Hereditary angioedema involving the duodenum. An unusual cause of upper abdominal pain

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A 20-years-old patient presented to the emergency department with acute upper abdominal pain, vomiting and diarrhea. She suffered for similar recurrent episodes of unexplained abdominal pain since early adolescence. Two years ago, she underwent right colectomy for ileocecal intussusception. On physical examination, there was epigastric tenderness and intense crampy pain. Investigation revealed leukocytosis around 13,700/mm$^3$ and normal C-reactive protein level. Contrast-enhanced CT of the abdomen showed a circumferential wall thickening of the 3rd–4th duodenal portions and the proximal jejunum, adjacent fat stranding and intraperitoneal fluid effusion (figure 1). Superior venous mesenteric vein was patent. According to her medical history past, hereditary angioedema (HAE) involving the duodenum was strongly suspected and confirmed by low concentration of C1 esterase inhibitor level. Symptoms resolved spontaneously two days later.

Comment

HAE is a rare autosomal dominant disease occurring in 1/50,000 to 100,000 persons of any ethnic group. Two types of HAE exist according to a quantitative (type 1, 85%) or a functional (type 2, 15%) deficiency of C1 inhibitor due to genetic mutations. HAE affects skin, gastrointestinal tract and upper airways. Symptoms are recurrent including self-limited subcutaneous edema without pruritus, life-threatening pharyngolaryngeal edema or acute abdominal attacks with crampy abdominal pain, vomiting and diarrhea [1]. CT is the modality of choice for diagnosis of digestive manifestations of HAE. Both small and large intestine can be involved with a segmental or diffuse mural thickening and a typically target appearance, so-called double-halo sign [2,3]. Ascite can be present. The differential diagnoses of HAE on imaging are mesenteric ischemia, vasculitis or Crohn’s disease. The mainstay of treatment for HAE attack whatever the symptoms is based on...
intravenous injection of C1 inhibitor concentrate. Early treatment reduces intensity of pain and duration of attacks.

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References


Figure 1
Axial (A) and coronal reformatted (B, C) computed tomography images showing symmetric oedematous mural thickening with a larger appearance of the 3rd–4th portions of the duodenal wall (white arrow) extended to the Treitz’s angle. Noted the intraperitoneal fluid effusion (black arrowhead)