

Catastrophic antiphospholipid syndrome presenting as renal thrombotic microangiopathy in an elderly man: rituximab-based remission and 28-month aspirin monotherapy maintenance: a case report

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Case Report

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Abstract

Background: Thrombotic microangiopathy (TMA) encompasses a group of disorders characterized by endothelial injury and microvascular thrombosis, with complex pathogenesis triggered by various factors. Catastrophic antiphospholipid syndrome (CAPS) is a rare and life-threatening manifestation of antiphospholipid syndrome (APS), representing a form of TMA driven primarily by anti-phospholipid antibody-mediated thromboinflammation that results in rapid multi-organ failure.

Case presentation: We report the case of a 79-year-old male patient who presented with acute kidney injury, thrombocytopenia, hemolytic anemia, and disturbed consciousness. Renal biopsy revealed thrombotic microangiopathy. Laboratory findings showed elevated inflammatory markers, proteinuria, a positive direct antiglobulin test, and high titers of anticardiolipin antibody (aCL) and anti- β 2-glycoprotein I antibody (anti- β 2GPI). Initial treatment included aspirin, pulsed intravenous immunoglobulin (IVIG), and glucocorticoids. Anticoagulation with low-molecular-weight heparin was later initiated, and rituximab was added due to refractory thrombocytopenia. The patient's organ function improved progressively. During 28 months of follow-up on maintenance aspirin monotherapy, the patient remained stable with normal platelet counts, renal function, and negative antiphospholipid antibodies.

Conclusions: CAPS is characterized by acute onset, rapid progression, and severe clinical manifestations; early diagnosis and prompt initiation of standardized treatment are critically important. Management involves trigger elimination and triple therapy comprising anticoagulation, glucocorticoids, and plasma exchange and/or IVIG. For refractory cases, rituximab should be considered. This case also highlights that individualized maintenance therapy—such as aspirin monotherapy—may be a viable option for elderly CAPS patients in whom standard anticoagulation carries substantial bleeding risk.

Background

Antiphospholipid syndrome (APS) is an autoimmune disease driven by circulating antiphospholipid antibodies (aPL) [1]. It presents as primary, secondary, or catastrophic forms [2]. Among these, catastrophic antiphospholipid syndrome (CAPS) is the rarest and most severe, occurring in approximately 1% of APS patients and carrying a high mortality rate [3, 4]. CAPS is characterized by the rapid onset of widespread microvascular and/or macrovascular thrombosis leading to acute multi-organ dysfunction in the presence of aPL [5]. Despite its low prevalence, CAPS remains difficult to diagnose and treat in a timely manner due to its complex pathogenesis and fulminant clinical course. This article reports the diagnosis and treatment of an elderly patient with CAPS, with the aim of improving clinicians' understanding of this disease and offering practical insights for managing similar cases.

Case presentation

A 78-year-old male patient was admitted to our nephrology department on June 26, 2022, with complaints of abdominal distension and dyspnea for one month and bilateral lower limb edema for half a month. Prior to admission, he visited the first hospital on June 10, 2022, due to abdominal distension.

Laboratory findings included serum creatinine (SCr) of 138 $\mu\text{mol/L}$, hemoglobin (Hb) of 128 g/L, and platelet count (PLT) of $130 \times 10^9/\text{L}$. Urinalysis showed proteinuria (2+). Non-enhanced thoracoabdominal CT revealed scattered chronic infection foci in both lungs, thickening of the fascia adjacent to the psoas muscle around both kidneys (suggestive of possible infection), and multiple enlarged lymph nodes in the peritoneum and bilateral inguinal regions. The patient received antibiotic therapy.

He was referred to a second hospital on June 15, 2022. Blood tests showed SCr 162 $\mu\text{mol/L}$, Hb 115 g/L, PLT $125 \times 10^9/\text{L}$; 24-hour urine protein quantification was 0.5 g. Bone marrow smear revealed a decreased erythroid-to-myeloid ratio, slight thrombocytopenia, and reduced neutrophil granulation, raising suspicion for myelodysplastic syndrome. The patient underwent a renal biopsy. Light microscopy revealed 19 glomeruli, three of which showed ischemic global sclerosis on PAS staining (Fig.A). The remaining glomeruli exhibited increased numbers of endothelial cells, with compressed and occluded capillary loops. On HE staining (Fig.B), the tubular epithelial cells showed granular degeneration, with some tubules displaying luminal dilatation and loss of brush borders. Small focal tubular atrophy (affecting approximately 5% of the tubular area) was observed. The renal interstitium showed edema, focal inflammatory cell infiltration, and mild focal fibrosis. On PASM staining (Fig.C), pale-staining pseudo-microthrombi were observed within the capillary loop lumina. On Masson staining (Fig.D), capillary loops were dilated, with bright red, amorphous fibrin-platelet microthrombi visible within the capillary lumina; the subendothelial spaces were widened and showed pale red staining. No fuchsinophilic deposits were observed in the mesangial area, along the epithelial side of the basement membrane, or within the basement membrane itself. Immunofluorescence (Fig.E) revealed no definitive immune complex deposition. Electron microscopy (Fig.F) showed marked vacuolar degeneration of glomerular capillary endothelial cell hyperplasia was present, with compressed capillary loops. Parietal epithelial cells showed vacuolar degeneration. The capillary walls displayed markedly widened subendothelial spaces containing flocculent material. Visceral epithelial cells (podocytes) were swollen with vacuolar degeneration and partial foot process fusion. In the mesangial area, there was proliferation of mesangial cells and matrix, with decreased density of the mesangial matrix. Tubular epithelial cells showed vacuolar degeneration, with a few detached tubular epithelial cells. Mild inflammatory cell infiltration was observed in the renal interstitium. In the interstitial vessels, occasional capillary lumina showed aggregation of red blood cells. These findings indicated renal injury due to thrombotic microangiopathy (TMA). Non-enhanced chest CT revealed worsening bilateral pneumonia and pleural effusions; upright abdominal X-rays indicated partial intestinal obstruction. The doctor prescribed enemas and antibiotic therapy.

The patient was referred to our hospital on June 26, 2022. He denied experiencing diarrhea, generalized skin rash, photosensitivity, arthralgia, oral ulcers, or acral Raynaud's phenomenon during the illness. His past medical history, personal details, and family history were all unremarkable.

Upon admission, laboratory tests (Table 1) and a non-contrast CT scan of the chest and abdomen were performed, revealing: (1) blurred fat planes in the abdomen and pelvis with scattered moderate fluid

accumulation and subcutaneous edema; (2) minor perirenal exudates, slight fluid accumulation and dilation of the right renal pelvis and calyces, and possible right ureteral inflammation; (3) significant bilateral pleural effusion with incomplete expansion of the lower lung lobes. The patient was treated with fasting, intravenous cefoperazone-sulbactam, nutritional support, thoracentesis and paracentesis for drainage, and albumin supplementation.

Table 1
Laboratory values of the patient at baseline

Variable	Value	Normal values
White cell count (10 ⁹ /L)	9.08	4–10
Hemoglobin (g/L)	97	120–160
Platelet count (10 ⁹ /L)	112	100–300
Creatinine (μmol/L)	273	44–133
Urea nitrogen (mmol/L)	24.0	2.8–7.1
Total bilirubin (μmol/L))	10.4	5.0–21.0
Prothrombin time (second) (s)	12.9	9.2–13.9
Activated partial thromboplastin time (second))	29.0	21.2–34.8
Fibrinogen (second)	3.4	2–4
International normalized ratio	1.12	0.8–1.2
D-Dimer (μg/ml)	4.25	0-0.55
Lactate dehydrogenase (U/L)	176	109–250
C3 (g/L)	1.18	0.79–1.52
C4 (g/L)	0.22	0.2–0.4
Antinuclear antibodies (ANA)	Negative	Negative
Anti-DNA antibodies (IU/ml)	Negative	< 10
ANA panel spectrum	Negative	Negative
Lupus anticoagulant	Negative	Negative
Cytoplasmic anti-neutrophil cytoplasmic antibody	Negative	Negative
Perinuclear anti-neutrophil cytoplasmic antibody	Negative	Negative
Anti-proteinase 3 antibody	Negative	Negative
Anti-myeloperoxidase antibody	Negative	Negative
Anti-phospholipase A2 receptor antibody (RU/ml)	Negative	≤ 2
Anti-glomerular basement membrane antibody	Negative	Negative
Serum immunofixation electrophoresis	IgG-κ band equivocal	Negative
Stool occult blood test	Positive	Negative
Urine red blood cells (per μL)	430	0–12

Variable	Value	Normal values
Urine protein-to creatinine ratio (mg/gcr)	1034	0-100

On June 30, follow-up laboratory tests showed Hb 93 g/L, PLT 70×10^9 /L, prothrombin time 66 seconds, activated partial thromboplastin time 43.9 seconds, international normalized ratio 5.64, fibrinogen 4.0 g/L, D-dimer 2.86 μ g/mL, and peripheral blood schistocytes 0.3%. Results from July 1 further demonstrated anticardiolipin antibody immunoglobulin G (aCL-IgG) 152 CU (normal < 20) and anti- β 2-glycoprotein I antibody immunoglobulin G (anti- β 2GPI-IgG) 1821 CU (normal < 20); ADAMTS13 activity was normal. Treatment was adjusted by discontinuing cefoperazone-sulbactam and switching to meropenem, along with vitamin K1 supplementation and fresh frozen plasma transfusion. However, the patient developed drowsiness on July 1. Cranial CT showed no obvious hyperdense lesions but suggested bilateral basal ganglia lacunar infarcts and bilateral frontotemporal subdural effusions.

Given the presence of neuropsychiatric symptoms, thrombocytopenia, acute kidney injury, positive aPL, and renal biopsy findings indicating TMA, CAPS could not be ruled out. Oral aspirin and hydroxychloroquine were started on July 2. The patient's condition changed on July 3, when 200 mL of bloody fluid was drained from the ascites. Aspirin was discontinued, and IVIG 20 g once daily was administered for three days. Methylprednisolone 40 mg was given intravenously daily starting from July 4. Laboratory tests on July 8 showed PLT 58×10^9 /L, Hb 63 g/L, normal coagulation function, SCr 180 μ mol/L, and a positive direct antiglobulin test. The peritoneal drainage fluid remained bloody. Starting from July 9, pulse therapy was administered with methylprednisolone 250 mg daily for 3 days and IVIG 20 g daily for 3 days, followed by oral prednisone 40 mg daily. On July 10, low-molecular-weight heparin 4000 U was administered subcutaneously every 12 hours. Nevertheless, PLT continued to decrease to 38×10^9 /L on July 12. Rituximab 500 mg was administered on July 13, and the patient was transferred to a local hospital on July 14 for continued anticoagulation therapy.

On July 22, the patient received another dose of rituximab 500 mg at the local hospital and had the thoracic and abdominal drainage tubes removed. Laboratory tests on August 9 showed PLT 67×10^9 /L, Hb 95 g/L, and SCr 76 μ mol/L. The patient was readmitted to our hospital on August 12, where anticoagulation therapy was continued. Repeat laboratory tests showed PLT 59×10^9 /L, Hb 114 g/L, SCr 76 μ mol/L, normal coagulation function, proteinuria 3+, urine red blood cells 178/ μ L, 24-hour urinary protein 2.63 g, aCL-IgG 20.5 CU, anti- β 2GPI-IgG 206 CU, and a negative direct antiglobulin test. Rituximab 500 mg was administered on August 19 and August 26, respectively. The aPL were rechecked on August 27 and were negative for aCL-IgG, with anti- β 2GPI-IgG 105 CU. The patient was discharged on daily oral aspirin 100 mg.

Follow-up monitoring on November 26, 2022, showed normal laboratory results (PLT 155×10^9 /L, Hb 145 g/L, white blood cell 6.17×10^9 /L, neutrophil 4.26×10^9 /L, SCr 80 μ mol/L, negative aPL and negative serum immunofixation electrophoresis). During the subsequent 28-month follow-up, the patient remained stable on daily aspirin 100 mg for maintenance therapy. His platelet counts and renal function remained normal, and urinary protein and aPL remained consistently negative (Fig. G).

Discussion

Thrombotic microangiopathy encompasses a group of diseases characterized by endothelial cell injury and microvascular thrombus formation, with complex pathogenesis involving multiple mechanisms. These include thrombotic thrombocytopenic purpura, Shiga toxin-mediated hemolytic uremic syndrome, complement-related thrombotic microangiopathy, or secondary conditions such as malignant hypertension, pregnancy, connective tissue diseases, malignancies, or drugs [6–9]. The patient was an elderly male with a subacute course and rapid multi-organ involvement, presenting with: (1) a systemic inflammatory state with elevated inflammatory markers and imaging findings suggesting pulmonary infection and perirenal exudation; (2) central nervous system involvement manifested by consciousness disturbance; (3) renal involvement evidenced by elevated serum creatinine and proteinuria; (4) hematologic involvement presenting as anemia and thrombocytopenia; and (5) vascular involvement demonstrated by microthrombus formation on renal biopsy. Laboratory tests were positive for aPL. The patient had no prior medication use, no diarrhea, normal ADAMTS13 activity, and no imaging evidence of tumors. Based on the revised Sapporo (Sydney) 2006 APS classification criteria, this case was preliminarily diagnosed as APS. The presence of microvascular thrombosis and multi-organ involvement within a short period strongly suggested CAPS. Unfortunately, complement-related laboratory tests were not performed, leaving it unclear whether complement factors contributed to the pathogenesis.

CAPS is a rare manifestation of APS, with an incidence of approximately 1%. It represents one form of TMA, driven by an aPL antibody-mediated thromboinflammatory cascade as its core mechanism, leading to widespread vascular lesions and multi-organ dysfunction. Clinical features include: renal involvement often presenting as acute kidney injury, hematuria, proteinuria, and hypertension; neurological involvement manifesting as stroke, epilepsy, or cognitive impairment; respiratory involvement showing pulmonary embolism or alveolar hemorrhage; cardiac involvement frequently presenting as myocardial infarction or heart failure; cutaneous manifestations such as livedo reticularis, skin ulcers, or purpura; peripheral vascular involvement potentially involving venous or arterial thrombosis; and hematologic involvement typically presenting as thrombocytopenia or microangiopathic hemolytic anemia [10–14]. CAPS has an acute onset, rapid progression, and high mortality rate, making early diagnosis critical but often delayed due to the complexity of its clinical presentations.

The currently widely adopted diagnostic criteria for CAPS are: (1) involvement of three or more organs, systems, or tissues; (2) simultaneous or sequential development of diverse clinical manifestations within one week; (3) histopathological confirmation of small vessel occlusion in at least one organ or tissue; and (4) laboratory evidence of persistently positive aPL (two consecutive positive tests at intervals exceeding 12 weeks). Meeting all four criteria confirms a diagnosis of CAPS. Alternative criteria for highly suspected CAPS include: (1) fulfillment of the above four criteria but affecting only two organs or systems or tissues; (2) all four of the above criteria present except for the absence of laboratory-confirmed aPL; (3) compliance with criteria 1, 2, and 4; and (4) compliance with criteria 1, 3, and 4, with a third thrombotic event occurring within 1 week to 1 month after onset despite anticoagulant therapy [15].

CAPS has a poor prognosis, with a case fatality rate of 33% reported in international cohort studies [10, 16]. Currently, prospective clinical trials are lacking, and treatment recommendations remain empirical. The preferred regimen is triple therapy consisting of anticoagulation, corticosteroids, and plasma exchange and/or IVIG. For patients with concomitant autoimmune diseases such as systemic lupus erythematosus, quadruple therapy (triple therapy combined with cyclophosphamide) may be considered; refractory cases may be treated with rituximab or eculizumab [17, 18]. In this case, the patient received a regimen of anticoagulation, corticosteroids, IVIG, and rituximab, which proved successful.

It is noteworthy that anticoagulation therapy is a pivotal step in CAPS management due to its critical impact on patient prognosis. On day 4 of hospitalization, the patient exhibited significantly reduced platelet levels, presenting a therapeutic dilemma between platelet transfusion and antiplatelet therapy—two conflicting approaches. Given the presence of bloody drainage fluid and the higher risks associated with low platelet levels, initial anticoagulation was withheld, and only aspirin was administered for antiplatelet therapy. Subsequently, considering that the platelet depletion was likely due to disease-related consumptive reduction, low-molecular-weight heparin anticoagulation was initiated. Despite a 3-day pulse therapy with 250 mg of methylprednisolone daily followed by sequential 40 mg of prednisone daily and two pulse courses of 20 g of IVIG daily over 3 days, platelet levels continued to decline. Rituximab therapy was promptly initiated, ultimately achieving recovery of organ function and clinical complete remission. This further underscores the importance of early identification and standardized treatment in improving the prognosis of CAPS patients.

Rituximab, an anti-CD20 monoclonal antibody, exerts precise targeting mechanisms in CAPS treatment: it specifically eliminates mature B lymphocytes, thereby blocking the production of pathogenic aPL at their source and interrupting a key pathway of autoimmune damage; at the same time, it lowers inflammatory cytokine levels, mitigating subsequent injury reactions mediated by these cytokines [19, 20]. A study from the CAPS registry analyzed 20 patients treated with rituximab, including 8 who received it as first-line combination therapy and 12 with refractory CAPS who received it as second-line combination therapy; 15 patients (75%) achieved successful recovery post-treatment [21]. The Bucciarelli cohort reported a recovery rate of 65% for CAPS patients treated with rituximab [16]. Furthermore, several case reports have documented successful use of rituximab for initial or refractory CAPS treatment [22–26]. Owing to the rarity of this disease and the lack of cohort studies on rituximab therapy, no consensus has been reached regarding the optimal dosing regimen. The most commonly used regimens are four weekly doses of 375 mg/m² or two infusions of 500–1000 mg at 7- or 14-day intervals. In this case, the patient's platelet counts and renal function returned to normal after 5 months of rituximab treatment, and aPL tested negative after 7 months.

In addition to acute-phase treatment, long-term patient management is equally critical. According to EULAR recommendations for the management of antiphospholipid syndrome in adults, most APS patients are advised to receive long-term standardized anticoagulant maintenance therapy, with warfarin as the first-line option to maintain an international normalized ratio range of 2.0–3.0 [27]. This recommendation theoretically applies equally to CAPS patients, the most severe subtype of APS. In this

case, after achieving complete remission during the acute phase through aggressive treatment, the patient did not receive anticoagulation but instead opted for aspirin as antiplatelet therapy, which maintained stability for two years without adverse events. Most evidence-based clinical guidelines are developed based on large-scale population studies and often inadequately address special populations such as elderly patients, those with comorbidities, or those in frail conditions. Standardized anticoagulation regimens in elderly CAPS patients frequently carry risks that outweigh potential benefits. While conventional anticoagulation effectively prevents thrombotic recurrence, long-term anticoagulation in elderly patients may lead to bleeding-related complications, which significantly contribute to poor long-term outcomes. No definitive guidelines exist for long-term maintenance therapy in CAPS to date; however, the successful two-year maintenance experience with aspirin monotherapy in this 78-year-old patient provides valuable clinical reference for individualized long-term management of elderly CAPS patients.

Conclusions

CAPS is characterized by acute onset, high heterogeneity, rapid progression, severe clinical manifestations, significant challenges in early diagnosis, and poor prognosis. For patients presenting with acute kidney injury and pathological features of TMA accompanied by thrombocytopenia, CAPS should be highly suspected, and comprehensive differential diagnostic evaluations should be promptly performed. Upon diagnosis or strong suspicion of CAPS, immediate initiation of standardized triple or quadruple therapy, along with other symptomatic treatments during the early stages, is essential to significantly improve patient survival rates.

Strengths and limitations: This case report provides detailed documentation of the diagnostic workup, treatment course, and long-term (28-month) follow-up of an elderly CAPS patient, offering valuable insights into individualized management. The main limitation is the lack of complement-related laboratory tests, which precludes assessment of whether complement factors contributed to the pathogenesis. Additionally, as a single case report, the findings may not be generalizable to all CAPS patients.

Abbreviations

TMA: Thrombotic microangiopathy

CAPS: Catastrophic antiphospholipid syndrome

APS: Antiphospholipid syndrome

aPL: Antiphospholipid antibodies

aCL: Anticardiolipin antibody

anti-β2GPI: Anti-β2-glycoprotein I antibody

IVIg: Intravenous immunoglobulin

SCr: Serum creatinine

Hb: Hemoglobin

PLT: Platelet count

MDS: Myelodysplastic syndrome

Declarations

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None.

Authors' contributions

Xiong LX and Xu XC contributed equally to this work. Xu XC was responsible for the initial patient data analysis. Xiong LX wrote the manuscript. Zhou JF provided critical feedback. Zhang YM designed the work. All authors read and approved the final manuscript.

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Availability of data and materials

The datasets used in the study are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate

The study was approved by the ethical committee of Sixth Affiliated Hospital, Sun Yat-sen University in Guangzhou, China.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Competing interests

The authors declare no competing interests.

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Figures

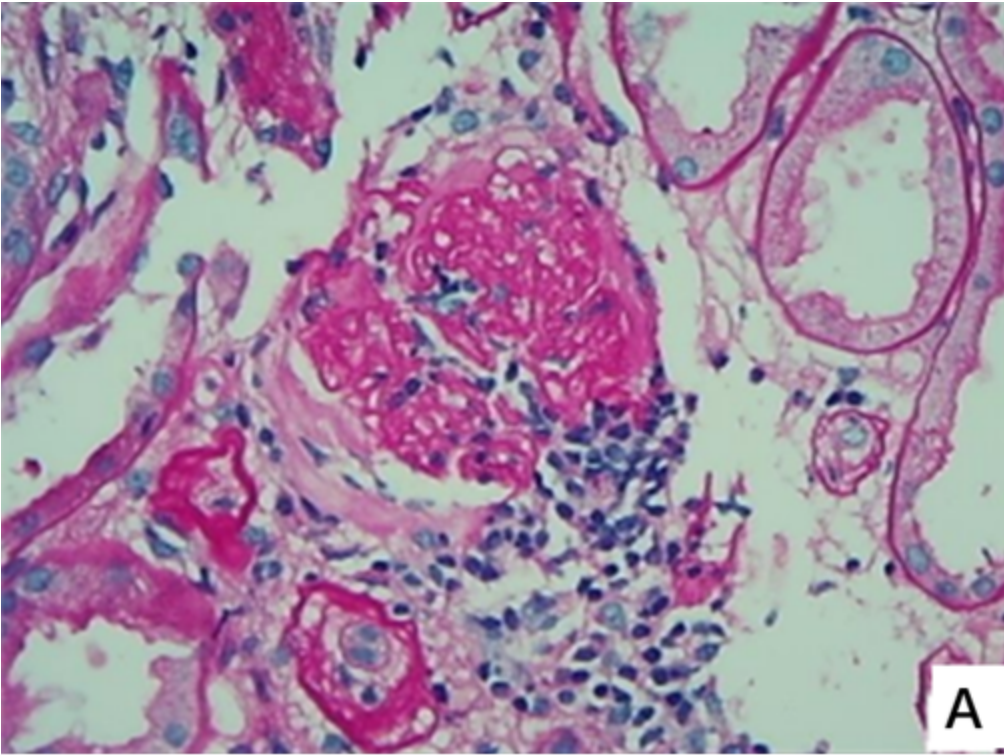


Figure 1

A (PAS staining): Ischemic global glomerulosclerosis.

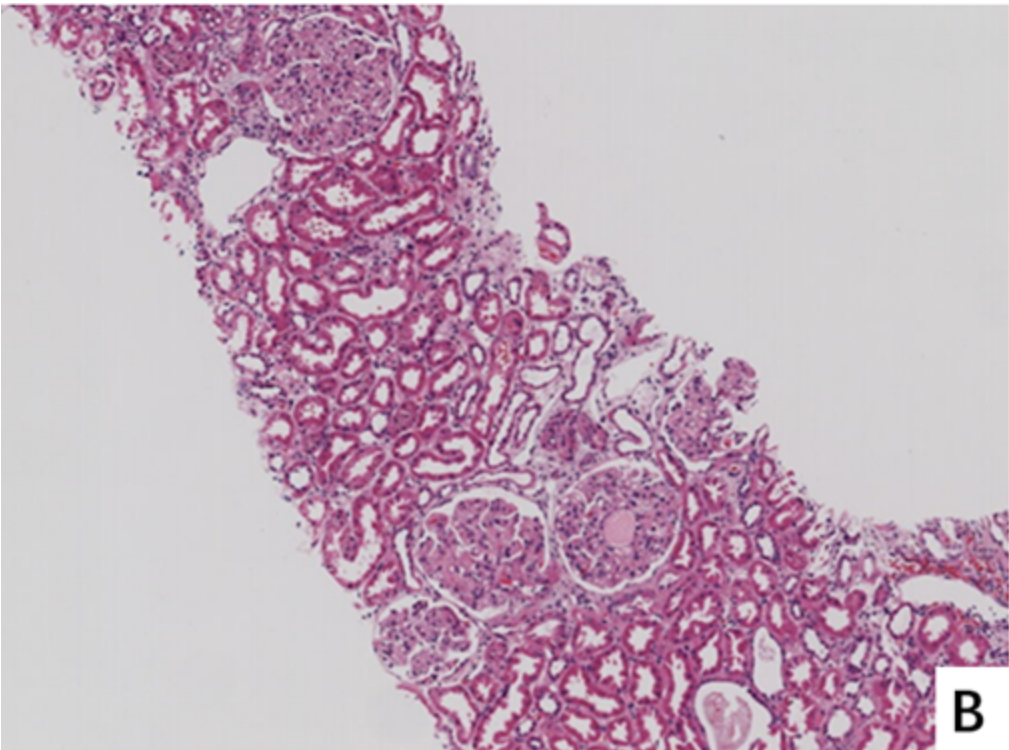


Figure 2

B (HE staining): The glomeruli show increased numbers of endothelial cells, with compressed and occluded capillary loops. The renal interstitium displays edema, focal inflammatory cell infiltration, and mild focal fibrosis.

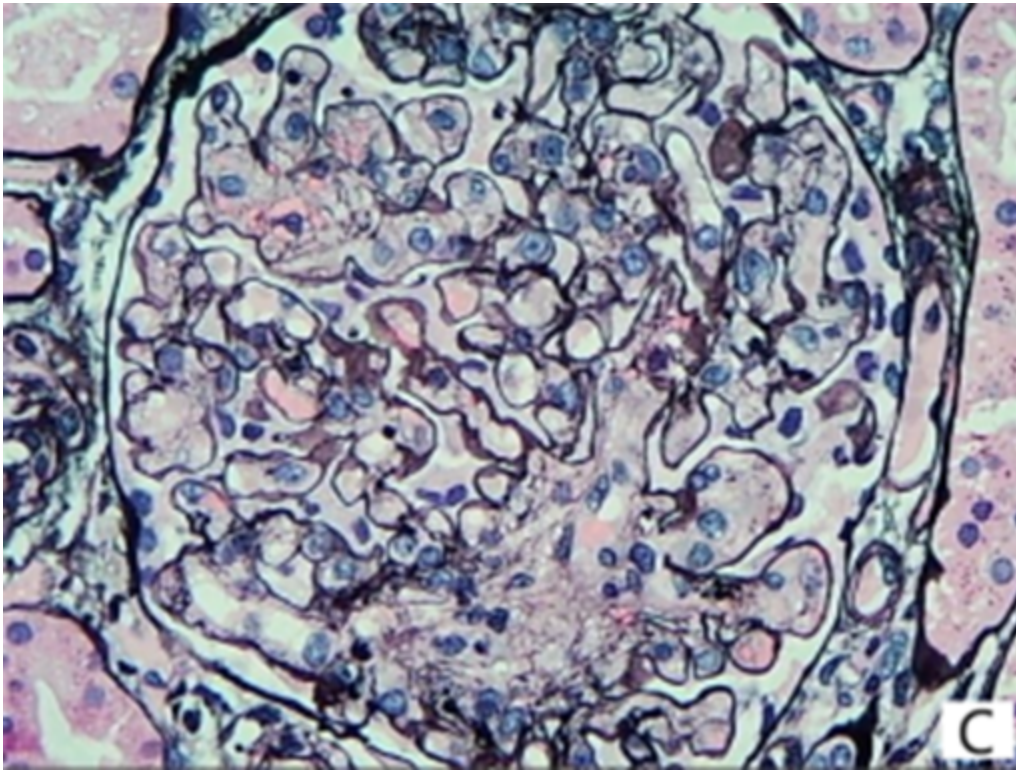


Figure 3

C (PASM staining): Intraluminal pale-staining pseudothrombi in the capillary loops.

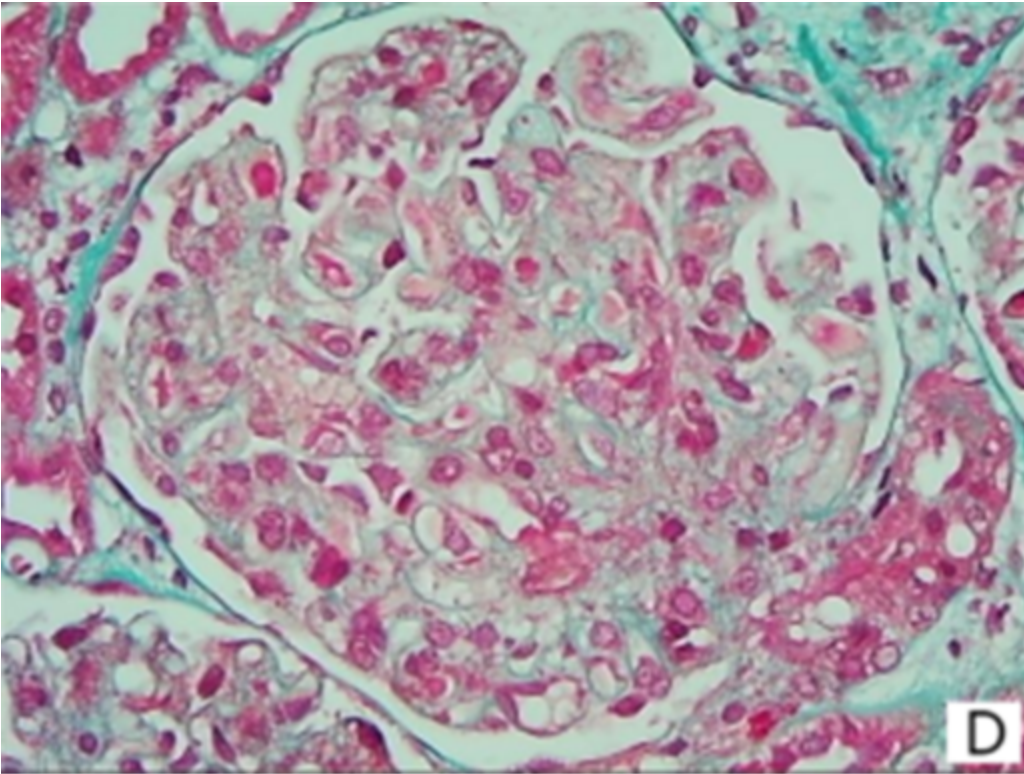


Figure 4

D (Masson staining): Capillary loops are dilated. Amorphous fibrin-platelet microthrombi are visible within the capillary lumina.

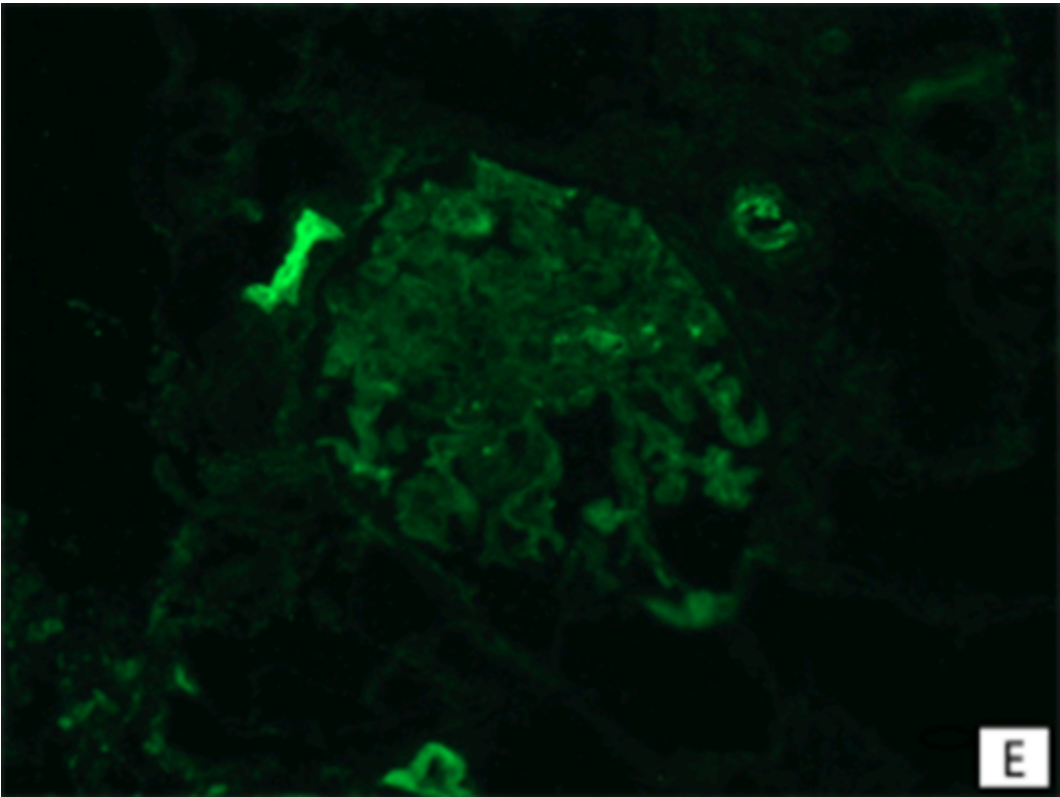


Figure 5

E (Immunofluorescence): No definitive immune complex deposits were identified.

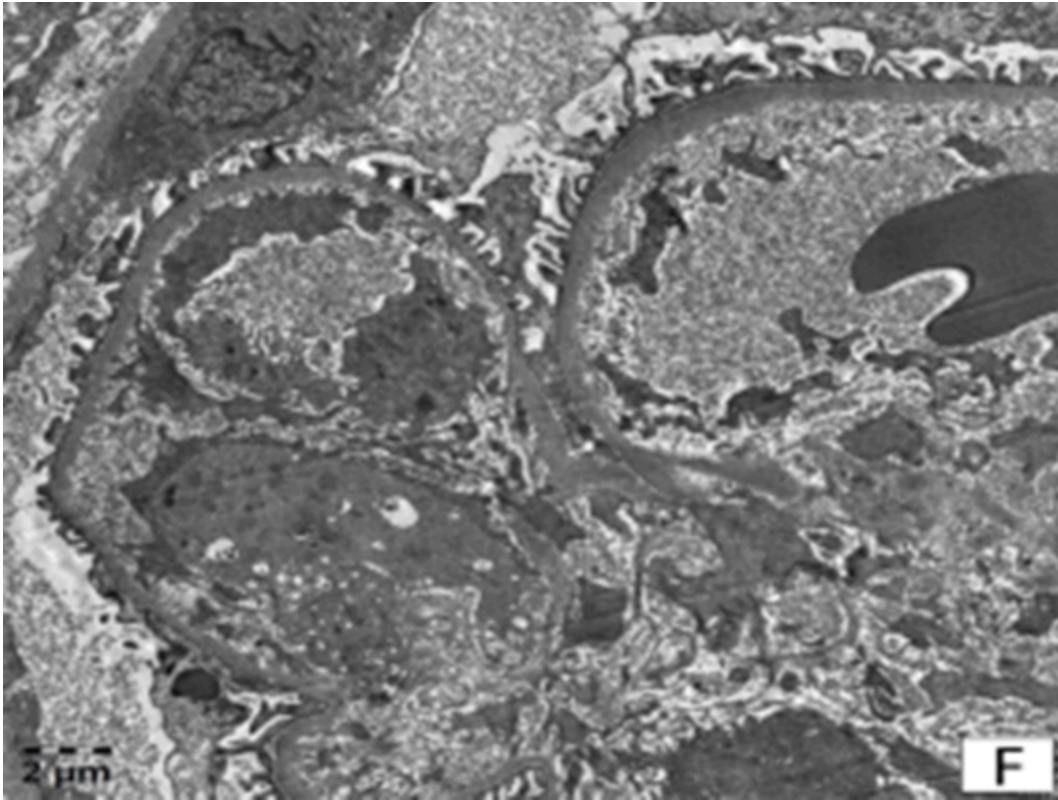


Figure 6

F (Electron microscopy): The glomerular capillary endothelial cells show marked vacuolar degeneration. In the mesangial area, there is proliferation of mesangial cells and matrix, with decreased density of the mesangial matrix.

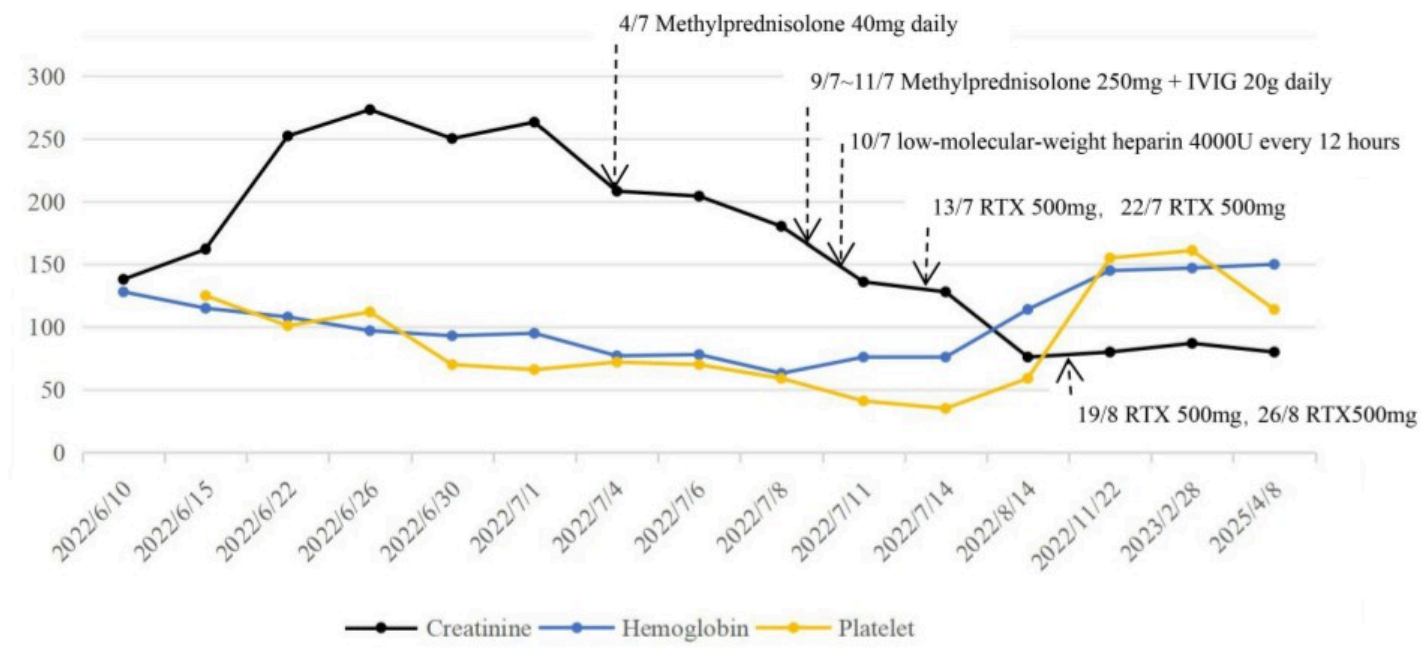


Figure 7

G. Therapeutic regimen and trend changes of key indicators