

THE CLINICAL PICTURE

SUPAKANYA WONGRAKPANICH, MD

Department of Medicine, Einstein Medical Center,
Philadelphia, PA

MOIZ SALAHUDDIN, MD

Department of Medicine, Einstein
Medical Center, Philadelphia, PA

PRIYANKA MITTAR, DO

Department of Hematology and Oncology,
Einstein Medical Center, Philadelphia, PA

An abnormal peripheral blood smear and altered mental status

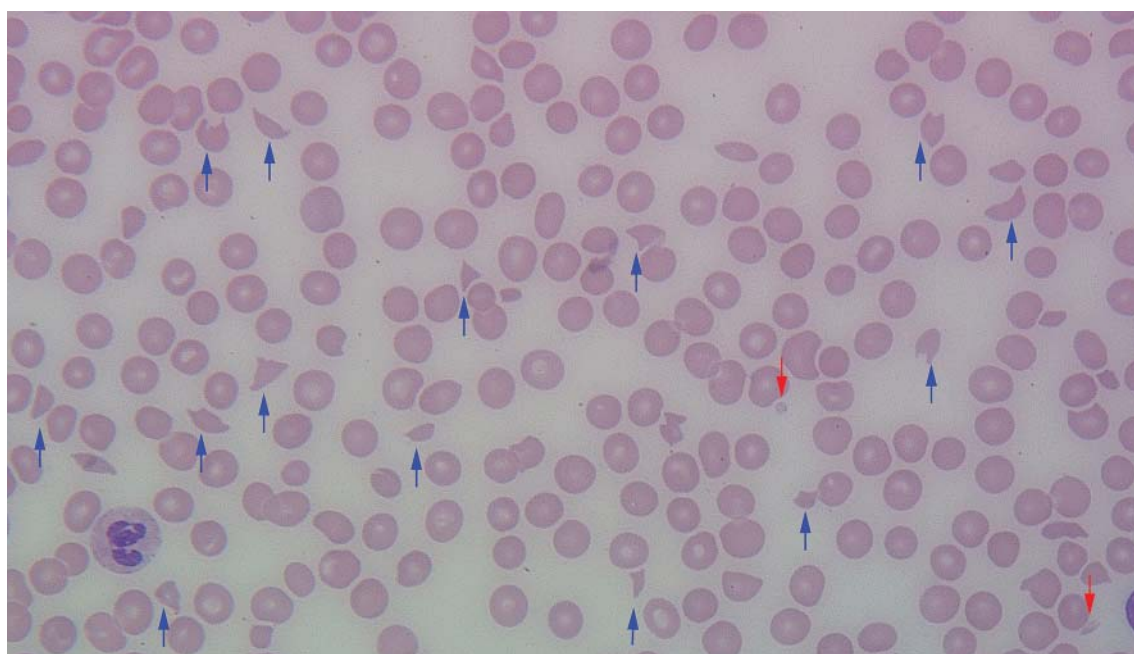


FIGURE 1. A peripheral blood smear showed abundant schistocytes (blue arrows) and a virtual absence of platelets, ie, one or two per field (red arrows) (Wright-Giemsa stain, $\times 600$).

Plasma exchange has greatly increased the survival rate in thrombotic thrombocytopenic purpura

A 72-YEAR-OLD WOMAN with type 2 diabetes mellitus, hypertension, and atrial fibrillation on anticoagulation was brought to the emergency department by her husband after 1 day of altered mental status with acute onset. Her husband reported that she had been minimally arousable, and the physical examination revealed that she was stuporous and withdrew extremities only from noxious stimuli.

Results of initial laboratory tests revealed a creatinine level of 2.4 mg/dL (reference range 0.7–1.4), hemoglobin 12.1 g/dL (12–16), platelet count $16 \times 10^9/L$ (150–400), white blood cell count of $7.7 \times 10^9/L$ (3.7–11), and

international normalized ratio of 2.1. A peripheral blood smear is shown in **Figure 1**.

Computed tomography showed evidence of chronic small vascular ischemia. Magnetic resonance imaging of the brain showed numerous foci of restricted diffusion within the supratentorial and infratentorial areas, suggesting microembolic phenomena.

The peripheral blood smear was compatible with microangiopathic hemolytic anemia, which can occur in thrombotic thrombocytopenic purpura (TTP), hemolytic uremic syndrome, malignant hypertension, scleroderma, antiphospholipid antibody syndrome, systemic lupus erythematosus, eclampsia, renal allograft rejection, hematopoietic stem cell transplant, and severe sepsis.^{1,2}

doi:10.3949/ccjm.83a.15152

In addition to hemolytic anemia, the patient also had neurologic abnormalities, renal involvement, and thrombocytopenia. The hemolytic anemia and thrombocytopenia were sufficient to raise our suspicion of TTP and to consider initiation of plasma exchange. Only 5% of patients with TTP demonstrate the classic pentad of clinical features,¹ ie, thrombocytopenia, microangiopathic hemolytic anemia, fluctuating neurologic signs, renal impairment, and fever.

In 1991, when plasma exchange was introduced for TTP, the survival rate of patients increased from 10% to 78%.^{1,3} Thus, the diagnosis of TTP is an urgent indication for plasma exchange. We normally do plasma exchange daily until the platelet levels improve.

Our patient received methylprednisone

125 mg intravenously every 12 hours and plasma exchange daily. After three cycles of plasma exchange, she regained normal consciousness, and her platelet count had increased to $20.5 \times 10^9/L$ on the day of discharge from our hospital.

TTP is a life-threatening hematologic disorder. Evidence of microangiopathic hemolytic anemia on a peripheral blood smear is vital to the suspicion of TTP. The diagnosis should be confirmed by ADAMTS13 testing, which should show decreased activity (< 10%) or increased inhibition, or both. Rapid management with plasma exchange and steroids can lead to a satisfactory outcome. ■

ACKNOWLEDGMENT: We are particularly grateful to Dr. Vivian Arguello (Director of Flow Cytometry, Department of Pathology, Einstein Medical Center, Philadelphia) for her kind support with the blood smear image.

REFERENCES

1. George JN. How I treat patients with thrombotic thrombocytopenic purpura: 2010. *Blood* 2010; 116:4060–4069.
2. Sadler JE. Von Willebrand factor, ADAMTS13, and thrombotic thrombocytopenic purpura. *Blood* 2008; 112:11–18.
3. Rock GA, Shumak KH, Buskard NA, et al. Comparison of plasma exchange with plasma infusion in the treatment

of thrombotic thrombocytopenic purpura. *N Engl J Med* 1991; 325:393–397.

ADDRESS: Supakanya Wongrakpanich, MD, Department of Medicine, Albert Einstein Medical Center, 5501 Old York Road, Philadelphia, PA 19141; WongrakS@einstein.edu