A 52-year-old woman presented with abdominal pain, dysphagia, and fatigue. She had gastric adenocarcinoma treated with lower esophago-gastroduodenectomy without splenectomy and chemoradiotherapy 6 years prior. CT angiography demonstrated a thrombus in the abdominal aorta with bilateral embolic kidney infarcts, but no tumor recurrence nor organomegaly. Laboratory findings included: hemoglobin 7 g/dL, MCV 61 fL, platelet count 750 thou/μL, normal WBC, reticulocytes 2.5%, serum ferritin 6 ng/mL, serum iron 12 μg/dL, and transferrin 400 μg/dL. Peripheral smear showed thrombocytosis (normal morphology), hypochromia, and anisopoikilocytosis (main panel). Oral iron absorption test showed no absorption, presumed secondary to extensive surgery. Upper endoscopy demonstrated upper esophageal webs (inset).

It is challenging to diagnose a myeloproliferative neoplasm (MPN) in the setting of severe iron deficiency and possible reactive thrombocytosis. Plummer-Vinson syndrome is a manifestation of severe, long-term, iron deficiency anemia causing dysphagia because of esophageal webs. The diagnosis of MPN was made based on the increased and sustained platelet count of >1 million after adequate intravenous iron repletion, the presence of the JAK2-V617F mutation, and an otherwise unexplained arterial thromboses. Treatment with hydroxyurea and periodic iron infusions has resulted in a persistently normal platelet count and hemoglobin values between 11 and 12 g/dL. Her dysphagia has resolved.

Many BloodWork images are provided by the ASH IMAGE BANK, a reference and teaching tool that is continually updated with new atlas images and images of case studies. For more information or to contribute to the Image Bank, visit http://imagebank.hematology.org.
Plummer-Vinson syndrome and reactive thrombocytosis mask a JAK2-V617F positive myeloproliferative neoplasm

Emma Catherine Scott and Stephen Schuster