CLINICAL IMAGE 449

Quiz: Abdominal pain in a patient with Paroxysmal Nocturnal Hemoglobinuria

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Key words: spontaneous intestinal intramural hematoma, paroxysmal nocturnal hemoglobinuria, thrombocytopenia, anticoagulation, complication.

Quiz:

A 42-year old man presented to emergency room with abdominal pain, nausea and vomiting for the last 4 days. He was diagnosed as Paroxysmal Nocturnal Hemoglobinuria (PNH) a year before and he had thrombocytopenia due to PNH. Physical examination revealed abdominal distention and abdominal sensitivity with rebound tenderness. His international normalized ratio (INR) was 1.12 and platelet count was 14000/mm³. What would you recommend for the evaluation of abdominal pain and what is your diagnosis?

Answer:

Abdominal Computed Tomography (CT) ordered during the Emergency Room stay showed large intramural hematoma of the jejunum (Fig. 1a). Enteroscopy was performed for the evaluation of intractable nausea and vomiting after a normal EGD. Single Balloon Enteroscopy (SBE) examination showed there was a 4-5 cm long segment of aperistalsis and distorted architecture that affected the 1/3rd of the lumen with loss of normal mucosa and plicae, which were replaced by hyperemia and friability (Fig. 1b) located at the proximal jejunum.

Spontaneous intestinal intramural hematoma (SIIH) may occur as a result of blunt trauma to the abdomen or an adverse event of anticoagulant therapy. Other diseases causing thrombocytopenia may also present with SIIH, which include leukemia, lymphoma, myeloma, hemophilia, chemotherapy and idiopathic thrombocytopenic purpura, as well as diseases causing hemorrhage such as pancreatitis, pancreatic carcinoma and polyarteritis nodosa.

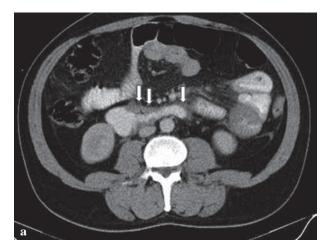




Fig. 1. — (a) Abdominal Computed Tomography of the patient showed a large intramural hematoma of the jejunum (arrows), almost obstructing the lumen. (b) The enteroscopy at 4th day of the presentation was performed via Single Balloon Enteroscope. Between the 4th portion of the duodenum and proximal jejunum, there was a 4-5 cms long segment of aperistalsis and distorted architecture that affected the 1/3rd of the lumen with loss of normal mucosa and plicae, which were replaced by hyperemia and friability consistent with ischemia.

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E-mail: tkav@hacettepe.edu.tr Submission date: 27/07/2015 Acceptance date: 07/09/2015