Giant Colonic Diverticulum Presenting Clinically as a Probable Gastrointestinal Stromal Tumor (GIST)

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Although colonic diverticular disease is relatively common in the Western hemisphere, giant colonic diverticulum (GCD) is an unusual finding, often not included in the initial differential consideration. We present a case of GCD with radiographic features suggestive of an infiltrating neoplasm involving the sigmoid colon, clinically suspicious for gastrointestinal stromal tumor (GIST). Due to its gross appearance and location, GCD can be distinguished from other diseases of the gastrointestinal tract which can have similar characteristics on physical exam and imaging studies. The severe complications of the giant colonic diverticulum and its surgical significance make this condition important to recognize.

CASE PRESENTATION

An 85 year old male was hospitalized complaining of constipation and acute lower gastrointestinal bleeding. Past medical history included: severe arteriosclerotic ischemic heart disease, three-vessel aortocoronary by-pass surgery, diabetes mellitus type II, hypertension, hyperlipidemia, benign prostatic hyperplasia, probable adrenal gland tumor, and osteoarthritis of the lumbosacral spine. Significant laboratory findings included an elevated CEA of 16.89 ng/ml. Colonoscopy revealed severe diverticulosis of the descending and sigmoid colon and a 3.5 cm polyp which was surgically excised. Pathologic examination of the latter revealed benign polypoid colonic mucosa with mild nonspecific inflammation associated with a reactive intramucosal lymphoid nodule without additional significant diagnostic abnormalities.

Approximately one month later, the patient presented with similar recurrent symptoms warranting a second colonoscopy which revealed a submucosal mass in the distal sigmoid colon. A CT scan of the abdomen and pelvis showed a 5 cm, pear-shaped, thick-walled mass at the distal sigmoid colon (Fig.1) associated with a distended bowel lumen proximal to the lesion (Fig.2), clinically suspicious for gastrointestinal stromal tumor (GIST). During the next six months, the patient was regularly followed by a colorectal surgeon.
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Figure 2
Figure 2: CT of pelvis proximal to the lesion: “A dilated segment of colon, proximal to the lesion.

Approximately six months later a third colonoscopy was done, when similar complaints persisted. There was difficulty passing the scope beyond the sigmoid colon. Subsequently, a barium study demonstrated narrowing and irregularity of the proximal sigmoid colon suggestive of diverticular disease. Due to the progressive size increase of the mass, the elevated CEA, and the malignant potential of a GIST, a sigmoid colectomy was performed. The preoperative physical examination demonstrated tenderness over the left lower abdomen with no evidence of a palpable mass. Bowel sounds were present and normoactive. A rectal exam revealed no masses and was negative for occult blood.

PATHOLOGY
The colectomy specimen consisted of a segment of sigmoid colon 12.5 cm long and 4.0 cm in diameter (Figs. 3, 4, & 5). Protruding from the mesenteric surface of the colon was a relatively smooth ovoid to cylindrical mass-like structure 5 cm long by 2 cm in diameter (Fig. 4) consistent with a giant colonic diverticulum (GCD). On section, the lumen of the intestine was patent, with the wall variably thickened up to 1 cm. The cut-surface of the diverticular lesion revealed a lumen approximately 4 cm long, varying up to 2.0 cm in diameter (Fig. 5), narrowly communicating with the intestinal lumen through a stenotic ostium less than 1 mm in diameter (Fig. 5). The diverticular lumen contained minimal amount of fecal debris. The mucosal surface of this large saccular structure was roughened with friable ulcerated red to grey brown necrotic debris. Multiple histopathologic sections of the saccular lesion consisted predominantly of a densely fibrotic wall exhibiting mucosal ulceration and extensive acute and chronic inflammation (Fig. 5), partially involving the mesocolon. Apart from the GCD, multiple grossly discernible diverticula invaginated through the full thickness of the colonic wall.

Figure 3
Figure 3: The surgically resected segment of sigmoid colon viewed from its serosal surface appearing essentially normal.

Figure 4
Figure 4: The mass (GCD) viewed after 90° rotation of the sigmoid segment.
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Figure 5
Figure 5: Longitudinal transmural dissection of the mass (GCD) revealing a lumen consistent with a giant colonic diverticulum. The latter communicates with the sigmoid lumen (shown in transverse xs) through a narrow ostium.

Figure 6
Figure 6: Histopathologic section of the giant colonic diverticulum wall reveals mucosal erosion, severe acute and chronic inflammation, and extensive fibrosis of the remaining subjacent underlying colonic wall.

DISCUSSION
A giant colonic diverticulum (GCD) is a rare complication of diverticular disease involving the colon. Though diverticulosis is highly prevalent in Western countries, there are only slightly over 130 GCD cases reported (1), including two cases describing an associated intestinal obstruction (2, 3). Giant colonic diverticula have been identified throughout the colon, with the majority (81%) occurring in the sigmoid colon (1). While giant colonic diverticulum is the general term used, the condition is often referred to as a giant sigmoid diverticulum due to its predominant location.

In the literature, 90% of reported cases of GCD are associated with diverticulosis. Most patients present at over 60 years of age with initial clinical findings essentially similar to our reported patient(1). The most common symptoms were abdominal pain, constipation, and rectal bleeding. Other reported symptoms include diarrhea, vomiting, nausea, and abdominal distention. Physical signs most noted were the intermittent presence of an abdominal mass and tenderness.

A diverticulum is considered as “giant” when it reaches over 4 cm in greatest dimension (3). Sizes vary but very few cases of GCD have reached over 25 cm. Three different histologic types of giant colonic diverticulum have been described (1, 2). Type I is a pseudodiverticulum with mural granulation tissue, fibrous tissue, and inflammatory cells. Despite lacking a normal smooth muscle layer, a small component of the bowel wall muscle layer may be observed in pseudodiverticula. Type II GCD is an inflammatory diverticulum lined with scar tissue without evidence of any distinct normal bowel wall layers. Localized perforation of the GCD may lead to abscess formation. This is the most common type of giant colonic diverticulum, with 66% of cases reported as histologically consistent with an inflammatory diverticulum (1). Type III GCD is a true diverticulum that involves all three layers of the bowel wall and is likely a congenital defect. A true diverticulum contains distinguishable smooth muscle layers from normal bowel wall (3,4). A more recent classification divides giant colonic diverticula into two types (5); type I is the pseudodiverticulum and type II is the true diverticulum.

Gender distribution among the types of giant colonic diverticula shows equal occurrence in men and women of pseudodiverticula and inflammatory diverticula and a predominantly male occurrence of true diverticulum (1).

Two theories have been suggested to explain the formation of a giant colonic diverticulum (6). A flap-valve mechanism forms from inflammation at the ostium of the diverticulum creating stenosis at its neck trapping bowel gas in its lumen. As intracolonic gaseous pressure increases, the communicating ostium allows more air-trapping, which progressively expands diverticular volume. This relationship between intracolonic and the giant colonic diverticulum pressure also explains the size variability observed in giant colonic diverticula. Alternatively, gas-producing bacteria in a diverticulum may produce enough gas pressure to create a
giant colonic diverticulum. In either theory, the large, extraluminal invagination can develop toward the anti-mesenteric side of colon (3, 4) or the mesenteric side where penetrating blood vessels are present (3, 5) as was the situation in our case.

Preoperative diagnosis of giant colonic diverticulum can usually be established by using CT and barium enema (3, 4). While CT may show a mass and barium enema can identify diverticular disease and locate strictures in the colon, definitive diagnosis of a giant colonic diverticulum by imaging may still be difficult (6). The differential diagnostic consideration of a relatively large mass protruding from the colonic serosal surface, identified on CT, may include giant colonic diverticulum (5) or gastrointestinal stromal tumor (GIST) (11, 12), as seen in our case. Barium studies are contraindicated in cases where perforation is of concern, which limits its use (7). Additionally, if a giant colonic diverticulum is suspected, a barium enema may adversely increase the size of the giant colonic diverticulum during insufflation in the study (8). Although our patient had radiographic evaluations with both CT and barium enema, the final diagnosis of giant colonic diverticulum was established only after the sigmoid resection.

For an adult male over age 60 with diverticulosis, probable differential diagnoses for an increasingly obstructive abdominal mass, abdominal pain, and lower gastrointestinal bleeding would include histologically and grossly distinct conditions such as malignancy, GIST, and volvulus. Colorectal carcinoma would grossly exhibit a partially ulcerative, fungating mass protruding into the bowel lumen. Primary lymphoma of the gastrointestinal tract (MALToma) arises mainly in the mucosal and submucosal layers, which are infiltrated with a monotonous proliferation of monoclonal lymphocytes. Conversely, an inflammatory giant colonic diverticulum protrudes away from the bowel lumen, leaving the healthy bowel patent, and is associated with a heterogeneous inflammatory infiltration of the stroma, consisting of granulocytes, macrocytes, and plasma cells. GIST’s are abdominal masses that are rarely found in the colon and are histologically composed of spindle-cells with epithelioid mesenchymal features. Inflammatory giant colonic diverticulum, commonly located in the sigmoid colon, morphologically shows only inflammatory cells without neoplastic mesenchymal components. Volvulus, the twisting of redundant loops of sigmoid colon, can also resemble a large mass on diagnostic studies. The gross appearance of the resected sigmoid from our patient showed signs of diverticulosis and a lack of redundant bowel.

Ultimately, the giant colonic diverticulum could become an obstructive mass as it enlarges with inflammatory changes, resulting in stricture of the colon (13). Of the other complications reported, 28% include abscess formation, perforation from a thinned diverticulum wall, and cancer, all of which necessitate invasive treatment (14, 15). Treatment of choice, as suggested in the review of reported cases, has been surgical resection of the sigmoid including the giant colonic diverticulum (11, 12). Despite the rarity of fatal complications of diverticular disease, due to the selection of patients with a giant colonic diverticulum and their comorbidities, operative mortality may reach 20% (16).

However, in documented giant colonic diverticular cases, non-fatal post-operative complications such as pneumonia and wound infection occurred at the same rate as fatal complications, each at 5%. Therefore, to prevent further abscess formation, perforation, and recurrence of the giant colonic diverticulum (16), surgical intervention is the definitive treatment.

CONCLUSION

While diverticular disease is relatively common in the Western hemisphere, a giant colonic diverticulum is an unusual occurrence. Due to their gross appearance and location, giant colonic diverticula can be distinguished from other diseases of the gastrointestinal tract which can have similar characteristics on physical exam and imaging studies. Although incidence of the condition is yet to be determined, the severe complications of the giant colonic diverticulum and its surgical significance make this condition important to recognize.

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