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Open Access Case Report

A Case of Thrombocytopenia, Anasarca, Fever, Reticulin Fibrosis, and Organomegaly (TAFRO) Syndrome Initially Presenting as Systemic Lupus Erythematosus: Diagnostic Reconsideration Based on Long-Term Clinical Course

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Review began 10/07/2026
Review ended 11/04/2026
Published 11/09/2026

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DOI: 10.7759/cureus.119338

Abstract

We report the case of an 80-year-old man who was initially diagnosed with systemic lupus erythematosus (SLE) based on thrombocytopenia, serositis, hypocomplementemia, and autoantibody positivity and achieved remission following glucocorticoid therapy. At that time, bone marrow examination showed no reticulin fibrosis.

Four and a half years after discharge, he developed recurrent thrombocytopenia, anasarca, organomegaly, and renal dysfunction, all of which were refractory to conventional immunosuppressive therapy. Reevaluation revealed disappearance of autoantibodies, normalization of C3 and C4 levels according to the institutional reference ranges, and newly detected mild bone marrow reticulin fibrosis (MF-1), supporting a clinical diagnosis of thrombocytopenia, anasarca, fever, reticulin fibrosis, and organomegaly (TAFRO) syndrome. Clinical improvement followed the introduction of tocilizumab as part of treatment that also included glucocorticoids, intravenous immunoglobulin, and romiplostim.

This case highlights the diagnostic challenge in distinguishing SLE from TAFRO syndrome in patients with overlapping clinical features. The longitudinal clinical course, including changes in serological findings and comparison of bone marrow pathology at two admissions, illustrates the importance of reassessing the diagnosis. Whether the initial episode represented SLE or TAFRO syndrome with lupus-like features remains uncertain. Careful diagnostic reassessment is essential when the clinical course deviates from the expected trajectory.

Categories: Hematology, Rheumatology, Internal Medicine
Keywords: bone marrow fibrosis, systemic lupus erythematosus, tafro syndrome, thrombocytopenia, tocilizumab

Introduction

Thrombocytopenia, anasarca, fever, reticulin fibrosis, and organomegaly (TAFRO) syndrome is an extremely rare systemic inflammatory syndrome that overlaps with the TAFRO clinical subtype of idiopathic multicentric Castleman disease (iMCD). It was first described by Takai et al. in 2010, based on three Japanese cases that presented with thrombocytopenia, anasarca, fever, mild bone marrow fibrosis, and organomegaly [1]. The Japanese diagnostic criteria permit a clinical diagnosis of TAFRO syndrome without lymph node histology [2,3], whereas iMCD-TAFRO requires compatible lymph node histopathology [4]. The acronym 'TAFRO' reflects its key features: thrombocytopenia, anasarca (generalized edema with pleural effusions and ascites), fever, reticulin fibrosis of the bone marrow, and organomegaly [5].

Particularly in elderly patients, TAFRO syndrome can be difficult to distinguish from other disorders. Its clinical manifestations may mimic those of autoimmune diseases such as systemic lupus erythematosus (SLE) [6-8]. In cases where TAFRO syndrome presents with apparent overlap features of a collagen vascular disease, the initial therapeutic response and the pattern of organ involvement may deviate from expectations, often prompting diagnostic reconsideration.

We report the case of an elderly male who was initially diagnosed with SLE and achieved apparent remission following treatment. He later developed a clinical picture suggestive of SLE flare; however, the disease proved refractory to conventional therapy, prompting a thorough reassessment that led to a diagnosis of TAFRO syndrome. Distinguishing TAFRO syndrome from autoimmune diseases such as SLE remains challenging, particularly in cases with overlapping clinical and serological features, and longitudinal

How to cite this article
Wada T T, Yokota K, Ueno T, et al. (November 09, 2026) A Case of Thrombocytopenia, Anasarca, Fever, Reticulin Fibrosis, and Organomegaly (TAFRO) Syndrome Initially Presenting as Systemic Lupus Erythematosus: Diagnostic Reconsideration Based on Long-Term Clinical Course. Cureus 18(11): e119338. DOI 10.7759/cureus.119338

comparisons of clinical, serological, and pathological findings are limited. This case highlights the importance of diagnostic reconsideration in patients with atypical clinical courses. The prolonged interval between episodes and comparison of the two marrow biopsies provide the principal educational contribution beyond previously reported SLE-TAFRO overlap cases.

Case Presentation

An 80-year-old man developed fever and lower leg edema five years earlier and visited a nearby clinic. Laboratory tests revealed marked thrombocytopenia (platelet (PLT)count 2.3 × 10^4/μL), and he was referred to our hospital for further evaluation. Plain CT of the chest, abdomen, and pelvis revealed bilateral pleural effusion, pericardial effusion, ascites, and enlargement of the left hepatic lobe (Figure *1*, panels A and B). Bone marrow biopsy was performed to evaluate hematologic disorders, including TAFRO syndrome, but showed no reticulin fibrosis and only mild hypercellularity (Figure *2*, panel A). The absence of fibrosis initially reduced the clinical suspicion of TAFRO syndrome but did not exclude it.

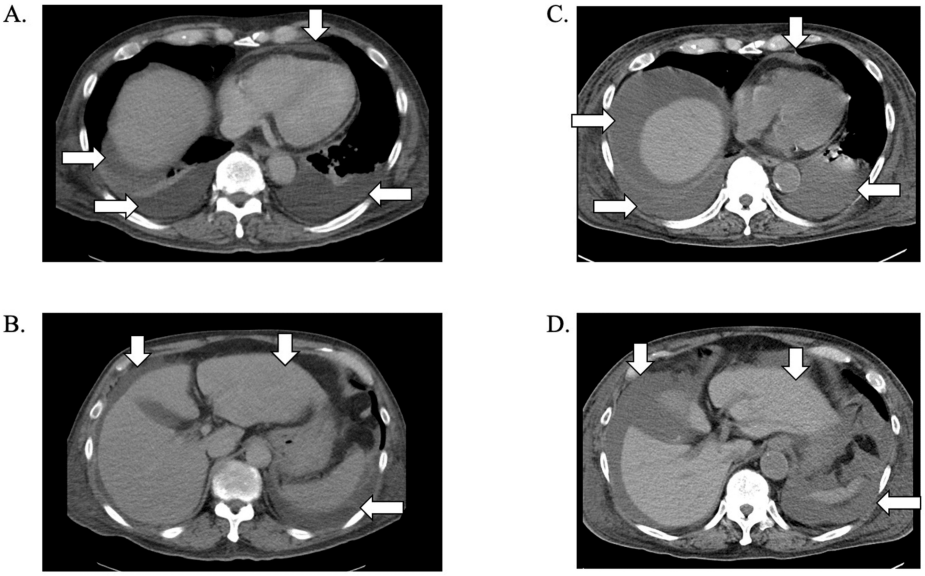


FIGURE 1: CT findings at initial presentation and relapse

A and B: Plain chest-abdominal CT at the time of SLE diagnosis shows bilateral pleural effusions, pericardial effusion, ascites, and enlargement of the left hepatic lobe. C and D: CT at relapse demonstrate recurrence of massive pleural, pericardial, and peritoneal effusions with left hepatic lobe enlargement.

In panels A and C, the lower right-pointing and left-pointing arrows indicate pleural effusions, the downward-pointing arrow indicates pericardial effusion, and the upper right-pointing arrow indicates ascites. In panels B and D, the downward-pointing arrow on the left and the left-pointing arrow indicate ascites, while the downward-pointing arrow on the right indicates enlargement of the left hepatic lobe.

SLE: Systemic lupus erythematosus

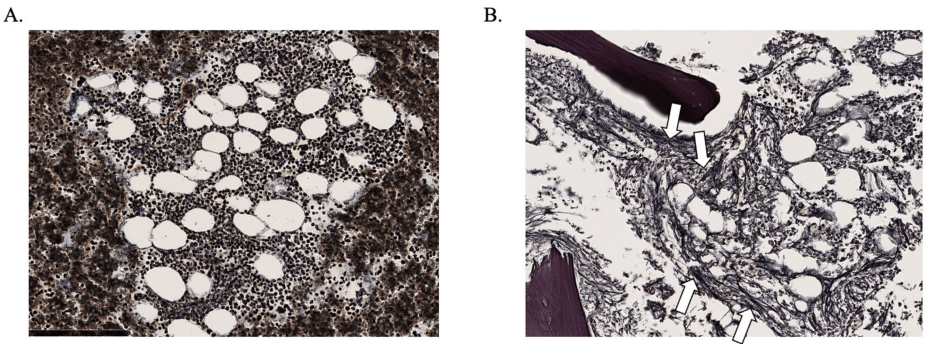


FIGURE 2: Bone marrow findings at the initial and second admissions (silver staining)

A: Bone marrow biopsy at the time of SLE diagnosis shows no fibrosis and only mild hypercellularity (×400); B: Repeat biopsy at relapse reveals mild reticulin fibrosis (MF-1 according to the European consensus grading system; arrows) and increased megakaryocytes (at least seven per high-power field), supporting the clinical diagnosis of TAFRO syndrome (×400).

SLE: Systemic lupus erythematosus, MF-1: Myelofibrosis grade 1, TAFRO: Tthrombocytopenia, anasarca, fever, reticulin fibrosis, and organomegaly

Laboratory findings demonstrated thrombocytopenia, positive antinuclear antibody (ANA), hypocomplementemia, and positivity for anti-U1-RNP and anti-cardiolipin antibodies. Thoracentesis revealed an exudative effusion (pleural fluid/serum protein ratio = 0.51), which was interpreted as serositis. Other laboratory findings are shown in Table *1*. Based on these findings, he fulfilled the 1997 American College of Rheumatology (ACR) criteria (serositis, thrombocytopenia, antinuclear antibody (ANA) positivity, and anticardiolipin antibody positivity) [9] and 2012 Systemic Lupus International Collaborating Clinics (SLICC) classification criteria for SLE (serositis, thrombocytopenia, ANA positivity, and low complement) [10] and was diagnosed accordingly. However, the ANA titer was low (1:40; laboratory reference, <1:40), and SLE-specific autoantibodies such as anti-dsDNA and anti-Sm antibodies were absent. The clinical course is shown in Figure *3*.

Category	Variable	Reference range	Results
Biochemistry	Total protein (g/dL)	6.0-8.3	6.5
	Albumin (g/dL)	3.9-4.9	3.1
	Aspartate aminotransferase (U/L)	10-37	19
	Alanine aminotransferase (U/L)	5-40	12
	Lactate dehydrogenase (U/L)	124-222	202
	Gamma-glutamyl transferase	0-50	22
	Blood urea nitrogen (mg/dL)	8-20	17.6
	Creatinine (mg/dL)	0.34-0.79	0.77
	Uric acid (mg/dL)	2.6-6.0	5.5
	Sodium (mEq/L)	138-147	138
	Potassium (mEq/L)	3.3-4.8	3.9
	Calcium (mg/dL)	8.8-10.4	8.5
	CRP (mg/dL)	0-0.25	6.04
	IgG (mg/dL)	870-1,700	2,548
	IgG4 (mg/dL)	11.0-121.0	86.4
	IgA (mg/dL)	93-393	337
	IgM (mg/dL)	50-269	76

Immunology	soluble IL-2 receptor (U/mL)	121-613	1,062
	Helicobacter pylori IgG (U/mL)	<9.0	<3.0
	Haptoglobin (mg/dL)	19-170	191
	C3 (IU/mL)	73-138	71
	C4 (IU/mL)	11-31	7
	CH50 (U/mL)	31.6-57.6	25.7
	ANA titer	<1:40	1:40 (H+Sp)
	Anti-dsDNA antibody (IU/mL)	<12.0	<1.2
	Anti-Sm antibody (U/mL)	<10.0	<1.0
	Anti-U1-RNP antibody (U/mL)	<10.0	28.8
	Anti-SS-A antibody (U/mL)	<10.0	<1.0
	Anticardiolipin(anti-CL)-IgG antibody (U/mL)	<10.0	31.4
	Anti-CL·b2GPI (U/mL)	<3.5	0.9
	PA-IgG (ng/10^7 cells)	0-46.0	3,040
Complete blood count	White blood cell count (/μL)	3,250-8,570	9,090
	Neutrophils (%)	40.3-71.7	75.8
	Lymphocytes (%)	20.9-50.5	18
	Monocytes (%)	2.0-10.0	5.6
	Eosinophils (%)	0-8.5	0.3
	Basophils (%)	0-2.5	0.3
	Hemoglobin (g/dL)	11.1-15.5	11.7
	PLT count (/μL)	14.8×10^4-33.6×10^4	2.3×10^4
Coagulation test	Activated partial thromboplastin time (APTT) (sec)	24-39	36
	Prothrombin time and international normalized ratio (PT-INR)	0.90-1.10	1.05
	D-dimer (mg/mL)	<10.00	11.59
Urinalysis	Protein		2
	Occult blood		+-
	Granular cast (whole field)		10-19
Pleural fluid	Appearance		Yellow and turbid
	Total protein (g/dL)		3.3
	Lactate dehydrogenase (U/L)		88

TABLE 1: Laboratory findings at the initial admission (diagnosed as SLE)

The results show thrombocytopenia, positive ANA, hypocomplementemia, and positivity for anti-U1-RNP and anti-CL antibodies, which supported the initial diagnosis of SLE but were not individually disease-specific. The institutional reference ranges are C3 73–138 mg/dL and C4 11–31 mg/dL.

SLE: Systemic lupus erythematosus, CRP: C-reactive protein, ANA: Antinuclear antibody, CL: Cardiolipin, PLT: Platelet

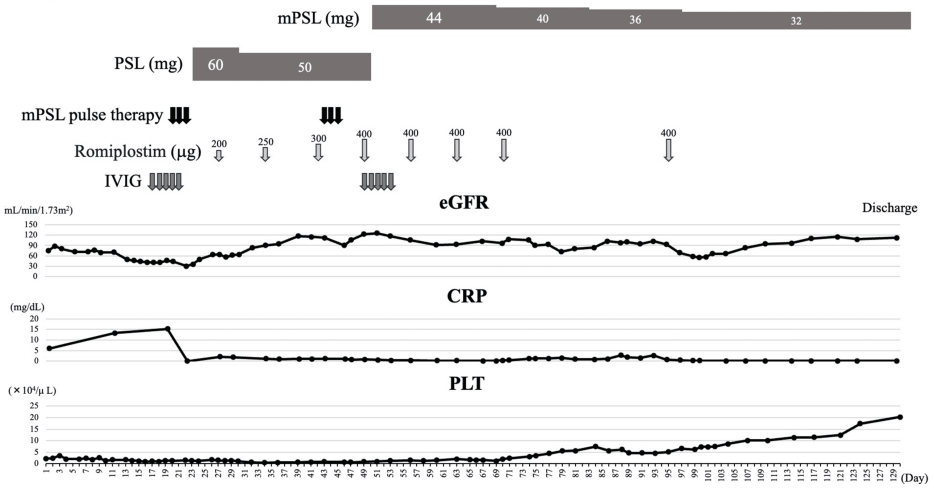


FIGURE 3: Clinical course of the first admission

This graph captures the changes in PLT, CRP, and estimated glomerular filtration rate (eGFR) during treatment for SLE. The patient received two courses of IVIG and mPSL pulse therapy, followed by PSL and romiplostim. Thrombocytopenia gradually improved, and the patient achieved remission.

PLT: Platelet, CRP: C-reactive protein, IVIG: Intravenous immunoglobulin, mPSL: Methylprednisolone, PSL: Prednisolone

Echocardiography at the first admission showed a left ventricular ejection fraction (LVEF) of 60%, no regional wall motion abnormalities, and no aortic stenosis. The E/e′ was 14.2, the estimated right ventricular systolic pressure was 41.5 mmHg, and the inferior vena cava (IVC) measured 12.7 mm with respiratory variation. Serum creatinine and proteinuria findings before the first admission were unavailable.

He was treated with two courses each of intravenous immunoglobulin (IVIG) and methylprednisolone (mPSL) pulse therapy, followed by prednisolone (PSL) 60 mg/day (later changed to an equivalent dose of mPSL considering fluid retention) and romiplostim. Thrombocytopenia gradually improved, and he was discharged on day 161. Thereafter, mPSL monotherapy was continued and tapered. Two and a half years after discharge, mPSL was discontinued, and he remained in remission without relapse.

At an outpatient visit four years and six months after discharge, the PLT count had again decreased to 2.5 × 10^4/μL, accompanied by lower leg edema and abdominal distension. Plain CT revealed marked pleural, pericardial, and peritoneal effusions with enlargement of the left hepatic lobe, similar to the previous findings (Figure *1*, panels C and D). He was admitted to our department for further examination of thrombocytopenia and fluid retention.

On admission, he was alert and oriented. His body height was 156 cm, weight 63.6 kg, temperature 37.0°C, blood pressure 137/72 mmHg, pulse 90/min, respiratory rate 20/min, and oxygen saturation 96% (on 1 L/min oxygen). The palpebral conjunctiva showed no anemia, and the sclera showed no jaundice. No thyroid enlargement or cervical lymphadenopathy was observed. Breath sounds were clear without rales. Heart sounds were normal, with a grade II/VI systolic murmur best heard along the right sternal border in the second intercostal space. The abdomen was distended but soft, with normal bowel sounds. No joint swelling or tenderness was noted. Bilateral pitting edema of the lower legs was present, pedal pulses were palpable, and there was no skin rash. His medical history included type 2 diabetes mellitus and aortic valve stenosis. There was no family history of collagen disease. He had smoked 40 cigarettes a day for five years until he was 25 and occasionally consumed alcohol. His only medication was repaglinide 0.75 mg/day. Laboratory data showed negative antinuclear, anti-cardiolipin, and anti-U1-RNP antibodies, with C3 of 74 mg/dL and C4 of 12 mg/dL, both within the institutional reference ranges (C3, 73-138 mg/dL; C4, 11-31 mg/dL). Other findings are summarized in Table *2*.

Category	Variable	Reference range	Results
	Total protein (g/dL)	6.0-8.3	5.1
	Albumin (g/dL)	3.9-4.9	2.3
	Aspartate aminotransferase (U/L)	10-37	18
	Alanine aminotransferase (U/L)	5-40	8

Biochemistry	Lactate dehydrogenase (U/L)	124-222	195
	Gamma glutamyl transferase	0-50	45
	Blood urea nitrogen (mg/dL)	8-20	27.1
	Creatinine (mg/dL)	0.34-0.79	0.88
	Uric acid (mg/dL)	2.6-6.0	7.7
	Sodium (mEq/L)	138-147	142
	Potassium (mEq/L)	3.3-4.8	3.8
	Calcium (mg/dL)	8.8-10.4	7.4
Immunology	CRP (mg/dL)	0-0.25	4.32
	IgG (mg/dL)	870-1,700	1258
	IgG4 (mg/dL)	11.0-121.0	64.1
	IgA (mg/dL)	93-393	228
	IgM (mg/dL)	50-269	67
	C3 (IU/mL)	73-138	74
	C4 (IU/mL)	11-31	12
	ANA titer	<1:40	<1:40
	Anti-dsDNA antibody (IU/mL)	<12.0	<1.2
	Anti-Sm antibody (U/mL)	<10.0	<1.0
	Anti-U1-RNP antibody (U/mL)	<10.0	9.2
	Anti-SS-A antibody (U/mL)	<10.0	<1.0
	Anti-CL-IgG antibody (U/mL)	<10.0	8.7
	Anti-CL·β2GPI (U/mL)	<3.5	0.9
	Lupus anticoagulant	0-1.2	0.9
Complete blood count	White blood cell count (/μL)	3,250-8,570	11,680
	Neutrophils (%)	40.3-71.7	79.6
	Lymphocytes (%)	20.9-50.5	14
	Monocytes (%)	2.0-10.0	5
	Eosinophils (%)	0-8.5	0.8
	Basophils (%)	0-2.5	0.6
	Hemoglobin (g/dL)	11.1-15.5	9
	PLT count (/μL)	14.8×10^4-33.6×10^4	2.5×10^4
Coagulation test	APTT(sec)	24-39	33.8
	PT-INR	0.90-1.10	1.11
	D-dimer (μg/mL)	<10.00	8.8
Urinalysis	Protein		3
	Protein-creatinine ratio (g/g·creatinine)		1.55
	Occult blood		+
	Granular cast (whole field)		1-4

TABLE 2: Laboratory findings at the second admission (diagnosed as TAFRO syndrome)

The results show marked thrombocytopenia and elevated CRP without autoantibody positivity. The C3 (74 mg/dL) and C4 (12 mg/dL) were within the institutional reference ranges of 73-138 mg/dL and 11-31 mg/dL, respectively.

TAFRO: Thrombocytopenia, anasarca, fever, reticulin fibrosis, and organomegaly; SLE: Systemic lupus erythematosus; CRP: C-reactive protein; ANA: Antinuclear antibody; CL: Cardiolipin; PLT: Platelet; APTT: Activated partial thromboplastin time; PT-INR: Prothrombin time and international normalized ratio

Before the second admission, serum creatinine was 0.79 mg/dL, and proteinuria was absent. Echocardiography showed an LVEF of 77%, no regional wall motion abnormalities, aortic stenosis with a mean transvalvular gradient of 20.7 mmHg and a valve area of 1.22 cm^2, and mild aortic regurgitation. The estimated right ventricular systolic pressure was 31.6 mmHg, and the IVC measured 7.8 mm with respiratory variation. Renal biopsy was not performed because of marked thrombocytopenia.

An SLE relapse was initially suspected, and PSL 20 mg/day was started on hospital day two. Intravenous immunoglobulin was administered from day three for thrombocytopenia. The PSL was increased to 30 mg/day on day four, and mPSL pulse therapy (1,000 mg/day for 3 days) was performed from day six, followed by an increase in PSL to 60 mg/day. However, thrombocytopenia and serous effusions persisted and worsened, and renal dysfunction progressed. Because of refractory thrombocytopenia, intractable pericardial, pleural, and peritoneal effusions, progressive renal impairment, enlargement of the left hepatic lobe, fever, and elevated serum C-reactive protein (CRP) levels, TAFRO syndrome was suspected. A bone marrow biopsy was performed on day 14, and tocilizumab (TCZ) 500 mg was initiated intravenously. The biopsy later demonstrated mild reticulin fibrosis (myelofibrosis grade 1 (MF-1) according to the European consensus grading system [11]) and increased megakaryocytes (at least seven per high-power field; Figure *2*, panel B). The patient fulfilled all three major categories of the Japanese TAFRO criteria: anasarca, pretreatment thrombocytopenia (25,000/μL), and systemic inflammation (CRP, 4.32 mg/dL). The minor categories were reticulin fibrosis with increased megakaryocytes, hepatomegaly, and progressive renal impairment during hospitalization [2,3]. These findings supported a clinical diagnosis of TAFRO syndrome.

Blood, urine, pleural fluid, and ascitic fluid cultures were negative, as were mycobacterial tests of pleural and ascitic fluid. No schistocytes were detected on peripheral blood examination. Lymph node biopsy was not performed because of marked thrombocytopenia; therefore, iMCD-TAFRO could not be histologically confirmed.

The subsequent clinical course is shown in Figure *4*. During hospitalization, the patient received two additional courses of IVIG and a total of eight doses of TCZ. He developed cholangitis twice, and both episodes responded to piperacillin/tazobactam. Thrombocytopenia, generalized edema, and renal dysfunction gradually improved, and he was discharged on day 154. Tocilizumab was continued after discharge, and PSL was gradually tapered. By day 250 after discharge, PSL had been reduced to 1 mg/day. He has since remained clinically stable without recurrence.

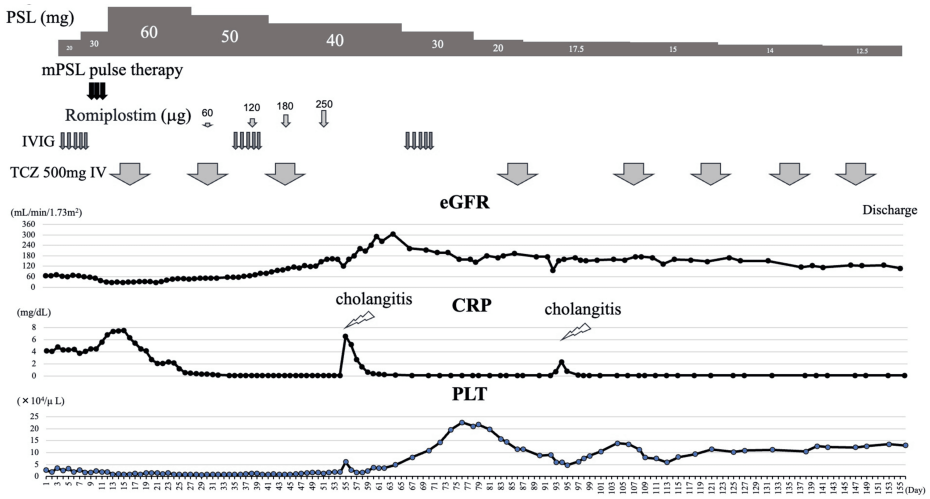


FIGURE 4: Clinical course of the second admission

After relapse, thrombocytopenia and systemic edema were refractory to glucocorticoids and IVIG. Tocilizumab 500 mg was introduced, and PLT count, renal function, and fluid retention subsequently improved during treatment that also included glucocorticoids, IVIG, and romiplostim. The improvement cannot be attributed solely to TCZ. Episodes of cholangitis were effectively treated with antibiotics. The patient remained stable after discharge under continued TCZ therapy with tapering PSL.

IVIG: Intravenous immunoglobulin, PLT: Platelet, TCZ: Tocilizumab, PSL: Prednisolone

Discussion

This case illustrates the difficulty in distinguishing SLE from TAFRO syndrome in patients presenting with overlapping clinical features. Importantly, the diagnosis required reconsideration based on the long-term clinical course and evolving laboratory and pathological findings. In retrospect, the diagnosis of SLE at the initial presentation remains uncertain. Although the patient fulfilled classification criteria, the ANA titer was low (1:40), and SLE-specific autoantibodies such as anti-dsDNA and anti-Sm were negative. The presence of anti-U1-RNP and anticardiolipin antibodies at relatively low titers may reflect nonspecific immune activation rather than a disease-specific autoimmune process, particularly in an elderly patient. In addition, several clinical features were atypical for SLE, including advanced age, male sex, marked anasarca, and hepatomegaly, which are uncommon in SLE in the absence of severe cardiac or renal involvement.

The TAFRO syndrome is an uncommon clinicopathological entity characterized by thrombocytopenia, anasarca, fever, reticulin myelofibrosis, and organomegaly and should be distinguished from histologically confirmed iMCD-TAFRO [4]. In the present case, TAFRO syndrome was considered early in the clinical course; however, the initial bone marrow biopsy showed no fibrosis, and the accompanying serological findings favored the working diagnosis of SLE. These findings did not definitively exclude TAFRO syndrome. The absence of reticulin fibrosis in the initial bone marrow biopsy does not exclude TAFRO syndrome, because reticulin fibrosis is not an individually mandatory diagnostic criterion [2,3]. It is therefore possible that the initial presentation represented TAFRO syndrome with lupus-like features rather than true SLE, although this distinction cannot be definitively established.

Notably, the clinical manifestations at initial presentation and relapse were highly similar, including thrombocytopenia, anasarca, organomegaly, and systemic inflammation. This similarity emphasizes the value of longitudinal diagnostic reassessment but does not establish a shared inflammatory process or a transition between diseases. The patient's demographic characteristics, advanced age and male sex, are atypical for SLE but have been reported in cases of TAFRO syndrome with lupus-like features, suggesting that such features may provide important diagnostic clues.

Several alternative interpretations should be considered, including TAFRO syndrome initially presenting with lupus-like features, an incomplete or early TAFRO phenotype at the first presentation, and the coexistence of overlapping immune-mediated processes. The present case cannot definitively distinguish among these possibilities.

Cardiac and renal contributions to fluid retention were considered. The LVEF was preserved, and the IVC was not dilated and showed respiratory variation at both admissions. Nevertheless, preserved LVEF did not exclude abnormal filling pressures, and the first-admission E/e′ of 14.2 and subsequent aortic valve disease did not permit complete exclusion of a cardiac contribution. The absence of proteinuria and a creatinine of 0.79 mg/dL before the second admission made established proteinuric diabetic kidney disease a less likely

explanation for the subsequent renal deterioration. However, nonproteinuric diabetic kidney disease, lupus nephritis, and other renal causes could not be definitively excluded without biopsy. Normal C3 and C4 and negative anti-dsDNA antibodies did not exclude lupus nephritis. Negative cultures and mycobacterial tests did not comprehensively exclude infection, and the absence of schistocytes alone did not exclude thrombotic microangiopathy. The absence of lymph node histology also limited the evaluation of lymphoproliferative disorders.

From a pathophysiological perspective, type I interferon (IFN-I) signaling provides a possible hypothesis for overlapping inflammatory features. Systemic lupus erythematosus is characterized by a prominent IFN-I signature [12], and IFN-I can enhance IL-6 production under specific experimental conditions [13]. However, neither pathway was directly investigated in this patient. A shared mechanism or a shift from autoantibody-associated disease to IL-6-mediated inflammation therefore remains hypothetical and cannot be established by this case or its treatment response.

At relapse, the patient presented with recurrent fever, effusions, hepatomegaly, and thrombocytopenia. Disappearance of previously detected autoantibodies, normalization of C3 and C4 according to institutional reference ranges, and newly detected marrow fibrosis prompted diagnostic reassessment; these findings did not independently exclude SLE activity. There are no established, unified treatment guidelines for TAFRO syndrome, and management is largely based on case series and expert opinion. High-dose glucocorticoids are generally used as first-line therapy, with additional agents such as TCZ, cyclosporine A, or rituximab recommended for refractory cases [14,15]. In the present case, improvement followed the introduction of TCZ during treatment that also included glucocorticoids, IVIG, and romiplostim, consistent with previous reports of improvement during IL-6-targeted therapy [16]. The contributions of individual agents and possible delayed effects of preceding treatments cannot be separated, and the response cannot be attributed solely to TCZ.

Previous studies have described cases of TAFRO syndrome mimicking SLE or coexisting with autoimmune conditions [6-8]. In a comparative study, Hasegawa et al. reported that four out of 46 patients (8.7%) with SLE also fulfilled the diagnostic criteria for TAFRO syndrome [8]. These patients were predominantly older males and exhibited higher inflammatory markers, more severe organ dysfunction, and lower platelet counts compared to typical SLE cases. Notably, they also tended to lack high-titer SLE-specific autoantibodies. These findings suggest that demographic characteristics and clinical phenotype may provide important diagnostic clues. In particular, an older patient diagnosed with SLE who presents with marked systemic inflammation, refractory edema or effusions, thrombocytopenia, and low-titer or absent SLE-specific autoantibodies should prompt consideration of TAFRO syndrome.

Conclusions

Whether the initial presentation represented SLE or TAFRO syndrome with lupus-like features remains uncertain. This case does not establish disease evolution or a shared inflammatory mechanism. This case underscores the importance of maintaining diagnostic flexibility and reconsidering the diagnosis when the clinical course deviates from expectations. Accumulation of similar cases will be essential to further clarify the relationship between TAFRO syndrome and autoimmune diseases and to refine diagnostic and therapeutic strategies.

Additional Information

Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

Concept and design: Takuma T. Wada, Kazuhiro Yokota, Toshihide Mimura, Yuji Akiyama

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Critical review of the manuscript for important intellectual content: Kazuhiro Yokota, Hiroshi Yamaguchi, Toshihide Mimura, Yuji Akiyama, Nobuhito Okumura, Takehito Ueno, Yoshihiro Yoshida

Supervision: Kazuhiro Yokota, Hiroshi Yamaguchi, Toshihide Mimura, Yuji Akiyama

Disclosures

Human subjects: Informed consent for treatment and open access publication was obtained or waived by all participants in this study. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support

was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

Acknowledgements

Author contributions were as follows: Takuma Tsuzuki Wada was involved in patient care, conceptualization of the case report, literature review, and drafting of the manuscript. Kazuhiro Yokota provided overall supervision and guidance. Takehito Ueno, Nobuhito Okumura, and Yoshihiro Yoshida were involved in the patient's inpatient care and preparation of the case summary. Hiroshi Yamaguchi provided supervision and guidance regarding the pathological findings. Toshihide Mimura and Yuji Akiyama provided overall supervision and guidance.

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