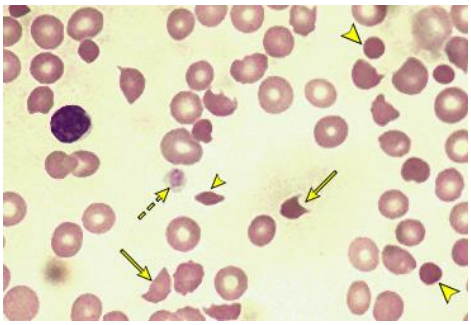
A. Whole blood transfusion

B. Anti-motility agents and whole blood transfusion

C. Plasmapheresis

D. Supportive therapy

E. Splenectomy



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Peripheral smear showing schistocytes, typical in micro-angiopathic hemolytic anemias.

Hemolytic Uremic Syndrome (HUS) is characterized by progressive renal failure, Microangiopathic hemolytic anemia, and thrombocytopenia.

EM Case of the Week is a weekly "pop quiz" for ED staff.

The goal is to educate all ED personnel by sharing common pearls and pitfalls involving the care of ED patients. We intend on providing better patient care through better education for our nurses and staff.

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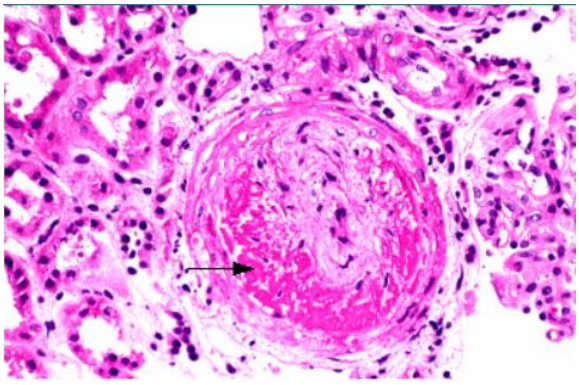
Answer

The answer is D. This patient has Hemolytic Uremic Syndrome (HUS) due to the triad of microangiopathic hemolytic anemia, thrombocytopenia, and acute kidney injury. HUS associated with diarrhea has a high likelihood of being shiga toxin related. In this instance, only supportive therapy is needed. Anti-motility agents should not be used to symptomatically treat his diarrhea because it has been shown to worsen HUS by increasing the toxin exposure in the gut (Choice B). Thrombotic thrombocytopenic purpura (TTP) can present similarly to HUS but does not present with diarrhea. In this instance, emergent plasmapheresis is implicated. Plasmapheresis has not been shown to improve outcomes in shiga toxin-mediated HUS (choice C). This patient is not experiencing active bleeding nor is his hemoglobin low enough to warrant a whole blood transfusion (choice A). Splenectomy would not help this patient as HUS is not due to splenic sequestration (choice E).

Discussion

HUS is a form of Thrombotic Microangiopathy (TMA) that is caused by an infection by enteric bacteria with Shiga toxin. Most patients are under 5 years old, but HUS occurs in adults. In the US, *E. coli* O157:H7, O111, or O104:H4 are organisms that cause this disease. Cases can occur in large outbreaks or sporadic due to improperly handled foods, particularly beef.[2,3]

TMAs are a group of disorders that cause disruption of the microvasculature of capillaries and arterioles. Disruption of the walls of tiny vessels lead to platelet aggregation, leading to microthrombi in the microvasculature. This consumption of platelets leads to a thrombocytopenia. In addition, as red blood cells flow through these vessels, they are mechanically sheared and fragmented leading to the formation of Microangiopathic Hemolytic Anemia (MAHA). Many organs may be affected by this pathology, primarily the kidney and the Central Nervous System (CNS) due to tissue necrosis produced by the occlusion of small vessels. Additionally, Shiga toxin directly damages renal podocytes, mesangial cells, and endothelial cells producing severe acute kidney injury.[2,3]



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Light microscopy of renal arteriole displaying subintimal fibrin deposition and narrowing of the lumen.

Diagnosis and Treatment

Diagnosis of HUS is primarily a clinical diagnosis. Patients will often have a history of animal exposure, under-cooked meats, contaminated vegetables, or contaminated water. Initial presenting symptoms include severe abdominal pain, nausea, vomiting, and bloody diarrhea. This is often mistaken for ischemic colitis or appendicitis in adults. Over the next several days, patients develop the characteristic MAHA, thrombo-cytopenia, and kidney injury. Initial workup includes CBC, LDH, serum creatinine, immunoassay for Shiga toxin, and stool culture for enterohemorrhagic *E. coli* and *Shigella* sp.[2]

Treatment of diarrhea-associated HUS is supportive care. This includes fluid administration, electrolyte management, and the cessation of nephrotoxic medications. RBC transfusions may be given for hemoglobin <7 g/dL. Platelet transfusions may be given for significant bleeding. Dialysis therapy may be initiated in patient who have symptomatic uremia, azotemia, or severe fluid overload. The use of anti-motility agents for diarrhea is contraindicated as this has been associated with hindering the gut's removal of the toxin.[4]

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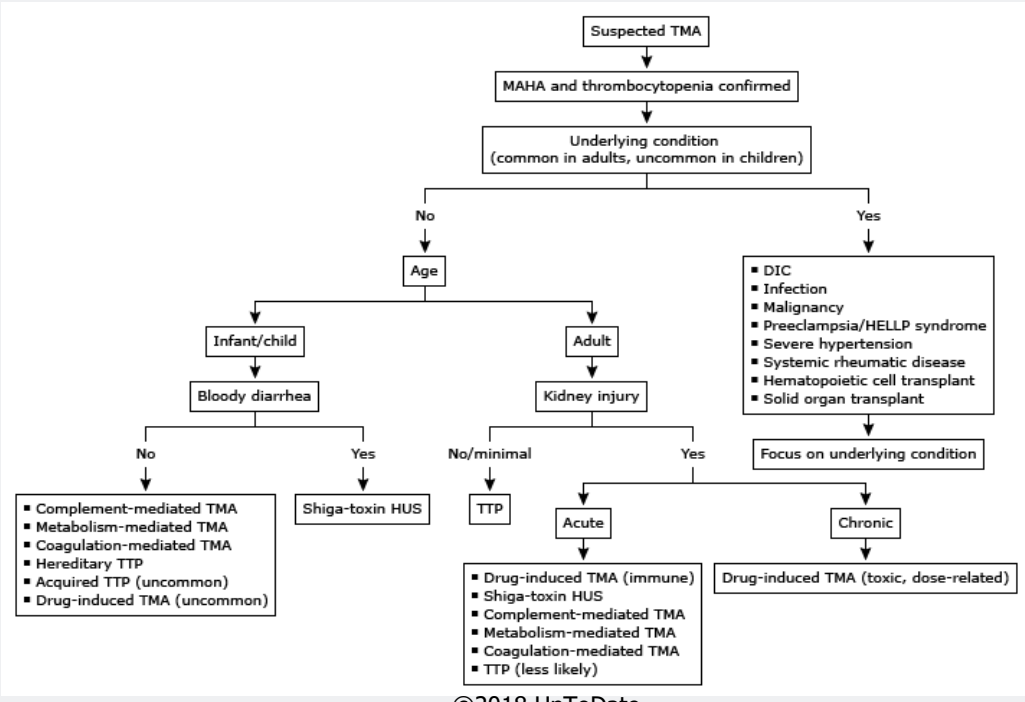
Algorithm to diagnosis in a patient with MAHA and thrombocytopenia



Prognosis

Complications of HUS include renal failure, stroke, coma, seizures, and hemorrhage. With supportive therapy, 85% of patients recover renal function. About 25% of patients suffer from cognitive decline. Risk factors that are associated with a poorer prognosis are: severe hypertension, involvement of medium sized arteries, severe involvement of CNS, extensive glomerular involvement (>80%), persistent consumption of clotting factors, and age >5 years old. Mortality rate is between 3%-5%, with older adults having a poorer prognosis.[4]

Take Home Points

- Hemolytic Uremic Syndrome (HUS) is characterized by progressive renal failure, microangiopathic hemolytic anemia, and thrombocytopenia.
- HUS is most often diagnosed in pediatric patients but can occur in adults, whom have worse prognoses.
- The best treatment for diarrheal-associated HUS due to shiga toxin is supportive care.

ABOUT THE AUTHOR

This month's case was written by Gina Furicchia. Gina is a 4th year medical student from FIU-COM. She did her emergency medicine rotation at BHMC in Dec 2018. Gina plans on pursuing a career in Pathology after graduation.

REFERENCES

[1]Kouzy R, et al. A Case of Typical Hemolytic Uremic Syndrome in an Adult. Cureus. 2018;10(9):e3289. Published 2018 Sep 11. doi:10.7759/cureus.3289

[2]UpToDate: Approach to patient with suspected TTP, HUS, or other TMAs

[3]UpToDate: Pathophysiology of acquired TTP and other TMAs

[4]Medscape: Hemolytic Uremic Syndrome