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Gastrointestinal Fistulae in Chronic Granulomatous Disease

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To the Editor: We present two patients with findings of fistulizing inflammatory bowel disease (IBD) associated with chronic granulomatous disease (CGD). Both patients had the diagnosis of X-linked CGD confirmed by dihydrorhodamine oxidation, superoxide production, immunoblotting, and molecular sequencing.

A 37-year-old male with CGD and a prior history of IBD presented with a 1-week history of pneumaturia and fecaluria. On examination he was afebrile and there was no abdominal tenderness.

A computed tomography scan of the abdomen and pelvis was performed (Figure 1a and b). The image (Figure 1a) shows continuity between the posterior abdominal wall and anterior rectal wall with posterior displacement and tethering of the bladder wall and air in the bladder. There is significant thickening in the rectum (Figure 1b). Colonoscopy showed ulcerated mucosa within a stenotic region in the rectum (Figure 1c). The image is characteristic of a rectovesical fistula and is an example of a gastrointestinal fistula related to CGD-associated IBD.

Another example of a gastrointestinal fistula related to CGD (Figure 1d and e) is from a 21-year-old patient with CGD-associated IBD who was admitted with fever and left-side buttock and thigh pain. This computed tomography shows a collection of enhancing cavities, note the three air fluid levels (Figure 1d) in continuity, consistent with abscess and fistulae involving the left pelvis extending posterior to the ischium and down the left thigh (Figure 1e) in the region of the gluteus and semimembranosus muscles to 7 cm below the level of the ischial tuberosity. Note the air-fluid level between the ischium and left femur (Figure 1e). Colonoscopy showed ulcerated and inflamed mucosa (Figure 1f).

Chronic granulomatous disease is a rare inherited disorder of the nicotinamide dinucleotide phosphate (NADPH) oxidase complex, in which phagocytes are defective in generating reactive oxidant superoxide and its metabolites. As a result, CGD patients suffer from recurrent bacterial and fungal infections with persistent tissue granuloma formation. IBD has also been associated with CGD (1,2). In our experience, CGD patients may often have unrecognized IBD and may develop dramatic fistulae as shown by the presented cases.

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CONFLICT OF INTEREST

The authors declare no conflict of interest.

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Figure 1.

Images **a**, **b**, and **c** from a 37-year-old patient with CGD. (**a**) CT image of a rectovesical fistula. (**b**) CT image of rectal thickening. (**c**) Colonoscopy of the rectum showed ulcerated mucosa within a stenotic region. Images **d**, **e**, **f** from a 21-year-old patient with CGD. (**d**) CT image of the pelvis with abscess and fistula. (**e**) CT image with extension of the fistulae tract between the ischium and left femur. (**f**) Colonoscopy with ulcerated and inflamed mucosa. 604 × 384 mm (72 × 72 d.p.i.). CGD, chronic granulomatous disease; CT, computed tomography.