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Original Research Paper

Internal Medicine

BILATERAL SYMMETRICAL DIGITAL GANGRENE, A RARE PRESENTATION OF HEMOLYTIC UREMIC SYNDROME(HUS) - A CASE STUDY

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ABSTRACT Hemolytic uremic syndrome (HUS) is a thrombotic microangiopathy characterized by intravascular hemolysis, thrombocytopenia, and acute kidney failure. HUS is usually categorized as typical, caused by Shiga toxin–producing Escherichia coli (STEC) infection, as atypical HUS (aHUS), usually caused by uncontrolled complement activation, or as secondary HUS with a coexisting disease. In recent years, a general understanding of the pathogenetic mechanisms driving HUS has increased. Typical HUS (STEC-HUS) follows a gastrointestinal infection with STEC, whereas aHUS is associated primarily with mutations or autoantibodies leading to dysregulated complement activation. Among the 30% to 50% of patients with HUS who have no detectable complement defect, some have either impaired diacylglycerol kinase enzyme (DGKε) activity, cobalamin C deficiency, or plasminogen deficiency. Some have secondary HUS with a coexisting disease or trigger such as autoimmunity, transplantation, cancer, infection, certain cytotoxic drugs, or pregnancy. The common pathogenetic features in STEC-HUS, aHUS, and secondary HUS are simultaneous damage to endothelial cells, intravascular hemolysis, and activation of platelets leading to a procoagulative state, formation of microthrombi, and tissue damage. In this review, the differences and similarities in the pathogenesis of STEC-HUS, aHUS, and secondary HUS are discussed. Common for the pathogenesis seems to be the vicious cycle of complement activation, endothelial cell damage, platelet activation, and thrombosis. This process can be stopped by therapeutic complement inhibition in most patients with aHUS, but usually not those with a DGKε mutation, and some patients with STEC-HUS or secondary HUS. Therefore, understanding the pathogenesis of the different forms of HUS may prove helpful in clinical practice.

KEYWORDS : HUS(HEMOLYTIC UREMIC SYNDROME),ADAMTS13,AKI(Acute Kidney Injury), Gangrene

CASE HISTORY

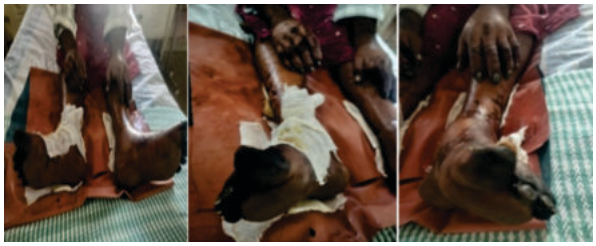
A 60-year-old chronic male smoker presented with a week-long low-grade fever, body pain, generalized weakness, blackening of fingers and toes, numbness, and swelling up to the mid part of limbs. Petechiae rash appeared on trunk and limbs. Dark-colored, decreased urine for two days. Rashes enlarged, yellowish scleral and leg discoloration, and facial swelling developed. No history of nausea, vomiting, diarrhea, limb pain, breathlessness, or signs of central nervous system involvement like altered behaviour, severe headache, blurring of vision, Convulsion, loss of consciousness etc.

Investigation

- CBC: Anaemia, thrombocytopenia with peripheral smear showing schistocytes and fragmented RBCs.
- LFT: Indirect hyperbilirubinemia and raised Serum LDH.
- RFT: Raised Serum creatinine and Serum Urea
- Urine routine examination: shows few RBCs and epithelial cells and Albumin +1 by dipstick.
- Coagulation profile, electrolytes, USG Abdomen pelvis and KUB, Chest XRay, Brain imaging were normal.
- Dengue NS1 and IgM and tests for Malaria are negative. USG Bilateral Upper and Lower limb Arterial Doppler was normal. Serum ANA was negative.

Investigations	On admission	After 3 cycles of plasmapheresis	On diascharge
CBC			
Hb(gm/dl)	10.90	9.8	9.7
TLC	9600	14000	11800
TPC	9000	1,80,000	3,84,000
LFT			
Total Bilirubin(mg/dl)	1.39	0.08	0.90
Ind. Bilirubin(mg/dl)	0.71	0.63	0.68
LDH(U/L)	547	444	<200
RFT			
Sr Urea(mg/dl)	227	64	30
Sr Creatinine(mg/dl)	2.76	1.34	0.77
Coagulation Profile			
PT(seconds)	15.20	21.40	16.70
INR	1.09	1.58	1.21
aPTT(seconds)	31.0	>140	31.6

Diagnosis: Bilateral Symmetrical Digital Gangrene in Case of HEMOLYTIC UREMIC SYNDROME



DISCUSSION

HUS typically manifests as a triad: microangiopathic hemolytic anaemia (MAHA), uremia (AKI), and thrombocytopenia. It has two types: Typical HUS, preceded by diarrhea, caused by Shiga toxin-producing E.coli; and Atypical HUS, linked to acquired or inherited complement dysregulation. Diagnosis involves detecting hemolysis, thrombocytopenia, screening for Shiga toxin-producing E.coli, ADAMTS 13, and genetic studies for complement dysregulation. Treatment includes plasmapheresis until platelet count normalizes, steroids, and anti-complement therapy like ECULIZUMAB.

Management and Out Come

Treatment involved plasmapheresis, resulting in improved laboratory parameters after six cycles. Discharge showed normal levels of laboratory parameters and clinical improvement, with follow-up recommended.