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Letter to Editor

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Thrombotic thrombocytopenic purpura in a HIV-positive patient: A hematological emergency

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Dear editor,

Thrombotic thrombocytopenic purpura (TTP) is an acute non-immune acquired microangiopathic hemolytic anaemia. It is characterized by the classic pentad features of fever, thrombocytopenia, hemolytic anemia, neurological symptoms and renal dysfunction, although all five clinical features are seen in less than 10% of patients[1]. The diagnosis of TTP is frequently neglected since thrombocytopenia is a common hematological finding in patients with TTP and human immunodeficiency virus (HIV) infection. The peripheral blood film composed of schistocytes (fragmented red blood cells) plays a crucial role in diagnosing TTP associated with a chronic infection such as HIV.

Here, we present a case of a 45-year-old female previously diagnosed with HIV. All appropriate consent forms were obtained from the patient. In the forms, the patient had given consent for her images and other clinical information to be reported in the journal. She presented to the emergency department with 10 d of fever, headache, and 3 d of vaginal bleeding. She also had an episode of tonic-clonic seizure that was generalized and lasted about 1 min and urinary incontinence. Her complete blood showed severe thrombocytopenia of 10 000/mm^3 (normal: 150 000-410 000/mm^3), anemia of 5.3 g/dL (normal: 12-15 g/dL) and a normal total leukocyte count of 8 600/mm^3 (normal: 4 000-10 000/mm^3). A peripheral blood smear examination showed schistocytes (helmet-shaped and triangular erythrocytes) (Figure 1).

The patient's reticulocyte count was 20% (normal: 0.5%-2.0%) and her direct and indirect Coombs tests were negative. Serum lactate dehydrogenase level was significantly high at 1 227 U/L (normal: 120-246 U/L). The absolute CD lymphocyte count was 211 cells/L (normal: 356-1 335 cells/L). Brain CT ruled out intracranial hemorrhage. Diagnosis of TTP was made in the HIV-positive patient based on clinical and hematologic findings. Plasmapheresis was performed with 0.9% normal saline, human albumin and cryofree plasma. In the following days, two more cycles of plasmapheresis were performed (three cycles in total). Her platelet count gradually improved after the second session and lactate dehydrogenase level began to decrease after the third session of plasmapheresis. She was discharged after the platelet count returned to normal.

The molecular mechanism related to TTP is the deficiency of Von Willebrand factor-cleaving protease. The cleaving protease is required to break down ultra-large Von Willebrand multimers and the cleavage is carried out by ADAMTS13, a disintegrin and

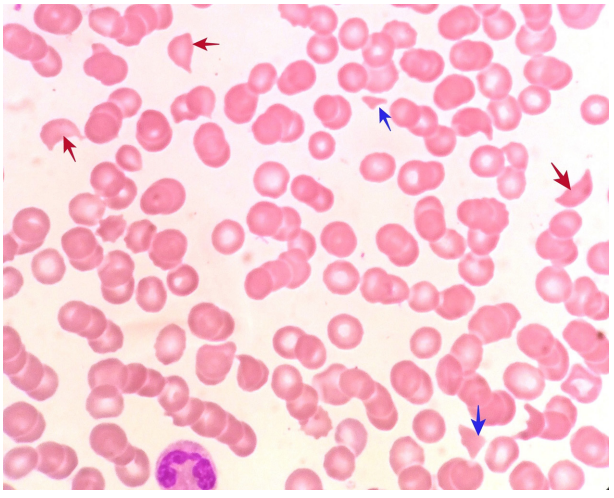

Figure 1. Peripheral smear (Leishman stain 40×) schistocytes; helmet-shaped (red arrows) and triangular red blood cells (blue arrows).

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metalloproteinase with a thrombospondin type 1 motif, member 13[2]. The patients ADAMTS13 activity was <3% (normal: 68%-163% activity) and ADAMTS13 inhibitor was 0.9 BEU (normal: <0.4 BEU). Jokela *et al*. reported the first case of HIV-related TTP in 1987, since then few cases have been reported in the literature[3]. The hematological hallmark of TTP is the development of helmet and triangular fragmented erythrocytes (schistocytes) in a peripheral blood sample. The International Council for Standardization in Hematology Schistocyte Working Group has agreed that a percentage of schistocytes greater than 1% in a peripheral blood smear in adults is a robust cytomorphological indication for the diagnosis of microangiopathic hemolytic anaemia[4]. Early plasmapheresis therapy holds a promising role in the treatment regardless of the etiology of TTP. Outcome and survival in HIV-related TTP are related to the stage of HIV infection and the severity of immunosuppression at the time of diagnosis and CD4 cell counts[5]. In our case, the patient revealed four clinical features of the classic pentad of TTP. The presence of schistocytes served as a clue to initiate plasmapheresis early and thus lead to successful treatment.

In conclusion, TTP is a hematological emergency and its association with HIV poses a challenge for the physician to rule out HIV-related cytopenia and immediate discussion between physicians and laboratory physicians is required to diagnose TTP.

Conflict of interest statement

The authors report no conflict of interest.

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Authors' contributions

MABM designed the study. MABM and RRDS developed the concepts and prepared the manuscript. MABM and RH edited the manuscript. All authors performed literature search and reviewed the manuscript.

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