

Angioneurotic oedema of the small bowel

Published on 04.02.2017

DOI: 10.1594/EURORAD/CASE.14439

ISSN: 1563-4086

Section: Abdominal imaging

Area of Interest: Abdomen

Procedure: Diagnostic procedure

Procedure: Radiobiology

Imaging Technique: CT

Special Focus: Oedema Congenital Case Type: Clinical Cases

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Patient: 32 years, male

Clinical History:

A 32-year-old man presented to the emergency department with a 3-day history of severe epigastric pain associated with vomiting and constipation. A blood test at presentation showed elevated CRP of 20.49 mg/L, high WBC of $13.9 \times 10^9/L$. Further investigations revealed that C1 Esterase inhibitor was 53 mg/L.

Imaging Findings:

A CT abdomen with iodine showed oedema of the bowel wall, stomach antrum and duodenum, with surrounding free fluid. There was enhancement of the mucosa and serosa as well as fluid accumulation in the bowel lumen. The mesenteric vessels were prominent.

No bowel loop obstruction or pneumoperitoneum was observed.

Discussion:

Angioedema is a noninflammatory disease characterized by episodes of increased capillary permeability resulting in extravasation of intravascular fluid and subsequent angioedematous swelling. Angioedema was first described by J.L. Milton in 1876 and first named angioneurotic oedema by Quincke in 1882 [1, 2].

There are various cases of angioedema, including hereditary angioedema, acquired C1 esterase inhibitor deficiency (like in our case), drugs and food. Some cases are idiopathic [3, 4].

It is characterized by localized and transient edematous swelling of superficial regions such as the face, upper airways, genitals and limbs [5]. Angioedema, however, may affect any part of the body, and intra-abdominal involvement may occur with or without cutaneous or respiratory involvement [3, 4].

Angioedema of the upper airways may result in acute respiratory distress, airway obstruction, and asphyxia [5, 6]. Gastrointestinal tract involvement sometimes mimics acute abdomen resulting in unnecessary laparotomy [2] or rarely causes potentially life threatening hypovolemic shock [5, 7]. Gastrointestinal complaints may be the initial symptoms in patients with angioedema, as in our case. Angioedema is seen during infancy or early adolescence, although it may occur until the sixth decade [7]. Clinical diagnosis of angioedema of the gastrointestinal tract is rarely considered until repeated attacks occur, like in our case.

Radiographic features during the acute phase may show multiple dilated small bowel loops with regularly thickened

mucosal folds, wall thickening. Air-fluid level may also be seen [8, 9].

Ultrasound usually reveals mucosal thickening, edematous bowel wall with increased intraluminal fluid and decreased motility, and it may demonstrate peritoneal fluid in dependent areas of the abdomen [10, 11].

Enhanced CT usually demonstrates segmental or diffuse thickening of the small bowel wall, low attenuation of the edematous submucosa, and markedly enhanced thickened mucosa and serosa [12].

Diagnosis of angioedema is mainly clinical, nevertheless imaging findings can help greatly, warranting further clinical workup and avoiding unnecessary surgical laparotomy.

Differential Diagnosis List: Angioedema of the small bowel secondary to C1 esterase inhibitor deficiency, Ischemia, Shock bowel, Henoch Schonlein purpura (vasculitis)

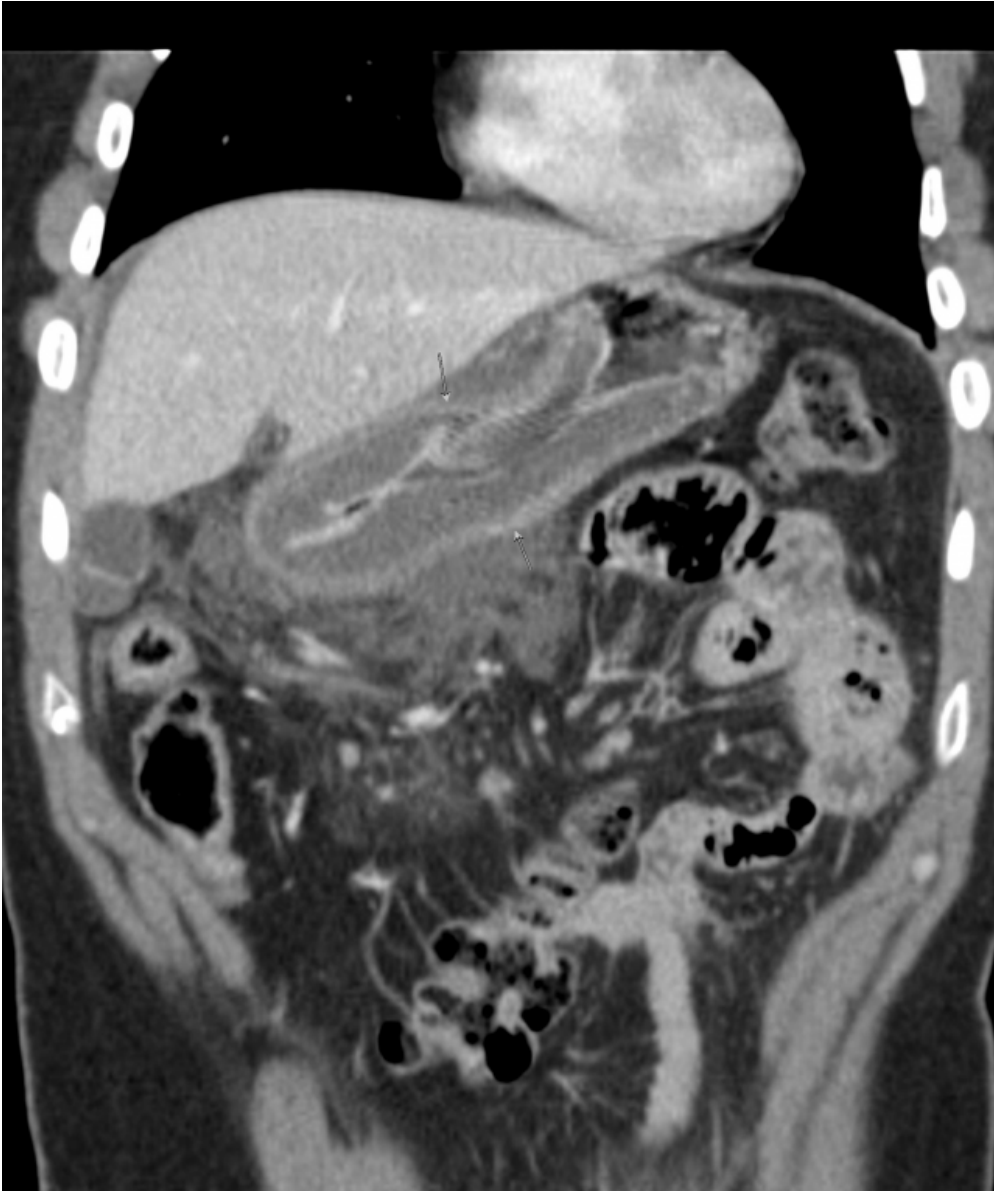
Final Diagnosis: Angioedema of the small bowel secondary to C1 esterase inhibitor deficiency

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Figure 1

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Description: Enhanced CT - Coronal reformat - soft tissue window reveals mural thickening and submucosal oedema of the gastric antrum with surrounding free fluid and prominent mesenteric vessels

Origin: Radiology department - SKMC - Abu Dhabi - UAE

Figure 2

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Description: Enhanced CT abdomen - axial view - soft tissue window **Origin:** Radiology department - SKMC - Abu Dhabi - UAE

Figure 3

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Description: Enhanced CT abdomen - coronal reformat - soft tissue window - clear delineation of different layers of duodenal wall and fluid accumulation within dilated duodenum. There is surrounding free fluid. **Origin:** Radiology department - SKMC - Abu Dhabi - UAE

Figure 4

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Description: Coronal reformat of enhanced CT abdomen - reveals free fluid accumulation. **Origin:** Radiology department - SKMC - Abu Dhabi - UAE