



## MASSIVE ENDOMETRIOSIS MIMICKING TUMOR – UNUSUAL CAUSE OF APPENDICEAL INTUSSUSCEPTION

BERNARDICA JURIC<sup>1</sup>, AMIR IBUKIĆ<sup>2</sup>, SLAVEN ČIČEK<sup>2</sup>, IVAN PENAVIĆ<sup>2</sup>,  
DAVOR TOMAS<sup>1,3</sup> and ALMA DEMIROVIĆ<sup>1,4</sup>

<sup>1</sup>Department of Pathology and Cytology, Sestre milosrdnice University Hospital Center, Zagreb, Croatia;

<sup>2</sup>Department of Surgery, Sestre milosrdnice University Hospital Center, Zagreb, Croatia;

<sup>3</sup>School of Medicine, University of Zagreb, Zagreb, Croatia;

<sup>4</sup>School of Dental Medicine, University of Zagreb, Zagreb, Croatia

### Summary

Appendiceal intussusception is a rare clinical entity that, in women of reproductive age, may be caused by endometriosis. We report a case of a 36-year-old woman presenting with abdominal pain. The patient underwent laparoscopic surgery, during which a suspicion of a cecal tumor was raised. A laparoscopic right hemicolectomy was performed. The pathological exam showed an intussuscepted appendix, and the cause of the intussusception was massive endometriosis involving the colon, ileum, and appendix. It is important not to mistake appendiceal intussusception and appendiceal endometriosis with other intraabdominal pathologies whose symptoms they can mimic.

KEYWORDS: *appendix; endometriosis; intussusception*

### INTRODUCTION

Endometriosis is the presence of functional endometrial tissues outside the lining of the uterine cavity, which commonly involves genital organs but can be found in other areas of the peritoneal cavity as well(1,2). It is a common condition among reproductive-aged women that can be asymptomatic, but it can also cause chronic pelvic pain and infertility, and sometimes mimic symptoms of acute appendicitis(2,3). The diagnostic method of choice for endometriosis is laparoscopy. Endometrial implants in the wall of the appendix can cause appendiceal intussusception, which is difficult to diagnose preoperatively. Surgical treatment of intussusception is necessary since it can cause secondary involvement of other bowel

segments(4). We present a case of intussuscepted appendix with histologically confirmed endometriosis with a preoperative suspicion of a tumor.

### CASE REPORT

A 36-year old female patient was admitted to the emergency department with diffuse abdominal pain over the last seven days, predominantly in the periumbilical and epigastric areas. During physical examination, the patient manifested right lower quadrant tenderness but no rigidity. Organomegaly was not found, peristalsis was normal, and lumbar succussion was negative. Urin analysis showed high leukocyte esterase levels (3+), leukocyturia (L>300), erythrocyturia (E 7), and the presence of bacteria (3+) and squamous epithelium (>30). Other laboratory findings were within normal limits. Native abdominal X-rays showed bowel meteorism, with a few air-fluid lev-

*Corresponding author:* Alma Demirović, Department of Pathology and Cytology, Sestre milosrdnice University Hospital Center, Vinogradska cesta 29, 10000 Zagreb, Croatia, e-mail: [alma.demirovic@kbcsm.hr](mailto:alma.demirovic@kbcsm.hr)



Figure 1. Ultrasonography showed small bowel loop distension filled with liquid content.

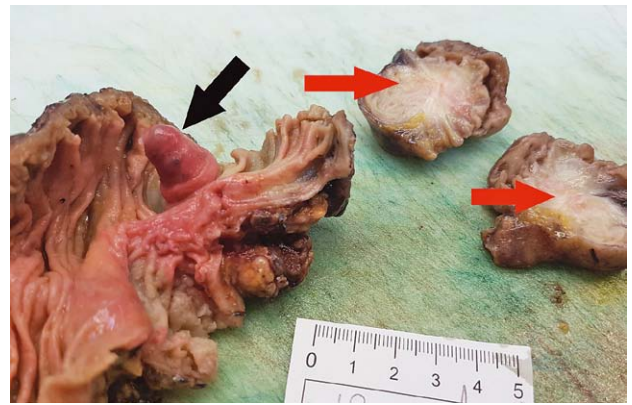


Figure 2. Macroscopically, the appendix was completely inverted (black arrow). Inside the bowel wall there were foci of endometriosis (red arrows).

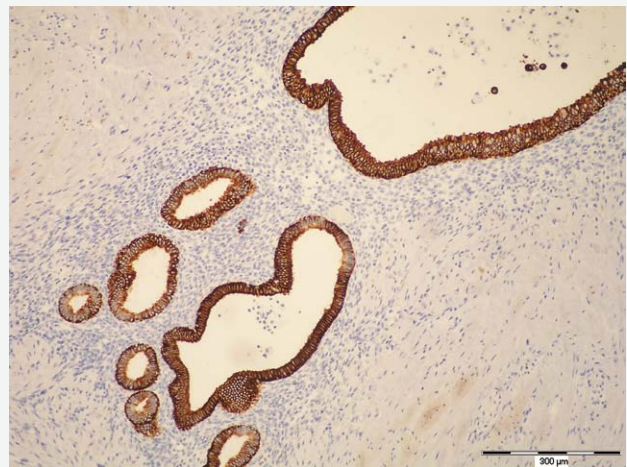
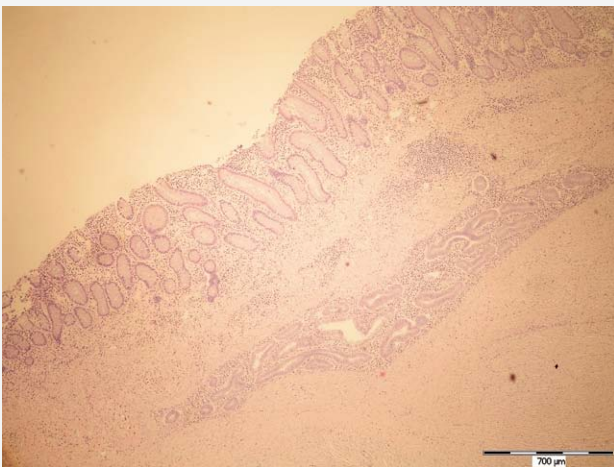


Figure 3. Microscopic slide showing endometrial glands and stroma inside the bowel wall, HE x 40 (A); positive immunohistochemical reaction of glandular epithelium for CK7, x100 (B); positive immunohistochemical reaction of stroma for CD10, x40 (C).

els in the bowel loops inside the pelvis without clearly visible distension. The X-ray image of the urinary tract was normal. Ultrasonography revealed small bowel loop distension up to 25 mm filled with liquid content (Fig. 1). Distal ileal loops and cecum were edematous, with a hypoechogenic area approximately 2 cm in diameter. MSCT of the abdomen and pelvis was recommended. Native and contrast-enhanced MSCT showed distended small bowel loops in the lower abdomen and pelvis up to 3,6 cm, filled with content. There was a tubular structure in the right periumbilical area leading to an area 3,9 x 3 cm in size, which shows post-contrast imbibition. Proximally to the described structure, a distended ileal loop was identified, indicating intussusception. MSCT of the pelvis revealed two ovarian cysts: the right ovarian cyst was 4,5 cm in diameter, and the left ovarian cyst 2 cm in diameter. The described findings suggested bowel obstruction, and laparoscopic exploration was indicated. During laparoscopy, free abdominal fluid and signs of metastatic disease were not found. A suspected tumor was identified in the cecum, which macroscopically didn't penetrate the bowel wall. The appendix wasn't visualized during the surgery. A small scar tissue was visible on the surface of the cecum. A laparoscopic right hemicolectomy was performed. Bowel continuity was restored by a latero-lateral ileocolic anastomosis using a surgical stapler. The specimen was sent for pathohistological analysis. Postoperatively, there were no complications, and regular bowel functions were restored. The patient was discharged on the seventh postoperative day in good condition. The gross specimen was a segment of terminal ileum and colon without a clearly identifiable appendix on the outside surface. Parts of the ileum were impassable. The cecal and ileal walls were focally thickened, firm, and white on the cut surface. Macroscopically, inside the colon, a completely inverted appendix was found. The appendix was 2,2 cm long with a maximal diameter of 0,8 cm. (Fig. 2). Microscopically, hemalaun and eosin (HE) stained sections of the colon, ileum, and appendix, including the area of stricture, endometrial glands and stroma were found. This was confirmed by immunohistochemical analysis. Endometrial stroma showed a positive reaction for CD10, and the glandular epithelium was positive for CK7 (Figs. 3A, B, and C). Foci of endometriosis were found in the submu-

cosa, muscularis propria, subserosa, and surrounding fatty tissue. The appendix was lined by normal colonic epithelium. Six lymph nodes that measured from 0,6 cm to 1,2 cm were found in the surrounding fat. Histologically, lymph nodes showed signs of sinus histiocytosis.

## DISCUSSION

Intussusception of the appendix is a rare clinical entity that, according to a study done by Collins in 1963(5) has an incidence of 0.01%. The first case of appendiceal intussusception was reported by McKidd in 1858(6), and since then, around 200 cases have been described(7). Some studies suggest that it might occur more frequently than is generally believed since transient appendiceal intussusception has been proven in asymptomatic patients using barium enema(8). According to McSwain(9), there are five anatomic types of appendiceal intussusception: Type I, the appendiceal tip is invaginated into the mid portion of the appendix; Type II, the middle portion is invaginated into the proximal appendix; Type III, the appendiceal base is invaginated into the cecum; Type IV, the proximal appendix is invaginated into the distal appendix; and Type V, the appendix is completely inverted into the cecum(4,7). Appendiceal intussusception can be caused by different pathological conditions such as inflammation, endometriosis, adenoma, adenocarcinoma, mucocele, carcinoid, etc(10). The most common histopathological finding in the intussuscepted appendix in pediatric and adult patients is inflammation and endometriosis, respectively(7,10). There have been fewer than forty cases of appendiceal intussusception caused by endometriosis reported in the literature(10). Anatomical factors predisposing an individual to intussusception of the appendix include a fully mobile appendix with a thin mesoappendix, a large proximal appendiceal lumen, and hyperperistalsis(4). This condition may be asymptomatic, or it can clinically mimic acute or chronic abdominal diseases, such as acute appendicitis(7). Sometimes it manifests as recurrent, cramping pain in the right lower quadrant of the abdomen(11). Preoperative diagnosis of appendiceal intussusception is difficult since clinical symptoms are nonspecific or non-existent and routinely done imaging studies, such as ultrasound and CT,

and colonoscopy may be misleading(12). It is important not to mistake an inverted appendix for a cecal polyp since an attempt at endoscopic polypectomy can cause iatrogenic complications, and a biopsy may lead to a bacterial infection(7). An inverted appendix may serve as a leading point for compound ileocolic intussusception, and in these cases, appendectomy is recommended(11). The extent of the surgical resection depends on the underlying pathology and the severity of the invagination(4). Endometriosis is defined as the presence of ectopic endometrial glands and stroma outside the lining of the uterine cavity and uterine musculature(1,2). It is not uncommon among women of reproductive age, affecting up to 15% of postmenarchal, premenopausal women and 2-5% of postmenopausal women(3). It is most frequently diagnosed in the fourth decade(13). Endometriosis can cause pelvic pain, dysmenorrhea, dyspareunia, heavy, irregular menstrual periods, and result in infertility(14). One third of patients are asymptomatic(3). Endometriosis commonly affects the genital organs and pelvic peritoneum, but it can also involve the umbilicus, surgical scars, and rarely the kidney, lung, skin, and diaphragm. The most commonly involved sites are the ovaries, posterior broad ligament, anterior and posterior cul-de-sac, and the uterosacral ligament. The gastrointestinal tract is affected in 3-37% of cases of pelvic endometriosis, and most commonly, endometriosis is found in the recto-sigmoid colon(3). The appendix is involved in only 2,8% of patients diagnosed with endometriosis(2). The prevalence of appendiceal endometriosis in the general population, calculated based on the review of the literature, is proposed to be 0.4%(1). Von Rokitansky was the first to describe the appendiceal endometriosis in 1860(15). Endometrial deposits of the appendix usually develop in the serosa, sometimes causing fibrosis and adhesions between neighboring bowel segments and pelvic structures. These implants may infiltrate the subserosa and the muscular layer, causing hypertrophy or hyperplasia of the muscularis propria that can constrict the lumen and create a mass that can act as a lead point for appendiceal intussusception(8,10). Endometrial implants inside the bowel wall can cause severe localized fibrosis leading to strictures(8). The muscular and seromuscular layers are involved in two thirds of cases, and the serosa alone in one third of cases(16). The submu-

cosa and mucosa may also be affected, particularly in symptomatic patients(3). Appendiceal endometriosis can cause symptoms of acute or chronic appendicitis, chronic right lower quadrant pain, sometimes cyclic in nature, lower intestinal haemorrhage and melena(1,14,17). It may clinically present as appendicitis or an appendicular mass that can mimic a neoplasm(2,18). Endometriosis also increases the risk of colonic and appendiceal perforation, particularly in pregnant women in their first two trimesters, which is believed to be caused by hormonal changes that lead to the expansion of the endometriotic foci(2,16). Endometrial implants can cause complete or partial luminal obstruction of the appendix, which is thought to lead to appendicitis, while some propose that symptoms may arise from the compression of the neural plexi in the appendiceal wall(10,19,20). There are several theories explaining the pathogenesis of endometriosis, including implantation or retrograde menstruation theory, direct transplantation and dissemination theories, coelomic metaplasia theory, induction theory, embryonic rest theory, and cellular immunity theory(14,16). Some suggest involvement of the lymphatic system in the pathogenesis and/or progression of endometriosis(12). On laparoscopic examination, endometriosis appears as raised flame-like patches, yellow-brown discoloration, reddish-blue irregularly shaped islands, or whitish opacifications(3). Numerous adhesions may be present in the peritoneal cavity and pelvis, with red, white, or black nodules(20). Ultrasonography may be of use in detecting endometriosis in atypical sites, including the appendix, where it is described as a solid, usually well-defined lesion(19). Magnetic resonance imaging may be useful in some cases, but it is of limited value in the right colon, particularly for smaller lesions, because of peristaltic artifacts and bowel content(21). In some cases, there are no signs of appendiceal endometriosis on laparoscopy, so some propose incidental appendectomy during surgical treatment of pelvic endometriosis in cases of severe endometriosis, which is controversial(2,3). Some even suggest appendectomy regardless of the laparoscopic appearance of the appendix in female patients of reproductive age with right lower quadrant abdominal pain(20,22,23). Between 1970 and 2008, ninety-eight cases of appendiceal endometriosis were described in the literature(24). There have been less

than thirty cases of appendiceal intussusception caused by endometriosis reported in the last fifty years(25). A definitive diagnosis is based on pathohistological analysis, which can sometimes be difficult in long-lasting cases in which the tissue is obscured by secondary fibrosis(3). If endometrial tissue cannot be found on H&E stain, appendiceal endometriosis can be suspected by the increased number of mast cells in the muscular layer of the appendix, so-called catamenial appendicitis(2). The treatment of endometriosis includes surgery and hormone therapy, depending on the patient's age and severity of symptoms(11). Laparoscopy is the recommended procedure for the clinical diagnosis and treatment of endometriosis(2). Surgical treatment of appendiceal endometriosis may resolve acute clinical symptoms, but chronic lower abdominal pain might recur, most likely because of the associated pelvic endometriosis(26). Laparoscopic ablation combined with medical treatment improves fertility(2). It's important to do a gynecological assessment and postoperative follow-up of the patient diagnosed with appendiceal endometriosis(16).

In conclusion, endometriosis and intussusception of the appendix are both rare clinical entities, but they should always be considered in the differential diagnosis of nonspecific recurrent lower abdominal pain in reproductive-aged women. Preoperative diagnosis is difficult since the clinical symptoms can mimic other intra-abdominal pathology, and imaging techniques are often not specific enough, so a definitive diagnosis is based on pathohistological analysis(27,28). Delayed treatment of endometriosis, and especially intussusception, may cause a severe outcome for the patient.

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## Sažetak

MASIVNA ENDOMETRIOZA KOJA KLINIČKI OPONAŠA TUMOR  
– NEUOBIČAJEN SLUČAJ INTUSUSCEPCIJE CRVULJKA

*B. Jurić, A. Ibukić, S. Čiček, I. Penavić, D. Tomas, A. Demirović*

Intususcepcija crvuljka je rijedak klinički entitet koji u žena reproduktivne dobi može biti uzrokovan endometriozaom. U članku je prikazan slučaj 36-ogodišnje žene koja se javila u hitnu službu s bolovima u abdomenu. Učinjena je laparoskop-ska operacija tijekom koje je postavljena sumnja na tumor cekuma te je napravljena desnostrana hemikolektomija. Patohi-stološki nalaz je pokazao intususcepciju crvuljka čiji je uzrok bio opsežna endometrioza koja je zahvatila debelo crijevo, ileum i crvuljak. Važno je prepoznati intususcepciju i endometrioza crvuljka, jer ih je lako zamijeniti za drugu intraabdomi-nalnu patologiju, čije simptome mogu oponašati.

KLJUČNE RIJEČI: *crvuljak; endometrioza; intususcepcija*