







## **Unusual Variations and Atypical Presentations** of Diverticulitis

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Clin Colon Rectal Surg

### **Abstract**

### **Keywords**

- diverticulitis
- segmental colitis associated with diverticulosis

In this article, we describe four unusual variations of diverticulitis: nonsigmoid colonic diverticulitis, giant colonic diverticulum, segmental colitis associated with diverticulosis, and small bowel diverticulitis. We discuss the epidemiology, presentation, and treatment of these types and how they differ from the presentation of typical sigmoid diverticulitis. We also review unusual presentations of typical sigmoid diverticulitis, including hematogenous liver abscess, necrotizing soft-tissue infection, and genitourinary fistula. Diverticulitis is a heterogeneous disease, and understanding the range of its presentations will facilitate early diagnosis and treatment.

Diverticulitis most commonly occurs in the sigmoid colon, presenting as localized inflammation of diverticula with or without colonic perforation. However, less common variations of diverticulitis exist. These include diverticulitis in other areas of the colon, giant colonic diverticula, segmental colitis associated with diverticulitis (SCAD), and small bowel diverticulitis. It is important to understand the distinctions between the pathophysiology, the presenting signs and symptoms, and the treatment of these variations.

Although diverticulitis usually presents with abdominal pain localized to the inflamed segment, atypical presentations are possible for both sigmoid and nonsigmoid diverticulitis. These atypical presentations include hematogenous liver abscess, necrotizing fasciitis, and extraperitoneal fistula formation. The sequelae of these presentations often require additional management compared to diverticulitis presenting only with colonic inflammation.

In this article, we discuss the epidemiology, presentation, and treatment of unusual variations of diverticulitis. We also review atypical presentations of diverticulitis and their treatment. Given its heterogeneity, diverticulitis may present a diagnostic and treatment challenge.

> Issue Theme Maguire Diverticulitis; Guest Editor: Lillias Maguire, MD

DOI https://doi.org/ 10.1055/s-0044-1791553. ISSN 1531-0043.

## **Nonsigmoid Diverticulitis**

While diverticulitis most commonly occurs in the sigmoid or left colon, patients may have diverticulosis anywhere throughout the colon. Diverticular inflammation of other locations in the colon often presents with atypical symptoms, including vague abdominal discomfort and milder pain compared to left-sided diverticulitis.

## **Right-Sided Diverticulitis**

Compared to diverticula in the left colon, right-sided diverticula are more often true diverticula, involving all layers of the bowel wall, and are thought to be congenital in this instance.<sup>1</sup> However, when multiple right-sided diverticula are present, they are generally false diverticula, thought to be from increased intraluminal pressure. The typical teaching is that left-sided disease mostly affects patients in western countries, while patients from Asian countries have a higher predominance of right-sided diverticulitis. Indeed, rightsided diverticulitis accounts for only 1 to 5% of diverticulitis cases in western countries compared to 38 to 75% of diverticulitis cases in Asian countries.<sup>1,2</sup> A meta-analysis of 21

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Thieme Medical Publishers, Inc., 333 Seventh Avenue, 18th Floor, New York, NY 10001, USA

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studies that included 2,811 patients found that patients with right-sided diverticulitis tended to be younger than those with left-sided disease, with a median presenting age of 37 to 54 years.<sup>3</sup>

Right-sided diverticulitis may present similarly to acute appendicitis, although some studies suggest that right-sided diverticulitis may have a longer duration of pain and lower incidence of nausea and vomiting typical of appendicitis. <sup>4-6</sup> The diagnosis is most often made on computed tomography (CT) scan, showing localized inflammation of the ascending colon with single or multiple diverticula. <sup>7</sup> Right-sided diverticulitis presents with complicated disease in 6.2 to 12.4% cases, similar to the 12% rate of complicated left-sided diverticulitis. <sup>3,8</sup>

Recurrence of right-sided disease is thought to be fairly low, with one meta-analysis of 11 studies finding a recurrence rate of 12% after nonoperative management with a median follow-up of 34 months. Only 10% of those who recurred required urgent surgery at the first recurrence, and there was no mortality. Another meta-analysis of 21 studies found a pooled recurrence rate of 11% compared to 15 to 30% for patients with left-sided disease. 3,10,11

Unlike left-sided diverticulitis, there are no consensus guidelines for management of right-sided diverticulitis. Given low recurrence rates, multiple studies support nonoperative management, but ileocolic resection or right colectomy may be required for complicated, refractory, or recurrent disease. 1,12–15

One distinct clinical subtype of right-sided diverticular disease is solitary cecal diverticula. Solitary cecal diverticula are generally located within 1 cm proximally and 2 cm distally to the ileocecal valve on the anterior surface of the cecum. <sup>16,17</sup> They are true diverticula, involving all layers of the bowel wall, and are thus suspected to be congenital in nature. <sup>1,16–18</sup> Overall, they have an incidence of 0.04 to 3.6% of reported cases of diverticulitis. <sup>17,19</sup> They present at an average age of 43.6 years and with predominance in men. <sup>16,17</sup> Most of the recent literature on solitary cecal diverticula consists of case reports or small case series.

Cecal diverticulitis most often presents with right lower quadrant pain, and before the routine use of CT was often misdiagnosed as appendicitis or cecal malignancy. <sup>16–18</sup> Only 3 to 9% of patients with solitary cecal diverticulitis are correctly diagnosed preoperatively, most of whom have had prior appendectomy. <sup>16,18,20</sup> If diagnosed preoperatively, the treatment options for cecal diverticulitis range from nonoperative management to surgery including local diverticulectomy with or without appendectomy, ileocecectomy, or formal right colectomy. <sup>16–18,21,22</sup> Some studies suggest high failure rates with medical management. <sup>17,18,23</sup> In one series from 1991 of 16 patients who underwent laparotomy, patients with a cecal phlegmon underwent right colectomy, while 4 patients who only had diverticular inflammation underwent a diverticulectomy. <sup>21,22,24</sup>

In another series of 49 patients, 80% underwent right colectomy, 14% underwent diverticulectomy, and 6% underwent appendectomy and drain placement (and later required right colectomy).<sup>25</sup> In summary, management of cecal diverticulitis should be tailored to the patient's presentation; if

surgery is required, diverticulectomy can be performed if tissue quality is acceptable, but in the case of more severe or phlegmonous inflammation, ileocecectomy or right colectomy may be needed to incorporate healthy tissue into an anastomosis.

#### Other Locations of Diverticulitis

Diverticulitis in the transverse colon or the rectum is even rarer than in the right colon, with only case reports of either type in the literature. Transverse colon diverticula are usually asymptomatic and found incidentally on colonoscopy, and diverticulitis in the transverse colon accounts for less than 1% of cases of colonic diverticulitis. Transverse colon diverticulitis may present similarly to right-sided diverticulitis with periumbilical pain or with focal tenderness over the inflamed segment, but it may also mimic symptoms of acute pancreatitis. Treatment can be approached similarly to right-sided diverticulitis.

Rectal diverticula are very rare, likely as the rectum lacks discrete taenia coli and as such does not have areas of weaker wall strength compared to the colon. The incidence of rectal diverticula is estimated to be 0.08 to 2% of all colorectal diverticula and may be true congenital diverticula or false acquired diverticula. Rectal diverticula have been associated with scleroderma and rectal prolapse and are typically larger—2.5 cm on average—than colonic diverticula. They are usually asymptomatic and found incidentally, but rectal diverticulitis may present with hematochezia, constipation, pelvic pain, pain with defecation, weight loss, or tenesmus. Surgery is usually only needed in the setting of perforation or intractable symptoms, and surgical management ranges from abscess drainage, diverting colostomy, diverticulectomy, or low anterior resection.

### **Giant Colonic Diverticulum**

A giant colonic diverticulum, defined as a large, air-filled outpouching of the colon greater than 4 cm in diameter, is a rare manifestation of colonic diverticular disease (**Fig. 1**).<sup>36</sup> A systematic review of 166 cases from 2015 found the most common clinical presentation was abdominal pain (69% of cases), followed by a palpable abdominal mass (28%), fever (20%), and constipation (17%).<sup>37</sup> The largest reported giant colonic diverticulum measured 40 cm in diameter.<sup>38</sup> The majority of these are pseudodiverticula from a walled-off perforation or abscess, while others are pulsion diverticula from high intraluminal pressures over time. The rarest type are true diverticula, thought to be congenital and possibly existing on a spectrum with duplication cysts.<sup>39</sup>

Treatment of giant colonic diverticulum includes nonoperative management, local resection with diverticulectomy, or segmental resection including the diverticulum. In the previously mentioned systematic review, the most frequent treatment was segmental resection with primary anastomosis (66%), sigmoid colectomy with end colostomy (11%), and diverticulectomy (10%).<sup>37</sup> Nonoperative management was used in 8% of patients but in many cases was followed by delayed elective sigmoid colectomy. In a subgroup analysis of



Fig. 1 CT of a patient with a giant diverticulum of the sigmoid colon.

the 15 cases reported in the most recent 5 years, 1 patient underwent diverticulectomy and 1 patient received nonoperative management; all other patients underwent resection with either anastomosis or colostomy. Segmental resection appears to be a safe option that minimizes the risk of recurrence, while diverticulectomy is not recommended given the risk of recurrence or leak.<sup>36</sup>

# Segmental Colitis Associated with Diverticulitis

SCAD is characterized by inflammation of the sigmoid colon in the presence of sigmoid diverticulosis.<sup>40</sup> Initially described in the 1980s, SCAD is considered a distinct clinical entity from sigmoid diverticulitis. On endoscopy, mucosal inflammation between the diverticula can be seen, but the orifices of the diverticula themselves are not inflamed, distinguishing SCAD from diverticulitis. 41,42 Endoscopic findings may also include mucosal ulcers and erosions, resembling inflammatory bowel disease (IBD), although SCAD is thought to be distinct from IBD as well.<sup>40</sup> Patients with SCAD are generally in their sixth decade of life and are typically male. 43,44 SCAD is rare, with a prevalence of 1.2 to 11.4% of patients with diverticulosis based on case series ranging from 10 to 129 patients. 40,42,45 Its etiology is not well understood, but one hypothesis is that fecal stasis within diverticula may lead to bacterial overgrowth and an inflammatory response in the affected colonic segment. 43 Another hypothesis is that mucosal redundancy between diverticula leads to mucosal shearing from stool, inducing mucosal hyperemia and inflammation.<sup>46</sup>

SCAD has a heterogeneous presentation, and patients present most commonly with rectal bleeding (present in 76–93% of patients). Bleeding is thought to come from the inflamed mucosa adjacent to diverticula, as opposed to arterial bleeding within the diverticulum itself in typical diverticular bleeding. Diarrhea is the next most common symptom (44–96% of patients) followed by abdominal pain. Systemic signs including fever or leukocytosis are

rare.<sup>41</sup> This presentation differs from typical sigmoid diverticulitis, in which bleeding is uncommon and fever and leukocytosis are typical. In SCAD, the clinical course is generally benign, with most cases resolving with antibiotics (generally ciprofloxacin and metronidazole) and a high-fiber diet. Salicylates such as mesalamine can be used for patients with persistent symptoms, but steroids are rarely needed (in contrast to IBD).<sup>43</sup> Surgery is reserved for refractory cases.

### **Small Bowel Diverticulitis**

Small bowel diverticulitis is a rare condition that can mimic traditional colonic diverticulitis. The reported incidence of small bowel diverticulosis is between 0.6 and 2.3%. <sup>47,48</sup> Small bowel diverticula are twice as common in men than in females, with a peak incidence between the sixth and seventh decades of life. <sup>49</sup> Diverticula are more frequent in the duodenum and least frequent in the ileum. <sup>50,51</sup> Diverticulitis can result from stasis of intestinal contents in the diverticulum and/or mucosal edema resulting in intradiverticular microbial growth. <sup>47,52</sup>

In general, the location of diverticulitis in the small bowel does not necessarily change how a patient may present. Patients often have nonspecific symptoms such as abdominal pain, nausea, vomiting, bloating gastrointestinal bleeding, or changes in bowel habits, which make diagnosis difficult. The diagnosis can usually be made by CT scan. Treatment may include antibiotics to address infection, dietary modifications, and pain management. In the setting of perforation or obstruction, percutaneous drainage or surgery to remove the affected portion of the intestine may be needed. Diverticulectomy and inversion of a perforated diverticulum should be avoided due to risk of recurrent disease or leak. Prompt medical attention and appropriate management are crucial in managing small bowel diverticulitis and preventing potential complications.

A Meckel's diverticulum is a specific type of small bowel diverticula that can manifest with diverticulitis. Meckel's diverticulum is a true, congenital diverticulum that is a remnant of an incompletely obliterated omphalomesenteric or vitelline duct, the fetal connection between the yolk sac and the developing embryonic midgut. While it is usually clinically silent, approximately 4 to 6% can present with symptoms such as gastrointestinal bleeding or acute abdominal symptoms related to bowel obstruction, diverticulitis, or perforation. 58,59 Meckel's diverticulitis can often be mistaken for acute appendicitis. Often, Meckel's diverticulitis can arise due to fecalith obstruction, inflammatory tissue, or foreign body leading to inflammation, necrosis, and possible perforation.<sup>59-61</sup> Meckel's diverticulitis should be managed either with diverticulectomy or with local small bowel resection.61,62

## **Atypical Presentations of Diverticulitis**

Typical symptoms of diverticulitis include abdominal pain, tenderness over the affected segment, fever, and changes in bowel habits. However, atypical presentations of diverticulitis can occasionally perplex clinicians, leading to diagnostic challenges and delays in appropriate management.

### **Hematogenous Liver Abscess**

Liver abscess can occur in diverticulitis via bacterial translocation into the portal venous system through disruption of the colonic mucosal barrier during the acute inflammatory process. These patients may present with sepsis, bacteremia, or symptoms similar to cholangitis, namely, fever, right upper quadrant pain, jaundice, nausea, biliary obstruction, and weight loss. Suspicion for a colonic source of a liver abscess should be raised with polymicrobial infections, including enteric flora or species from the *Streptococcus milleri* group. *Streptococcus milleri* are anaerobic bacteria that are normal members of the gastrointestinal flora, but are opportunistic in nature if seeded elsewhere.

Hematogenous liver abscess can be treated with intravenous antibiotics alone if it is less than 5 cm in size, but source control, by percutaneous drainage, operative drainage, or even liver resection, is required if abscesses are greater than 5 cm or cause persistent systemic illness refractory to antibiotics. After the abscess has been treated and the patient recovers from any deconditioning or infectious complications, resection of the colonic diverticulitis should be pursued to prevent any future recurrences of liver abscess.

### **Extraperitoneal Manifestations**

Fistula formation following diverticulitis occurs in 2% of patients with complicated diverticulitis, which can manifest outside the peritoneal cavity.<sup>64</sup> While colovesical (50%), colovaginal (25%), and coloenteric (7%) are the most common fistulas, there are other atypical fistulous tracts that can obscure the primary diagnosis of perforated diverticulitis.<sup>65</sup> One such fistula is a colocutaneous fistula, which can lead to necrotizing soft-tissue infection (NSTI).<sup>66,67</sup> This may present with signs and symptoms such as subcutaneous emphysema, crepitus, or erythema. In contrast, it may lack more classic features of diverticulitis such as pneumoperitoneum or peritonitis.<sup>66,67</sup> NSTI from diverticulitis may involve the retroperitoneum, abdominal wall, scrotum, or even lower extremity through contiguous spread of infection from enteric and gas-forming flora.<sup>66,67</sup>

Development of an NSTI as a sequela of perforated diverticulitis carries significant morbidity and mortality; however, rates are not known due to the rarity of this presentation. Failure of early debridement may lead to rapid progression of the infection and sepsis. Treatment necessitates soft-tissue debridement as well as definitive source control with resection and typically end colostomy. 66,67

Fistula formation to the urologic or gynecologic systems can also cause an atypical presentation of diverticulitis. Cases have been reported in men in which the patient presented with signs and symptoms of an acute scrotum such as scrotal pain, swelling, and scrotal air on ultrasound. Patients may also have urinary frequency, urgency, dysuria, or nocturia. In these presentations, it is important to keep neighboring anatomy in mind, such as a patent processus

vaginalis or indirect inguinal hernia, that may connect the intra-abdominal cavity to the scrotum.<sup>67,68</sup> The presence of scrotal involvement may delay diagnosis as primary scrotal pathologies are ruled out. CT scan often aids in prompt diagnosis. Additionally, fistulas from the colon to the tubes, ovaries, uterus, or vagina (in the setting of a prior hysterectomy) can form in women and present as tubo-ovarian abscess, air in the uterus, salpingitis, vaginal bleeding, and pelvic pain, with or without more classic symptoms of fecal discharge from the vagina.<sup>64,65</sup>

### **Conclusion**

Diverticulitis may occur throughout the small bowel, colon, and rectum, and has rarer variations including giant diverticulum and SCAD. These variations represent distinct pathophysiology and benefit from a nuanced diagnostic and treatment approach, taking into account patient factors, risk of recurrence, and effects on quality of life. Even typical colonic diverticulitis can have a heterogenous presentation, with liver abscess, fistula between the colon and the urogenital tract or reproductive organs, or even NSTI of the retroperitoneum or abdominal wall. These presentations pose diagnostic challenges and clinicians must be vigilant in recognizing atypical symptoms stemming from diverticulitis as a source.

Funding None.

Conflict of Interest None declared.

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