

# Obstructive Mucocele of the Appendix Secondary to Endometriosis

## - A Case Report -

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Appendiceal mucoceles are usually associated with hyperplastic or neoplastic mucosal proliferation and obstructive lesions such as postinflammatory scarring, fecalith, carcinoid tumor, and endometriosis. Among these, an association with endometriosis is known to be very exceptional. We herein report on a rare case of obstructive mucocele of the appendix that was secondary to endometriosis in a 42-year-old patient with pelvic endometriosis. A computed tomography scan demonstrated a periappendiceal abscess-like lesion with a left adnexal mass that was suggestive of endometriosis. On gross examination, the periappendiceal lesion consisted of a mucin-filled cavity (the so-called mucocele) that was 1.8 cm in diameter, and it protruded into the cecal lumen. Microscopically, the lining epithelium of the cavity was almost totally denuded. A small amount of mucus spilled over outside the mucocele, but pseudomyxoma peritonei was not present. The wall of the mucocele showed the characteristic multiple foci of endometriosis involving predominantly the muscularis propria and the serosa of the appendix and adjacent cecal walls.

**Key Words** : Appendix; Mucocele; Endometriosis

For the appendiceal disease traditionally designated as mucocele, the appendix shows a localized or diffuse globular enlargement that is associated with excess glairy mucin production. A few instances of appendiceal mucoceles are caused by mucous distention secondary to the obstruction of the appendix, and these are the so-called obstructive, retention, or simple mucoceles. Obstruction can be caused by fecaliths, postinflammatory scarring, carcinoid tumor, or rarely, endometriosis.<sup>1</sup> We report here on a peculiar case of obstructive appendiceal mucocele secondary to periappendiceal endometriosis that presented as a polypoid mass protruding into the cecal lumen. To the best of our knowledge, this is only the eighth case of such type in the English literature since the first description by Hapke and Bigelow in 1977,<sup>2</sup> and it is the first case to be reported in Korea.

### CASE REPORT

A 42-year-old woman (2-2-0-2) was admitted because of progressive lower abdominal pain and backache of 4 months' duration. She had been told that she had a left ovarian cystic mass

that was discovered by ultrasonography (US) performed at the local hospital three months ago. Otherwise her medical history was also unremarkable. The routine laboratory tests were normal, including the C-reactive protein and white blood cell count. A computed tomography (CT) scan demonstrated an ill-defined, multiseptated left adnexal mass that showed adherence to such adjacent organs as the urinary bladder, rectum and uterus. Additionally, a periappendiceal abscess-like lesion was also noted. The radiologic diagnosis was pelvic endometriosis, involving the left ovary and its adjacent organs, and periappendiceal abscess. She underwent an operation. The surgeons noticed that there were several blood-filled cystic lesions in both the cul-de-sac and left ovary. The appendiceal region was replaced by fibrotic tissue with a gross loss of the appendiceal outline. The frozen diagnosis of the left ovarian lesion was endometriosis. Ileocecal resection, total abdominal hysterectomy and bilateral salpingo-oophorectomy were performed.

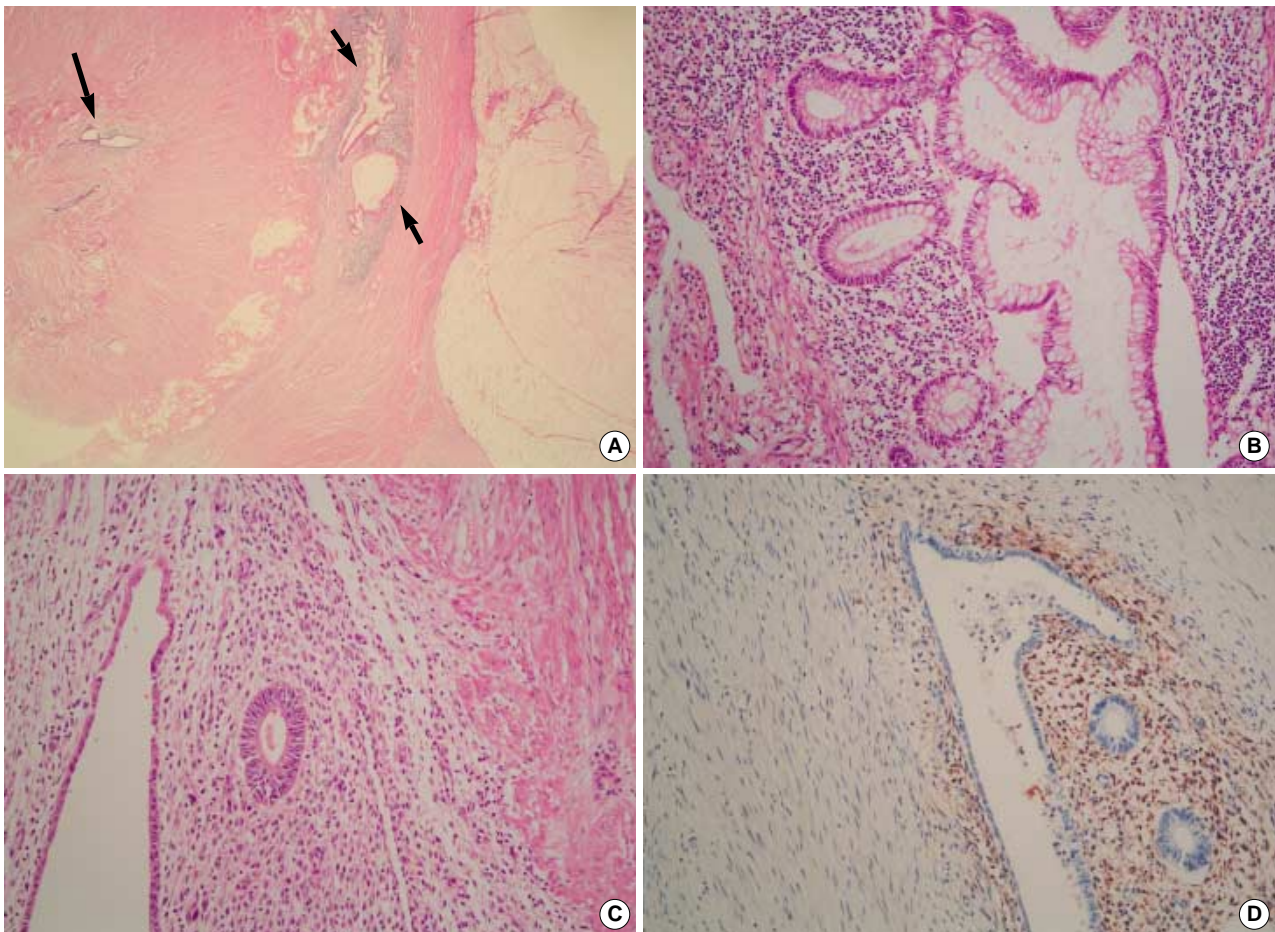
Upon gross examination, the opened cecal portion of the ileocecal resection showed a conically protruding mass, measuring 3.5 × 3.5 × 3.0 cm for its dimensions, and it was covered with intact mucosa. On sectioning, the lower center of the cecal mass

consisted of a well-circumscribed cavity, 1.8 cm in diameter, and it was filled with yellow mucoid materials (Fig. 1). The mucin-



**Fig. 1.** Appendiceal mucocele is conically protruding into cecal lumen and shows a well circumscribed, mucin-filled cavity bordered on the serosa.

filled cavity was surrounded by fibrotic tissue with minute hemorrhagic spots. The proximal lumen of the appendix was almost completely obliterated. The overlying cecal and ileal mucosa was unremarkable. Microscopically, the mucocele was filled with mucus and some inflammatory cells, and its lining epithelium was almost totally denuded (Fig. 2A). A small amount of mucus had spilled over from the mucocele into the surrounding tissue and resulted in extravasated mucus pools, but there was no evidence of proliferating epithelium in the mucus pools. Small irregular lumina were found in some areas around the cavity. Those lumina were lined by tall, columnar, mucous cells showing hyperplastic changes, and the lumina were supported by lymphocytic cells (Fig. 2B). Additionally, around the mucocele, there were multiple foci of endometriosis predominantly involving the muscularis propria and the serosa of the appendix and adjacent cecal wall. The endometriotic glands were surrounded by sheets of small, round to oval stromal cells (Fig. 2C). Some of the glandu-



**Fig. 2.** (A) The cavity of the mucocele on the right side shows a nearly total loss of epithelial lining and the wall reveals residual appendiceal mucosa (short arrows) and scattered islands of endometriosis (long arrow). (B) Residual appendiceal mucosa shows hyperplastic colonic-type epithelium supported by lymphoid tissue. (C) Endometrial glands of proliferative feature are surrounded by endometrial stromal cells. (D) Diffuse immunoreactivity for CD10 is noted in endometrial stromal cells of the endometriotic lesion.

lar lumina contained blood. On immunohistochemistry the hyperplastic, columnar mucous epithelium mentioned earlier was occasionally immunoreactive to Ki-67 antibody (K-3, Oncogene, San Diego, CA, USA; 1:100). Therefore, this tissue was regarded as entrapped appendiceal mucosa. The stromal cells surrounding the endometriotic glands were diffusely immunoreactive to CD10 antibody (56C6, NeoMarkers, Fremont, CA, USA; 1:50) (Fig. 2D). The final pathologic diagnosis was obstructive mucocele of the appendix, secondary to periappendiceal endometriosis. The uterine serosa, left ovary and salpinx that were examined together also revealed endometriosis. The right ovary showed luteal hemorrhage.

After the operation, the patient received pharmacological suppressive treatment with gonadotropin releasing hormone analogues, and she recovered uneventfully. At 3 months of postoperative follow-up, she is still doing very well.

## DISCUSSION

The term *mucocele*, as applied to the appendix and other organs or sites, denotes the macroscopic appearance of localized mucus accumulation. As a whole, appendiceal mucoceles are known to be found in only 0.2-0.3% of all appendectomy materials. The male:female ratio is 1:4 and the mean age of these patients is 55 years. The clinical presentation is usually nonspecific and up to 50% of these lesions are incidental findings at surgery. Related symptoms include vague abdominal distress or pain, or a chronic or intermittent colicky pain caused by intussusception of the mucocele.<sup>3</sup> In our case the appendiceal mucocele was incidentally recognized as a periappendiceal abscess-like lesion during the radiologic workup for the pelvic endometriosis. Radiologically, the reported US findings include a purely cystic mass with anechoic fluid, and a hypoechoic mass with variable internal echogenicity according to internal contents (watery or thick gelatinous). Those findings may help to confirm the lesions, but often they pose difficulties for the differential diagnosis with periappendiceal abscess. The typical CT aspect is a cystic, well-encapsulated mass, yet sometimes there are mural calcifications in the expected location of the appendix and this can cause extrinsic pressure on the cecal wall. Because of these detailed findings, CT appears as a mandatory examination since it allows an accurate preoperative diagnosis.<sup>4</sup> The lesion itself is not a disease entity because it may be related to a variety of pathologic conditions, ranging from nonneoplastic to malignant neoplasms. Most of the cases occur secondary to hyperplastic or neoplastic alterations of

the appendiceal lining epithelium; these lesions can also include hyperplastic polyps, adenomas, adenocarcinomas, and the so-called mucinous tumors of undetermined malignant potential.<sup>5</sup> Such hyperplastic and neoplastic changes in a mucocele can be missed if insufficient sections are microscopically examined. In the present case we submitted the entire mucocele lesion for histologic evaluation in 6 sections, but we could not find any neoplastic changes except for focal hyperplastic changes in the entrapped appendiceal mucosa. Approximately 6% of appendiceal mucoceles are associated with gelatinous mucinous ascites. When this ascites contains epithelial cells, it is termed as pseudomyxoma peritonei.<sup>3</sup> The term itself does not allude to the pathogenesis, which includes rupture, or spillage of mucus from an appendiceal mucocele or from a mucinous neoplasm arising in an abdominal organ. Actually, most important for the surgeons are the mucoceles that are caused by mucinous cystadenomas and cystadenocarcinoma. In the latter case, a possible rupture of the mucocele, either spontaneous or accidental, during surgery may result in the spread of malignant cells throughout the entire peritoneal cavity. In the present case a small mucus spillage from appendiceal mucocele occurred, but the phenomenon was limited just around periappendiceal tissue, and there was no gross mucus accumulation. Additionally, there was no evidence of proliferating epithelium in the microscopic mucus pools. A minority of the lesions, however, can develop secondary to obstruction by fecaliths, postinflammatory scarring, carcinoid tumors, or endometriosis, and so they are named as obstructive, retention, or simple mucoceles.<sup>1</sup>

Endometriosis involves the intestines in up to 37% of cases.<sup>6</sup> The appendix is involved less often than the rectosigmoid colon and small intestine, with the incidence of appendiceal involvement varying from 0.8% to 20%.<sup>7,8</sup> The clinical and pathologic findings differ according to the site of involvement. Colonic endometriosis is usually symptomatic with pain, bleeding, or obstruction; luminal obstruction is common and is caused by endometriotic deposits in the muscularis propria, mucosa, or both with secondary muscular hypertrophy and fibrosis. As contrasted with colonic lesion, the appendiceal involvement is rarely symptomatic and it is usually found incidentally in patients with pelvic endometriosis; our present case was an example of this. Appendiceal endometriosis usually involves the serosa or the serosa and muscularis propria together.<sup>7</sup> As for the incidence of association between appendiceal mucocele and endometriosis, there have been only eight well-documented reports in the English literature since the first description by Hapke and Bigelow in 1977,<sup>2</sup> although these reported cases probably don't reflect the true frequency of the association. Six of these cases, except for two with localized pseu-

domyxoma peritonei,<sup>1</sup> were very similar to our case.

In conclusion we describe here a rare occurrence of appendiceal mucocele secondary to endometriosis, and this report highlights one of the events relevant to the pathogenesis of appendiceal obstructive mucocele. To the best of our knowledge, this case is the first documented case reported in Korea.

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