Acute appendicitis in Crohn's disease: CT appearance and clinical significance

Clinical History:

Young female with long-standing ileo-colonic Crohn's disease treated with steroids, azathioprine and adalimumab, the latter recently restarted after childbirth.
Progressive clinical deterioration with increased stool frequency, abdominal discomfort and weight loss, followed by acute lower right abdominal pain with positive Blumberg and McBurney signs, leukocytosis and mildly increased C-reactive protein.

Imaging Findings:

Months earlier, colonoscopy described atrophied ileal mucosa, sparse hyperaemia and aphitoid ulcers in the colon, and MR-enterography (Fig.1) showed moderate circumferential mural thickening of caecum and ascending colon, without mural and extraluminal signs of Crohn's disease (CD) activity, and normal-appearing appendix.
Currently, emergency CT (Fig.2) showed development of mural stratification at caecum and ascending colon with oedematous submucosa and thin hyperenhancing mucosa, consistent with CD reactivation; the minimally distended appendix showed hyperenhancing mural thickening with associated periappendiceal inflammatory fat stranding nearby the appendiceal orifice. As measured sonographically (Fig.3), mural thickness was normal at distal ileum, mild (5-6 mm) at caecum, maximum (7-10 mm) along appendix.
Endoscopy (Fig.4) confirmed appearance of marked congestion and inflammation centred at the appendicular orifice, focal hyperaemic mucosal changes in the caecum, without signs of ileal activity; biopsies revealed periappendicular inflammatory infiltration.
According to the European Crohn's and Colitis Organisation guidelines, she was treated with high-dose steroids and promptly improved.
Discussion:

According to its original description, Crohn’s disease (CD) was traditionally believed to stop at the ileocecal valve with sparing of the appendix; however, this early theory was soon disproved. In the setting of known CD, appendiceal involvement is currently reported in 12.8–21% of patients, almost invariably associated with terminal ileitis and more frequent in those with widespread colonic disease. Pathology on surgical specimen from ileocecal resections in CD identified appendiceal involvement in up to 40% of cases, with characteristic histological features including mucosal erosions, ulceration with crypt abscesses, mural thickening, focal or discontinuous transmural histiocytic-lymphocytic inflammation, non-caseating epithelioid granulomas [1, 2]. Alternatively, approximately one hundred cases of primarily appendiceal CD have been reported, diagnosed on the basis of granulomatous histopathology on appendectomy specimens and characterized by favourable outcome with low recurrence rate compared to CD in other sites of the bowel [2-5].

In the hereby presented patient the combined endoscopic and imaging findings were consistent with CD reactivation involving mainly the appendix and minimally the caecum, with spared terminal ileum; a similar occurrence has been occasionally described following adalimumab therapy [6]. Therefore, when interpreting urgent abdominal CT and elective CT- or MR-enteroclysis studies in patients with chronic inflammatory bowel disease, the appendix should be always scrutinized: a recent study revealed that inflammatory thickening and hyperenhancement of the appendiceal wall is found in nearly 19% of patients with active disease and has absolute (100%) specificity for differentiating between active and inactive CD. Compared to acute suppurative appendicitis in the general population, in CD appendicitis the lumen is generally obliterated and not dilated, and abscesses are rarely encountered [7]. According to the European Crohn’s and Colitis Organisation (ECCO) guidelines, on the basis of CT and endoscopic findings, in absence of obstruction this situation should be managed medically as a localised active ileocecal CD including initial induction of remission by systemic steroids [8].

**Differential Diagnosis List:** Crohn’s disease reactivation in the appendix, Acute suppurative appendicitis, Complicated appendicitis with abscess formation, Appendiceal diverticulitis, Fibrostenosing Crohn's disease, Intestinal tuberculosis, Intestinal amoebiasis

**Final Diagnosis:** Crohn’s disease reactivation in the appendix

**References:**


Description: Despite poor ingestion of polyethylenglycole solution, the ascending colon and caecum (arrowheads) showed moderate (6-7 mm) circumferential mural thickening, without active inflammation and hypervascularity of surrounding fat (*). Origin: Tonolini M, Radiology Department, "Luigi Sacco" University Hospital – Milan (Italy)
Description: On post-gadolinium fat-suppressed T1-weighted images minimal enhancement was seen in the thickened caecal wall, without hypervascularity of the surrounding fat (*). Origin: Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
**Description:** The appendix (arrows) showed normal, collapsed appearance on coronal (c) and sagittal (d) T2-weighted images. Note moderate caecal wall thickening (arrowheads), unremarkable pericaecal fat (*). **Origin:** Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
Description: The appendix (arrows) showed normal, collapsed appearance on coronal (c) and sagittal (d) T2-weighted images. Origin: Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
Description: The ascending colon and caecum showed moderate mural thickening (arrowheads) with stratified appearance due to oedematous submucosa and thin mucosal hyperenhancement (thin arrows). Note mesenterial lymphadenopathies (thick arrow). Origin: Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
Description: The minimally distended appendix showed development of hyperenhancing mural thickening (arrows) associated with peri-appendiceal inflammatory fat stranding (*) near the appendiceal orifice. Origin: Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
Description: The minimally distended appendix showed development of hyperenhancing mural thickening (arrows) associated with peri-appendiceal inflammatory fat stranding (*) near the appendiceal orifice. Origin: Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
**Description:** Coronal images (d,e) confirmed stratified mural thickening (arrowheads) of ascending colon and caecum with oedematous submucosa and thin mucosal hyperenhancement (thin arrows), appendiceal thickening (arrows) and periappendiceal fat stranding (*). **Origin:** Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
Description: Coronal images (d,e) confirmed stratified mural thickening (arrowheads) of ascending colon and caecum with oedematous submucosa and thin mucosal hyperenhancement (thin arrows), appendiceal thickening (arrows) and periappendiceal fat stranding (*). Origin: Tonolini M, Radiology Department, "Luigi Sacco" University Hospital – Milan (Italy)
Description: Compared to the stratified caecal wall (arrowheads) with oedematous submucosa and thin mucosal hyperenhancement (thin arrows), the distal ileum (*) did not show significant mural thickening. Origin: Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
Description: Compared to the stratified caecal wall (arrowheads) with oedematous submucosa and thin mucosal hyperenhancement (thin arrows), the distal ileum (*) did not show significant mural thickening. Origin: Tonolini M, Radiology Department, "Luigi Sacco" University Hospital – Milan (Italy)
**Description:** The caecum showed minimal, sparse hyperaemic mucosal changes. **Origin:** Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
Description: The appendicular orifice showed marked congestion and inflammation. Origin: Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
Description: With patent ileocaecal valve, the distal ileum (25 cm) showed no visible abnormalities consistent with Crohn's disease. Origin: Tonolini M, Radiology Department, "Luigi Sacco" University Hospital – Milan (Italy)
Description: Focused ultrasound confirmed normal mural thickness of terminal ileum (3.5 mm, between calipers). Origin: Tonolini M, Radiology Department, "Luigi Sacco" University Hospital – Milan (Italy)
Description: The minimally distended appendix showed thickened stratified wall (arrows) measuring 7-8 mm in thickness (in b), approximately 10 mm at the appendicular orifice (in c).**Origin:** Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
**Description:** The minimally distended appendix showed thickened stratified wall (arrows) measuring 7-8 mm in thickness (in b), approximately 10 mm nearby the appendicular orifice (in c). **Origin:**
Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)