If you’re preparing for the United States Medical Licensing Examination® (USMLE®) Step 2 exam, you might want to know which questions are most often missed by test-prep takers. Check out this example from Kaplan Medical, and read an expert explanation of the answer. Also check out all posts in this series.

This month’s stumper

A 40-year-old woman is admitted to the hospital because of fever, headache, confusion and jaundice for one week. She underwent hysterectomy two months ago and began estrogen replacement therapy with ethinyl estradiol and a progestin. On admission, her temperature is 38.7°C (102°F), blood pressure is 140/90 mm Hg, pulse is 98/min, and respirations are 20/min. She is disoriented to time and place. Physical examination reveals jaundiced sclerae and skin, purpura on the trunk and bleeding gums. A stool guaiac test is positive for occult blood. Blood and urine cultures are negative, but urinalysis reveals hemoglobinuria. Blood studies show:

- Hematocrit 28%
- Red blood cells 2.5 million/mm3
- Leukocytes 10,000/mm3
- Platelets 15,000/mm3
- BUN 40 mg/dL
- Creatinine 2.8 mg/dL
- LDH 800 U/L
Bilirubin (Total)  4.0 mg/dL
Bilirubin (Direct)  0.8 mg/dL

Coagulation tests are within normal limits; fibrin-split products and Coombs test are negative. A peripheral blood smear shows schistocytes, helmet cells and tri-angle cells.

Which of the following is the most likely diagnosis?

A. Disseminated intravascular coagulation (DIC).
B. Evans syndrome.
C. Hemolytic-uremic syndrome (HUS).
D. Idiopathic (autoimmune) thrombocytopenic purpura (ITP).
E. Malignant hypertension.
F. Thrombotic thrombocytopenic purpura (TTP).

The correct answer is F.

Kaplan Medical explains why

The key data to make a correct diagnosis include the following: severe thrombocytopenia, which results in a bleeding diathesis; elevated indirect bilirubin and high LDH with schistocytes in the blood.
smear, indicating microangiopathic hemolytic anemia; renal dysfunction (high creatinine); and neurologic and systemic symptoms (headache, confusion and fever).

Negative findings important to rule out similar conditions include a negative Coombs test and absence of fibrin split products. TTP is a disorder of unclear pathogenesis, perhaps related to circulating platelet-agglutinating factors. It presents with a characteristic combination of microangiopathic hemolytic anemia, fever without infection, neurologic symptoms, bleeding diathesis secondary to thrombocytopenia, and renal impairment. This condition may be precipitated by pregnancy or use of estrogens.

**Why the other answers are wrong**

**Choice A:** Disseminated intravascular coagulation can be differentiated from TTP because of abnormal coagulation tests. In DIC, microangiopathic hemolysis is also present, but prothrombin time (PT) is prolonged, fibrinogen levels are reduced, and fibrin split products are elevated.

**Choice B:** Evans syndrome refers to coexistence of autoimmune hemolytic anemia (positive Coombs test), and autoimmune thrombocytopenic purpura.

**Choice C:** Hemolytic-uremic syndrome is not significantly different from TTP. The two conditions, in fact, are considered manifestations of the same pathogenetic spectrum. However, the vascular bed of the CNS is not involved in HUS; thus, mental status changes are not part of the clinical picture.

**Choice D:** Idiopathic (autoimmune) thrombocytopenic purpura is an immune disorder caused by autoantibodies to platelet antigens. Systemic illness is not present in ITP, which is characterized by isolated thrombocytopenia without other hematologic abnormalities. Ten percent of cases will manifest in association with autoimmune hemolytic anemia (Evans syndrome).

**Choice E:** Malignant hypertension may cause microangiopathic hemolytic anemia. However, blood pressure values would be extremely elevated.

For more prep questions on USMLE Steps 1, 2 and 3, view other posts in this series.

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