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 42-year-old woman comes to the physician because of a four-month history of intermittent abdominal cramps and diarrhea. She has had episodes of skin flushing lasting from two to 15 minutes, most pronounced in the head and neck area. She denies nausea, vomiting, constipation or blood in her stools. Her medical history is unremarkable, and she takes no medications. A grade 3/6 murmur is heard along the mid left sternal border. The lungs are clear to auscultation. Abdominal examination shows no abnormalities. Laboratory studies show urinary excretion of 5-hydroxyindoleacetic acid of 75 mg/day (normal: 0.5-9.0 mg/day). CT scan of the liver demonstrates a 2-cm lesion. Which of the following is the most likely diagnosis?

A. Carcinoid tumor, metastatic

B. Cholangiocarcinoma, primary

C. Hepatocellular carcinoma, primary

D. Mucinous adenocarcinoma of the colon, metastatic

E. Squamous cell carcinoma of the esophagus, metastatic
The correct answer is A.

**Kaplan Medical explains why**

Carcinoid syndrome is the term used for the cluster of cutaneous flushing, abdominal cramps, bronchospasm and diarrhea that is seen in patients who have excess amounts of circulating vasoactive substances, including serotonin. The usual cause is an intestinal carcinoid tumor that has metastasized to the liver.

Gastrointestinal carcinoids that have not yet metastasized are much less likely to produce carcinoid syndrome, because the liver metabolizes and clears the portal blood of the vasoactive substances. Much less commonly, carcinoids at nongastrointestinal sites, such as lung or ovary (whose venous blood does not pass through the liver before entering the systemic circulation), can cause carcinoid syndrome without metastatic disease.

Right-sided endocardial fibrosis, with pulmonary stenosis and tricuspid regurgitation, is common in carcinoid patients and is the result of toxic damage to the heart. 5-hydroxyindoleacetic acid is a urinary metabolite of serotonin that can be used to confirm the clinical impression of carcinoid syndrome. Carcinoid tumors are often indolent, and 10- to 15-year survivals after liver metastasis can occur.

**Why you shouldn’t choose the other answers**

Read these explanations to understand the important rationale for each answer.

**Choice B:** Cholangiocarcinoma arises from the epithelial cells of the bile duct. Symptoms of cholangiocarcinoma include painless jaundice caused by obstruction of the biliary system, pruritus, abdominal pain, weight loss and fever. This tumor does not secrete hormonally active substances.

**Choice C:** Hepatocellular carcinoma usually arises in the setting of cirrhosis or other risk factors, such as hepatitis B, hepatitis C, aflatoxin exposure or hemochromatosis. This tumor does not secrete hormonally active substances.

**Choice D:** Mucinous adenocarcinoma of the colon typically causes iron-deficiency anemia caused by
blood loss, bowel obstruction or change in bowel habits. This tumor does not secrete hormonally active substances.

**Choice E:** Squamous cell carcinoma of the esophagus usually causes symptoms with swallowing; it is unlikely to cause diarrhea. This tumor does not secrete hormonally active substances.

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**Key tips to remember**

- Carcinoid syndrome is described as cutaneous flushing, abdominal cramps and diarrhea caused by excessive circulating serotonin.
- Metastasis of primary intestinal carcinoid to the liver, or primary lung or ovarian carcinoid tumor, results in symptoms of carcinoid syndrome.
- The small bowel is the most common site of carcinoid tumor; however, small bowel carcinoid tumors do not cause carcinoid syndrome because the liver metabolizes and clears the portal circulation of the excessive circulating serotonin.

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

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