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Review Article

Recurrent Atypical Hemolytic Uremic Syndrome Presenting with Acute Kidney Injury in an Adolescent: A Case Report

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Atypical hemolytic uremic syndrome (aHUS) is a rare condition that can cause thrombocytopenia, hemolysis, and acute kidney injury. We report the case of a 17-year-old male with a previous episode of hemolytic uremic syndrome in 2023 who presented with high-grade fever, hematuria, generalized weakness, presyncope, angioedema, and vomiting. Initial investigations showed severe thrombocytopenia with a platelet count of 44 × 10^3/µL, elevated serum creatinine of 4.6 mg/dL, reduced estimated glomerular filtration rate of 17.4 mL/min/1.73 m^2, and markedly elevated lactate dehydrogenase (LDH) of 2191.9 U/L. Urinalysis revealed significant proteinuria and hematuria, while peripheral smear findings were suggestive of microangiopathic hemolytic anemia. Investigations for infectious and autoimmune causes were noncontributory. Based on the clinical and laboratory findings, recurrent aHUS with acute kidney injury was diagnosed. The patient was treated with plasma exchange, hemodialysis, intravenous fluids, blood-product transfusions, antibiotics, and other supportive measures. Following treatment, the platelet count improved to 350 × 10^3/µL, serum creatinine decreased to 2.0 mg/dL, and LDH declined to 451 U/L. The patient showed clinical and laboratory improvement and was discharged with advice for regular follow-up and monitoring of hematological and renal parameters.

Keywords: Atypical hemolytic uremic syndrome, recurrent aHUS, thrombotic microangiopathy, acute kidney injury, thrombocytopenia, plasma exchange

INTRODUCTION:

Hemolytic uremic syndrome (HUS) is a rare but serious thrombotic microangiopathy characterized by a triad of microangiopathic hemolytic anemia, thrombocytopenia, and acute kidney injury. The kidneys are among the most commonly affected organs, and the severity of renal involvement can vary considerably between patients.(1) Among the different forms of HUS, atypical hemolytic uremic syndrome (aHUS) is an uncommon and potentially life-threatening condition that is mainly associated with uncontrolled activation of the alternative complement pathway.(2)

In aHUS, genetic abnormalities affecting complement regulatory proteins or the presence of acquired autoantibodies can interfere with the normal regulation of the complement system. This results in excessive complement activation, which can damage vascular endothelial cells and promote the formation of microvascular thrombi. These changes ultimately contribute to hemolysis, thrombocytopenia, and injury to various organs.(3) Although the kidneys are predominantly affected, aHUS can also involve other organs. Patients may present with hematuria, proteinuria, hypertension, reduced renal function, and, in severe cases, acute kidney injury requiring renal replacement therapy.(2)

Diagnosing aHUS can be challenging because its clinical presentation may resemble other thrombotic microangiopathies, particularly Shiga toxin-associated HUS and thrombotic thrombocytopenic purpura (TTP). Therefore, diagnosis generally involves careful evaluation of the clinical presentation, laboratory findings, and appropriate investigations to exclude

other possible causes. Complement-related investigations and genetic or antibody testing may also help support the diagnosis.(3)

Severe renal involvement in aHUS may progress rapidly and can require interventions such as hemodialysis. Early recognition and appropriate management are therefore important to limit further organ damage and improve patient outcomes.(1,2) Even after the acute episode, patients may remain at risk of persistent or recurrent renal complications, emphasizing the importance of regular clinical and laboratory monitoring during follow-up.(1)

Case study:

A 17-year-old male presented with high-grade continuous fever, hematuria, generalized weakness, presyncope, angioedema, and two episodes of vomiting. He had a previous history of hemolytic uremic syndrome (HUS). Given his previous episode of HUS in 2023 and current clinical presentation, recurrence was suspected. Initial investigations showed a platelet count of 44 × 10^3/µL, serum creatinine of 4.6 mg/dL, and an estimated glomerular filtration rate (eGFR) of 17.4 mL/min/1.73 m^2, indicating significant renal impairment. His lactate dehydrogenase (LDH) was markedly elevated at 2191.9 U/L. Urinalysis showed proteinuria and hematuria, and the peripheral smear showed features suggestive of microangiopathic hemolysis. To identify the possible cause of the condition, several investigations were performed. Tests for malaria, dengue, leptospirosis, HIV, hepatitis B did not reveal any significant findings. Autoimmune evaluation, including ANCA testing, was also negative. Based on the clinical presentation and laboratory findings, together with his previous history of HUS, the patient was diagnosed with recurrent atypical hemolytic uremic syndrome (aHUS) associated with acute kidney injury. Treatment was started promptly with therapeutic plasma exchange. The first plasma exchange was performed with approximately 1400 mL of plasma exchanged, followed by another session. Due to worsening renal function, the patient also required hemodialysis. He received intravenous fluids, antibiotics, blood-product support, and other supportive treatment during his hospital stay. His blood counts, LDH, renal function, and electrolytes were closely monitored to assess his response to treatment. Over the course of treatment, the patient gradually improved. His platelet count increased from 44 × 10^3/µL to 350 × 10^3/µL, while LDH decreased from 2191.9 U/L to 451 U/L, suggesting improvement in hemolysis. Renal function also showed recovery, with serum creatinine decreasing from a peak of 9.1 mg/dL to 2.0 mg/dL. After his clinical condition and laboratory parameters improved, the patient was discharged with advice for regular follow-up. Continued monitoring of complete blood count, LDH, renal function, and electrolytes was advised because of the recurrent nature of his condition. This case highlights the importance of recognizing aHUS early in patients presenting with thrombocytopenia, hemolysis, and renal impairment. In this patient, timely plasma exchange, hemodialysis, and supportive management were followed by improvement in both hematological and renal parameters.

Table 1: Relevant Laboratory Findings During Hospitalization

Laboratory parameter	At presentation	During hospitalization / peak abnormality	At discharge
Platelet count	44 × 10^3/µL	—	350 × 10^3/µL
Serum creatinine	4.6 mg/dL	9.1 mg/dL	2.0 mg/dL
eGFR	17.4 mL/min/1.73 m^2	—	Improved
LDH	2191.9 U/L	—	451 U/L
Peripheral smear	Microangiopathic hemolytic picture	—	Improved

DISCUSSION:

Hemolytic uremic syndrome (HUS) is a thrombotic microangiopathy that typically presents with thrombocytopenia, microangiopathic hemolytic anemia, and renal impairment.(2,3) Atypical HUS (aHUS) is mainly associated with uncontrolled activation of the alternative complement pathway, resulting in endothelial injury and formation of microvascular thrombi. The patient's previous episode of HUS in 2023 is particularly important because aHUS can follow a relapsing course.(4,5) Complement abnormalities may remain clinically silent until an appropriate trigger initiates endothelial injury and

thrombotic microangiopathy.(4) Pediatric studies have also shown that some complement abnormalities are associated with recurrent disease, while the clinical course can vary considerably between patients.(5) In our patient, the recurrence of a similar clinical picture several years after the first episode raises the possibility of an underlying complement-mediated susceptibility.

Diagnosis of aHUS can be challenging because its clinical findings overlap with other thrombotic microangiopathies, particularly Shiga toxin-associated HUS and thrombotic thrombocytopenic purpura (TTP).(6,7) Therefore, evaluation generally involves excluding other causes of thrombotic microangiopathy, including Shiga toxin-producing *Escherichia coli* infection and severe ADAMTS13 deficiency associated with TTP.(6) In our patient, investigations for several infectious and autoimmune causes were noncontributory, while the combination of thrombocytopenia, hemolysis, renal impairment, and previous HUS supported the clinical diagnosis of recurrent aHUS. Renal involvement is one of the most important manifestations of aHUS, and severe acute kidney injury may require dialysis or other forms of renal replacement therapy.(2,3) A recent prospective national pediatric cohort from Poland demonstrated that aHUS continues to be associated with considerable renal morbidity, although outcomes have improved with advances in disease-specific treatment.(1) Our patient developed severe renal impairment and subsequently required hemodialysis, highlighting the potential severity of renal involvement during an acute episode. The improvement in serum creatinine from a peak of 9.1 mg/dL to 2.0 mg/dL suggests substantial renal recovery following treatment. Plasma therapy has historically played an important role in the initial management of aHUS, particularly when complement-directed treatment is unavailable or while the diagnostic evaluation is ongoing.(2,6) Previous pediatric literature has reported clinical improvement following intensive plasma therapy in selected patients with aHUS.(5) A pediatric study evaluating plasma exchange in aHUS also reported its usefulness in achieving hematological remission, particularly in settings where access to complement inhibitors is limited.(7) In our patient, plasma exchange was followed by a marked improvement in platelet count, which increased from 44 × 10^3/μL to 350 × 10^3/μL, together with a reduction in LDH from 2191.9 U/L to 451 U/L, suggesting improvement in the thrombotic microangiopathy and hemolysis. The management of aHUS has changed considerably with the introduction of complement inhibitors, particularly eculizumab, which blocks terminal complement component C5.(2,4,6) Current literature considers complement inhibition an important disease-specific treatment for confirmed complement-mediated aHUS and reports improved hematological and renal outcomes compared with historical plasma-based treatment alone.(2,6,8) Recent recommendations also emphasize complement genetic testing and assessment for complement-related autoantibodies when aHUS is suspected, although genetic testing may not always be positive or immediately available.(8) In resource-limited settings, plasma exchange and supportive renal therapy may therefore remain important components of acute management while definitive diagnostic evaluation and access to complement-directed therapy are considered.

Overall, this case demonstrates the importance of recognizing the combination of thrombocytopenia, hemolysis, and acute kidney injury, particularly in a patient with a previous episode of HUS.(3,6) The patient's improvement following plasma exchange, hemodialysis, and supportive management highlights the potential for significant hematological and renal recovery when treatment is initiated promptly. Long-term follow-up remains important because recurrent disease and persistent renal complications can occur in aHUS.(1,4)

CONCLUSION:

This case highlights the importance of early recognition and timely management of recurrent atypical hemolytic uremic syndrome (aHUS), particularly when thrombocytopenia, hemolysis, and acute kidney injury occur together. The patient's previous history of HUS, along with the current clinical and laboratory findings, raised strong suspicion of a recurrent episode. Prompt initiation of plasma exchange, supported by hemodialysis and appropriate supportive care, was followed by progressive improvement in both hematological and renal parameters. The platelet count increased from 44 × 10^3/μL to 350 × 10^3/μL, while LDH decreased substantially, indicating improvement in hemolysis. Renal function also improved, with serum creatinine decreasing from a peak

of 9.1 mg/dL to 2.0 mg/dL. These findings demonstrate the potential for significant recovery when severe thrombotic microangiopathy is recognized and managed promptly. However, aHUS can have a recurrent course and may result in long-term renal complications. Therefore, continued follow-up with regular monitoring of complete blood count, LDH, renal function, and electrolytes is important. Complement studies and genetic evaluation, when available, may further clarify the underlying cause and help guide long-term management.

REFERENCES

1. Zagożdżon I, Szczepańska M, Leszczyńska B, Jarmużek W, Miklaszewska M, Tkaczyk M, et al. Changing Epidemiology and Outcomes of Hemolytic Uremic Syndrome in Children: A Prospective National Cohort Study from the Polish Pediatric HUS Registry and the Polish Registry of Renal Replacement Therapy in Children. J Clin Med. 2024 Oct 30;13 (21):6499. doi:10.3390/jcm13216499 PubMed PMID: 39518638; PubMed Central PMCID: PMC11546500.
2. (PDF) Atypical Hemolytic-Uremic Syndrome: An Update on Pathophysiology, Diagnosis, and Treatment. ResearchGate [Internet]. 2026 Aug 2 [cited 2026 Sep 24]. Available from: https://www.researchgate.net/publication/328143509_Atypical_Hemolytic-Uremic_Syndrome_An_Update_on_Pathophysiology_Diagnosis_and_Treatment
3. Sridharan M, Go RS, Willrich MAV. Atypical hemolytic uremic syndrome: Review of clinical presentation, diagnosis and management. J Immunol Methods. 2018 Oct;461:15–22. doi:10.1016/j.jim.2018.07.006 PubMed PMID: 30031798.
4. Campistol JM, Arias M, Ariceta G, Blasco M, Espinosa L, Espinosa M, et al. An update for atypical haemolytic uraemic syndrome: diagnosis and treatment. A consensus document. Nefrol Publicacion Of Soc Espanola Nefrol. 2015;35 (5):421–47. doi:10.1016/j.nefro.2015.07.005 PubMed PMID: 26456110.
5. Loirat C, Noris M, Fremeaux-Bacchi V. Complement and the atypical hemolytic uremic syndrome in children. Pediatr Nephrol. 2008 Nov;23 (11):1957–72. doi:10.1007/s00467-008-0872-4 PubMed PMID: 18594873; PubMed Central PMCID: PMC6904381.
6. Franchini M. Atypical hemolytic uremic syndrome: from diagnosis to treatment. Clin Chem Lab Med. 2015 Oct;53 (11):1679–88. doi:10.1515/cclm-2015-0024 PubMed PMID: 25803082.
7. Khandelwal P, Thomas CC, Rathi BS, Hari P, Tiwari AN, Sinha A, et al. Membrane-filtration based plasma exchanges for atypical hemolytic uremic syndrome: Audit of efficacy and safety. J Clin Apheresis. 2019 Oct;34 (5):555–62. doi:10.1002/jca.21711 PubMed PMID: 31173399.
8. Java A. Atypical hemolytic uremic syndrome: diagnosis, management, and discontinuation of therapy. Hematol Am Soc Hematol Educ Program. 2024 Dec 6;2024 (1):200–5. doi:10.1182/hematology.2024000543 PubMed PMID: 39644051; PubMed Central PMCID: PMC11665500.

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