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Case Report

Giant Extraluminal Leiomyoma of the Colon: Rare Cause of Symptomatic Pelvic Mass

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Leiomyomas (LMs) may appear throughout the entire gastrointestinal tract but are rarely seen in the colon-rectum and only 5 of those measured greater than 15 cm in diameter. Pain and palpable abdominal mass are the most common symptoms. Surgical resection is the treatment of choice for most LMs. We here describe a case of a 46-year-old woman who presented with a 3-month history of abdominal pain associated with worsening constipation and abdominal distension. A pelvic solid, polylobulate, left-sided mass was noted on examination. Preoperative findings revealed a dishomogeneous sigmoid mass with calcified spots compressing small intestine and bladder. At laparotomy, a large polylobulate and well-circumscribed mass arising from the descending colon mesentery and displacing small intestine, uterus, and ovaries. A segmental colon resection was performed. An extraluminal 18- × 12- × 5-cm paucicellular sigmoid colon leiomyoma was histologically diagnosed. Our case is one of the few giant (>15 cm) sigmoid colon LMs reported in the literature. Although rare and benign in nature, LMs of the colon can cause life-threatening complications that could require emergency treatment and they should be included in the differential diagnosis of large abdominopelvic masses. Follow-up after surgery is necessary for tumors with any atypia or mitotic activity.

Key words: Leiomyoma – Digestive tract – Bowel obstruction – Pelvic mass

Primary leiomyomas (LMs) present most commonly in the female genital tract and skin but they are rarely seen in the colon-rectum. Macroscopically, LMs may be intraluminal, intramural, extramural, or dumbbell.[1,2] The majority of LMs remain asymptomatic until they have reached a large size: most common symptoms are pain and palpable abdominal mass.[1,2] Symptomatic left colon LMs are uncommon and only 5 of those reported in the literature measured greater than 15 cm in diameter.[1] Histologically, most of them arise from the muscularis propria.[1,3] The most important

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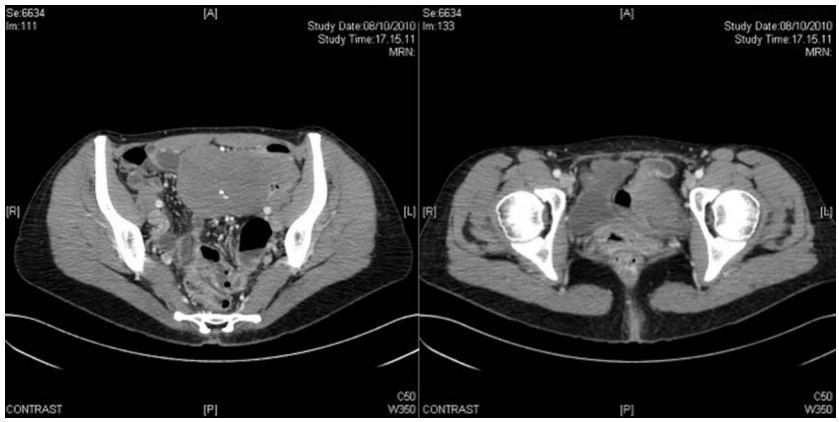

Fig. 1 CT scan showing a solid dishomogeneous mass with calcified spots arising from the mesentery with compressive effect on small intestine and bladder.

differential diagnoses are malignant leiomyosarcoma and gastrointestinal stromal tumors (GIST).[1,3] Surgical resection is the treatment of choice for large LMs: complete excision should be always attempted and follow up is necessary for tumors with any atypia or mitotic activity.[1,2] Although rare and benign in nature, LMs of the colon can cause life-threatening complications that require emergency treatment.[1–3] We here describe a case of large extraluminal sigmoid colon LM causing pain and worsening bowel obstruction.

Case Report

A 46-year-old woman presented with a 3-month history of abdominal pain associated with worsening constipation and abdominal distension. On examination, a pelvic solid, polylobulate left-sided mass was noted. Gynecological examination was negative. Laboratory, including CA-125 and CEA levels, was normal. An abdominal US revealed a polylobulate mass close to the left ovary and left colon. Colonoscopy revealed an external sigmoid stricture. A computed tomography scan showed a dishomogeneous mass with calcified spots compressing small intestine and bladder and probably arising from the mesentery with a compressive effect on the small intestine and bladder (Fig. 1). Thus, the patient underwent exploratory laparotomy: small intestine, uterus, and ovaries were displaced by a large polylobulate and well-circumscribed mass arising from the descending colon mesentery (Fig. 2a). A segmental colon resection was performed with an end-to-end hand-sewing colocolic anastomosis. The surgical specimen consisted of a 12-cm colon with an extraluminal capsulate 18- × 12- × 5-cm mass (Fig. 2b). The neoplasia involved full thickness in the colon wall without mucosal infiltration. Histologically, the tumor was composed of spindle cells with oval to moderately elongated, blunt-ended nuclei and eosinophilic cytoplasm, resembling well-differentiated smooth muscle cells. The neoplastic cells were arranged to form intersecting fascicles (Fig. 3a, 3b) and contained multiple eosinophilic globules, well-formed round hyaline bodies, markedly highlighted by brightly periodic Schiff stain (PAS), mimicking the skeinoid fibers commonly seen in gastrointestinal stromal tumors (GIST; Fig. 3c). In the tumor, mitotic figures (0 mitoses/50 HPF), necrotic areas, or nuclear atypia were consistently undetectable. Immunohistochemically, the neoplastic cells were markedly positive for α-smooth-muscle actin and desmin (Fig. 3d) and were consistently negative for

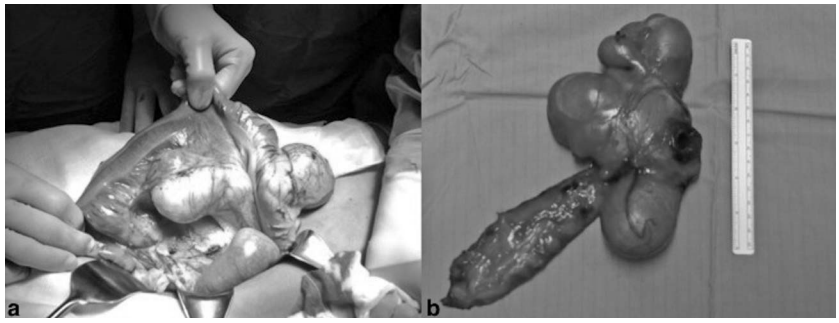

Fig. 2 Surgical findings. (a) Intraoperative photograph showing the large, polylobulate, and well-circumscribed mass arising from the descending colon mesentery. (b) View of the gross specimen: 12 cm of colon with a capsulate extraluminal 18- × 12- × 5-cm mass, arising from the mesentery and compressing the colic lumen.

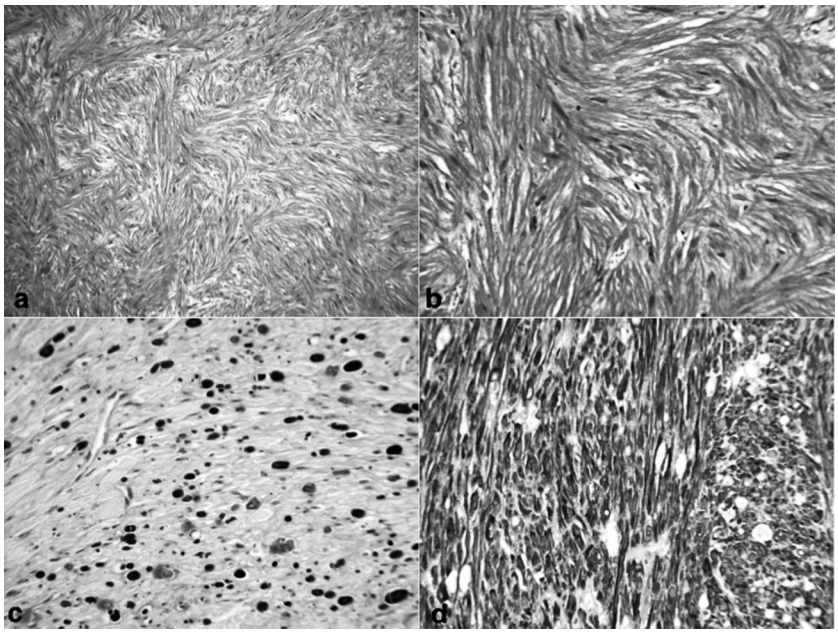

Fig. 3 Microscopic examination of the tumor. (a, b) Fascicles of spindle cells with regular "cigar-shaped" nuclei and eosinophilic cytoplasm, without evidence of mitotic figures, necrotic areas, nor nuclear atypia (hematoxylin and eosin; ×63 and ×160, respectively). (c) Different-shaped well-formed hyaline bodies as eosinophilic blobs mimicking extracellular skeinoid fibers of GIST (PAS stain; ×160). (d) Intense positive desmin immunostain in the cytoplasm of neoplastic cells (×160).

c-Kit (CD117), CD34, DOG-1, S-100 protein, and β-catenin; proliferative index evaluated by Ki67 immunostaining accounted for less than 1%. Based on the histologic and immunohistochemical features, a paucicellular sigmoid colon leiomyoma was diagnosed. The patient is disease free after 36 months of follow-up.

Discussion

Leiomyomas arising from the colon are extremely rare and represent only 3% of all gastrointestinal leiomyomas.[1–5] The descending and sigmoid colon seem to be the most frequent sites of its occurrence in the colon, typically presenting as incidental small intraluminal polyps.[1–3] Skandalakis *et al*,[2] originally reviewing the world literature between 1875 and 1959 while Tarasidis[3] and Hatch[1] updated these data: a total of 331 colon LMs is reported between 1875 and 1996. Hatch *et al* found that 43% of colon LMs were less than 5 cm in greatest diameter and only 5 of the 25 descending/sigmoid colon LMs measured greater than 15 cm in diameter.[1] Our case measured 18 cm in greatest diameter: in the recent years, only Sayer *et al*[4] reported a case of similar size, but located in the rectal wall; while Anagnostou[5] and Weston[6] reported cases of larger masses, 26 cm and 27 cm, respectively. Macroscopically, LMs may be intraluminal, intramural, extramural, or dumbbell: the intraluminal type can be detected earlier because of the early presentation of symptoms.[1,3,7] Imaging findings are nonspecific: calcification and varying degrees of necrosis or cystic changes on CT scan can be seen in both benign and malignant lesions.[1,7] Often, as in our case, endoscopy can demonstrate only an external stricture, although an associated ulceration could be detected.[1,4,7,8] Preoperative biopsy, moreover, is not always possible and clarifying.[1] So, in most cases, a combination of endoscopy with complementary investigations such as CT scan, magnetic resonance imaging, endoscopic ultrasonography, strongly corroborates the diagnosis.[1,3–7] Pain and palpable abdominal mass are the most common symptoms.[1–3] Rarely acute surgical abdomen may be due to complications as ulceration,[10] intussusception,[11] and perforation.[12] Our patient presented with abdominal pain associated to worsening obstructive symptoms with diagnosis of pelvic mass probably arising from the mesentery but utero-adnexal origin could not be excluded, although CA-125 and gynecological examination were negative.

Histologically, LMs arise from the muscularis propria and distinction with leiomyosarcomas is difficult and mainly based on the presence of necrosis, nuclear pleomorphism, cellularity, tumor size, and the number of mitotic figures: to avoid a misdiagnosis, due to considerable variability in different areas of the same tumor, adequate sampling is essential, while immunohistochemistry distinguish them from GISTs.[1–3,13]

About treatment, a variety of operative techniques have been employed to treat these tumors, from simple endoscopic excision for small endolu-

minal LMs to subtotal colectomy.[1–3] Surgical resection is the treatment of choice for most LMs. The extent of resection depends on the nature, size, and location of the tumor. For a small, apparently benign tumor, excisional biopsy is possible while for larger lesions, as our case, wedge colon resection is necessary to obtain complete mass removal.[9]

Regarding long-term results, Hatch *et al* analyzed survival in 89 patients with LMs: 7 died from the disease without having surgery and 77 had surgery; of these, 1 patient died immediately postoperatively and 2 died of unrelated causes. Thus, the overall prognosis is good and recurrence has never been reported in the literature.[1]

Finally, our case is one of the few giant sigmoid colon LMs reported in the literature. Although rare and benign in nature, LMs of the colon may cause life-threatening complications that require emergency surgery and they should be included in the differential diagnosis of large abdominopelvic mass; a gynecological, urological, or retroperitoneal origin is more likely, but a colic origin should not be excluded. Ensuring the complete removal and follow-up are necessary precautions for tumors with any atypia or mitotic activity.

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