

## Extrapelvic Uterus-like Masses Presenting as Colonic Submucosal Tumor: A Case Study and Review of Literature

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A uterus-like mass (ULM) is a central cavity lined by endometrial glands and stroma and surrounded by thick-walled smooth muscles. To date, 31 cases of ULM have been reported in the English literature. ULM typically presents as a single mass and is located in the pelvic cavity. We report here a very rare case of multiple extrapelvic ULMs involving the cecum, descending colon, and mesocolon. After extensive literature research, our case appears to be the first case of multiple ULMs found in extrapelvic sites and the first case of ULM in the colon. The present case suggests that ULM should be included in the differential diagnosis of colonic submucosal tumors in female patients with chronic abdominal pain or menstruation-associated symptoms.

**Key Words:** Uterus-like mass; Endomyometriosis; Adenomyoma; Colon

Uterus-like mass (ULM) is a rare benign tumorous lesion. It presents as a central cavity lined by endometrial glands and stroma and surrounded by thick-walled smooth muscles. ULM was first described in a 31-year-old woman by Cozzutto,<sup>1</sup> and 30 subsequent cases have been reported to date in the English literature.<sup>2-15</sup> Typically, ULM presents as a single mass and is located in a variety of organs in the pelvic cavity, including the ovary,<sup>2</sup> uterus,<sup>3</sup> uterine ligament,<sup>4</sup> pelvic wall,<sup>5</sup> conus medullaris,<sup>6</sup> obturator lymph node,<sup>7</sup> inguinal soft tissue,<sup>8</sup> and vagina.<sup>9</sup> We herein report a very rare case of multiple extrapelvic ULMs involving the cecum, descending colon, and mesocolon in a 39-year-old woman with chronic abdominal pain and iron deficiency anemia. To the best of our knowledge, multiple extrapelvic ULMs of the colon have not yet been reported.

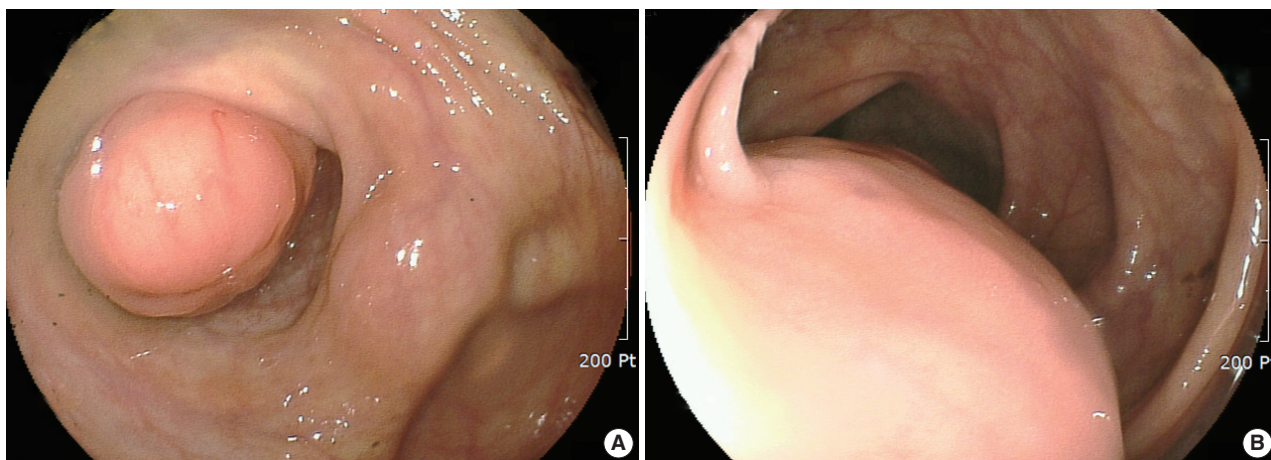
### CASE REPORT

A 39-year-old woman was admitted to the gynecology department for continuous abdominal pain in the right lower quadrant and which radiated to the right thigh. She denied dyspepsia or bowel habit changes. Her medical history was remarkable for chronic abdominal pain and ovarian endometriosis two years ago, which were managed by total hysterectomy and left salpingo-oophorectomy. Physical examination revealed mild tenderness on the suprapubic area and right lower quadrant. Laboratory data included a white blood cell count of 6,200/mm<sup>3</sup>, a hemoglobin concentration of 7.8 g/dL, and a platelet count of 397,000/mm<sup>3</sup>. The work-up for anemia revealed serum iron of 59 µg/dL, total iron-binding capacity of 454 µg/dL,

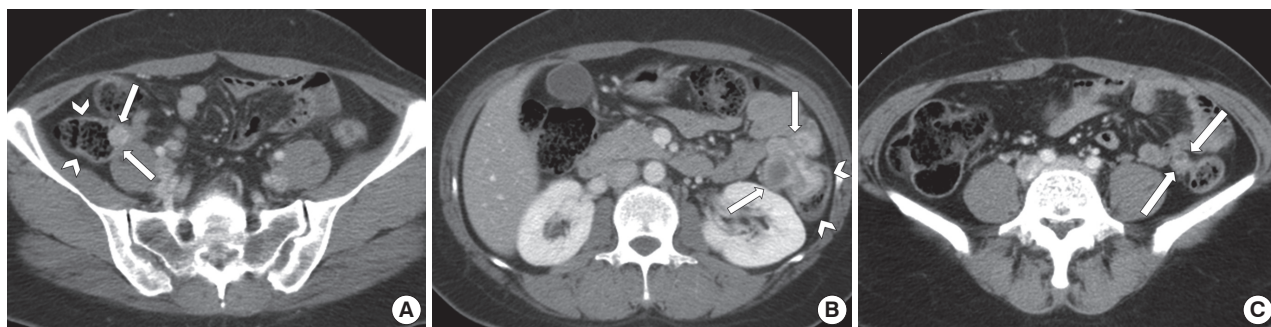
serum ferritin of 70 ng/mL, and transferrin saturation of 12%, which were consistent with iron deficiency anemia. Pelvic ultrasonography revealed a 6×4 cm-sized multilocular cystic mass in the right adnexa. Ovarian endometriosis was suspected, and laparoscopic right salpingo-oophorectomy was performed. The histologic diagnosis was a hemorrhagic corpus luteum. Despite the surgery, her symptoms persisted. Considering the possibility of gastrointestinal bleeding, colonoscopy was performed. Colonoscopy demonstrated a protruding, firm submucosal mass at the appendiceal orifice of the cecum that was covered by intact mucosa (Fig. 1A). In addition, another lesion suggestive of an extrinsic compression rather than a broad based submucosal tumor was noted in the descending colon (Fig. 1B). Contrast-enhanced computed tomography of the abdomen and pelvis showed a small luminal protruding mass with enhancement in the medial wall of the cecum and another lobulating mass with contrast enhancement that contained a small internal area of low attenuation in the descending colon (Fig. 2A, B). The latter

lesion was located both inside and outside of the colon and appeared dumbbell-like in shape. In addition, another small enhancing mass containing an area of low attenuation was observed in the descending mesocolon (Fig. 2C). Tumor marker levels, including alpha-fetoprotein, carcinoembryonic antigen, and carbohydrate antigen 19-9, were within normal ranges. With a presumptive diagnosis of gastrointestinal stromal tumor or intestinal endometriosis, laparoscopic exploration was performed, and the three masses were resected by cecectomy and left hemicolectomy.

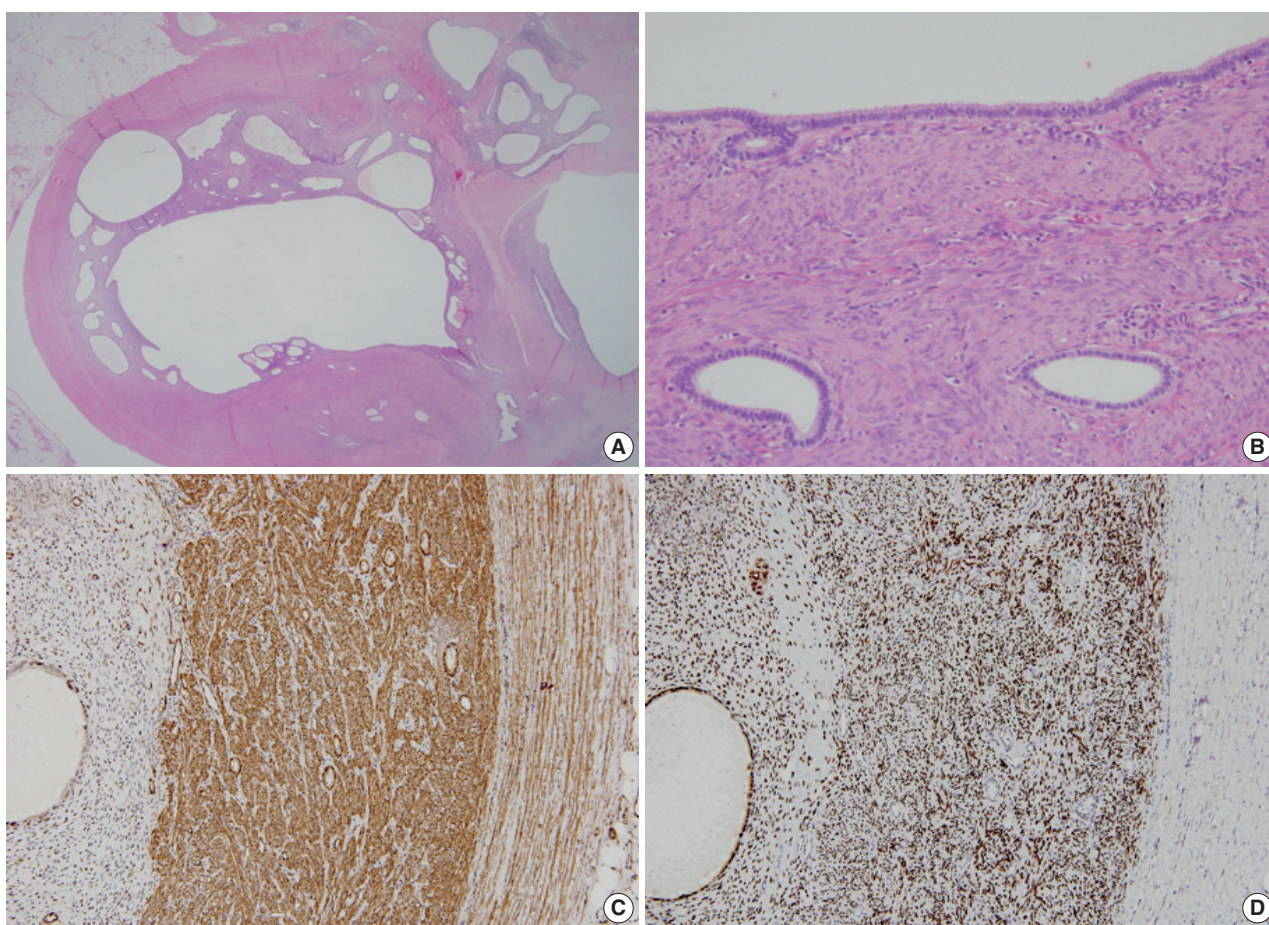
Grossly, the cut surfaces of these masses showed a central cavity containing serosanguinous fluid, lined by a thick, soft, tan-yellow leiomyomatous wall. Histologically, the central cavity was lined by endometrial glands and stroma, which were surrounded by smooth muscles. In addition, small endometrial glands and stroma were scattered within the masses (Fig. 3A, B). On immunostaining, the endometrial glandular cells within the masses were positive for estrogen receptor (ER; 1:25, Novo-



**Fig. 1.** Colonoscopy shows polypoid masses covered by intact mucosa in the cecum (A) and descending colon (B).



**Fig. 2.** Computed tomography findings of uterus-like masses. A small, ovoid, enhancing mass (arrows) is shown in the lumen of the cecum (arrowheads) (A). A dumbbell-shaped, lobulating solid mass containing a cystic portion (arrows) is observed in the descending colon (arrowheads) (B). A round, solid mass containing a cystic portion (arrows) is noted in the medial aspect of the descending colon (C).



**Fig. 3.** (A, B) Histologically, the mass shows a cavity lined by endometrial glands and stroma and surrounded by smooth muscles. (C) Immunostaining with smooth muscle actin is positive in the smooth muscle cells and focally positive in the endometrial stromal cells of the mass. (D) Immunostaining with estrogen receptor is positive in all components of the mass (left), whereas it is negative in the colonic smooth muscles (right).

castra, Newcastle upon Tyne, UK) and progesterone receptor (PR; 1:200, Novacastra). The endometrial stromal cells were positive for ER, PR, and CD10 (1:500, Novacastra) and focally positive for smooth muscle actin (SMA; 1:4,000, Dako, Glostrup, Denmark). The smooth muscle cells of the masses were positive for ER, PR, and SMA and focally positive for CD10. Colonic smooth muscles adjacent to the masses were negative for ER, PR, and CD10 (Fig. 3C, D). These findings were consistent with a previously-reported case of ULM.<sup>2</sup>

The patient was discharged two weeks after surgery without postoperative complication. On follow-up, she reported no pain or shown evidence of anemia for 15 months.

## DISCUSSION

In the first report of ULM, Cozzutto<sup>1</sup> suggested the use of the terms 'ULM' or 'metriosis' based on the distinctive morpholog-

ic features. Grossly, the lobular architecture and central cavity of the mass resembled that of the uterus. Histologically, the mass had a well-organized endometrium and myometrium. To date, 31 cases of ULM have been reported in the English literature with variable terms such as endomyometriosis<sup>4</sup> and ULM with features of an extra-uterine adenomyoma.<sup>9</sup>

The clinical features of these cases and the present case are summarized in Table 1. The ages of the patients ranged from 11 to 59 years, with most patients in their 40s and 50s. The ovary was most frequently involved (n=9), followed by the uterus (n=6), uterine ligament (n=4), and pelvic walls (n=3). The most common presenting symptom was abdominal pain (n=18), frequently associated with menstruation-related symptoms. One-third of the patients had endometriosis (n=8) or adenomyosis (n=2).

In this case report, we describe multiple extrapelvic ULMs involving the cecum, descending colon, and mesocolon. Most

**Table 1.** Clinical features of previously reported cases of uterus-like mass

Authors	Age/sex	Location	Size (cm)	Clinical presentation	Treatment	Associated conditions
Cozzutto <sup>1</sup>	32/F	Ovary	6	LAP	Mass excision	Unilateral renal agenesis, endometriosis
Pai <i>et al.</i> <sup>2</sup>	38/F	Both ovaries <sup>a</sup>	4 and 3	Incidental finding	BO	Invasive ductal carcinoma
	43/F	Ovary	11.5	Pelvic mass	Mass excision	None
	39/F	Ovary	13	LAP, vaginal spot	TH+UO	Adenomyosis
Tijani <i>et al.</i> <sup>3</sup>	35/F	Uterus	21	LAP, dysmenorrhea	Mass excision	None
Matsuzaki <i>et al.</i> <sup>4</sup>	29/F	Uterosacral ligament	7.5	LAP, dysmenorrhea	Mass excision	None
Carinelli <i>et al.</i> <sup>5</sup>	46/F	Pelvic wall and mesentery <sup>a</sup>	Up to 10	LAP, constipation	Partial colectomy	Endometriosis, leiomyomatous nodules
	39/F	Ovary and pelvic wall <sup>a</sup>	Up to 3.5	LAP, dysmenorrhea	Mass excision	Endometriosis, leiomyomatous nodules
Sharma <i>et al.</i> <sup>6</sup>	33/F	Conus medullaris	3	Lower back mass	Mass excision	Spina bifida, subcutaneous lipoma, tethered cord
	24/F	Conus medullaris	NA	Lower back pain	Mass excision	Tethered cord
Rohlfing <i>et al.</i> <sup>7</sup>	35/F	Obturator lymph node	4.5	Lymph node enlargement	TH+mass excision	Squamous cell carcinoma of cervix
Seki <i>et al.</i> <sup>8</sup>	44/F	Inguinal soft tissue	3.8	LAP	Mass excision	Endometriosis
Redman <i>et al.</i> <sup>9</sup>	50/F	Vagina	8	LAP, dysuria	Mass excision	Ureteral stricture
Peterson <i>et al.</i> <sup>10</sup>	12/F	Ileum	2.5	Incidental finding	Segmental resection	Lower intestinal and urinary tract anomaly
Horie and Kato <sup>11</sup>	59/F	Ileal mesentery	14	Abdominal mass	Mass excision	None
Shin <i>et al.</i> <sup>12</sup>	31/F	Sigmoid mesocolon	3	LAP	Partial colectomy	None
Rougier <i>et al.</i> <sup>13</sup>	18/F	Conus medullaris	2.5	Paresthesia	Mass excision	Spina bifida
Jung <i>et al.</i> <sup>14</sup>	43/F	Uterus	3	Menorrhagia, dysmenorrhea	TH	Adenomyosis
	52/F	Uterus	8	Vulvar itching	TH+USO	Endometriosis
Pai <i>et al.</i> <sup>15</sup>	25/F	Broad ligament	20	Abdominal mass	Mass excision	None
Present case	39/F	Cecum, colon, and mesocolon <sup>a</sup>	5	LAP, anemia	Partial colectomy	Endometriosis

F, female; LAP, low abdominal pain; TH, total hysterectomy; UO, unilateral oophorectomy; BSO, bilateral salpingo-oophorectomy; BO, bilateral oophorectomy; USO, unilateral salpingo-oophorectomy; NA, not available.

<sup>a</sup>Multiple uterus-like masses.

ULMs have been reported in the pelvic cavity, except for three cases involving the ileum,<sup>10</sup> ileal mesentery,<sup>11</sup> and sigmoid mesocolon.<sup>12</sup> To the best of our knowledge, ULM of the colonic segment has not yet been reported. Together with the three previous cases of extrapelvic ULMs, the present case supports the notion that ULMs can involve regions outside of the pelvic cavity. Another unique finding of the present case is the multiplicity of the lesions. Twenty-eight of the 31 ULMs presented as a single mass. Three previous cases of multiple ULMs were reported to involve the ovary<sup>2,5</sup> and pelvic wall.<sup>5</sup> We report for the first time the extrapelvic involvement of ULM, without accompaniment of the usual sites of involvement.

In this case, a part of the mass was abutting the smooth muscles of the colon. Several histological and immunohistochemical findings distinguish the smooth muscles of the ULM and the colon. As previously mentioned by Cozzutto,<sup>1</sup> the smooth muscle components of ULMs are histologically identical to myometrium, which have irregularly arranged muscle fibers, and are different from the inner circular and outer longitudinal smooth muscles of the colon, which have parallel arrangements. In ad-

dition, immunostaining demonstrated that the smooth muscles of the ULM were positive for ER and PR, two markers that are not found in colonic smooth muscles.

Three theories have been suggested regarding the histogenesis of ULMs: 1) the congenital anomaly theory, 2) the heterotopia theory, and 3) the metaplasia theory. The congenital anomaly theory is supported by several reports showing the coexistence of developmental disorders in the lower gastrointestinal tract and the urinary tract in patients with ULMs. Rosai<sup>16</sup> suggested that ULMs result from either a Müllerian duct fusion defect or a partial duplication of the Müllerian system. The heterotopias theory was suggested by Peterson *et al.*,<sup>10</sup> who reported an ileal ULM in a patient with multiple anomalies in the lower genital and intestinal tracts, as well as a sacrococcygeal anomaly. In addition, based on reports of ULMs found in the spinal cords of patients with spina bifida, Rougier *et al.*<sup>13</sup> and Sharma *et al.*<sup>6</sup> suggested that heterotopic Müllerian duct tissue within the neural tube can result in ULMs.

The metaplasia theory has been most widely accepted in reports of patients with ULMs who have no other identifiable

malformations. In their report of an ovarian ULM, Cozzutto<sup>1</sup> postulated that ovarian endometriosis elicits the transformation of ovarian stromal cells to smooth muscle cells via myofibroblasts. This hypothesis is linked to the observation that smooth muscle metaplasia can be associated with endometriosis. Given the reports showing the various locations of ULMs, peritoneal mesothelial cells and endometrial stromal cells have been speculated to be the sites of origin of the metaplastic cells.<sup>14</sup> In the present case, the patient did not have any other anomalies, and therefore the metaplastic theory seems to be the most appropriate. In addition, our findings of multiple endometriotic foci outside of the mass suggest the possibility of smooth muscle metaplasia from pre-existing endometriosis. Our immunohistochemical findings of CD10-positive smooth muscle cells and SMA-positive endometrial stromal cells within the mass may reflect the close association between these cells.

In conclusion, we have reported the first case of multiple ULMs with extrapelvic involvement in a patient with chronic abdominal pain and iron deficiency anemia. Although ULM is rare, extrapelvic ULM should be considered as a differential diagnosis for colonic submucosal tumors in female patients who have chronic abdominal pain or menstruation-associated symptoms.

### Conflicts of Interest

No potential conflict of interest relevant to this article was reported.

### REFERENCES

1. Cozzutto C. Uterus-like mass replacing ovary: report of a new entity. *Arch Pathol Lab Med* 1981; 105: 508-11.
2. Pai SA, Desai SB, Borges AM. Uteruslike masses of the ovary associated with breast cancer and raised serum CA 125. *Am J Surg Pathol* 1998; 22: 333-7.
3. Tijani el H, Meryem T, Lamy GA, Abdelouahed J. Giant uterus-like mass of the uterus. *Indian J Pathol Microbiol* 2010; 53: 793-5.
4. Matsuzaki S, Murakami T, Sato S, Moriya T, Sasano H, Yajima A. Endometriosis arising in the uterosacral ligament: a case report including a literature review and immunohistochemical analysis. *Pathol Int* 2000; 50: 493-6.
5. Carinelli S, Motta F, Frontino G, Restelli E, Fedele L. Multiple extra-uterine adenomyomas and uterus-like masses: case reports and review of the literature. *Fertil Steril* 2009; 91: 1956.e9-11.
6. Sharma MC, Sarkar C, Jain D, Suri V, Garg A, Vaishya S. Uterus-like mass of müllerian origin in the lumbosacral region causing cord tethering: report of two cases. *J Neurosurg Spine* 2007; 6: 73-6.
7. Rohlfing MB, Kao KJ, Woodard BH. Endometriosis: possible association with leiomyomatosis disseminata and endometriosis. *Arch Pathol Lab Med* 1981; 105: 556-7.
8. Seki A, Maeshima A, Nakagawa H, *et al.* A subserosal uterus-like mass presenting after a sliding hernia of the ovary and endometriosis: a rare entity with a discussion of the histogenesis. *Fertil Steril* 2011; 95: 1788.e15-9.
9. Redman R, Wilkinson EJ, Massoll NA. Uterine-like mass with features of an extrauterine adenomyoma presenting 22 years after total abdominal hysterectomy-bilateral salpingo-oophorectomy: a case report and review of the literature. *Arch Pathol Lab Med* 2005; 129: 1041-3.
10. Peterson CJ, Strickler JG, Gonzalez R, Dehner LP. Uterus-like mass of the small intestine: heterotopia or monodermal teratoma? *Am J Surg Pathol* 1990; 14: 390-4.
11. Horie Y, Kato M. Uterus-like mass of the small bowel mesentery. *Pathol Int* 2000; 50: 76-80.
12. Shin SY, Kim HJ, Kim YW, Lee KY. CT characteristics of a uterus-like mass in the sigmoid mesocolon. *Br J Radiol* 2011; 84: e1-3.
13. Rougier A, Vital C, Caillaud P. Uterus-like mass of the conus medullaris with associated tethered cord. *Neurosurgery* 1993; 33: 328-31.
14. Jung WY, Shin BK, Kim I. Uterine adenomyoma with uterus-like features: a report of two cases. *Int J Surg Pathol* 2002; 10: 163-6.
15. Pai RR, Sharma L, Pinto PJ, Raghuvveer CV. Endometriosis. *J Indian Med Assoc* 1995; 93: 326.
16. Rosai J. Uteruslike mass replacing ovary. *Arch Pathol Lab Med* 1982; 106: 364-5.