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From Atypical Suspect to Typical Diagnosis: An Adult Woman Shiga Toxin HUS Case

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Background: Thrombotic microangiopathies (TMAs) are disorders marked by microvascular clots, thrombocytopenia, microangiopathic hemolytic anemia, organ injury. Hemolytic uremic syndrome (HUS), belongs to this group and may have infectious or autoimmune origins: the typical form (tHUS) follows Escherichia Coli infection, the atypical form (aHUS) stems from dysregulated complement activation.

Case History: A 62-year-old woman presented to the emergency department with a 7-day history of asthenia, diarrhea, vomiting, self-treated with antibiotic. At home, she developed aphasia and confusion. Admitted to Internal Medicine, laboratory tests showed acute renal failure, severe thrombocytopenia, anemia, low haptoglobin, mild hyperbilirubinemia, raised inflammatory markers, and high LDH; brain CT/MRI were normal. ADAMTS-13 activity was normal; peripheral smear revealed schistocytes, suggesting HUS. After expanded stool culture, considering the patient's age, empiric treatment for suspected aHUS was started with a 2700 mg dose of ravulizumab and supportive care with fluids and ceftriaxone 2 g/day. Culture later identified Shiga toxin-producing E. Coli, establishing tHUS. After ten days: platelet count rose, creatinine dropped, transfusions were no longer needed. Patient was stable, though mild neurological symptoms persisted (slowed speech, temporal-spatial disorientation).

Differences between Typical HUS, Atypical HUS, and TTP

Characteristic	Typical HUS	Atypical HUS	TTP (Thrombotic Thrombocytopenic Purpura)
Etiology	Shiga toxin (E. coli O157:H7)	Complement dysregulation	ADAMTS13 deficiency (acquired or congenital)
Typical age of onset	Children (especially <5 years)	All ages, more common in young adults	Young adults (more common in women)
Triggers	Gastrointestinal infection	Sporadic or familial, various triggers	Infections, pregnancy, drugs, autoimmune diseases
Main symptoms	Triad: hemolytic anemia, thrombocytopenia, renal failure	Similar to typical HUS but often more severe and persistent renal involvement	Pentad: hemolytic anemia, thrombocytopenia, fever, neurological symptoms, renal failure
Renal involvement	Marked	Marked	Less prominent than in HUS
Neurological involvement	Rare	Possible	Common (confusion, seizures, coma)
Laboratory findings	Schistocytes, ↑ LDH, ↓ haptoglobin, anemia, thrombocytopenia, renal failure, positive E. coli	Similar, but no evidence of infection; ↓ C3/C4 sometimes	Similar, but ADAMTS13 activity <10%
Complement (C3, C4)	Normal	Decreased in some cases	Normal
ADAMTS13 activity	Normal	Normal	<10%
Treatment	Supportive, dialysis, avoid antibiotics	Eculizumab (anti-C5), plasma therapy	Urgent plasma exchange, corticosteroids, Rituximab

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Prognosis	Generally good	Worse, risk of relapse	Potentially fatal if untreated Discussion: TMA prognosis is often poor; clinical course and therapeutic su

Discussion: TMA prognosis is often poor; clinical course and therapeutic success highly depend on timely diagnosis and early initiation of targeted treatments

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