

Images in Clinical Hematology

Assessing treatment response in thrombotic thrombocytopenic purpura: Beyond the platelet count



Cilomar Martins de Oliveira Filho ^{a,*}, Ibidunni Bode-Sojobi ^b,
Barbara D. Lam ^b, Stephanie Conrad ^b, Jonathan Berry ^b, Brian J. Carney ^b

^a Mass General Brigham, Salem Hospital, Salem, Massachusetts, USA

^b Beth Israel Deaconess Medical Center, Boston, USA

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A 50-year-old male with systemic lupus erythematosus presented with fever, abdominal pain, and diarrhea. Hemoglobin was 10 g/dL, near patient's baseline, and there was new thrombocytopenia with a platelet count $109 \times 10^9/L$. The following day, platelets dropped to $24 \times 10^9/L$. Hemolysis parameters were unremarkable. Treatment for immune thrombocytopenic purpura with IV dexamethasone was started but there was no improvement in the platelet count. On hospital Day 7, he developed a seizure, and was intubated. He evolved with shock and renal failure requiring dialysis. Lactate dehydrogenase rose to 1054 U/L and haptoglobin became undetectable. A peripheral blood

smear revealed a large population of schistocytes. The PLASMIC score was 6.¹ ADAMTS13 assay was done, the patient received fresh frozen plasma (FFP) and was transferred to a tertiary center for daily plasmapheresis with full FFP replacement. The platelet counts initially rose, then remained at around $50 \times 10^9/L$ on subsequent days. Notably, hemolysis parameters rapidly normalized. The population of schistocytes steadily decreased. Antiphospholipid antibodies and enterohemorrhagic *E. coli* tests were negative. On Day 5 of plasmapheresis, ADAMTS13 activity was undetectable, confirming a diagnosis of acquired thrombotic thrombocytopenic purpura. By Day 11 on

* Corresponding author.

E-mail address: cmartins5@mgb.org

(C. Martins de Oliveira Filho).

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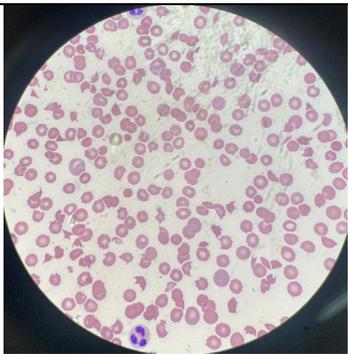
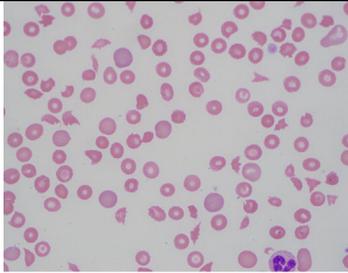
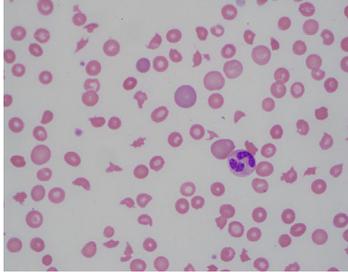
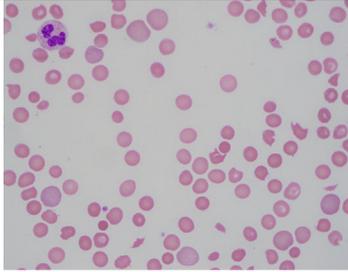
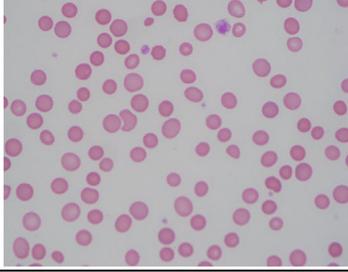
	Peripheral blood smear (x50)		
Day -1		Hemoglobin (g/dL)	7.7
		MCV (fL)	74
		Platelet count (x 10 ⁹ /L)	13
		Haptoglobin (mg/dL)	<14
		LDH (U/L)	1054
		Reticulocyte (M/μL)	0.13
Day +1		Hemoglobin (g/dL)	7.4
		MCV (fL)	73
		Platelet count (x 10 ⁹ /L)	51
		Haptoglobin (mg/dL)	<10
		LDH (U/L)	5640
		Reticulocyte (M/μL)	0.11
Day +3		Hemoglobin (g/dL)	6.8
		MCV (fL)	78
		Platelet count (x 10 ⁹ /L)	58
		Haptoglobin (mg/dL)	127
		LDH (U/L)	476
		Reticulocyte (M/μL)	0.18
Day +5		Hemoglobin (g/dL)	7.9
		MCV (fL)	85
		Platelet count (x 10 ⁹ /L)	64
		Haptoglobin (mg/dL)	150
		LDH (U/L)	177
		Reticulocyte (M/μL)	N/A
Day +8		Hemoglobin (g/dL)	6.8
		MCV (fL)	93
		Platelet count (x 10 ⁹ /L)	119
		Haptoglobin (mg/dL)	108
		LDH (U/L)	162
		Reticulocyte (M/μL)	0.35

Figure 1 – Days on plasmapheresis and changes in blood smears (x50), blood counts, and hemolysis parameters.

plasmapheresis the patient improved consistently and was extubated (Figure 1).

Conflicts of interest

None.

REFERENCE

1. Bendapudi PK, Hurwitz S, Fry A, et al. Derivation and external validation of the PLASMIC score for rapid assessment of adults with thrombotic microangiopathies: a cohort study. *Lancet Haematol.* 2017;4(4):e157–64. [https://doi.org/10.1016/S2352-3026\(17\)30026-1](https://doi.org/10.1016/S2352-3026(17)30026-1).