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ORIGINAL ARTICLE

Real-World Outcomes of First-Line Rituximab in Immune Thrombotic Thrombocytopenic Purpura

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ABSTRACT

Background: **Immune thrombotic thrombocytopenic purpura (iTTP) is an acute and life-threatening rare disease. While plasma exchange (PEX) and steroids have improved patient survival, mortality and relapse rates remain high.**
Methods: **We performed a retrospective analysis of newly diagnosed iTTP patients admitted to the Second Affiliated Hospital of Anhui Medical University from May 2009 to May 2025. The treatment group received first-line rituximab combined with PEX and steroids, while the control group received PEX and steroids alone. Primary efficacy endpoints included clinical remission rate and 3-month mortality. Secondary efficacy endpoints comprised relapse rate, number of PEX sessions, ADAMTS13 activity recovery rate, cellular immune function, and overall survival (OS). The primary safety outcomes were infusion-related reactions, allergic reactions, and infection rates.**
Results: **The clinical remission rates were 91.7% vs. 40% (p = 0.037) in the treatment and control groups, respectively. The 3-month mortality rates were 8.3% vs. 46.7% (p = 0.043). Relapse rates were 9.1% vs. 62.5% (p = 0.041). The median number of PEX sessions was 10 vs. 18 (p = 0.049). ADAMTS13 activity recovery rates were 75% vs. 26.7% (p = 0.021). During rituximab treatment, B cells showed a progressive decline, reaching 0.8% (0% - 3.12%) after the fourth infusion. By the follow-up cutoff in August 2025, 75% of patients had normalized B-cell counts. No effects were observed on CD4+ T cells, CD8+ T cells, or immunoglobulin (IgG, IgA, IgM) levels. The 5-year OS rates were 70.7% vs. 26.7%. Median survival times were 54.1 months (95% CI: 36.791 - 71.369) vs. 39.9 months (95% CI: 8.11 - 71.69) (p = 0.007). No increased infection rates were recorded in the treatment group during hospitalization or follow-up.**
Conclusions: **First-line rituximab treatment can reduce mortality and relapse rates in iTTP patients, improve survival outcomes, and demonstrates good safety.**
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KEYWORDS

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INTRODUCTION

Immune thrombotic thrombocytopenic purpura (iTTP) is an acute, life-threatening disorder characterized by thrombocytopenia, microangiopathic hemolytic anemia, and signs of organ dysfunction such as typical neurological, cardiac, renal, or abdominal symptoms. In most

cases, no obvious underlying causative factors are identified. In iTTP cases, there is a severe deficiency of von Willebrand factor cleaving protease (ADAMTS13) due to the presence of anti-ADAMTS 13 antibodies, predominantly IgG. The primary treatment remains plasma exchange (PEX) and corticosteroids. However, patients often require longer periods to achieve normal platelet counts, need more plasma therapy, and face relapse rates as high as 30% - 50%, necessitating additional immunosuppressive therapies such as cyclosporine, cyclophosphamide, or vincristine [1]. In recent years, rituximab has been reported for the treatment of iTTP, though its use has been limited to refractory or relapsed cases. To date, the use of rituximab as first-line therapy remains controversial [2,3]. We present outcomes using rituximab in combination with standard treatment (PEX and corticosteroids). The primary objective was to determine the safety and efficacy of first-line rituximab in iTTP. Additionally, we aimed to investigate whether early administration of rituximab at presentation could shorten the time to remission, thereby preventing end-organ damage caused by microvascular thrombosis and reducing early mortality.

MATERIALS AND METHODS

Subjects

A retrospective analysis was conducted on newly diagnosed TTP patients admitted to the Second Affiliated Hospital of Anhui Medical University from May 2009 to May 2025. Inclusion criteria: ≥ 18 years old, iTTP patients, no prior rituximab treatment. Exclusion criteria: pregnant or lactating women, HIV-positive patients, hereditary TTP, hemolytic uremic syndrome, and transplant-associated thrombotic microangiopathy. All participants provided written informed consent in accordance with the Declaration of Helsinki. For unconscious patients or those unable to provide consent, approval was obtained according to current regulatory requirements.

Laboratory tests

Plasma ADAMTS13 activity was measured by collagen-binding assay, expressed as a percentage of normal plasma activity [4]. ADAMTS13 inhibitors were measured using enzyme-linked immunosorbent assay (ELISA) [5]. Cellular immune function was assessed by flow cytometry before and after treatment. Method: 2 - 5 mL of peripheral blood was collected in heparinized tubes and tested within 4 hours. 100 μL of peripheral blood was incubated with specific monoclonal antibodies at 25℃ for 15 minutes, followed by red blood cell lysis. Analysis was performed using an FC-500 flow cytometer with CXP 2.0 software [6].

Treatment protocol

All patients received daily fresh frozen plasma exchange (PEX) starting from admission, 1,500 mL - 2,000 mL per session or 40 mL/kg body weight, once daily until platelets remained ≥ 150 x 10^9/L for 2 consecutive days [7]. Concurrent steroids included methylprednisolone (80 - 120 mg/day IV) or dexamethasone (15 - 20 mg/day IV), transitioning to prednisone (1 - 2 mg/kg/day) during remission with gradual tapering (control group). The treatment group additionally received rituximab 375 mg/m^2 IV weekly for 4 weeks. Patients with persistently low ADAMTS13 activity or detectable anti-ADAMTS13 antibodies received up to 8 rituximab doses [7].

Efficacy evaluation

Clinical response: Platelet count ≥ 150 x 10^9/L for 2 consecutive days, with stabilization of organ dysfunction parameters if initially present. Clinical remission: Sustained response > 30 days after PEX discontinuation. Relapse: Platelet count < 150 x 10^9/L requiring urgent treatment, or ADAMTS13 activity < 20% without thrombocytopenia. Refractory iTTP: Persistent thrombocytopenia and elevated LDH despite adequate PEX and steroids [8].

Follow-up

Final follow-up: August 2025. Primary endpoints: Clinical remission rate, 3-month mortality. Secondary endpoints: Relapse rate, ADAMTS13 activity recovery, overall survival. Safety outcomes: Cellular immune function, infusion-related reactions, infection rate.

Statistical analysis

All continuous variables underwent normality testing. Continuous data presented as median or mean ± SD ($\bar{x} \pm s$). Categorical variables as counts/percentages. Comparisons used Mann-Whitney/*t*-tests (continuous) or χ^2/Fisher's exact tests (categorical). OS analyzed by Kaplan-Meier with log-rank test (SPSS 26.0). $p < 0.05$ considered significant.

RESULTS

Patient characteristics

Among 27 patients, 11 were male and 16 were female. Median age was 59 (range 20 - 86) years. Comorbidities included rheumatic connective tissue disease (9 cases), gastric cancer (1 case), and hepatitis B (1 case); 16 patients had no significant comorbidities. Twenty patients required ICU admission. Seventeen patients underwent endotracheal intubation and mechanical ventilation. 74.1% had varying degrees of consciousness impairment, 44.4% had fever, 77.8% had abnormal high-sensitive cardiac troponin I (hs-cTnI), and 74.1% had renal dysfunction. Patients requiring intubation at admission (n = 17) showed higher cardiac injury biomarkers. Baseline characteristics are detailed in Table 1.

Table 1. Patient characteristics.

Characteristics	Treatment group (n = 12)	Control group (n = 15)	*χ2/t*	p
Gender (female/male)	8/4	8/7	0.688	0.696
Age (Y)	52 (26 - 80)	69 (20 - 86)	0.553	0.585
Leukocyte (x 10^9/L)	8.18 ± 4.26	6.01 ± 3.18	1.520	0.141
Platelet (x 10^9/L)	6.5 (2 - 49)	8.0 (3 - 54)	0.543	0.592
Hemoglobin (g/L)	81.082 ± 3.89	72.73 ± 19.62	0.998	0.328
Lactic dehydrogenase (U/L)	1,116.925 ± 55.46	1,017.33 ± 731.44	0.390	0.700
Creatinine (μmol/L)	119.924 ± 9.12	128.825 ± 4.27	0.442	0.663
Indirect bilirubin (μmol/L)	47.632 ± 6.86	46.822 ± 6.01	0.080	0.937
ADAMTS13(%)	1.45 (0.1 - 5)	2.0 (0.8 - 9)	1.098	0.283
hs-cTnI (pg/mL)	121.6 (3.9 - 7,145)	154 (9.3 - 5,336)	0.253	0.803
Consciousness disorder (n)	10	10	0.964	0.408
Tracheal cannula (n)	8	9	0.350	0.726

Table 2. Efficacy evaluation.

Parameter	Remission rate (%)	Plasma exchanges (n)	Mortality (%)	Hospital stay (d)	ADAMTS13 activity (%)	Recurrence rate (%)
Treatment group	91.7	10 (5 - 15)	8.3	23 (11 - 61)	75	9.1
Control group	40	18 (4 - 38)	46.7	18 (4 - 52)	26.7	62.5
χ2/t	2.418	2.067	2.127	0.875	2.451	2.407
p	0.037	0.049	0.043	0.390	0.021	0.041

Table 3. Immune function of patients in the treatment group.

Variable	CD4 + T(%)	CD8 + T(%)	B cell (%)	IgG (g/L)	IgA (g/L)	IgM (g/L)
Pre-therapy	37.9 (19.3 - 58.5)	20.4 (14.4 - 41.3)	13 (4.5 - 37.8)	13.0 (8.5 - 18)	2.6 (1.0 - 4.9)	1.75 (0.6 - 2.9)
Post-therapy	39.1 (20.3 - 60.2)	21.8 (13.5 - 40.2)	0.8 (0 - 3.12)	12.9 (8.7 - 17.3)	2.5 (1.1 - 4.8)	1.65 (0.5 - 3.0)
t	0.672	0.578	5.046	0.540	0.561	0.923
p	0.516	0.575	< 0.001	0.600	0.586	0.376

Efficacy

In the treatment group, 10 patients received at least 4 doses of rituximab as per protocol. Two patients received 3 doses, while 2 others required 6 doses based on ADAMTS13 monitoring results after the initial 4 doses. No correlation was observed between ICU admission status (particularly intubation requirement at presentation) and the need for additional rituximab doses. Hemoglobin and reticulocyte counts required 29 days to 8 weeks to normalize post-treatment despite completion of both PEX and rituximab therapy. Bilirubin and LDH levels typically normalized before the second rituximab infusion. No cases of acute oliguric/anuric renal failure occurred.

Clinical remission was achieved in 11 treatment-group patients versus 6 controls. The median number of PEX sessions until remission was 18 (range 4 - 38) in controls versus 10 (5 - 15) in the treatment group, demonstrating significantly fewer PEX requirements with rituximab therapy. Mortality at 3 months was 7 cases in controls versus 1 in the treatment group. ADAMTS13 activity recovery (> 20%) occurred in 9 treatment-group patients versus 4 controls. Hospital stays were 23 (11 - 60) days (treatment) versus 18 (4 - 52) days (controls).

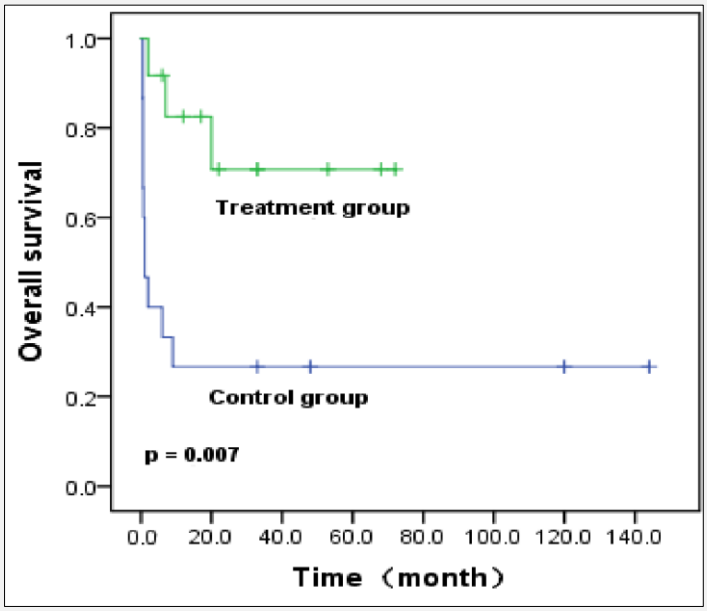

Figure 1. Comparison of overall survival in iTTP patients treated with or without rituximab.

During follow-up, 5 of 8 surviving controls relapsed compared to 1 of 11 treatment-group survivors (Table 2).

Survival analysis

As of the final follow-up in August 2025, the 5-year overall survival (OS) rates were 70.7% in the treatment group versus 26.7% in the control group. The median survival time was 54.1 months (95% CI: 36.791 - 71.369) in the treatment group compared to 39.9 months (95% CI: 8.11 - 71.69) in the control group (χ^2 = 7.363, p = 0.007), as shown in Figure 1.

Immune function

Baseline B-cell levels were 13% (range: 4.5% - 37.8%) at admission (normal range: 5% - 22%). During rituximab treatment, B-cells showed progressive depletion, declining to 0.8% (range: 0% - 3.12%) of CD19+ cells after the fourth infusion, which correlated with reduced ADAMTS13 IgG antibody levels and shorter time to remission. By the final follow-up in August 2025, 75% of patients had restored normal B-cell counts. CD4+ T-cells (normal range: 33% - 58%) and CD8+ T-cells (normal range: 13% - 39%) remained within normal limits throughout treatment, as did serum immunoglobulin levels. All patients maintained B-cell levels below normal range during therapy (Table 3).

Safety

During hospitalization and follow-up, the treatment group experienced 7 infection episodes (2 viral, 3 bacterial, and 2 unspecified), primarily respiratory (n = 5) and urinary tract (n = 2), with 1 pulmonary infection requiring hospitalization. The control group had 10 infection episodes (3 viral, 3 bacterial, 1 fungal, and 3 unspecified), with 2 cases requiring hospitalization. No significant increase in infections was observed in the treatment group (χ^2 = 0.437, p = 0.706). One patient developed mild arthralgia responsive to simple analgesics. We documented 1 case of transient fever related to rituximab infusion and 1 case of chest tightness during infusion that resolved after slowing the infusion rate. These reactions occurred primarily during the first infusion, with no adverse events reported during subsequent infusions.

DISCUSSION

The estimated annual incidence of clinically diagnosed TTP is 4 - 11 cases per million population, typically occurring in patients aged 36 - 51 years with a female predominance. Neurological involvement is present in 79% of cases, ranging from mild headache to severe manifestations such as delirium, confusion, personality changes, coma, quadriplegia, and seizures [9]. Cardiac involvement occurs in 91% of TTP cases [10]. In our study, the median age of onset was 59 (20 - 86) years, higher than

previously reported [3], with 74.1% of patients exhibiting varying degrees of consciousness impairment and 77.8% showing abnormal hs-cTnI levels. Acute myocardial infarction is a common and severe complication of TTP, with serum LDH levels > 1,000 U/L combined with troponin I levels > 0.20 ng/mL serving as reliable predictors of subsequent acute myocardial infarction [10]. iTTP patients are at risk of cardiovascular death, and cardiac troponin testing should be performed. Elevated troponin levels indicating ischemic heart disease warrant attention to sudden death risk [10].

When patients present with thrombocytopenia and unexplained hemolytic anemia, clinicians should measure ADAMTS13 activity. A level < 10% confirms TTP diagnosis, while positive anti-ADAMTS13 autoantibodies establish iTTP [7]. ADAMTS13 activity testing is essential for TTP diagnosis, demonstrating high sensitivity (97%) and specificity (100%) [8]. iTTP may be triggered by various conditions, including abnormal immune responses to viral infections, autoimmune diseases, malignancies, bone marrow transplantation, or chemotherapy drugs [11]. TTP associated with connective tissue disease typically has worse prognosis than idiopathic TTP and often requires combination therapy with glucocorticoids and other immunosuppressants due to poor response to plasma exchange alone. However, anti-CD20 monoclonal antibody therapy shows superior efficacy compared to other immunosuppressants [12]. Our study included 9 cases with rheumatic connective tissue disease, 1 gastric cancer case, 1 hepatitis B case, and 16 cases without identified comorbidities. Due to small sample size, subgroup analysis of connective tissue disease patients was not performed.

iTTP is a rare, life-threatening disorder associated with significant morbidity and mortality if untreated. A critical component of acute-phase treatment is early eradication of anti-ADAMTS13 antibody-producing B lymphocytes [13]. iTTP management should include therapeutic plasma exchange (TPE) to replace ADAMTS13-deficient plasma and corticosteroids to suppress pathogenic antibody production [14]. Additionally, rituximab, a human/mouse chimeric anti-CD20 monoclonal antibody, can be administered upon diagnosis to deplete B lymphocytes and inhibit ADAMTS13 inhibitor production [15].

Early mortality remains a major determinant of iTTP survival. While mortality has improved with increased disease awareness and optimized treatment [16], diagnostic delays appear associated with more severe neurological symptoms and higher mortality [17]. The degree of hemolysis and signs of organ ischemia, such as cardiovascular symptoms and focal neurological deficits, are indicators of mortality risk [17]. Real-world registry studies highlight diagnostic challenges in elderly iTTP patients (≥ 60 years), who exhibit greater renal injury risk, atypical neurological features, and less severe cytopenias, potentially leading to delayed diagnosis. The proportion of patients experiencing ≥ 2-day delays from symptom onset to PEX was 47.6% in those ≥ 60 years versus 31.9% in younger adults [18]. Microvascular thrombosis caused by anti-ADAMTS13 autoantibodies underlies the fatal pathology [19]. Our real-world study assessed 3-month mortality due to prolonged hospitalizations and found iTTP mortality rates substantially higher than literature reports [16], potentially attributable to older age (median 59 years) and delayed diagnosis/treatment. Notably, the rituximab treatment group showed significantly lower early mortality than controls ($p = 0.043$), as rituximab rapidly clears ADAMTS13 autoantibodies and restores activity (75% vs. 26.7% recovery rates), enabling faster clinical remission and preventing end-organ damage from microvascular thrombosis. The treatment group also required significantly fewer PEX sessions, indicating faster disease control and reduced burdens of invasive therapy. A recent meta-analysis demonstrated rituximab's high efficacy in reducing mortality in acquired TTP [20]. Thus, enhancing the awareness of TTP and early initiation of rituximab are critically important for reducing early mortality in patients with iTTP.

Relapse represents another major survival determinant. Reduced ADAMTS13 activity serves as a predictive and sensitive biomarker for impending relapse [21]. We used ADAMTS13 activity < 20% as the relapse threshold, as evidence suggests levels ≥ 20% prevent clinical recurrence [22]. Doyle et al. reported 40% relapse rates in iTTP, with ADAMTS13 activity < 20% being the primary factor; anti-CD20 therapy effectively reduces relapse rates [1]. B-cell counts typically normalize 6 - 12 months post-anti-CD20 therapy, though some patients subsequently show declining ADAMTS13 activity. Unlike other autoimmune diseases, iTTP patients don't relapse during B-cell reconstitution. TPE doesn't induce sustained ADAMTS13 activity recovery, and iTTP patients with initial clinical response to TPE may deteriorate shortly after discontinuation [23]. For remission-phase iTTP patients with low ADAMTS13 activity but no clinical symptoms, rituximab is recommended to reduce relapse risk [24]. Our cohort demonstrated significantly lower relapse rates with rituximab versus controls (9.1% vs. 62.5%, $p = 0.041$), primarily attributable to higher clinical remission ($p = 0.037$) and ADAMTS-13 activity recovery rates ($p = 0.021$). It is particularly noteworthy that the ADAMTS13 activity has recovered, which explains the improvement in the pathological mechanism of iTTP.

While rituximab shows high efficacy in preventing relapse and reducing mortality, its safety profile warrants examination. B-cell levels decreased to 0.8% (0% - 3.12%) after the fourth rituximab infusion, with 75% of patients achieving normal B-cell counts by August 2025. CD4+ T-cells, CD8+ T-cells, and serum immunoglobulin levels remained normal throughout. Despite prolonged B-cell depletion, no significant infection increase occurred, consistent with literature [25]. Compared to conventional immunosuppressants (steroids, cyclosporine, cyclophosphamide), rituximab demonstrates substantially improved safety [26]. Though en-

hanced preventive measures remain necessary for high-risk groups like connective tissue disease patients on long-term immunosuppression or hepatitis B carriers. Transient infusion-related reactions (chest tightness, fever) occurred but were manageable and primarily limited to the first infusion.

In conclusion, despite PEX and corticosteroids remaining foundational iTTP treatments, significant mortality and relapse risks persist. Rituximab halts anti-ADAMTS13 antibody production, enabling sustained remission and reducing post-remission relapse. Rapid ADAMTS13 activity recovery decreases early mortality in patients with end-organ dysfunction, particularly cardiac/neurological involvement. Our real-world study demonstrates substantial survival improvements with frontline rituximab. ADAMTS13 activity measurement proves valuable not only for diagnosis but also for guiding clinical management during response and remission phases.

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Statement of Ethics:
This study was conducted ethically in accordance with the World Medical Association Declaration of Helsinki and approved by the Institutional Review Board Institutional of the Affiliated Second Hospital of Anhui Medical University (PJ-YX2009-015). The patients or their families enrolled in the study signed an informed consent.

Declaration of Generative AI in Scientific Writing:
No AI assistance was used to write this article.

Declaration of Interest:
The authors have no relevant conflicts of interest.

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