Modern Rheumatology Case Reports, 2026, Vol. 10, Issue 1, rxaf075
doi:10.1093/mrcr/rxaf075
Advance access publication date 20 November 2025
Case Report

Polyarteritis nodosa presenting with TAFRO signs following COVID-19 infection: case report

Rina Ishigame[a,b], Shigeru Iwata[b,*], Kayoko Tabata[c], Hiromitsu Kita[d], Yuri Nakashima[d], Mizuki Nakatsuka[c], Shotaro Tabata[e], Ryuta Iwamoto[f], Masatoshi Jinnin[c], Shin-ichi Araki[d] and Takao Fujii[b]

[a]Department of Internal Medicine, Iwate Prefectural Chubu Hospital, Iwate, Japan; [b]Department of Rheumatology and Clinical Immunology, Wakayama Medical University, Wakayama, Japan; [c]Department of Dermatology, Wakayama Medical University, Wakayama, Japan; [d]Department of Nephrology, Wakayama Medical University, Wakayama, Japan; [e]Department of Hematology, Wakayama Medical University, Wakayama, Japan; [f]Department of Human Pathology, Wakayama Medical University, Wakayama, Japan

*Correspondence: Shigeru Iwata; iwatas@wakayama-med.ac.jp; Department of Rheumatology and Clinical Immunology, Wakayama Medical University, 811-1 Kimiidera, Wakayama 641-8509, Japan.

ABSTRACT

The diagnostic criteria for TAFRO syndrome exclude autoimmune diseases, and they have been considered to be mutually independent. However, several cases of autoimmune diseases with TAFRO signs have been reported in recent years. Also, similarities in cytokine profiles in COVID-19 and TAFRO syndrome have been previously reported. Our patient, a 53-year-old Japanese man, was diagnosed with COVID-19 and had a persistent fever. Two weeks later, pain, numbness, and oedema appeared, mainly in the right lower leg but gradually spreading to the distal extremities. Subsequently, purpura appeared on his forearms and lower legs, and 10 weeks after the COVID-19 diagnosis he presented to our hospital. On admission, in addition to fever, polyangiitis, and purpura of the extremities, he had splenomegaly, lymphadenopathy, and anasarca. Skin and renal histopathology revealed fibrinoid necrotising vasculitis of small and medium-sized arteries. In addition, his platelet count was low, Alkaline phosphatase (ALP) was elevated, and there was anasarca, fever, and renal failure. A diagnosis of polyarteritis nodosa with TAFRO signs was made. On the 20th day of admission, high-dose glucocorticoids and high-dose intravenous cyclophosphamide were started. The platelet count initially improved, with gradual improvement of vasculitis and symptoms of fever, purpura, and neuropathy. However, there was another decrease in platelets, progression of renal dysfunction, and worsening of fluid retention. Tocilizumab was added, but the disease could not be controlled, and on the 51st day, necrotising fasciitis developed and the patient died. This case suggests that COVID-19, TAFRO syndrome, and vasculitis may be interrelated in their pathogeneses.

KEYWORDS COVID-19; TAFRO syndrome; polyarteritis nodosa; cytokines; anasarca

Introduction

TAFRO syndrome is a systemic inflammatory disease of unknown aetiology that is characterised by thrombocytopenia, anasarca (oedema, pleural effusions, ascites), fever, reticulin fibrosis, renal failure, and organ enlargement (hepatosplenomegaly and lymphadenopathy). Our patient had polyarteritis nodosa with TAFRO signs after COVID-19 infection. The diagnostic criteria for TAFRO syndrome exclude autoimmune diseases, and they have been considered to be mutually independent. However, several cases of autoimmune diseases with TAFRO signs (especially Sjögren's syndrome and systemic lupus erythematosus) have been reported in recent years [1–7]. We suggest that clearer definitions of the pathogenesis and diagnosis of TAFRO syndrome are required. Meanwhile, there have also been reports of development of autoimmune disease after COVID-19 vaccination/infection [8, 9]. Although one case of cutaneous arteritis with TAFRO signs has been reported in the literature [10], here, we present what is thought to be the first reported case of polyarteritis nodosa with TAFRO signs after COVID-19 infection. Before treatment, our patient's serum IL-10, IP-10, and IL-18 concentrations, which are reported in both COVID-19 and TAFRO syndrome, were elevated, and this correlated with changes in TAFRO syndrome after treatment. This case is thought to be notable because it suggests that COVID-19, TAFRO syndrome, and vasculitis may be interrelated in their pathogeneses.

Case presentation

A 53-year-old man without notable medical history was diagnosed with COVID-19 and had a persistent fever in the 38°C range. Family history was unremarkable. Two weeks later, pain, numbness, and oedema appeared, mainly in the right lower leg and then gradually spreading to the periphery of both lower extremities. Subsequently, purpura appeared on his forearm and lower leg, and approximately 1 month after the COVID-19 diagnosis he visited his local doctor. Skin biopsy showed histological features consistent with small-vessel arteritis, and ∼10 weeks after the onset of diagnosis of COVID-19 infection he was referred to our hospital for further examination and treatment. Blood tests revealed anaemia (Hb 8.7 mg/dl), elevated ALP (895 IU/l), and renal dysfunction (Cre 1.18 mg/dl). His body temperature was 37°C, he had hypertension (blood pressure: 173/83 mmHg), and he had lymphadenopathy in the neck and right inguinal region. He also had pitting oedema on the face, dorsum of

Received 25 February 2025; Accepted 14 October 2025

Table 1. Laboratory data on admission.

<Haematology>			Normal range	<Immunology>			Normal range
White blood cells	4230	/μl	3300–8600	Antinuclear antibody	<40		
Red blood cells	3.18	$\times 10^6/\mu l$	3.86–4.92	Anti-dsDNA antibody	18.3	IU/ml	<12.0
Haemoglobin	8.7	g/dl	11.6–14.8	Anti-ssDNA antibody	82	AU/ml	<25.0
Platelet	17.2	$\times 10^4/\mu l$	15.8–34.8	Anti SS-A antibody	<1.0	IU/ml	<10.0
<Biochemistry/Serology>				Immunoglobulin G	1811	mg/dl	861–1747
Total protein	6,2	g/dl	6.6–8.1	IL-6	22.16	pg/ml	<7.0
Albumin	2,4	g/dl	4.1–5.1	C3	50	mg/dl	73–138
AST	47	IU/l	13–30	C4	3	mg/dl	11–31
ALT	38	IU/l	7–23	CH50	<15	mg/dl	31.6–57.6
LDH	240	IU/l	124–222	Anticardiolipin IgG	6.5	U/ml	<10
ALP	895	IU/l	106–322	Anti-beta 2 glycoprotein IgG	<1.3	U/ml	<3.5
γGTP	269	IU/l	<50	MPO-ANCA	(−)		
BUN	15.4	mg/dl	8–20	PR3-ANCA	(−)		
Creatinine	1.18	mg/dl	0.46–0.79	VEGF	121.47	Pg/ml	<38.3
eGFR	51.8	ml/min/1.73 m^2	>90	<Urinalysis>			
Na	131	mEq/l	138–145	Protein	(+1)		
K	3.6	mEq/l	3.6–4.8	Occult blood	(+1)		
Cl	97	mEq/l	101–108	Red blood cell	5–9	/HPF	
CRP	3.42	mg/dl	0.00–0.14	Granular casts	1–9	/HPF	
<Coagulation>				Protein–creatinine rate	0.56	g/gCr	
APTT	37.8	s	24.0–39.0	β2MG	52,863	mg/dl	0.9–1.9
PT	85.3	s	11.0–13.4	NAG	51	U/l	<11.5

the hands and feet, and purpura on the forearms and lower legs. Neurological examination revealed peripheral numbness in the upper and lower limbs, decreased pain and temperature sensation, reduced muscle strength in the distal limbs on the manual muscle testing scale, and decreased patellar/Achilles tendon reflexes.

Upon admission as an inpatient, blood tests (Table 1) showed Hb 8.7 mg/dl indicating anaemia, and a platelet count of 172 000/μl, which had previously dropped to 86 000/μl by the time treatment was initiated. Aspartate transferase (AST) and Alanine transaminase (ALT) were not elevated, but there was elevation of biliary enzymes ALP (895 IU/l) and Gamma-glutamyl transpeptidase (γ-GTP) (269 IU/l). There was renal dysfunction, and mild elevation of c-reactive protein (CRP) (3.42 mg/dl) and Immunoglobulin G (IgG) (1811 mg/dl). Immunological tests were negative for antinuclear antibodies, MPO-ANCA and PR3-ANCA. Normal anti-dsDNA antibody levels were normal (18.3 IU/l), but the patient had hypocomplementaemia (C3 50 mg/dl, C4 5 mg/dl, CH50 < 15 mg/dl). Additional tests showed serum IL-6 at 22.16 pg/ml and Vascular endothelial growth factor (VEGF) at 121.47 pg/ml. Urinalysis showed proteinuria (+1), haematuria (+1), a TP/Cre ratio of 0.56 g/g creatinine, and granular casts of 1–9/high powered field (HPF), which was suggestive of tubular damage. Simple computed tomography showed multiple enlarged lymph nodes, hepatomegaly, splenomegaly, ascites, and subcutaneous oedema (Figure 1). Nerve conduction studies indicated mononeuritis multiplex.

During hospitalisation, peripheral numbness and decreased pain and temperature sensation spread to the proximal parts of the upper and lower limbs, and there was further decline in muscle strength in the distal limbs on the manual muscle testing scale. Fluid retention gradually increased, and in addition to pitting oedema of the face and limbs, there was increase in pleural effusion. On the 11th day after admission, blood tests showed a further decrease in platelet count to 86 000/μl, progressive renal dysfunction, and elevation of ALP to 1 533 IU/l. Other causes of thrombocytopenia, such as drug-induced thrombocytopenia, *Helicobacter pylori* infection, disseminated intravascular coagulation, and thrombotic microangiopathy, were ruled out based on test results.

For further examination and diagnosis, we performed kidney, right axillary lymph node, and bone marrow biopsies. Pathological examination results showed 17 glomeruli in kidney tissue (Figure 2(a)), with two glomeruli showing global sclerosis and two showing collapse. There was no glomerular hypertrophy, but mild mesangial cell proliferation and increased mesangial matrix were observed in 80% of the glomeruli. There were no intraluminal lesions, no thickening of the basement membrane, no duplication, no spikes, no deposits, and no crescents. Approximately 30% of the interstitium showed chronic inflammation and fibrosis, and the tubules were atrophied. Red blood cells were seen in the tubular lumen, and fibrinoid necrosis was observed in interlobular arterioles and medium-sized vessels. Immunostaining was negative. There was no clear evidence of vascular wall destruction, inflammatory cell infiltration, or thickening of the vascular endothelium. In addition to a renal biopsy, a skin biopsy was performed from the purpuric lesion on the right thigh (Figure 2(b)). In several vessels at the boundary between the deep dermis and subcutaneous fat tissue, there was marked fibrinoid necrosis, destruction of the vascular wall with a small amount of nuclear debris, and reactive swelling of the vascular endothelium. There was infiltration of neutrophils, lymphocytes, and a small number of eosinophils around the vessels, suggesting vasculitis. There were no findings of leukocytoclastic vasculitis in the superficial dermis, but there was neutrophil infiltration around the large deep vessels. Elastica van Gieson staining revealed findings suggestive of vasculitis in small and medium-sized arteries, such as the course of the internal elastic lamina in some of the damaged vessels. Overall, the findings were consistent with polyarteritis nodosa. When combined with the renal pathological

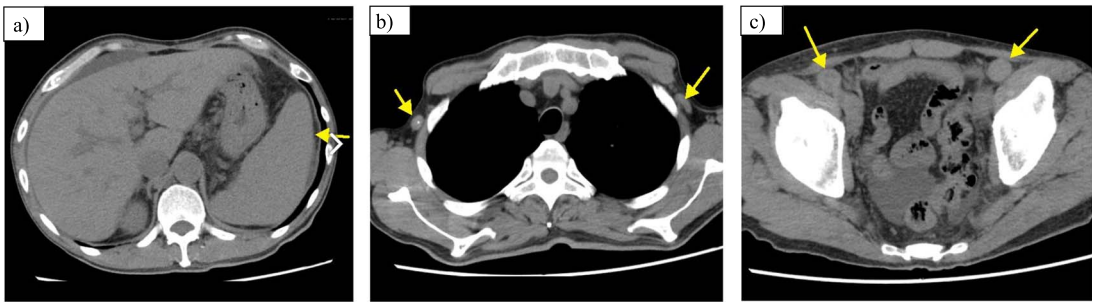

Figure 1. Imaging findings at admission. (a) Hepatomegaly and splenomegaly. (b) Enlarged axillary lymph nodes. (c) Enlarged inguinal lymph node.

findings, these findings are thought to strongly support the diagnosis of polyarteritis nodosa. Bone marrow tissue (Figure 2(c)) was hyperplastic, with no increases or maturation disorders in haematopoietic cells of the three lineages, and mature megakaryocytes increased to ∼10/HPF. Mild reticular fibrosis corresponding to MF-1 was observed on silver staining, which is a finding of TAFRO syndrome but which could also occur in autoimmune diseases. In the right axillary lymph node tissue (Figure 2(d)), there were no specific findings except for dense fibrosis in the central part of the lymph node. The possibility of an old infarction of the lymph node was suggested by the vasculitis findings in the skin.

Regarding the diagnosis, the clinical symptoms of vasculitis and the pathological findings of medium-sized arteritis met the diagnostic criteria for polyarteritis nodosa revised by the Ministry of Health, Labour and Welfare in 2006 and the American College of Rheumatology 1990 criteria for the classification of polyarteritis nodosa [11]. The major signs included fever, hypertension, rapidly progressive renal failure, mononeuritis multiplex, and purpura, along with the histopathological finding of fibrinoid necrotising vasculitis of medium and small arteries, leading to a diagnosis of polyarteritis nodosa. According to Watts's algorithm, it could also be classified as polyarteritis nodosa. However, the progression of thrombocytopenia, elevated ALP, and fluid retention trend could not be explained by polyarteritis nodosa alone. According to the 2015 diagnostic criteria for TAFRO syndrome (minor revision 2019) [12], the patient met all three required criteria: fluid retention (pleural/ascitic fluid, generalised oedema), thrombocytopenia (minimum platelet count $< 100\,000/\mu$l before treatment initiation), and unexplained fever ($\geq$37.5°C) or positive inflammatory response (CRP $\geq$ 2 mg/dl). Additionally, the patient met two of the four minor criteria: Castleman disease–like findings on lymph node biopsy, bone marrow fibrosis (reticular fibrosis or increased megakaryocytes), mild organ enlargement (hepatosplenomegaly, lymphadenopathy), and progressive renal dysfunction.

Although the exclusion criteria for TAFRO syndrome included autoimmune diseases, this case was comprehensively diagnosed as polyarteritis nodosa with TAFRO signs. Differential diagnoses for thrombocytopenia, including drug-induced thrombocytopenia, *H. pylori* infection, disseminated intravascular coagulation (DIC), and thrombotic microangiopathy, were carefully considered and systematically excluded. There was no history of new drug exposure, and *H. pylori* infection was not detected. Although fragmented red blood cells were present on peripheral smear, serum LDH and indirect bilirubin levels were within normal ranges, haptoglobin was not decreased, and there was no clinical or laboratory evidence of haemolytic anaemia. ADAMTS13 activity was normal, effectively ruling out thrombotic thrombocytopenic purpura. Both direct and indirect Coombs tests were negative, and the absence of preceding gastrointestinal symptoms made haemolytic uremic syndrome unlikely. Furthermore, thrombotic microangiopathy could not account for the systemic oedema, fluid accumulation, organomegaly, and bone marrow fibrosis observed in this case. We therefore excluded thrombotic microangiopathy from the differential diagnosis.

DIC was also considered, but prothrombin time (PT), activated partial thromboplastin time, and fibrinogen levels were all within normal ranges, and the findings indicative of coagulopathy were minimal. Fibrin degradation products and D-dimer levels were only mildly elevated, and the diagnostic criteria for DIC established by the Japanese Association for Acute Medicine and the Japanese Society on Thrombosis and Haemostasis were not met. We therefore ruled out DIC.

Macrophage activation syndrome (MAS) was also considered as a differential diagnosis. However, serum triglyceride and fibrinogen levels were within normal ranges. Bone marrow examination revealed hyperplasia without evidence of pancytopenia, maturation arrest, or haemophagocytosis, which are findings typically associated with MAS. Instead, plasma cell infiltration and mild fibrosis were observed, which were considered to be distinct from MAS. Based on the clinical course and the bone marrow findings, we also excluded MAS.

In addition, this patient presented with hypocomplementaemia, hypoalbuminaemia, and ascites. Noncontrast CT revealed hepatomegaly and splenomegaly. The Child–Pugh classification was B (7 points) based on hypoalbuminaemia and ascites, and there was a predominant elevation in biliary enzymes. These findings raised the possibility of acute liver injury resembling primary biliary cholangitis. However, antimitochondrial antibodies were negative, serum IgM was within the normal range, and there were no reductions in PT%, prolongations in Prothrombin Time and International Normalized Ratio (PT-INR), or elevations in total bilirubin, which are typically associated with acute liver injury. Additionally, AST was only mildly elevated at 47 IU/l, and ALT remained within the normal range, further supporting the exclusion of acute liver injury. In contrast, serum alkaline ALP is often markedly elevated in TAFRO syndrome. In this case, ALP alone was markedly elevated at 895 IU/l, which was considered to be supportive of a diagnosis of TAFRO syndrome.

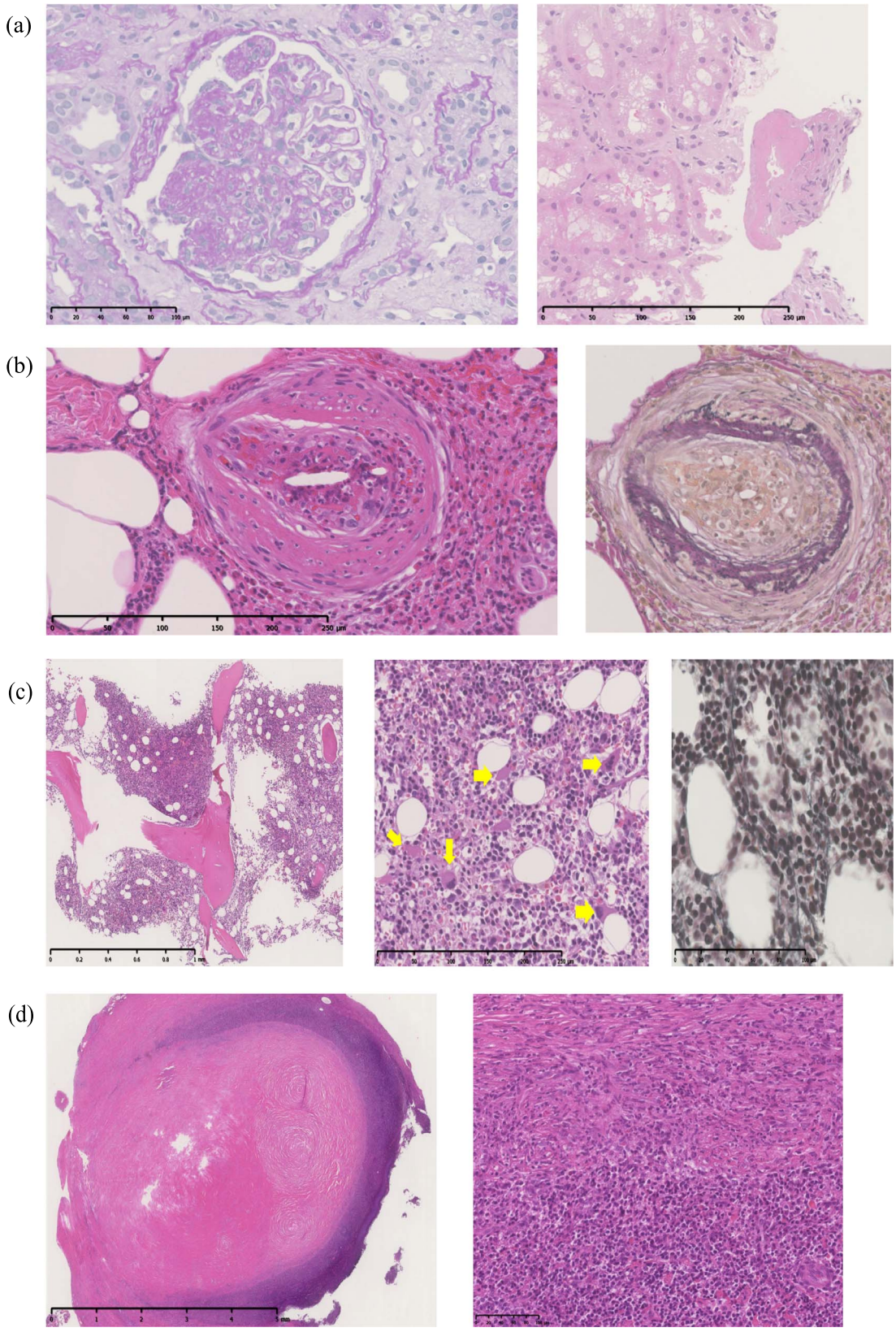

Figure 2. Pathological histopathology of this case. (a–d) Pathological findings of the kidney, the bone marrow, and the lymph node. There were no malignant findings in any of the tissues. Kidney (a): HE staining; there was no glomerular hypertrophy, but mild mesangial cell proliferation and increased mesangial matrix were observed in 80% of the glomeruli. Fibrinoid necrosis of medium-sized vessels was observed, a finding of vasculitis. Immunostaining was negative. Skin (b): HE staining; mixed inflammatory infiltrate of neutrophils and eosinophils. Intimal proliferation with fibrinoid change, resulting in luminal narrowing. Elastica van Gieson stain: intraelastic plate running on part of the injured vessel. Bone marrow (c): HE staining; bone marrow tissue was hyperplastic, and mature megakaryocytes increased to ∼10/HPF. Silver impregnation; mild reticular fibrosis corresponding to MF-1 was observed, a finding of TAFRO syndrome, but which may also occur in autoimmune diseases. Lymph node (d): HE staining; there was no specific finding except for dense fibrosis in the central part of the lymph node.

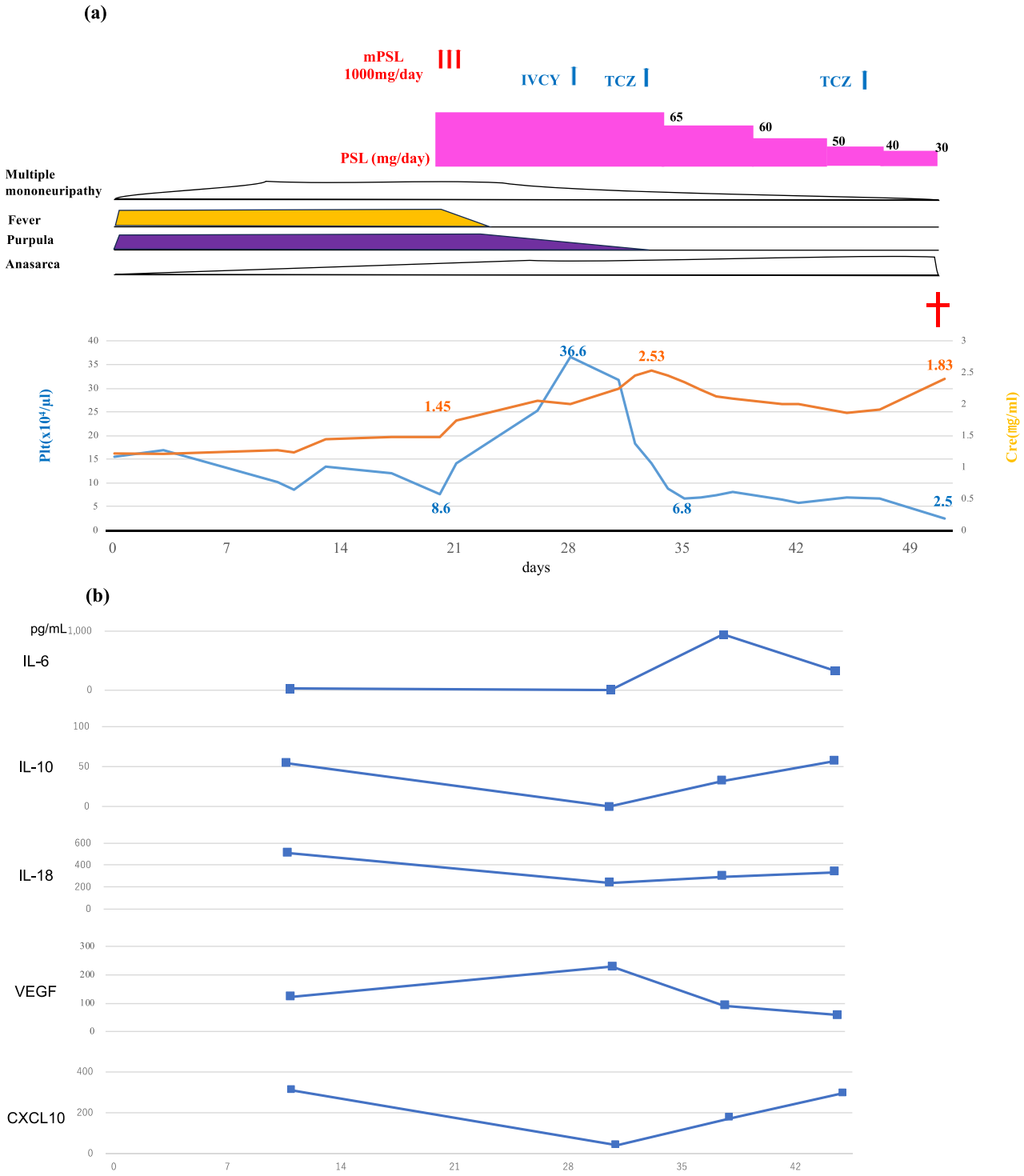

Figure 3. Clinical course of a patient with polyarteritis nodosa with TAFRO syndrome after COVID-19 infection. (a) Clinical course; (b) change of serum cytokines before and after treatment in the patient with polyarteritis nodosa with TAFRO syndrome after COVID-19 infection.

Regarding treatment progress (Figure 3(a)), high-dose glucocorticoid therapy combined with intermittent high-dose intravenous cyclophosphamide (500 mg/kg/2 weeks) was initiated. Platelet count temporarily increased to 366,000/μl, and the vasculitis symptoms of fever, purpura, and neuropathy showed gradual improvement. However, the platelet count decreased again, reaching 68,000/μl on the 35th hospital day. The similarity of the cytokine profile between COVID-19 infection and TAFRO syndrome has been previously reported [13]. This led to later measurement of 10 cytokines (measured by Luminex Discovery Assay, Human Premixed Multi-Analyte Kit, F-RD-LuminexHM-10, R&D Systems) that are confirmed to be elevated in TAFRO syndrome (C-X-C motif chemokine ligand (CXCL)10/IP-10/cytokine responsive gene 2 (CRG-2), IL-18/IL-1F4, IL-6, IL-10, IL-28A/Interferon (IFN)-λ2, Tumor Necrosis Factor

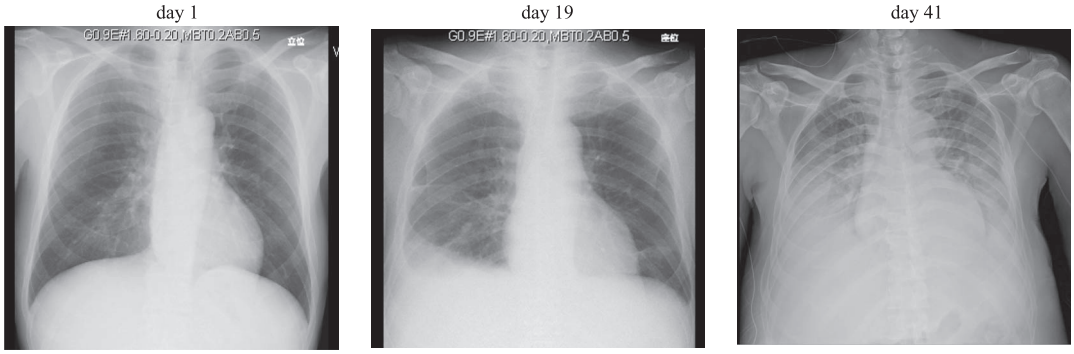

Figure 4. Changes in imaging before and after treatment. Chest radiography on admission and on Days 19 and 41. Lung field permeability tended to worsen.

(TNF)-α, IFN-γ, IFN-α, VEGF, and IL-1β/IL-1F2). Among them, CXCL10/IP-10/CRG-2, IL-10, and IL-18 were elevated (309.65, 54.33, and 511.04 pg/ml) before treatment and temporarily decreased with treatment (40.68 pg/ml, limit of quantitation, 238.26 pg/ml), but they increased again (295.97, 56.99, and 339.55 pg/ml) with the exacerbation of TAFRO symptoms such as thrombocytopenia, renal dysfunction, and increased pleural effusion. IL-28A/IFN-λ2, TNF-α, IFN-γ, IFN-α, and IL-1β/IL-1F2 were measured but below the sensitivity threshold (Figure 3(b)). Renal dysfunction further progressed, and fluid retention worsened over time (Figure 4). Based on a previous report regarding the efficacy of tocilizumab for TAFRO syndrome [14], the patient received two doses of tocilizumab (8 mg/kg every 2 weeks), but disease control was not achieved. On Day 51, the patient developed cytomegalovirus infection and necrotising fasciitis and subsequently died.

Discussion

In this case, polyarteritis nodosa complicated by TAFRO signs developed following COVID-19 infection. COVID-19 has been reported to trigger distinct immune responses and inflammatory patterns from those of conventional viral infections [15]. The clinical features observed in this case may therefore represent a novel disease entity or phenotype caused by COVID-19 that does not fit within existing disease classifications. However, after careful evaluation, if this case were to be classified according to existing disease categories, it would most closely resemble polyarteritis nodosa with TAFRO signs. The 2015 diagnostic criteria for TAFRO syndrome (minor revision 2019) [12] exclude autoimmune diseases because they are considered to be independent conditions. This made it impossible to accurately and definitively diagnose TAFRO syndrome in this case. Conversely, recent reports have documented multiple cases of autoimmune diseases with TAFRO signs [1–7]. In this case, the diagnosis of polyarteritis nodosa was definitive according to clinical symptoms and histopathological findings. However, thrombocytopenia, elevated ALP, and fluid retention cannot be adequately explained by polyarteritis nodosa. We therefore believe polyarteritis nodosa complicated by TAFRO signs is a more reasonable diagnosis in this case, and we suggest there should be further examination into whether TAFRO syndrome and autoimmune diseases are independent conditions.

To the best of our knowledge, there is only one previous report on vasculitis with TAFRO signs [10], making this the second reported case. However, this is thought to be the first reported case of polyarteritis nodosa with TAFRO signs to develop after COVID-19 infection. A relationship between COVID-19 infection and TAFRO syndrome has been suggested, with reports of viral and bacterial infections being related to the onset of TAFRO syndrome [12], severe exacerbation of TAFRO syndrome following COVID-19 infection [16], and development of TAFRO syndrome after SARS-CoV-2 vaccination or infection [8, 9]. The molecular mechanisms of TAFRO syndrome are not fully understood, but the main pathophysiology is thought to involve cytokine storms, with IL-6 and VEGF being particularly important [17, 18]. Other cytokines, such as IL-10 and IP-10, have also been reported to increase [19]. Meanwhile, COVID-19 has been reported to increase serum cytokine levels such as IL-1β, IL-2, IL-6, IL-10, IFN-γ, TNF-α, and IFN-γ IP-10. Cytokines IP-10 [20, 21], Granulocyte-macrophage colony-stimulating factor (GM-CSF), and Monocyte chemoattractant protein-1 (MCP-1) were reportedly correlated with the severity of COVID-19 infection [21]. The similarity of cytokine profiles between COVID-19 and TAFRO syndrome is therefore considered to include increased serum levels of IL-6, IL-10, and IP-10. In this case, serum levels of IL-6 at 22 ng/ml, IL-10 at 54 ng/ml, IL-18 at 511 ng/ml, and IP-10 at 309 ng/ml were elevated before treatment. Although it was difficult to evaluate IL-6 after tocilizumab administration, serum levels of IL-10, IL-18, and IP-10 were correlated with the severity of TAFRO symptoms. There have been reports examining changes in serum IL-10 and IP-10 with disease status in TAFRO syndrome [18]. However, there have been no previous reports on the possibility of serum IL-18, and serum IL-18 levels being related to the pathogenesis of COVID-19 infection and TAFRO syndrome. The number of cases of elevated cytokines in polyarteritis nodosa is small, so there are no previous reports on cytokines, and the association between COVID-19 infection and TAFRO syndrome remains unclear.

Conclusion

Our patient had polyarteritis nodosa with TAFRO signs following COVID-19 infection. The current diagnostic criteria for TAFRO syndrome exclude autoimmune diseases, but cases like this one suggest that neither autoimmune diseases nor TAFRO syndrome alone can fully explain the condition. Further discussion is needed regarding the pathology and diagnostic criteria for TAFRO syndrome. Additionally, there may be a correlation between COVID-19, TAFRO syndrome,

and autoimmune diseases, necessitating further investigation of their pathology, including cytokine profiling.

Acknowledgement

We acknowledge proofreading and editing by Benjamin Phillis, a Board-Certified Editor in the Life Sciences (BELS), at Wakayama Medical University.

Conflict of interest

S.I. has received consulting fees, speaking fees, and honoraria from Ono, Taisho, Janssen, UCB, AstraZeneca, Pfizer, GlaxoSmithKline, Astellas, Asahi Kasei, Teijin, Eli Lilly, Nippon Shinyaku, Daiichi Sankyo, AbbVie, Eisai, Mitsubishi Tanabe, and Chugai. All the other authors declare no conflicts of interest.

Funding

None declared.

Patient consent

Written informed consent for the publication of this report, including cytokine and chemokine analysis, was obtained from the patient's family.

Ethical approval

Cytokine and chemokine examination in sera from this patient was approved by our ethical committee as 'Research on the measurement of autoantibodies and soluble factors and the analysis of cellular immune functions for the early diagnosis, elucidation and establishment of new treatment strategies for systemic autoimmune rheumatic diseases' (#1799). This case report was approved by the ethical committee of our university.

References

1. Grange L, Chalayer E, Boutboul D *et al.* TAFRO syndrome: a severe manifestation of Sjögren's syndrome? A systematic review. *Autoimmun Rev* 2022;**21**:103137.
2. Ohta T, Oda N, Saito K *et al.* A case of repeated TAFRO syndrome-like symptoms and retroperitoneal hemorrhage in a patient with Sjögren syndrome. *Cureus* 2020;**19**;12:12175.
3. Fujimoto S, Kawabata H, Kurose N *et al.* Sjögren's syndrome manifesting as clinicopathological features of TAFRO syndrome. *Medicine (Baltimore)* 2017;**96**:9220.
4. Li ZY, Kim S, Huang S *et al.* Multicentric Castleman disease with TAFRO syndrome and Sjögren's. *Clin Case Rep* 2019;**7**: 2388–92.
5. Tokunaga M, Yamada M, Yoshikawa S *et al.* Systemic lupus erythematosus with marked eosinophilia and clinical features mimicking TAFRO syndrome. *Rinsho Ketsueki* 2018;**59**:688–94. [Japanese]
6. Hasegawa E, Sato H, Wada Y *et al.* Characterization of patients with systemic lupus erythematosus who meet the diagnostic criteria for TAFRO syndrome. *Lupus* 2018;**27**:417–27.
7. Kadoba K, Waki D, Nishimura K *et al.* Development of severe thrombocytopenia with TAFRO syndrome-like features in a patient with rheumatoid arthritis treated with a Janus kinase inhibitor. *Medicine (Baltimore)* 2020;**99**:22793.
8. Yamada M, Sada RM, Kashihara E *et al.* TAFRO syndrome with a fatal clinical course following BNT162b2 mRNA (Pfizer-BioNTech) COVID-19 vaccination: a case report. *J Infect Chemother* 2022;**28**:1008–11.
9. Tane M, Kosako H, Hosoi H *et al.* Severe systemic inflammation mimicking TAFRO syndrome following COVID-19. *Int J Hematol* 2023;**118**:374–80.
10. Maquet J, Bories E, Nogier MB *et al.* TAFRO syndrome and cutaneous necrotizing vasculitis. *Rev Med Interne* 2021;**42**: 134–9.
11. Lightfoot RW Jr, Michel BA, Bloch DA *et al.* The American College of Rheumatology 1990 criteria for the classification of polyarteritis nodosa. *Arthritis Rheum* 1990;**33**:1065–298.
12. Masaki Y, Kawabata H, Takai K *et al.* Japanese TAFRO Syndrome Research Team. 2019 updated diagnostic criteria and disease severity classification for TAFRO syndrome. *Int J Hematol* 2020;**111**: 155–8.
13. Tane M, Kosako H, Sonoki T. TAFRO syndrome and COVID-19. *Biomedicines* 2024;**12**:1287.
14. Miura K, Nishimaki-Watanabe H, Takahashi H *et al.* TAFRO syndrome: guidance for managing patients presenting thrombocytopenia, anasarca, fever, reticulin fibrosis, renal insufficiency, and organomegaly. *Biomedicines* 2024;**12**(6):1277.
15. Ramos-Casals M, Brito-Zerón P, Mariette X. Systemic and organ-specific immune-related manifestations of COVID-19. *Nat Rev Rheumatol* 2021;**17**(6):315–32.
16. Oshima K, Kanamori H, Takei K *et al.* Severe coronavirus disease 2019 in a patient with TAFRO syndrome: a case report. *Clin Infect Pract* 2022;**16**:100158.
17. Chen T, Feng C, Zhang X *et al.* TAFRO syndrome: a disease that known is half cured. *Hematol Oncol* 2023;**41**:310–22.
18. Iwaki N, Fajgenbaum DC, Nabel CS *et al.* Clinicopathologic analysis of TAFRO syndrome demonstrates a distinct subtype of HHV-8-negative multicentric Castleman disease. *Am J Hematol* 2016;**91**:220–6.
19. Iwaki N, Gion Y, Kondo E *et al.* Elevated serum interferon γ-induced protein 10 kDa is associated with TAFRO syndrome. *Sci Rep* 2017;**7**:42316.
20. Hawerkamp HC, Dyer AH, Patil ND *et al.* Characterisation of the pro-inflammatory cytokine signature in severe COVID-19. *Front Immunol* 2023;**14**:1170012.
21. Zanza C, Romenskaya T, Manetti AC *et al.* Cytokine storm in COVID-19: immunopathogenesis and therapy. *Medicina (Kaunas)* 2022;**58**:144.

Modern Rheumatology Case Reports, 2026, Vol. 10, Issue 1, rxaf075
doi:10.1093/mrcr/rxaf075
Case Report