Clinical Case Reports International

Case Report
Published: 30 May, 2026

Chronic Kidney Disease as a Progressive Long-Term Sequela of Thrombotic Thrombocytopenic Purpura: A Case Report

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Abstract

The case report describes a 39-year-old male patient who has Chronic Kidney Disease (CKD) stage 3b as a sequela of Thrombotic Thrombocytopenic Purpura (TTP). He was diagnosed in 2014. The patient had positive negative ADAMTS13 activity and biopsy-proven TTP. He did not respond to hypertensive emergency and microangiopathic hemolytic anemia. The acute management was comprised of the continuous veno-venous hemodiafiltration, plasmapheresis, rituximab and methylprednisolone. After 11 years' follow-up, the renal function reached the level of CKD 3b with an estimated glomerular filtration rate of 40 mL/min/1.73m2, whereas hypertension was treated by multidrug therapy. The assessment carried out recently showed no active symptoms and controlled blood pressure. This report highlights the possibility of long-term renal impairment after resolving TTP and suggests the use of long-term multidisciplinary monitoring.

Keywords: Thrombotic Thrombocytopenic Purpura (TTP); Chronic Kidney Disease (CKD); Thrombotic Microangiopathy; ADAMTS13 deficiency; Acute Kidney Injury (AKI)

Key Messages

- Thrombotic Thrombocytopenic Purpura (TTP) can result in long-term renal impairment, even after successful hematologic remissions.
- Chronic Kidney Disease (CKD) can develop gradually after TTP specifically in the patients having severe initial acute kidney injury.
- Long-term nephrology follow-up with daily monitoring of eGFR along with blood pressure is critical in TTP survivors.
- Multidisciplinary management has a notable role in preventing progression to advanced CKD and optimizing patient outcomes.

Introduction

TTP is a dysregulated thrombotic microangiopathy or deficiency of ADAMTS13, which results in microvascular thrombosis, haemolytic anemia, and thrombocytopenia [1]. Plasma exchange and immunosuppressive therapy [2,3] has helped to improve survival, but acute kidney injury is frequent [2]. Although renal involvement is normally mild, in some survivors, kidney disease develops into a chronic kidney disease [1-5]. The long-term renal outcomes are not well reported especially after 10 years [6]. This case is of CKD stage 3b that occurred 11 years after the TTP remission.

Case Presentation

The patient is a 39-year-old male who recently underwent nephrology follow-up. Also, he had a history of TTP diagnosed in October 2014 as a result of the hypertensive emergency complicated with microangiopathic hemolytic anemia. ADAMTS13 activity was critically deficient (negative) at the first presentation and renal biopsy revealed thrombotic microangiopathy which is associated with TTP-related renal involvement as reported in Figure 1. The acute treatment was made up of constant-veno-venous hemodiafiltration to support the renal system, four rounds of plasmapheresis, rituximab and Methylprednisolone, which resulted into hematologic remission. After recovery, the patient acquired chronic hypertension that needed multidrug treatment and gradual changes of

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Received Date: *05 May 2026*
Accepted Date: *20 May 2026*
Published Date: *30 May 2026*

Citation:
Shehata A, Shaikh J, Jasmin AS, Khan MM, Khanum IT, Al Nour HH. Chronic Kidney Disease as a Progressive Long-Term Sequela of Thrombotic Thrombocytopenic Purpura: A Case Report. Clin Case Rep Int. 2026; 10: 1771.

Table 1: Key Laboratory Parameters at Different Time Intervals.

Parameter	Unit	Reference Range	2014 (Initial Presentation)	2022–2025 (Stable Follow-Up)
Serum Creatinine	mg/dL	0.74–1.35	Elevated (requiring CVVHD)	2.0–2.2
Estimated Glomerular Filtration Rate (eGFR)	mL/min/1.73 m²	>90	Decreased (acute kidney injury)	40
Hemoglobin	g/dL	13.8–17.2	Decreased (due to hemolysis)	Elevated
Platelet Count	×10³/μL	150–450	Decreased (thrombocytopenia)	218 (normal)

Legend: Laboratory data indicates the renal and hematological condition at the first diagnosis and during the long-term follow-up. The reference values are on standard adult male values. CVVHD is the abbreviation of continuous veno-venous hemodiafiltration. The stage of chronic kidney disease was identified based on the KDIGO 2012 Clinical Practice Guidelines [17].

Table 2: Key Laboratory Parameters in TTP-Related CKD.

Parameter	Unit	Reference Range	2014 (Initial Presentation)	2022–2025 (Stable Follow-Up)	Status in 2014
Serum Creatinine	mg/dL	0.7-1.35	Elevated (requiring CVVHD)	2.0–2.2	Elevated (AKI)
Estimated Glomerular Filtration Rate (eGFR)	mL/min/1.73 m²	>90	Decreased (acute kidney injury)	40	Decreased (AKI)
Hemoglobin	g/dL	13.8-17.2	10.4 (decreased due to hemolysis)	15.1 (elevated)	Decreased (anemia)
Platelet Count	×10³/μL	150-450	80 (decreased thrombocytopenia)	222 (normal)	Decreased (thrombocytopenia)
WBC Count	×10³/μL	3.6-11.0	8.7	5.7	Normal
RBC Count	×10³/μL	4.50-6.00	3.58	5.2	Decreased
Hematocrit	%	40.0-52.0	29.6	46	Decreased
MCV	fL	77.0-92.0	82.9	82.9	Normal
MCH	pg	26.0-34.0	29.2	29.2	Normal
MCHC	g/dL	32.0-36.0	35.2	35.2	Normal
RDW	%	11.5-14.0	14.5	13.9	Elevated
Neutrophil %	%	54-62	78	58.2	Elevated (left shift)
Lymphocyte %	%	20-40	12	32.3	Decreased
Monocyte %	%	4-10	10	9	Normal
Eosinophil %	%	1–6	0	0.5	Decreased
Basophil %	%	0–2	0	0	Normal
Lactate Dehydrogenase (LDH)	U/L	135–225 (typical)	Elevated (indicating tissue damage and hemolysis)	Assumed normal post-remission	Elevated
Bilirubin (Total)	mg/dL	0.1-1.0	Elevated (indicating red blood cell destruction)	Assumed normal post-remission	Elevated

Legend: The laboratory values indicate both the initial diagnosis (2014, acute TTP episode with MAHA and AKI) and the final diagnosis (2022-2025, CKD stage 3b stable). Reference ranges are based on standard adult male values. CVVHD is an indicator of continuous veno-venous hemodiafiltration, whereas MAHA is an indicator of microangiopathic hemolytic anemia. KDIGO stage chronic kidney disease guidelines 2012 [17]. Acute phase Hemolysis is a confirmed elevation of bilirubin and LDH.

Chronic Kidney Disease (CKD) to stage 3b during the next years.

More recently, high hemoglobin levels led to the referral to hematology where JAK2 mutation testing was negative, and the patient did not present with the symptoms of polycythemia. There was no follow-up urinary, fatigue, edema, shortness of breath, chest pain, abdominal pain, dysuria, or headaches.

Current medications are telmisartan/amlodipine 80/5 mg/kg/day, hydralazine 50 mg thrice a day, labetalol 400 mg/kg/day, moxonidine 400 mcg/kg/day in the evening, prazosin 2 mg/kg/day three times a day, allopurinol 100 mg/kg/day, pantoprazole 40 mg/kg/day, allopurinol, alfuzosin as required for the urinary symptoms.

The examination found that the blood pressure was 123/94 to 135/91 mmHg (meaning that they have managed to control polypharmacy fairly well), the weight was between 84 and 85 kg, and the body mass index was 29.76 kg/m^2 (overweight/obesity category). The physical exam was normal with a normal cardiac, respiratory, abdominal, and neurological. Systems review negative regarding any acute complaints. Available records did not record family history and lifestyle issues. Ethnicity was not specified. Table 1 shows the lab parameters both at the time of diagnosis and follow up.

Diagnostic assessment

Diagnostic tests included ADAMTS13 activity test (negative in 2014) and renal biopsy to confirm TTP and JAK2 mutation (negative). Serial monitoring revealed stable and dysfunctional renal: elevated serum creatinine and lowered estimated glomerular filtration rate were signs of CKD stage 3b. The levels of hemoglobin were high and this ruled out polycythemia. Peripheral blood smear with polychromasia with many schistocytes (dying erythrocytes) as in Figure 2. Differential diagnoses were considered as hemolytic uremic syndrome, but biopsy was found to be in favor of TTP. No evidence of secondary causes of infectious, malignant, or other causes was found. The results of laboratory parameters are presented in Table 1 and 2.

Therapeutic intervention

In 2014, initial treatment included continuous veno-venous hemodiafiltration of acute kidney injury, plasmapheresis of ADAMTS13 deficiency, rituximab of immunomodulation and

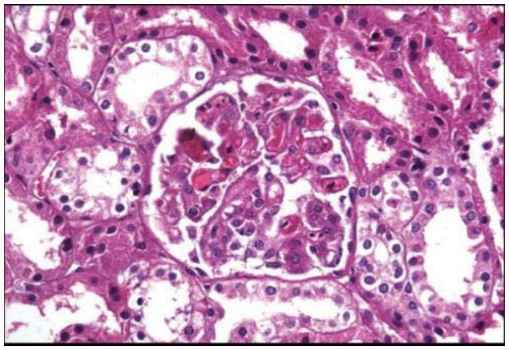

Figure 1: Renal biopsy with the classic features of thrombotic microangiopathy, including endothelial injury, capillary wall thickening, and hyperplasia of the intima of the arterioles (light microscopy, hematoxylin and eosin stain, × 400 magnification). These findings endorse the fact that TTP involves the kidneys.

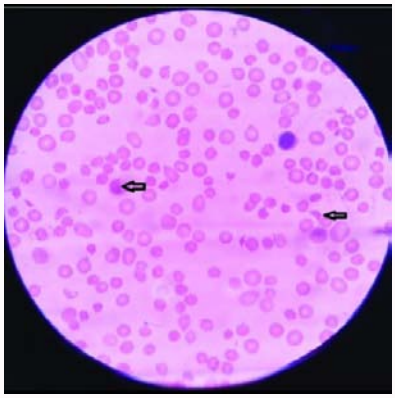

Figure 2: Peripheral blood smear (Wright-Giemsa stain, × 1000 magnification) of polychromasia and numerous patients of schistocytes (ruptured erythrocytes), which are characteristic of microangiopathic hemolytic anemia, a feature of TTP.

methylprednisolone of anti-inflammatory support resulting in resolution of the acute episode but renal residual damage. Hypertension control was addressed over a long period of time using several agents, which produced consistent blood pressure without any adverse effects reported. The supportive treatment involved allopurinol to treat uric acid and iron replacement. The patient was also recommended to avoid nonsteroid anti-inflammatory drugs, instead to use paracetamol as a pain reliever, and take antibiotic prophylaxis in case dental extraction was necessary.

Outcomes, prognosis, and follow up

The CKD progression of the patient was slow with an improvement to stage 3b but was maintained, with no recurrences of TTP or severe deteriorations within 11 years of follow-up (20142025). There was an improvement in blood pressure control and negative polycythemia examination. Recent examinations have proved the absence of contraindication to common procedures like dental hygiene. The condition prognoses well with further compliance with treatment, however, constant observance of CKD progression is required. The ultimate result is a predictive of a stable clinical state in the multidisciplinary care.

Discussion

The case indicates stage 3b CKD as chronic sequela of TTP and it is stable with respect to renal performance despite having severe Acute Kidney Injury (AKI) at admission. The significantly decreased activity of ADAMTS13 is in favor of the diagnosis of acquired TTP, where renal recovery can be partial despite hematologic remission. This observation agrees with the reports that a section of TTP survivors develop chronic renal failure [1,7].

Literature is pointing out that as many as 50 percent of TTP cases result in acute renal failure; but development into CKD is less prevalent in acquired TTP than in hereditary cases, recurrent episodes of TTP lead to accumulated renal damage [10,14]. Hereditary TTP is known to be linked with repeated microvascular trauma related to development of progressive renal pathology and adverse prognosis [6,10]. The lack of relapse during a period of over a decade, in the current case, can be seen as the possible reason why the renal trend in the case is comparatively stable, and the significance of the early and successful acute-phase treatment must be pointed out.

It has been identified that there are a number of risks of the long-term renal impairment, which includes the male sex, obesity, and hypertension, and all of them were present in this patient [8,12]. Stable hypertension in specific, can intensify the damage of the glomeruli and speed up the CKD development [9]. Therapeutic plasma exchange studies have shown increased survival and inconsistent long-term renal improvement, which indicates that survival gains may not always result in full renal recovery [11].

Pathophysiologically, due to the lack of ADAMTS13, ultra-large von Willebrand factor multimers accumulate, which favors platelet aggregation and microvascular thrombosis [13,14]. Subclinical endothelial damage and microthrombi can also remain even following clinical remission contributing to chronic kidney damage. Recent advances in treatment by introducing newer therapies like caplacizumab have further enhanced the acute outcomes although their long-term effects on renal prognosis are still being studied [15].

The existing management procedures are focused on early plasma replacement, immunosuppressive therapy, and close observation at the acute stage [16]. Nevertheless, the case shows the necessity of organized long-term follow-up, such as checking the estimated glomerular filtration rate, proteinuria, and blood pressure, in accordance with the guidelines of managing chronic kidney disease, such as KDIGO [17].

This case presents the significance of multidisciplinary care (nephrology and hematology) in order to maximize long-term outcomes. Future studies ought to aim at discovering early renal injury biomarkers and renal injury predictors among TTP survivors in order to implement interventions on high-risk groups.

Conclusion

As demonstrated in this case, Thrombotic Thrombocytopenic Purpura (TTP) despite receiving the proper treatment during the acute phase can be accompanied by chronic renal failure and development into chronic kidney disease. Even though renal activity in TTP is usually thought to be less significant than in other thrombotic microangiopathies, chronic follow up renal studies have found that there are still long-term renal impairments and progressions to CKD or end-stage renal disease in some patients. Even with the hematologic remission, renal dysfunction can remain, which suggests that the recovery of the renal system does not necessarily occur with the fixation of acute hematologic disorders.

Long term registry data indicates that residual renal impairment following TTP is more common than it has been previously considered, especially where systematic renal follow up is not applied. A number of causes have been suggested to play a role in chronic renal injury and they include severity of initial renal involvement, late diagnosis, repeated microvascular injury, and insufficient recovery of ADAMTS13 activity. Since better short-term survival has been achieved due to the growth in therapeutic plasma exchange and the use of caplacizumab, the long-term effects of these like CKD have become a growing concern. This case demonstrates that nephrology follow-up is essential after restoring normal hemoglobin levels in the event of TTP with renal functioning, blood pressure, and proteinuria. Strong cooperation between nephrology and hematology teams is such that it helps to identify and treat late renal complications in such patients. Additional prospective studies are required that will further specify predictors of long-term renal outcome in order to implement follow-up approaches in TTP patients.

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