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INTESTINAL ANGIONEUROTIC EDEMA

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Background: This 38-year-old female patient presented at the emergency department with subacute low abdominal pain for 3 days. There was little abdominal tenderness, but no signs of peritonitis. Her blood results showed little inflammation with mild leukocytosis 13.0 (3.5-11.0 x 10*3/µL) and mild CRP elevation 44 (< 5 mg/L).
The patient had a CT scan of the abdomen on the same day. The day after, a colonoscopy was performed which was perfectly normal. Because the discrepancy between the findings on CT scan, biochemistry and colonoscopy, the patient underwent an explorative laparoscopy, which showed a normal colon.
Two days after the initial CT scan, a control CT scan was performed.

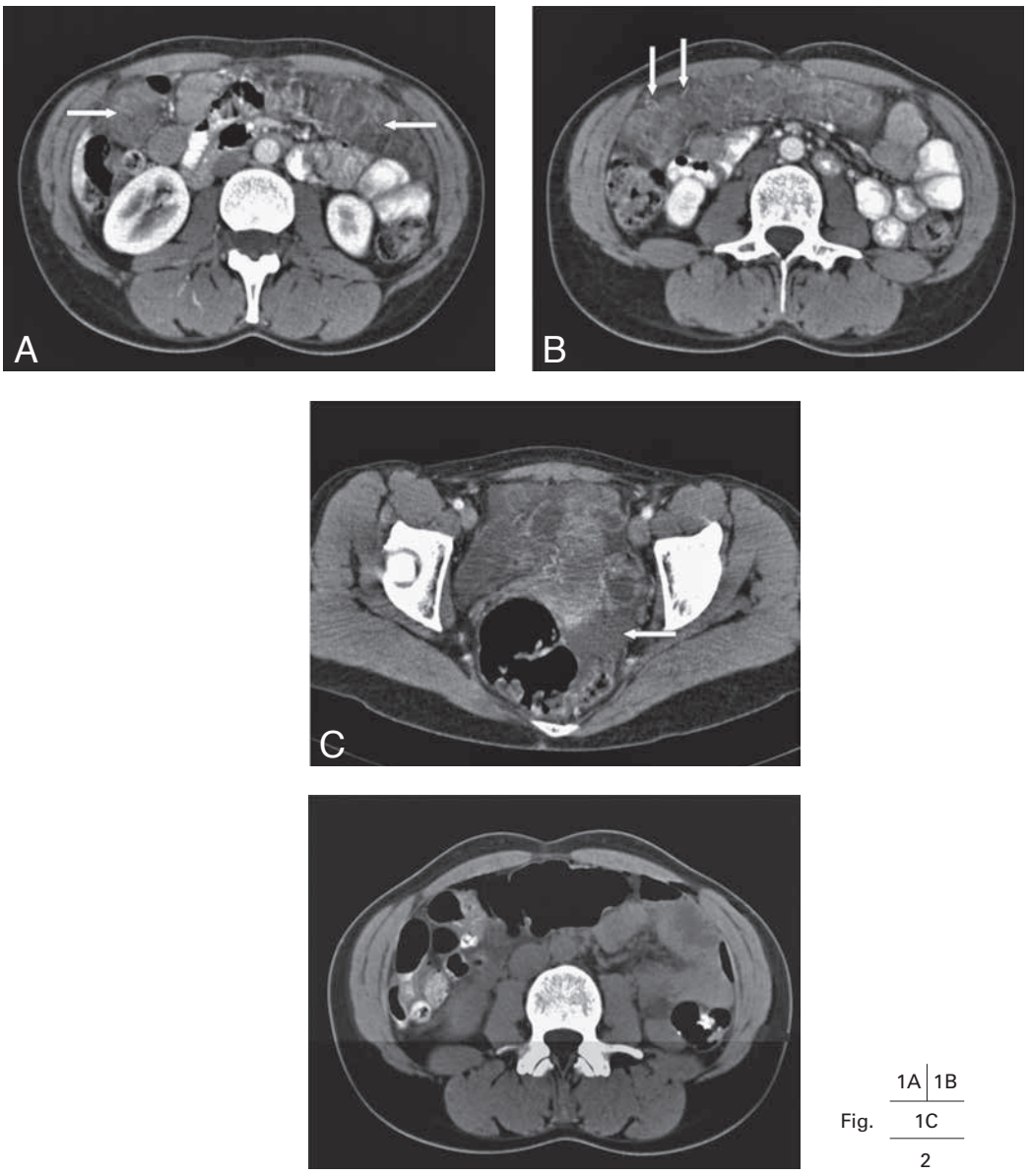

Fig. 1A | 1B / 1C / 2

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Work-up

CT scan of the abdomen following intravenous and oral administration of contrast medium, during the acute episode (Fig. 1) shows on section at the level of the upper abdomen (A) a regular wall thickening of the transverse colon with thickened mucosal folds (white arrows). On section at a lower upper abdominal level (B), prominent mesenteric vessels (white arrows) adjacent to the thickened transverse colon are seen.

On section at pelvic level (C), pelvic ascites is shown (white arrow).

Control CT scan of the abdomen, two days after the initial CT (Fig. 2), there is complete resolution of the mucosal thickening of the transverse colon and the prominent mesenteric vessels. Note the residual free intra-abdominal air due to laparoscopy.

Radiological diagnosis

On the basis of the transient appearance of colitis on CT with fast normalisation of CT findings in the periods of days, the diagnosis of *intestinal angioneurotic edema* was made.

Discussion

Angioedema or angioneurotic edema is a non inflammatory disease featured by recurrent edema of the cutis or mucosa of the upper airways and gastro-intestinal tract. It is caused by an elevated vascular permeability with extravasation of fluid in the interstitium. The condition can be hereditary (deficiency of serum C1 inhibitor) or acquired. The acquired form is usually associated with B-cell lymphoproliferative diseases or with the use of angiotensin-converting enzyme inhibitor (ACE-I).

Gastro-intestinal involvement is less frequent than angioedema of the face or the upper airways and may be the only manifestation of the disease in 21% of cases of hereditary angioneurotic edema. In the most cases of gastro-intestinal angioedema, the small bowel or duodenum is affected.

The symptoms are often nonspecific and consist of recurrent abdominal pain, abdominal distension, nausea and vomiting. Sometimes, angioedema can mimic a (sub)acute abdomen. Therefore making the right diagnosis can prevent unnecessary invasive interventions.

Diagnosis is suggested by discrepancy between heavy abdominal pain and the absence of peritoneal signs, fever and leukocytosis.

Small bowel follow-through and barium enema studies will show a "stacked coin" appearance, "thumb- printing" and separation of bowel loops secondary to mucosal thickening. Diffuse bowel wall thickening and ascites can also be visualised on ultrasonography, although these findings are nonspecific. However, the most requested examination in the acute phase is a CT scan of the abdomen. Contrast-enhanced CT scan shows symmetrical thickening of the mucosa, submucosa and muscularis propria, which may present as a "target sign". Other findings are intraluminal fluid accumulation, prominent mesenteric vessels, omental thickening and moderate ascites.

The differential diagnosis includes adverse reaction to contrast material, ischemia, Henoch-Schönlein vasculitis, intramural bleeding from trauma, anticoagulation therapy or hemophilia. However the transient appearance and rapid resolution of the symptoms are quite suggestive for angioneurotic edema. The diagnosis is confirmed with measurement of serum levels of C1-esterase inhibitor and C1-esterase inhibitor functional activity complement levels. These tests were not performed on the patient in the case, since she was lost for follow-up.

The initial treatment consists of supportive measures, certainly in case of dehydration and hypovolemic shock. If available a concentrate of C1 inhibitor can be administered in both hereditary and acquired angioedema. Androgens, such as danazol, are proven to be effective prophylactics in hereditary angioedema. In case of ACE-I induced angioedema cessation of the medication is sufficient as therapy.

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