Incidental finding of inflammatory fibroid polyp of the appendix in a patient with acute appendicitis

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Dear Editor,

Inflammatory fibroid polyps (IFPs), also known as Vanek's tumors, are rare benign lesions that are mostly found in the antrum of the stomach and rarely in the appendix. There are only five reported cases of this tumor in the appendix. Here, we present the case of a 57-year-old woman with IFP of the appendix who presented with acute appendicitis. We also present our study to review available literature.

A 57-year-old woman with no significant medical history came to the emergency department complaining of a 3-day history of right lower quadrant pain radiating to the umbilical area and accompanied by fever, chills and nausea. She denied vomiting, urinary symptoms, change in bowel habits, melena and hematochezia. Vital signs in the emergency room revealed blood pressure of 104/72 mmHg, heart rate of 105 beats per minute, respiratory rate of 19 breaths per minute and temperature of 103oF. Examination of the abdomen revealed tenderness to palpation at the right lower quadrant with rebound and guarding. The rest of the physical examination was unremarkable. Initial laboratory workup revealed white blood cells of 10.7 x 10^9/L with normal serum comprehensive metabolic panel and urinalysis. Computed tomography scan of the abdomen revealed marked inflammatory changes surrounding the distended appendix with discontinuity of the mucosa, concerning for a perforated appendicitis with possible abscess (Figure 1). She was empirically started on piperacillin-tazobactam and was scheduled for emergency laparoscopic appendectomy. Intraoperative finding revealed a gangrenous appendix. Macroscopic examination showed a fragmented appendix with smooth, congested and scattered fibrinopurulent exudate on the serosal surface. On microscopic examination, cryptitis, crypt abscesses and diffuse transmural neutrophilic infiltration were noted. In addition, an incidentally well-circumscribed, benign, hyalinized spindled-cell proliferation with prominent eosinophilic infiltrate and thin- and thick-walled blood vessels was identified within the submucosa (Figure 2A-D). Immunohistochemical stains revealed that the spindle cells were focally positive for CD34 and negative for desmin, S100, EMA and CD117. The characteristic submucosal location of the lesion and the distinctly prominent eosinophilic infiltrate, along with the immunohistochemical profile, were consistent with hyalinized inflammatory fibroid pol-

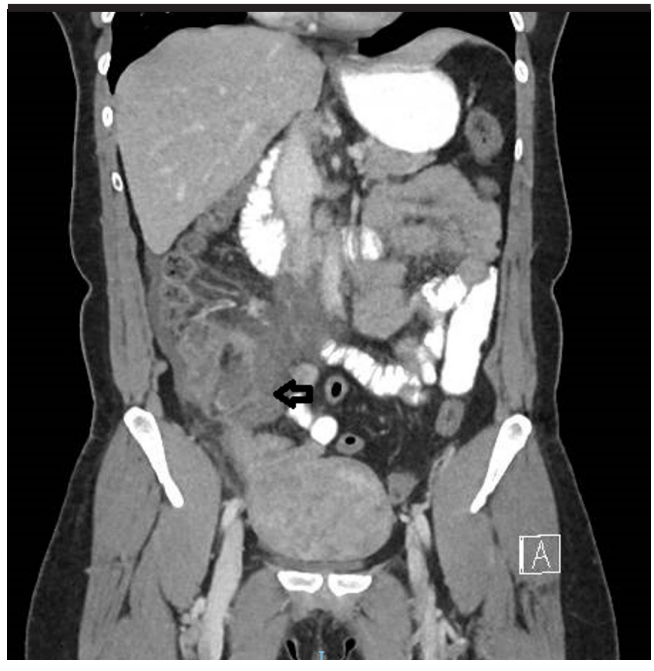

Figure 1. Coronal section of the abdominal computed tomography scan showing a distended appendix (arrow) with marked surrounding inflammatory changes.

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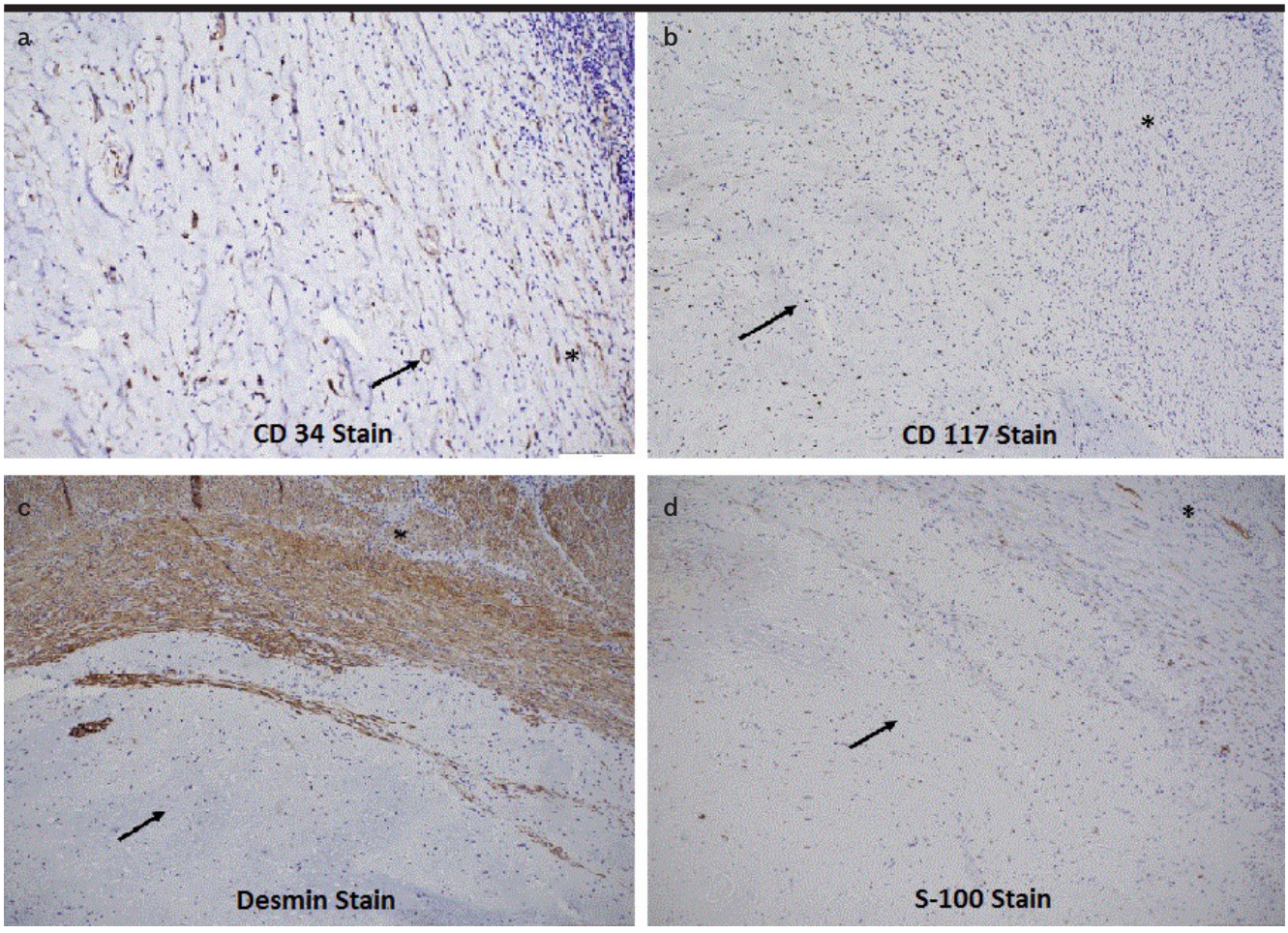

Figure 2. a-d. Hematoxylin & eosin staining. (a) Well-circumscribed polypoid lesion that involves the submucosa (arrow) and does not involve the mucosa (asterisk) and muscularis propria (arrowhead). (20x). (b) Characteristic prominent eosinophilic infiltration (100x). (c) Spindled stromal cells with hyalinized background. The nuclei are cytologically bland with no mitoses. In the center, the blood vessels have thick and thin walls. The stromal cells surrounding the thick-walled vessel have vague onion-skinning appearance (arrow). (100x). (d) Clear demarcation between the polyp (arrow) and muscularis propria (asterisk). (100x).

yp (Figure 3). The final diagnoses were inflammatory fibroid polyp and acute appendicitis with periappendicitis. The patient's postoperative course was uncomplicated, and she was discharged 3 days later.

IFPs, also known as Vanek's tumors are rare, mesenchymal lesions that arise in the submucosa of the gastrointestinal tract. They can be found anywhere in the gastrointestinal tract; 70% are seen in the antrum of the stomach, 20% in the small intestines, 5% in the colorectal area, 2% in the esophagus, and <1% in the appendix (1). The pathogenesis of this tumor remains unclear but immunohistochemical staining suggests that these tumor cells probably originate from the dendritic cell (2).

Most of the IFPs are asymptomatic and are incidentally found during endoscopic evaluation for other causes or on post-surgical specimens. However, large gastric IFPs are known to cause abdominal pain, early satiety, and gastric outlet obstruction while small intestinal IFPs can present as intussusception, or obstruction. Appendiceal IFPs, on the other hand, can present as right lower quadrant abdominal pain causing or mimicking acute appendicitis (3-5) or may appear as appendiceal masses mistaken as mucocele or carcinoid tumors (1). Microscopically, these polyps are characterized by mononuclear spindle-shaped cells that are typically arranged in a short fascicular pattern surrounding the blood vessels in a concentric "onionskin-like" fashion. Characteristic feature are the prominent infiltration of eosinophils and

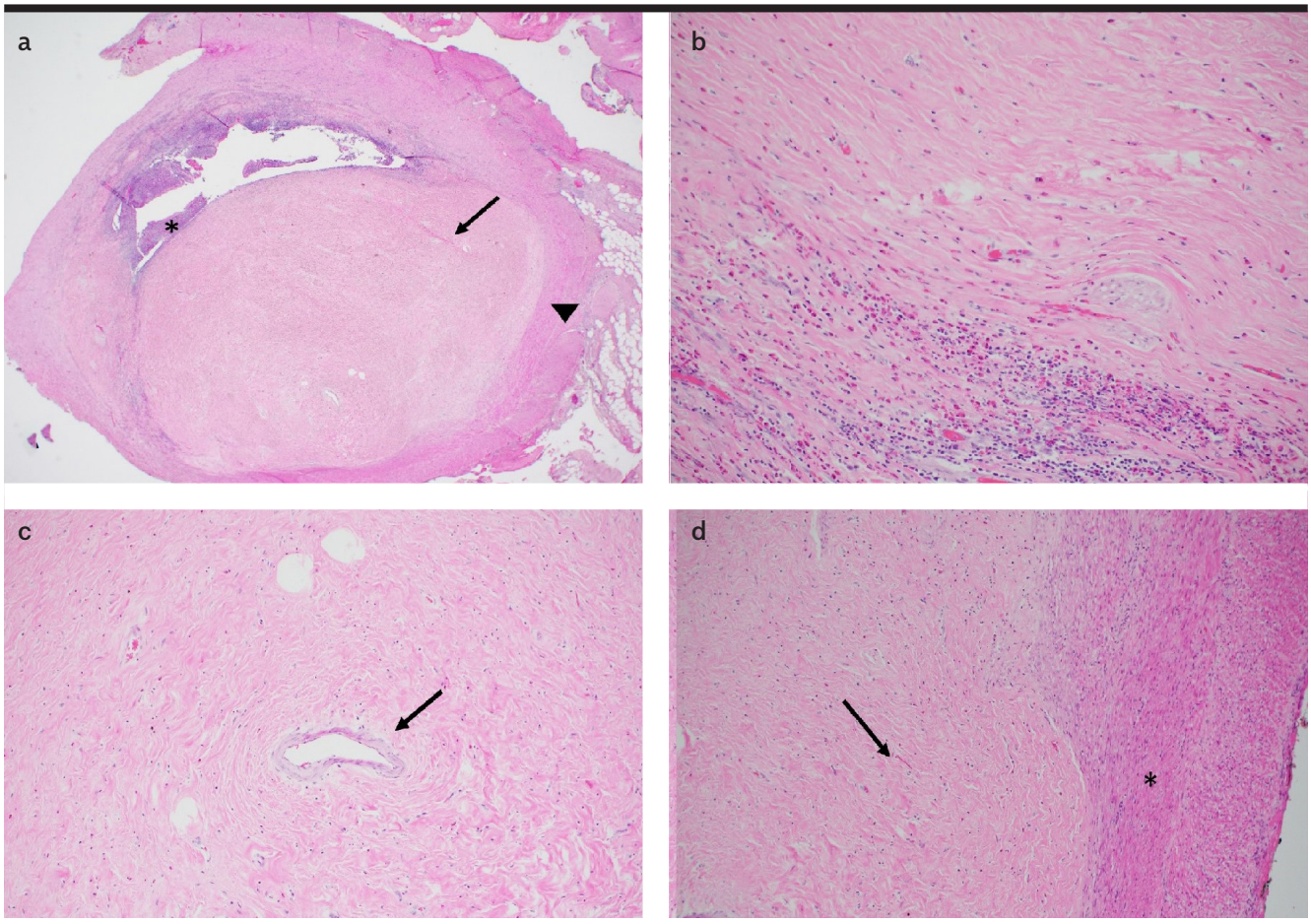

Figure 3. a-d. Immunohistochemical staining. (a) CD 34 stain is positive for both the vessels (arrow) and spindle cells within the polyp (asterisk). (200x). (b-d) CD117, Desmin and S-100 stains are negative for both the spindle cells within the polyp (arrow) and the smooth muscle of muscularis propria (asterisk). (100x).

vessels with surrounding concentric fibroblastic proliferation. Immunohistochemically, the stromal cells of IFP are positive for CD34 and negative for CD117 and S-100, differentiating them from other mesenchymal tumors (6). Unfortunately, laboratory testing and radiological imaging investigation have minimal or no value in the diagnosis of IFP, and in the colonoscopy finding it may be mistaken for an inverted appendix (7).

Symptomatic appendiceal IFPs are treated surgically by appendectomy, either laparoscopic guided or open. In general, IFPs have been considered to be non-neoplastic and do not recur or metastasize. However, a study found some IFPs to have gain-of-function mutations in the platelet-derived growth factor receptor alpha (PDGFRA) gene, similar to those found in CD117-negative gastrointestinal stromal tumors, suggesting the possibility of a neoplastic process needing further investigation (8).

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