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Case Study

Atypical Hemolytic Uremic (Ahus) Syndrome with Kidney Injury in Pediatric Patient: A Case Report

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ABSTRACT

Hemolytic uremic syndrome is a form of thrombotic microangiopathy (TMA) which is characterized by thrombocytopenia, microangiopathy hemolytic anemia and renal impairment. Approximately 10% of cases of the hemolytic–uremic syndrome is classified as atypical, since they are not caused by either Stx-producing bacteria or streptococci. Early recognition and initiation of appropriate treatment are essential in patient with a HUS which involves supportive care and inhibition of complement system. Plasmatherapy is recommended as first line treatment approach for aHUS. Timely initiation of plasma exchange, dialysis along with supportive treatment results in gradual improvement in 60- 70% of cases. There are other evolving approaches for the management of a HUS which shows promising results such as anti-C5 monoclonal antibody such as eculizumab, and treating complementary etiology

INTRODUCTION

Hemolytic uremic syndrome is a form of thrombotic microangiopathy (TMA) which is characterized by thrombocytopenia, microangiopathy hemolytic anemia and renal impairment.1

HUS is classified as typical hemolytic uremic syndrome and atypical hemolytic uremic syndrome (aHUS). Typical hemolytic uremic syndrome also called as STEC-HUS (for Shiga toxin–producing Escherichia coli HUS) Caused by Shiga toxin, E.coli, or shigella. aHUS caused by dysregulation of alternative pathway.5

Approximately 10% of cases of the hemolytic–uremic syndrome is classified as atypical, since they are not caused by either Stx-producing bacteria or streptococci. 3

Vascular endothelial cell injury is the key mechanism underlying all forms of HUS, these can

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be caused by inflammatory and non-inflammatory mechanism.2

The clinical presentation of aHUS in children and adults are often nonspecific including pallor, poor feeding, vomiting, fatigue, and drowsiness. Renal involvement may manifest as oliguria or anuria with or without peripheral edema. HTN is also an important manifestation that develop due to renal injury which can cause neurological complication or cardiac failure. 4

Early recognition and initiation of appropriate treatment are essential in patient with a HUS which involves supportive care and inhibition of complement system. Plasmatherapy is recommended as first line treatment approach for aHUS. It should be started within 24 hours of diagnosis with 20-30ml/kg body weight.3,7

Persistent hemolysis and or failure of platelet count improvement within 4-5 days after plasmatherpay is indication of failure of treatment hence should be stopped immediately, and start eculizumab it.4

Appropriate fluid and electrolyte management is a key component of HUS treatment. Dehydrated patients should be given appropriate fluid replacement, however excessive fluid overload should be avoided especially with patients developing oliguria. Hyponatremia occurs in renal injury hence it corrected carefully by hypertonic saline. Other electrolyte abnormalities (hyperkalemia, hypophosphatemia) in initially managed medically, if severity persist dialysis and hemofiltration may be required. HUS patient with high catabolic rate is managed by giving carbohydrate's and amino acid. If hemoglobin reduces below 6g/dl prbc transfusion is recommended. 2

Some patients with delayed diagnosis or irresponsive to therapy progress to ESRD. Renal replacement is done is such cases. Posttransplant HUS occurs in four clinical contexts that are inherited HUS, idiopathic HUS, de novo occurrence of HUS after renal transplant. Eculizumab is also used prophylactically to prevent reoccurrence of HUS after renal transplantation.2,4,

Case report:

A 7 yrs old male patient admitted in pediatric medicine department Karnataka Medical College and Research Institute (KMCRI) Hubballi, on 22 april 2026

Chief complaints:

This patient was referred from his native government hospital i/v/o vomiting, fever, headache, periorbital puffiness since 4 days, With h/o hematuria

Physical examination:

Pallor - +ve, Icterus - +ve, clubbing - -ve, cyanosis - -ve, lympedenopathy - -ve, edema - -ve.

Vital parameters:

Table : 1 Vital parameters:

Temperature	37.5 °c
Pulse rate	120bpm
Respiratory rate	26cpm
blood pressure	120/70mmHg
SpO2	99%

Anthropometry:

Anthropometry assessment revealed weight, height, BMI and other parameters were appropriate for age, gender with no evidence of malnutrition

Table : 2 Systemic examination:

Cardiovascular system	Pericardium normal, normal heart sound with no murmur
Respiratory system	NVBS +ve, A/E - +ve
Central nervous system	Conscious oriented, HMF intact,
Per abdomen	Tenderness present in right upper quadrant, bowel sound +ve

Table : 3 Neurological examination:

	Upper limb		**Lower limb**	
	Right	**Left**	**Right**	**Left**
Muscle	Normal	Normal	Normal	Normal
Power	4/5+	4/5+	4/5+	4/5+
Tone	Normal	Normal	Normal	Normal
Reflex	2+	2+	2+	2+

Table : 4 Hematological report:

Parameter	**21/04/2026**	**26/04/2026**	**1/05/2026**	**10/05/2026**	**Normal value**
Hemoglobin	6.3	8.3	8	9.3	12-15g/dl
WBC	10400	9030	4450	4840	5000-13000/ul
Lymphocytes	42.9	41.9	51.3	58.6	28-48%
Neutrophils	53	40.2	27.4	3.0	33-72%
PCV	18.0	-	23.9	27.6	35-50%
MCV	77.8	-	-	89.0	77- 95fl
MCH	27.3	-	-	30.0	25-33pg
MCHC	35.1	-	-	33.7	31-37%
PLT	0.32	0.43		1.55	1.7-4.5lakhs/cmm
S. urea	178.0	124.0	121.0	48.2	15- 45 mg/dl
S. creatinine	4.7	4.3	3.2	1.0	0.7-1.3mg/dl
S. Sodium	132.9	127.6	140.1	138.5	135-145mmol/l
S. Potassium	3.5	3.7	3.7	3.9	3.5-5.0mmol/l
S. Chloride	99.6	95.9	88.8	105.2	98-107mmol/l
LDH	3357.0	-	-	-	142-270U/L
Rc	30				2-6%
ASLO	negative				0-200IU/ml

Peripheral smear report:

RBC- microcytic hypochromic with macroovulocytes with few nucleated RBC's and good number of acanthocytes.

Impression:

Microcytic hypochromic anemia with relative lymphocytosis and thrombocytopenia.

Table : 5 LFT:

Parameter	Observed value	Normal value
Total bilirubin	1.4	0.3-1.2mg/dl
Direct bilirubin	0.2	0.03-0.18mg/dl
Indirect bilirubin	1.20	0.75mg/dl
Total protein	6.0	6-8.3g/dl
Serum albumin	3.0	3.5-5.0g/dl
Serum globulin	3	2.3-3.5g/dl
A/G ratio	1	1-2.5
AST	172.0	Upto 35U/L
ALT	52.1	Upto 45U/L
ALP	7.0	54 – 369U/L

Table : 6 Arterial blood gas analysis:

pH	7.45	7.35 – 7.45mmol/l
pCO2	23	35-45mmhg
PO2	34	80-100mmhg
HCO3	16	22-30mmol/l
TCO2	16.7	19-24mmol/l
BEecf	-8.0	-3 - 7 mmol/l

Urine routine analysis:

Table : 7 Chemical examination:

Urine glucose	nil	Absent
Urine protein	present	Absent

Table : 8 Microscopic examination:

parameter	result	Reference range
Pus cells (wbc's)	2-4	0-5
RBC	1-2	0-2

Urine culture and sensitivity test: no growth
Stool culture and sensitivity test: no enteric bacterial pathogen isolated

Ultrasonography reports:
-mild hepatosplenomegaly with increased echotexture of liver
-b/l kidney- bulky with increased echotexture and loss of CMD- likely s/o renal parenchymal diseases with RI values less than 0.8
-wedge shaped hypodensity seen in upper poles of right kidney? infract
-minimal interbowel loop fluid.

The patient was diagnosed with atypical hemolytic uremic(aHUS) syndrome. The patient underwent a detailed clinical examination including assessment of vital signs, systemic examination, physical examination, neurological examination, peripheral blood smear, hematological investigations, routine urinalysis, ASLO test, USG of abdomen, urine and stool culture and sensitivity test. The vital signs and anthropometry were normal, on physical examination pallor and icterus was seen. In systemic examination the tenderness was present in right upper quadrant, neurological examination

showed mild weakness with normal reflexes. The low hemoglobin level with marked LDH and peripheral blood smear raised suspicion of hemolytic anemia. Whereas elevated urea, creatinine, abnormal LFT and USG report demonstrated significant renal involvement, the arterial blood gas investigation showed metabolic acidosis these findings were considered supportive of the patient systemic and renal dysfunction. In view of the combination of anemia, thrombocytopenia, biochemical evidence of hemolysis (elevated LDH, indirect bilirubin, reticulocyte count) and renal dysfunction, a thrombotic microangiopathy was suspected. HUS was considered as differential diagnosis. Urine and stool culture sensitivity and ASLO test were negative for an infectious etiology making typical HUS less likely. The clinical presentation, laboratory evidence of TMA and predominant renal involvement subsequently supported the diagnosis of atypical hemolytic uremic syndrome after exclusion of other causes of TMA. For treatment four sessions of dialysis were performed to manage AKI and patient developed severe HTN after first dialysis hence he was treated with tablet nefidipine and later titrated and shifted to tablet amlodipine when BP subsequently shows decreasing trends. plasmapheresis was done to which patient responded well hence continued the same treatment. Other medications such as paracetamol, emeset, calcium, multivitamins, were given as supportive therapy.

Table : 9 Treatment chart:

SI NO	Drug	Dose	ROA	Frequency	Days given
01	Allowed orally				
02	T. paracetamol	500mg	PO	SOS	
03	Inj. Ranitidine	0.5cc	IV	BID	4days then stopped
04	Inj. emeset	2mg	IV	SOS	4days then stopped
05	T. Nifedipine	5mg	PO	BD	On 4^{th} day
06	T. Nifedipine	2.5mg	PO	BD	From 5^{th} – 11^{th} day
07	Inj. pantoprazole	20mg	IV	OD	From 5^{th} day
08	Syp. Calcium	5ml	PO	TID	From 6^{th} day
09	Syp. multivitamin	5ml	PO	BID	From 6^{th} day
10	T. Nifedipine	2.5mg	PO	TID	From 12^{th}- 15^{th} day
11	Syp.potklor	2ml	PO	QID	On 13^{th} day
12	T. Amlodipine	10mg	PO	HS	From 16^{th} day

Dialysis –4 cycles done on 23/4/26, 25/04/2026, 29/04/2026, 30/04/2026

After first dialysis patient developed BP hence titrated with nifedipine.

Plasmapheresis – 4 cycles done.

DISCUSSION

Atypical hemolytic uremic syndrome is rare life threatening TMA with a characteristic triad of thrombocytopenia, microangiopathy hemolytic anemia and renal impairment. Atypical HUS is mainly associated with dysregulation of complementary pathways unlike typical HUS.5

Differential diagnosis:

The differential diagnosis for aHUS were thrombotic thrombocytopenic purpura (TTP), typical HUS/ STEC – HUS and secondary TMA. The combination of anemia, thrombocytopenia, biochemical evidence of hemolysis (elevated

LDH, indirect bilirubin, reticulocyte count) and renal dysfunction, a TMA was suspected. However, ASLO test and antimicrobial test were negative, and the absence of diarrhea which exclude it from typical HUS. Clinical assessment and marked renal dysfunction favours a HUS over TTP. The clinical presentation, laboratory evidence of TMA and predominant renal involvement subsequently supported the diagnosis of atypical hemolytic uremic syndrome after exclusion of other causes of TMA.

The key management strategy for aHUS is plasmapheresis following which patient's hemoglobin, platelet were increasing gradually, dialysis was performed to reverse AKI and HTN was managed by nifedipine titration which was then switched to amlodipine. Other medications such as paracetamol, emeset, calcium, multivitamins, were administrated as supportive therapy.

CONCLUSION

Sever patients with aHUS progress to ESRD hence early diagnosis and proper differentiation from TTP, typical HUS/ STEC – HUS and secondary TMA is very important for proper management of a HUS. Identification of complentary abnormality helps to accurately manage aHUS. In this case timely initiation of plasma exchange, dialysis along with supportive treatment resulted in gradual improvement. There are other evolving approaches for the management of a HUS which shows promising results such as anti-C5 monoclonal antibody, eculizumab. Thus early recognition, accurate diagnosis and prompt initiation of appropriate therapy are important for preventing disease progression and improving long term patient outcome.

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