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CASE REPORT

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Concurrent myocardial infarction in the setting of thrombotic thrombocytopenic purpura secondary to systemic lupus erythematosus: A case report

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Key Clinical Message

In acute thrombotic thrombocytopenic purpura (TTP), apart from urgent treatment, assessing the patient's medical history, especially conditions like systemic lupus erythematosus that could trigger TTP, is crucial. Rarely, TTP patients may experience cardiac conditions as severe as a myocardial infarction.

Abstract

A 45-year-old woman manifested severe and acute thrombotic thrombocytopenic purpura (TTP) of unknown origin. The patient's symptoms, the laboratory data, the detection of the reduction in ADAMTS13 activity, and the presence of schistocytes on the peripheral smear confirmed the diagnosis. The patient was then planned for therapeutic plasma exchange (TPE). Prior to the scheduled TPE, she suddenly experienced extreme shortness of breath and chest pain. An electrocardiogram was obtained immediately after reporting signs of an inferior myocardial infarction. Further examinations to acquire information about the patient's underlying medical conditions in order to study the secondary causes of TTP, combined with the results of the laboratory tests, resulted in the patient being diagnosed with systemic lupus erythematosus.

KEYWORDS

case report, myocardial infarction, systemic lupus erythematosus, thrombotic thrombocytopenic purpura

1 | INTRODUCTION

Thrombotic thrombocytopenic purpura (TTP) is a rare thrombotic microangiopathy that manifests as microangiopathic hemolytic anemia, severe thrombocytopenia, and ischemia damage to important organs caused by platelet-rich thrombi produced in the microvasculature. TTP is caused by a congenital or acquired lack of the von Willebrand factor-cleaving protease, ADAMTS13 (A Disintegrin and Metalloproteinase with a Thrombospondin type 1 motif, member 13). Anti-ADAMTS13 autoantibodies are the primary cause of ADAMTS13 deficiency, which can be idiopathic or associated with various pathological processes such as neoplasias, infections, and autoimmune

disorders like systemic lupus erythematosus (SLE).[1] If left untreated, TTP is a dangerous illness with a high death rate. The prognosis has significantly improved with therapeutic plasma exchange (TPE), and mortality has dropped from 85%–100% to 10%–30%.[2,3] TPE is a technique that involves separating blood cells from plasma. When elements in the plasma are responsible for the pathophysiology of the disease, this approach is employed. This method is used as a treatment method for various diseases, but the reason for using TPE in a patient suffering from acquired TTP is to remove anti-ADAMTS13 autoantibodies. Relapse happens in 40% of patients, and there are cases that are resistant to treatment. Various immunosuppressive medications, including rituximab (RTX), have been used in these cases.[4] TTP is relatively uncommon in SLE patients. It is unknown how often TTP occurs overall in SLE patients; however, it has been estimated to be as low as 0.5%.[5] Instances of simultaneous TTP and SLE are often discussed in the medical literature in patients who have substantial renal dysfunction and lupus activity.[6] Here we present a clinical case of a woman who developed myocardial infarction[7] in the context of TTP. After further investigation, undiagnosed SLE disease was suggested as the cause of TTP.

2 | CASE PRESENTATION

2.1 | Case history/examination

A 45-year-old lady presented to the emergency room with a slew of symptoms, including lethargy, reduced urine production, and petechiae and purpura skin manifestations primarily affecting her lower limbs. The patient also exhibited mild icterus. She had never experienced a comparable incident before, and there was no family history of cancer or bleeding disorders. She did not take any drugs and had no known allergies. Her vital signs at the time of the visit were as follows: a pulse rate of 115, a respiratory rate of 19, an oxygen saturation of 96%, a temperature of 37.8°C, and a blood pressure of 135/80.

2.2 | Methods

2.2.1 | Differential diagnosis, investigations, and treatment

Laboratory investigations of the patient revealed anemia on a complete blood count (CBC) with a hemoglobin (Hb) of 4.1 g/dL, leukopenia with a white blood cell (WBC) count of 2600×10^6/L, thrombocytopenia with a platelet count of $85{,}000 \times 10^6$/L, elevated lactate dehydrogenase level at 707 U/L, and the presence of schistocytes on the peripheral smear, as well as an increase in bilirubin levels, with a total bilirubin level of 3.9 mg/dL and a direct bilirubin level of 0.6 mg/dL. With a highly probable diagnosis of TTP, which was later corroborated by the reduction in ADAMTS13 activity (activity level, 2.7%), she was planned for TPE. Notably, the chosen therapeutic strategy centered on TPE, and immunosuppressive therapy was not included in the treatment plan for TTP. In light of the potential presence of Catastrophic Antiphospholipid Syndrome (CAPS) as another differential diagnosis, initiation of therapy involved 500 mg methylprednisolone sodium succinate daily pulse therapy. In response to low Hb levels, three packed red blood cells (RBCs) were administered to the patient. Prior to the scheduled TPE, she suddenly experienced extreme shortness of breath and chest pain. In addition to the electrocardiogram (ECG) findings (Figure 1), confirming signs of an inferior myocardial infarction, the diagnosis was further supported by elevated high-sensitive Troponin I levels, measuring 1208 ng/L (normal range: <20 ng/L). Although cardiac catheterization was not performed, the notable Troponin elevation adds valuable corroboration to the MI diagnosis. The patient received prompt and comprehensive treatment, including

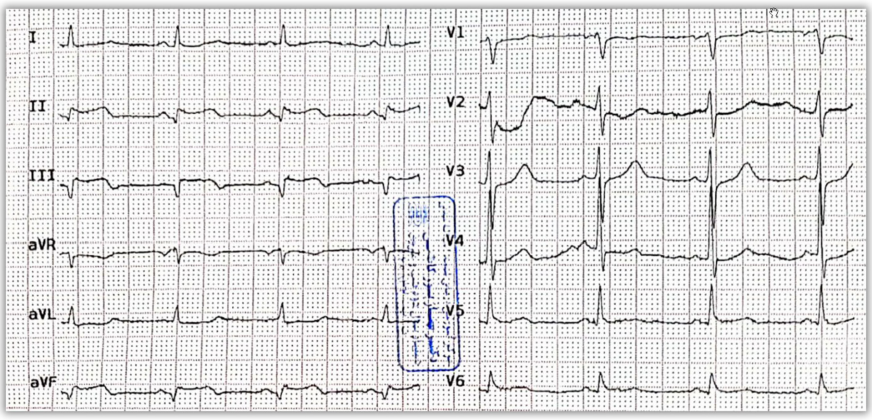

FIGURE 1 An electrocardiogram taken from the patient shortly after the onset of cardiac symptoms, indicating the presence of an inferior MI.

aspirin (a loading dose of 320 mg aspirin, followed by a daily maintenance dose of 75 mg), clopidogrel (a loading dose of 300 mg clopidogrel, followed by a daily maintenance dose of 75 mg), apixaban (a dosage of 2.5 mg every 12 h), and Nitroglycerin (0.4 mg administered sublingually every 5 min for chest pain relief; in this case, only three doses were needed to alleviate the pain). Furthermore, the administration of pulse corticosteroid therapy was discontinued. It is worth noting that, due to anemia and strong clinical suspicion of TTP, this patient did not receive reteplase treatment.

Due to the presence of the patient's companions, further attempts were made to gather a full medical history once the patient was stabilized and the emergency situation was resolved. The patient's companions provided important information that clarified the patient's recent hospitalization due to a hematologic problem. After checking the patient's medical data to determine the reason for the patient's admission, it was discovered that 3 weeks before, the patient had presented with anemia on a CBC test, revealing a WBC count of 5.6×10^3/μL (normal range: 4–10×10^3/μL), red blood cell count of 3.63×10^6/μL (normal range: 4.1–5.1×10^6/μL), Hb level of 8.8 g/dL (normal range: 12–15 g/dL), hematocrit (HCT) of 26.8% (normal range: 35–45%), mean corpuscular volume (MCV) of 73.8 fL (normal range: 80–100 fL), mean corpuscular hemoglobin (MCH) of 24.2 pg (normal range: 27–33 pg), mean corpuscular hemoglobin concentration (MCHC) of 32.8 g/dL (normal range: 31–36 g/dL), and a platelet count of 168×10^3/μL (normal range: 150–450×10^3/μL). Additionally, during the prior hospitalization, the patient underwent both upper and lower endoscopic examinations. Negative colonoscopy and endoscopy results were determined, and gastrointestinal endoscopy evaluations did not result in a diagnosis of any pathologic findings. In current hospitalization, pancytopenia on a CBC test ordered as a part of a normal checkup was discovered. The patient was then referred to a hematologist for additional examination and care. The patient underwent a bone marrow aspiration procedure, in which the results predominantly showed erythroid hyperplastic marrow. In addition, during the current hospitalization, serologic tests for autoimmune antibodies of rheumatologic diseases, especially SLE, were asked. The results of laboratory tests were as follows: total complement activity (CH50), 62 U (70–150); complement C3 protein, 0.3 mg/dL (0.9–1.8 mg/dL); complement C4 protein, 0.08 mg/dL (0.1–0.4 mg/dL); anti-La/SSB antibodies, 1.9 U/mL (<12 U/mL); anti-SSA/Ro, 283 U/mL (<18 U/mL); anti Centromere antibodies (CREST), negative; anti SCL-70 antibodies, 0.1 AU/ML (<3.2 AU/ML); anti-beta-2-Glycoprotein I IgG, 59 U/mL (<10 U/mL); anti-beta-2-Glycoprotein I IgM, 296 IU/mL (<10 IU/mL); lupus anticoagulant 88.8 s (31–47); anti cardiolipin IgG, 172 U/mL (<18 U/mL); anti cardiolipin IgM, 323 U/mL (<18 U/mL); antiphospholipid antibodies IgG, 72.4 U/mL (<10 U/mL); antiphospholipid antibodies IgM, 383 U/mL (<10 U/mL); antinuclear antibody, 4.1 Index (<0.8 Index); anti-double-stranded deoxyribonucleic acid antibody (anti-dsDNA), 783.6 IU/mL (<100 IU/mL); anti–cyclic citrullinated peptide (anti–CCP) antibody, 4.5 IU/mL (<12 IU/mL); rheumatoid factor (RF), negative.

3 | CONCLUSION

3.1 | Outcome and follow-up

According to the SLICC criteria for SLE classification, the diagnosis of SLE was confirmed, with clinical manifestations (leukopenia, hemolytic anemia, and thrombocytopenia) and immunologic manifestations (high ANA level, elevated Anti-dsDNA level, positive antiphospholipid tests, and low serum complement level). Therefore, the manifestation of TTP was probably indicated to be associated with SLE. The patient was finally discharged with a good general condition following hospitalization for 15 days. She regularly visits her rheumatologist for follow-up appointments.

TTP is one of the medical emergencies that necessitates rapid action, such as TPE. In addition to determining the best therapy and implementing it as quickly as possible, it is critical to review the patient's medical history to determine the existence or absence of underlying factors contributing to the onset of TTP. Investigation of the patient's underlying issues was so beneficial in our case study that it led to a diagnosis of overlooked SLE. Furthermore, while it is rare, it is critical to pay attention to the signs of cardiac involvement in TTP patients since this disease can cause serious cardiac conditions such as MI.

4 | DISCUSSION

TTP is an infrequent and lethal hematologic disorder described by the clinical pentad of microangiopathic hemolytic anemia (MAHA), thrombocytopenia, renal failure, fever, and neurologic abnormalities. TTP may coexist with SLE.[8,9] TTP was observed in sporadic cases of SLE, but there is not much clarity as to why these two conditions are associated.[7,10] TTP diagnosis in SLE patients can be confusing since both disorders have numerous overlapping symptoms such as anemia, thrombocytopenia, central nervous system symptoms, renal insufficiency, and fever. Early diagnosis of TTP and monitoring of the physical conditions may improve the prognosis. Therefore, it is critical to diagnose TTP in patients with SLE since there are divergent treatment options for them. The most efficient treatment for TTP is currently TPE,

although the efficacy of this therapy in SLE has been disputed.[8,10] Fragmentation of RBCs, which is rare in SLE, is an important observation for the determination of TTP. Thus, we were able to diagnose TTP with the recognition of fragmented RBCs, or schistocytes.[10] Furthermore, to discriminate the occurrence of TTP from disease exacerbation in SLE patients with MAHA, we need to measure ADAMTS13 activity and its inhibitor. Recent studies have shown that one of the contributing factors to TTP may be a reduced functional effect of ADAMTS13 which is caused by either inhibitory antibodies or a mutation in ADAMTS13 gene.[8] Additionally, the coexistence of acute MI along with TTP and SLE is a rare condition that occurred in our patient. The myocardial injury could lead to death in patients with TTP. Though the heart is one of the most frequently involved organs in autopsies on TTP patients, clinical evidence regarding the involvement of cardiac tissue is surprisingly limited[11,12]; however, several studies suggest that the pathogenesis of cardiac manifestations is based on platelet microthrombi discovered in the heart and many small infarctions affecting the microvascular circulation.[12,13]

AUTHOR CONTRIBUTIONS

Saleh Azadbakht: Conceptualization; project administration; supervision; validation. **Bardia Amidi:** Conceptualization; data curation; investigation; writing – original draft; writing – review and editing. **Narges Naderi:** Conceptualization; data curation; investigation; writing – original draft; writing – review and editing. **Anwar Sharifaskari:** Conceptualization; supervision; validation. **Mahtab Hatami:** Conceptualization; supervision; validation.

FUNDING INFORMATION

None.

CONFLICT OF INTEREST STATEMENT

The authors declare that there is no conflict of interest.

DATA AVAILABILITY STATEMENT

Data sharing is not applicable; no new data are generated.

ETHICS STATEMENT

This case report did not require ethical approval from ethics committee.

CONSENT

We obtained written informed consent from the patient to publish this case report in accordance with the journal's patient consent policy.

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REFERENCES

1. Knöbl P. Thrombotic thrombocytopenic purpura. *Memory*. 2018;11(3):220-226.
2. George JN. How I treat patients with thrombotic thrombocytopenic purpura: 2010. *Blood*. 2010;116(20):4060-4069.
3. Peyvandi F, Palla R, Lotta LA. Pathogenesis and treatment of acquired idiopathic thrombotic thrombocytopenic purpura. *Haematologica*. 2010;95(9):1444-1447.
4. Caramazza D, Quintini G, Abbene I, et al. Relapsing or refractory idiopathic thrombotic thrombocytopenic purpura-hemolytic uremic syndrome: the role of rituximab. *Transfusion*. 2010;50(12):2753-2760.
5. George P, Das J, Pawar B, Kakkar N. Thrombotic thrombocytopenic purpura and systemic lupus erythematosus: successful management of a rare presentation. *Indian J Crit Care Med*. 2008;12(3):128-131.
6. Letchumanan P, Ng HJ, Lee LH, Thumboo J. A comparison of thrombotic thrombocytopenic purpura in an inception cohort of patients with and without systemic lupus erythematosus. *Rheumatology (Oxford)*. 2009;48(4):399-403.
7. Brunner HI, Freedman M, Silverman ED. Close relationship between systemic lupus erythematosus and thrombotic thrombocytopenic purpura in childhood. *Arthritis Rheum*. 1999;42(11):2346-2355.
8. Kwok S, Ju J, Cho C, Kim H, Park SJL. Thrombotic thrombocytopenic purpura in systemic lupus erythematosus: risk factors and clinical outcome: a single centre study. *Lupus*. 2009;18(1):16-21.
9. Vasoo S, Thumboo J, Fong KJL. Thrombotic thrombocytopenic purpura in systemic lupus erythematosus: disease activity and the use of cytotoxic drugs. *Lupus*. 2002;11(7):443-450.
10. Sakarcan A, Stallworth J. Systemic lupus erythematosus and thrombotic thrombocytopenic purpura: a case and review. *Pediatr Nephrol*. 2001;16:672-674.
11. Hawkins BM, Abu-Fadel M, Vesely SK, George JN. Clinical cardiac involvement in thrombotic thrombocytopenic purpura: a systematic review. *Transfusion*. 2008;48(2):382-392.
12. Tasaki T, Yamada S, Nabeshima A, et al. An autopsy case of myocardial infarction due to idiopathic thrombotic thrombocytopenic purpura. *Diagn Pathol*. 2015;10:1-6.
13. Wahla AS, Ruiz J, Noureddine N, Upadhya B, Sane DC, Owen J. Myocardial infarction in thrombotic thrombocytopenic purpura: a single-center experience and literature review. *Eur J Haematol*. 2008;81(4):311-316.

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