A 25-year-old female with recurrent abdominal pain

F. Ufuk[1], M. Celik[2]

(1) Department of Radiology, University of Pamukkale, Kinikli, 20100 Denizli, Turkey; (2) Department of Gastroenterology, University of Pamukkale, Kinikli, 20100 Denizli, Turkey.

Question

A 25-year-old female patient, who had a history of medical treatment with the diagnosis of asthma for the last two years, presented with complaints of abdominal pain, nausea, and vomiting. Similar complaints recurred several times over the previous two months and regressed spontaneously in his medical history. These symptoms were acute onset and lasted for 6-8 hours. She had no history of weight loss or fever. Family history revealed the patient's mother had a history of recurrent swelling of the face and lips, which resolved spontaneously, and her father had coronary artery disease. Physical examination revealed epigastric tenderness without guarding. Laboratory test results were within normal limits, except for low hemoglobin level (13.2 g/dL; reference range 14-16 g/dL). Abdominal ultrasound showed mild peri-intestinal free fluid, and contrast-enhanced abdominal computed tomography (CT) was obtained for further evaluation. Abdominal CT showed an edematous thickening of the duodenum wall and peri-intestinal free fluid (Figure 1). The remaining abdominal CT findings were within normal limits.

What is the diagnosis?

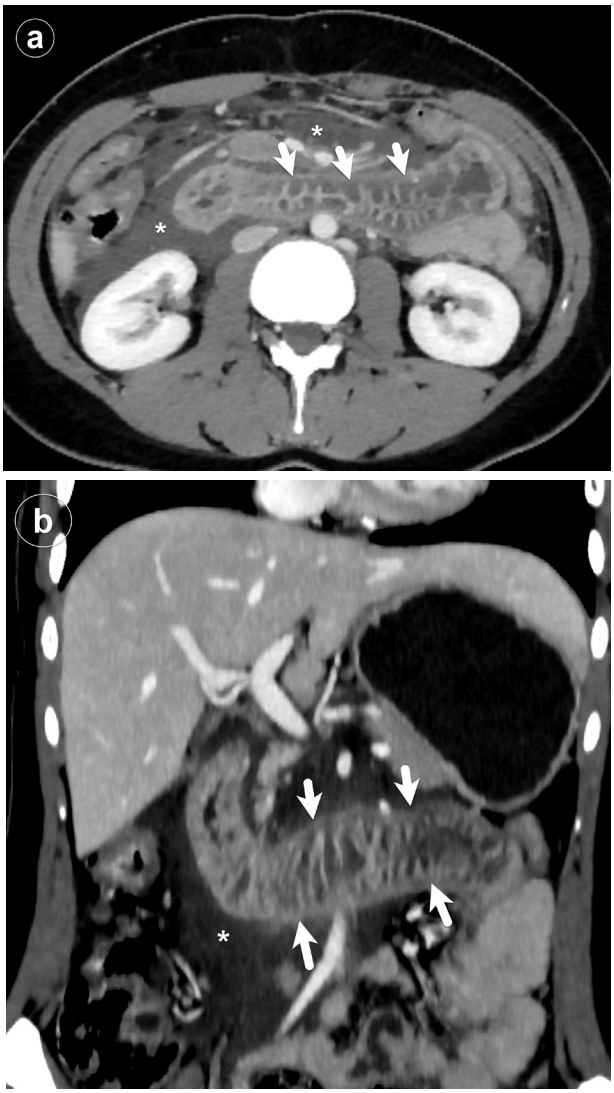

Figure 1. — **a)** Axial and **b)** coronal abdominal CT images show mucosal hyperenhancement and severe submucosal edema of the duodenum consistent with a "bowel target sign" (arrows). Note the peri-intestinal free-fluid (*).

Answer

The abdominal CT images revealed an edematous wall thickening in duodenum and peri-intestinal free fluid. The CT images showed a target sign characterized by the contrast-enhancing mucosa and muscularis propria layers and the submucosal edema. The differential diagnosis of "bowel target sign" includes ischemia, infection, the active phase of inflammatory bowel disease, and angioedema. On abdominal CT, the superior mesenteric artery and vein were patent, and there were no other signs of ischemia in the intestinal walls. The patient did not have a fever that could be compatible with infection, and laboratory values were within normal limits. The active phase of inflammatory bowel disease was also considered in the differential diagnosis, but other bowel segments other than the duodenum were normal on CT. The patient had recurrent abdominal pain and edematous bowel wall thickening on CT; the bowel angioedema was considered at the forefront in the differential diagnosis. Laboratory analysis revealed low serum C4 level and C1 esterase inhibitor deficiency, consistent with angioedema.

Correspondence to: Furkan Ufuk, MD., Assoc. Prof. of Radiology, Pamukkale University Medical School, Department of Radiology, Kinikli, 20100 Denizli, Turkey. Phone: +90 554 511 50 88. Fax: +90 258 444 0 728.
Email: furkan.ufuk@hotmail.com

Submission date: 28/12/2021
Acceptance date: 29/01/2022

Hereditary angioedema (HA) is an unusual disorder characterized by recurrent cutaneous or submucosal edema, and most patients have a family history (1). HA often presents due to C1 esterase inhibitor deficiency, and isolated bowel involvement is infrequent (2). Recurrent abdominal pain, nausea, vomiting, and watery diarrhea are the most common clinical findings in the bowel involvement of HA. Generally, clinical symptoms are not specific and may be ignored by patients as they are self-limiting (1). HA rarely mimics an acute abdomen, leading to unnecessary diagnostic laparotomy (3). Abdominal CT reveals the presence of peri-intestinal free fluid, bowel wall edema, and the "bowel target sign" and differentiates HA from other acute abdominal causes. In the presence of a history of recurrent abdominal pain, HA should be considered in the differential diagnosis when a "target sign" is seen in the small bowels on CT (1-3). Accurate diagnosis of HA prevents unnecessary invasive procedures and complications.

The patient's acute symptoms were treated with an intravenous plasma-derived C1 esterase inhibitor (CINRYZE 500IU/5ml, Takeda, Turkey), and she was discharged with Cinryze 500IU/5ml was prescribed as on-demand therapy.

Conflict of interest

The authors declare that they have no conflict of interest.

Funding

No funding was received for this study.

References

1. DE BACKER AI, DE SCHEPPER AM, VANDEVENNE JE, SCHOETERS P, MICHIELSEN P, STEVENS WJ. CT of angioedema of the small bowel. *AJR Am J Roentgenol*., 2001;**176**:649-52.
2. WEI SC, FITZGERALD K, MAGLINTE DD. CT findings in small bowel angioedema: a cause of acute abdominal pain. *Emerg Radiol*., 2007;**13**:281-3.
3. JAMIL B, NAEEM MS, ANACHEBE T, MAJEED MH. Hereditary angio-oedema as a rare cause of small-bowel obstruction. *BMJ Case Rep*., 2019; **12**:e231186.