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Therapeutic Plasma Exchange in a Child with Anti-Factor H Antibody-Associated Atypical Hemolytic Uremic Syndrome: A Case Report

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Abstract: ***Atypical hemolytic uremic syndrome (aHUS) is a rare thrombotic microangiopathy caused by uncontrolled complement activation, and anti-factor H (anti-FH) autoantibodies are an important cause in children. We report a 10-year-old girl with fever, hypertension, anemia, thrombocytopenia (55,000/µL), raised LDH (1101 U/L), schistocytes (6–7%) and renal failure (creatinine 6 mg/dL), who had a high anti-FH antibody titre (15,000). She received prednisolone, azathioprine, enalapril and hemodialysis; because there was no improvement after 2 days, five therapeutic plasma exchange (TPE) sessions were done on alternate days with plasma replacement. Platelets rose to 2.82 lakh/µL, LDH fell to 191 U/L, schistocytes fell to <2%, the anti-FH titre became negative, and creatinine, unchanged at 6 mg/dL up to the fourth session, fell to 4 mg/dL after the fifth. No adverse events occurred. These changes were temporally associated with TPE given with immunosuppression and dialysis; renal recovery was partial and follow-up was short.***

Keywords: atypical hemolytic uremic syndrome, anti-factor H antibody, therapeutic plasma exchange, thrombotic microangiopathy

1. Introduction

Hemolytic uremic syndrome (HUS) is a thrombotic microangiopathy defined by microangiopathic hemolytic anemia, thrombocytopenia and acute kidney injury. It is classed as typical (Shiga toxin-producing E. coli), atypical (complement dysregulation), or secondary to another condition [1]. Atypical HUS has no single confirmatory test, so diagnosis depends on excluding TTP and Shiga toxin-HUS and on complement studies. In children, autoantibodies against complement factor H cause a distinct form of aHUS [2], and plasma exchange with immunosuppression has been used to treat it [3]. We report a child with anti-FH-associated aHUS treated with TPE and immunosuppression.

2. Case Presentation

2.1 Patient information and findings

A 10-year-old girl (weight 45 kg, height 148 cm) was admitted with fever. She had elevated blood pressure and pallor.

2.2 Diagnostic assessment

Hb was 8.8 g/dL, platelets 55,000/µL, LDH 1101 U/L, and the peripheral smear showed 6–7% schistocytes. Serum creatinine was 6 mg/dL and urea 256 mg/dL. The anti-FH antibody titre was high at 15,000. Stool Shiga toxin testing and genetic studies were not done. A diagnosis of anti-FH-associated aHUS was made.

2.3 Treatment

Prednisolone, azathioprine, enalapril and hemodialysis were started. As there was no improvement after 2 days, TPE was begun. Five sessions were done on alternate days on the Spectra Optia device after priming of the circuit, exchanging 1 total plasma volume each time with plasma as replacement fluid and citrate anticoagulant. Calcium gluconate (1 ampoule IV) was given with every session as citrate prophylaxis. No adverse events (hypotension, citrate reaction, allergic reaction) were recorded. Procedure details are in Table 1.

Table 1: TPE procedure details

Cycle	Removed (mL)	Replaced (mL)	Run time (min)	AC used (mL)	Plasma vol. exchanged
1	2294	2181	131	296	1 TPV
2	2248	2136	165	316	1 TPV
3	2754	2625	183	392	1 TPV
4	2296	2234	181	286	1 TPV
5	2342	2321	185	321	1 TPV

TPV: total plasma volume; AC: anticoagulant.

2.4 Outcome

Hemolysis settled and platelets recovered over the course of TPE (Table 2). LDH fell from 1101 to 191 U/L, schistocytes

fell to <2%, and platelets rose from 55,000 to 2.82 lakh/µL. The anti-FH titre, 15,000 at diagnosis, became negative after the course. Serum creatinine stayed at 6 mg/dL up to the fourth session and fell to 4 mg/dL after the fifth session, so renal improvement lagged behind the hematological response.

Table 2: Laboratory trend during TPE

Parameter	Cycle 1	Cycle 2	Cycle 3	Cycle 4	Cycle 5
Hb (g/dL)	8.8	9.2	9.3	9.5	9.7
Platelets (×10³/µL)	55	67	103	248	282
LDH (U/L)	1101	1000	700	223	191
Schistocytes (%)	6–7	5	3	<2	<2
Calcium (mg/dL)	9	8.5	8.5	8.7	8.5
Reticulocytes (%)	10.4	8	7	8.3	2.1
Corrected retic. (%)	6.4	–	–	5.2	3.9
Potassium (mmol/L)	3.2	3.4	3.6	3.8	4.1
TRBC (million/µL)	3.14	3.2	3.26	3.2	3.2

All values are pre-session. –: not available. Retic. = reticulocyte count.

3. Discussion

Anti-FH autoantibodies impair factor H control of the alternative complement pathway, and this antibody-mediated form is seen mainly in children [2]. Plasma exchange removes the autoantibodies, and with immunosuppression it is used to induce remission and reduce relapse [3], [4]. Complement inhibitors are an alternative where available. In our patient, platelet recovery, falling LDH and clearance of schistocytes followed the start of TPE, and the anti-FH titre became negative. These changes were temporally associated with TPE, but she also received steroids, azathioprine, antihypertensives and dialysis, so the improvement cannot be attributed to TPE alone. This is in line with Indian pediatric experience, where prompt plasma exchange with immunosuppression has been associated with better outcomes in anti-FH-associated HUS [3].

4. Limitations

This is a single case on combination therapy, with partial renal recovery and short follow-up. Shiga toxin testing and genetic studies were not available, so the diagnosis rests on the clinical picture and the high anti-FH titre.

5. Conclusion

In anti-FH-associated aHUS, TPE with immunosuppression was followed by a hematological response and anti-FH titre clearance in this child. Renal outcome needs longer follow-up.

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