Practical Teaching Case – A Small Complaint

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A 60-year-old man with a past medical history of hypertension, hyperlipidemia, gastroesophageal reflux disease, Chronic Obstructive Pulmonary Disease, and Obstructive Sleep Apnea presents for a routine, previously scheduled visit with his primary care provider. He reports no specific chief complaint, but with prompting, he mentions a small complaint about malaise that has been going on for a few months and endorses some mild generalized abdominal pain. He then admits to occasional nausea and intermittent vomiting with no clear exacerbating or alleviating factors, which has progressively worsened. He also notes associated weight loss but is unable to quantify the amount. He denies any dysphagia or odynophagia. He denies any diarrhea, change in bowel habits, flushing, or diaphoresis. A review of his family history is significant for a father with lung cancer. He has smoked one pack of cigarettes daily for twenty years, but does not drink alcohol or use any illicit substances.

On physical exam, he is afebrile and vitally stable. He is obese and noted to have marked left supraclavicular fullness. His complete blood count and comprehensive metabolic panel are normal with a hemoglobin of 14.3 g/dL. Given concern for the left supraclavicular fullness, a Computed Tomography scan of the neck and chest with contrast is completed, which is significant for left cervical, mediastinal, and upper abdominal adenopathy as well as an irregular nodular density along the left side of the upper esophagus. It also demonstrates multiple sub-centimeter liver lesions (new since his last scan two years prior). An upper endoscopy shows a normal esophagus with no endoscopic evidence of an esophageal mass or extrinsic compression. The stomach is normal, but a large frond-like villous and infiltrative mass without overt bleeding is found in the third part of the duodenum (Figures A and B). Biopsies were obtained (Figures C and D). A colonoscopy from four years prior showed seven small (<10 mm) polyps; given the significant findings on the upper endoscopy, colonoscopy was not repeated. A position emission tomography (PET) scan confirms hypermetabolic wall thickening at the third and fourth part of the duodenum. Multiple other areas show hypermetabolic activity including a large para-esophageal lymph node and multiple liver lesions.

After reviewing the endoscopy images and biopsies, what is the most likely diagnosis?

A. Lymphoma
B. Sarcoma
C. Adenocarcinoma
D. Carcinoid tumor

Answer: C

This patient has metastatic small bowel adenocarcinoma (SBA) based on history, workup, and histology findings (option C). Small bowel malignancies are rare, accounting for only 2% of all gastrointestinal tumors in the United States.[1] SBAs make up almost 60% of duodenal histologic tumor subtypes.[2] They occur in men and women between the age of 40 and 70. They typically do not become symptomatic until they reach an advanced stage with deep wall penetration and nodal metastases, at which point gastric outlet obstruction is a common presentation. CT and MRI scans are useful to assess for local invasion and distant metastases, while endoscopy is useful for biopsy of proximal tumors. On histology, as seen in Figures C and D, cells are moderately well differentiated and are usually positive for acid mucin. Scattered endocrine cells can also be seen. The management for stage I to III SBA is primarily surgical with the specific procedure depending on the location.[3] Adjuvant therapy is also recommended due to risk of local and distant recurrence. The 5-year survival rate is around 85% for people in these early stages, with approximately 40% recurrence rate after resection (median time to recurrence is 25 months). For stage IV SBAs, which make up 32% of all cases at diagnosis, chemotherapy is the mainstay of treatment with first-line regimens such as FOLFOX (folinic acid, 5-fluorouracil, and oxaliplatin - the regimen selected for this patient).[3] The liver and peritoneal cavity are the most common sites of metastases and the 5-year survival is around 42%.[3]

Lymphomas (option A) can present with B-symptoms like fevers and night sweats, which was not the case here. The median age for non-immunoproliferative small intestinal disease (IPSID) lymphomas (which are more common in industrialized nations like the United States) is typically in the mid-30s. Enteropathy-associated T cell lymphoma can present in the sixth decade of life, but is associated with a history of celiac disease which this patient lacked. Sarcomas (option B) comprise only 10% of small bowel neoplasms and show pleiomorphic spindle cells with nuclear atypia, hypercellularity, and several mitotic figures on histology, which were not present here. Carcinoid tumors or neuroendocrine tumors (NETs) (option D) are usually found incidentally due to being asymptomatic. When symptomatic, they present as carcinoid syndrome - flushing, diarrhea, or wheezing. Morphologically, well-differentiated NETs have characteristic organoid arrangements of tumor cells with either solid, gyriform, trabecular, or glandular patterns. The cells are relatively uniform, and they have round to oval nuclei, coarsely stippled chromatin, and a finely granular cytoplasm.

SBAs are rare and insidious; a vigilant workup is warranted to attain the diagnosis and improve patient's prognosis.

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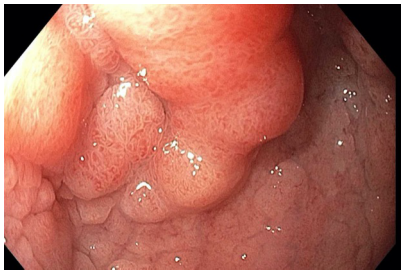

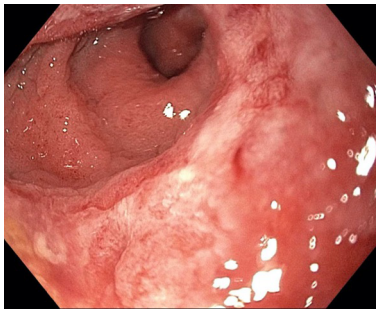

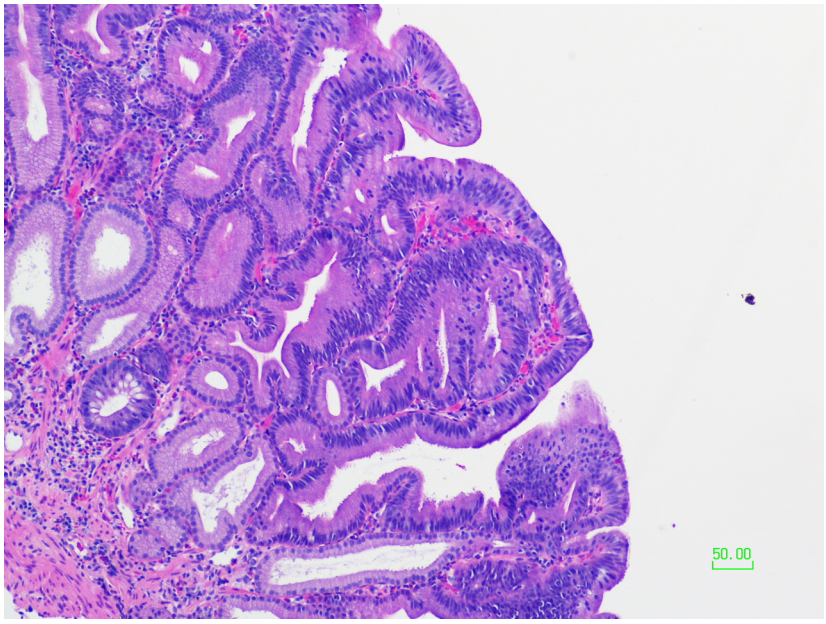
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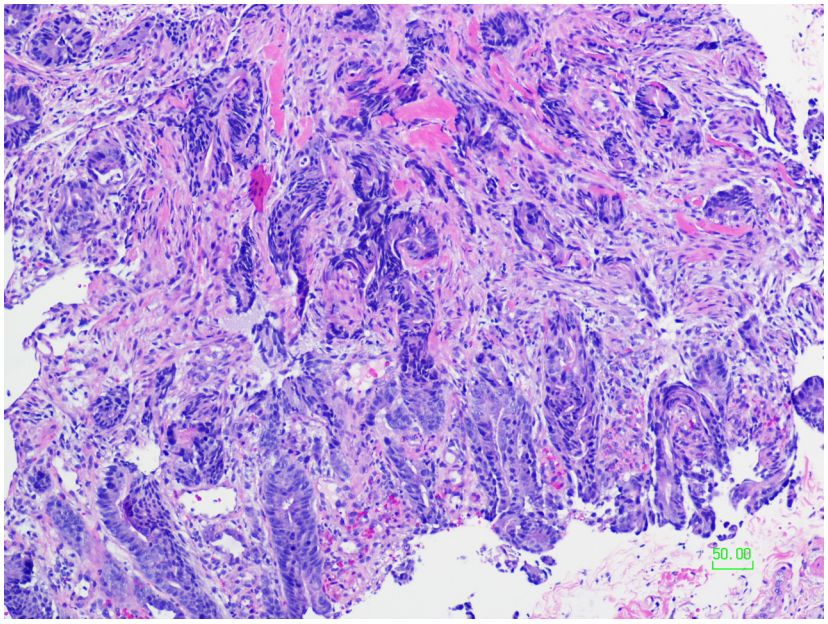
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