A unique case of abdominal pain and anemia

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A 38-year-old female presented with intermittent abdominal pain and rectal bleeding and found to have severe anemia. The computerized tomography of abdomen and pelvis with contrast showed a solid mass measuring 5.4 × 3.4 × 3.1 cm in the cecum (Figure 1, left). The colonoscopy revealed a large protruding lesion with ulceration in the right colon (Figure 1,right). Multiple biopsies of the ulcerated area were obtained.

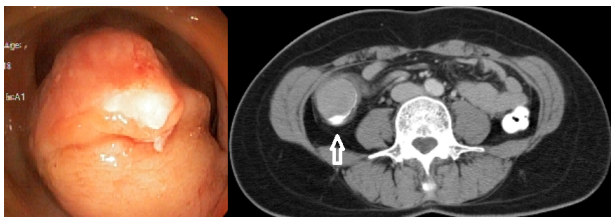

Figure 1. — Colonoscopy revealed a large protruding lesion in the right colon (Figure 1, left). Abdominal computed tomography demonstrated a solid mass (Figure 1, right arrow).

What is the etiology?

Biopsies of the ulcerated area were suggestive of mesenchymal neoplasm. The patient subsequently underwent robotic assisted right hemicolectomy with ileocolonic anastomosis. Microscopic examination showed tumor sheets of spindle to epithelioid cells with interspersed band-like collagen, mild atypia and 4 mitosis/10 high-power field. Immunohistochemical stains showed that tumor cells were diffusely positive for HMB-45 (Figure 2), cathepsin K and melan-A, focally positive for SMA, and negative for CD117, S100, myogenin, and MUC4. These findings were suggestive of perivascular epithelioid cell tumor (PEComa) with sclerosing features. Surveillance at 3 years did not show any evidence of recurrence.

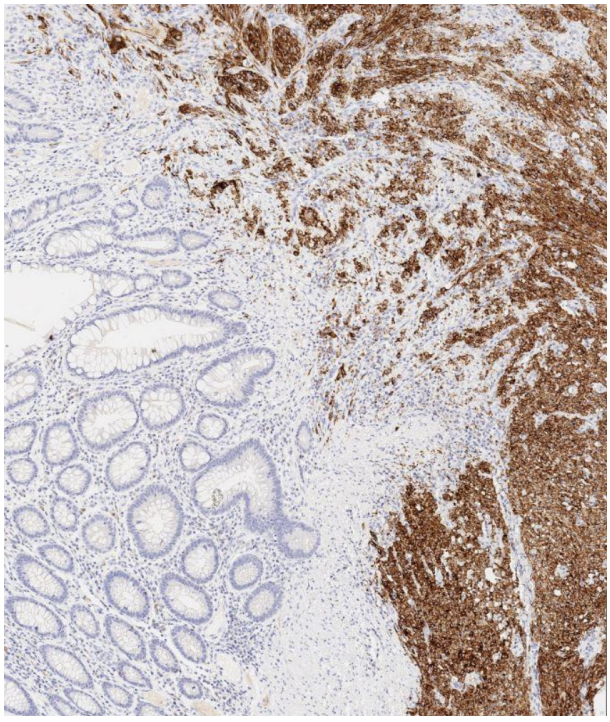

Figure 2. — HMB-45 staining of tumor cells.

PEComas are a group of rare mesenchymal tumors composed of perivascular epithelioid cells characterized by myomelanocytic differentiation (1). This group of tumors exhibit variable clinical behavior, ranging from benign to aggressive local tumor growth and seeding of metastasis. PEComas are most often diagnosed in middle-aged females and can arise at various sites such as visceral, abdominopelvic, retroperitoneal, bone and soft tissue. The gastrointestinal tract is the second most common site of PEComas and most commonly found in the colon (2). The clinical presentation is not specific and depends on the organ and size of the tumor. PEComas can be asymptomatic or can manifest as abdominal pain, obstruction and gastrointestinal bleeding. PEComas should be differentiated from other mesenchymal epithelial tumors using endoscopic ultrasound and immunohistochemistry. A correct diagnosis before surgical resection is hard to make because of non-specific imaging features. The PEComa is characterized by positivity with melanocytic (HMB-45 and/or Melan-A) and myogenic (actin and/or desmin) markers. Surgical resection of the tumor and regional lymph nodes seems to be the treatment of choice for most PEComas.

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Conflict of Interest Statement

All authors declare that no conflicts of interest or financial relationships exist

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