

Intussusception of The Appendix Duo To Endometriosis: A Case Report and Literature Review

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Abstract: Appendiceal intussusception happens when appendix segment is pulled into itself or into the cecum. This condition can mimic various chronic and acute abdominal conditions, with an important entity to be recognized, since it could be mistaken as a cecal mass. It is an extremely rare condition that ranges from partial invagination of the appendix to involvement of the entire colon. Endometriosis is defined as the presence of ectopic endometrial tissue outside the lining of the uterine cavity. Endometriosis is an exceptionally rare cause of appendiceal intussusception and only very few cases have been reported in the literature to date. We report here a case of appendiceal intussusception secondary to endometriosis.

Keywords: appendiceal intussusception- appendicitis- Endometriosis- Chronic and acute abdominal pain

I. Background

Appendiceal intussusception is a rare condition which has been diagnosed in case reports. (1,2, 3) The incidence rate of appendiceal intussusception is 0.01% in 11000 human appendix specimens. (1, 2) This condition primarily occurs in children. (1,2,3) In most cases, intussusception is limited to appendix but in some conditions cecocolic or ileocolic intussusception may occur. (4) A variety of underlying anatomical and physiopathological causes as lead point are described. (2-5) Endometriosis is a well-recognized gynecological condition in the reproductive age group. Endometriosis of the appendix is an entity of extragonadal endometriosis. The incidence of appendiceal endometriosis is lower than 1% among pathologies of pelvic endometriosis. Women can present with symptoms mimicking acute appendicitis or chronic pelvic pain. Diagnosis can be made only after a histopathological examination following the operation. We report here a case of appendiceal endometriosis, which were operated on for a prediagnosis of acute appendicitis, but postoperatively diagnosed as appendiceal endometriosis.

II. Case report

A 41-year-old woman single admitted to our surgical department for severe lower abdominal pain with a several week history of right iliac fossa pain. Apart from longstanding dysmenorrhea and menorrhagia, she did not have any other symptoms. She underwent laparoscopic cholecystectomy severe month ago and she had no family history of endometriosis. A clinical examination of the patient, with a complete gynaecological examination, was within normal limits. Initial blood investigations revealed an anemia with a hemoglobin level of 10.1 g/dl (iron deficiency). Abdominal ultrasound examination revealed an agglutination of the digestive loops with presence at the level of the right iliac fossa a picture in cockade of 22.5x16.5 mm (Photo 2). CT scan of the abdomen and pelvis showed: Nodular formation of the internal wall of the lower caecal base facing the proximal end of the appendix without signs of loco-regional infiltration (Photos 3,4,5). Right ovarian simple cyst, uterine cervix thickened and heterogeneous. A colonoscopy was performed and showed: Healthy ileo caecal valve. The ileum is healthy on 5 cm. The rest of the whole colon is without significant abnormalities. Tumor markers (carcino-embryonic antigen-alpha fetoprotein-CA 19-9) are within normal values. The diagnosis of acecal mass was retained and the patient underwent surgery. A laparoscopic assisted ileo-caecal resection was achieved. The appendix was found intussuscepted in the caecum. Last ileal loop appears inflammatory and stenotic in some places with granulation on the intestinal wall. The appendix was found into the caecum. The patient passed a smooth post-operative period and she was discharged on the seventh post-operative day. The histopathological result was: The pathological character is located at the level of the appendix which makes hernia in the cecal light. This appendix is the seat of multiple foci of transperietal endometriosis

III. Discussion

Intussusception of the appendix is a rare clinical event. Historically, intussusception of the appendix was found intra-operatively in patients with acute right lower quadrant pain and presumed appendicitis. There is an increasing number of case reports of intussusception of the appendix found in patients with chronic abdominal pain. As endoscopic and radiologic technology advances and becomes more prevalent in the workup of abdominal pain, it is possible that intussusception can be diagnosed pre-operatively, and patients treated with

a simple appendectomy. Endometriosis is a common condition that can affect up to 15% of women of child-bearing age and 2% to 5% of post-menopausal women. Endometriosis is seen at a rate of 3% to 37% in various parts of the gastrointestinal system, from the small intestine to the anal canal and 76% of these occur in the sigmoid colon and rectum [6]. Appendiceal endometriosis is a rarely seen condition comprising less than 1% of pelvic endometriosis cases [7] and preoperative diagnosis is difficult. The reported incidence in pre-menopausal women is in the order of 8–15%. Although the disease classically involves the pelvic organs and pelvic peritoneum, seeding has been observed in surgical scars, around the umbilicus, in the inguinal canal, intestines, bladder, heart and lungs. The transportation theory presumes that endometrial cells are transported to distant sites through surgical manipulation, menstrual shedding via the fallopian tubes or through lymphatic or vascular spread. Alternatively, the metaplastic theory suggests that embryonic coelomic mesothelium dedifferentiates into endometrial tissue in response to inflammation or trauma [8, 9]. The most common symptoms of endometriosis are dysmenorrhea, pelvic pain and infertility but patients can also be asymptomatic. The incidence of gastrointestinal endometriosis varies between 3–37% of those women who have proven disease. The rectum and sigmoid colon are most commonly involved, followed by the rectovaginal septum, small intestine, caecum and appendix. It usually takes the form of asymptomatic, small, serosal deposits. Under cyclical hormonal influences these deposits may proliferate and infiltrate the bowel wall. Cyclical haemorrhage from the endometrioma then leads to an intense, localized fibrosis within the bowel wall that can result in the formation of strictures.

In addition, serosal deposits can lead to the formation of adhesions between neighbouring pelvic structures or bowel loops [10]. Appendiceal intussusception is very uncommon, seen more in the first two decades of life. [11] The first case of appendiceal intussusception was reported in 1858. Intussusception of the appendix probably occurs by the same mechanism and pathogenesis as intussusception elsewhere. Appendiceal intussusception mostly simple (primary) form intussusceptions of appendicular mucosa (ie, only spontaneously invagination of appendix into cecum) in some instances might be associated with secondary cecocolic or ileocolic intussusception. [1, 11, 12] Primary type occur by either partial or complete invagination of the appendix into itself. Appendiceal endometriosis patients can be categorized into four groups in terms of symptomatology: 1/ patients who present with acute appendicitis; 2/ patients who present with appendix invagination; 3/ patients manifesting atypical symptoms such as abdominal colic, nausea and melena; and 4/ patients who are asymptomatic [13]. The most commonly seen group comprises patients who present with appendicitis. Acute appendiceal inflammation can arise because of partial or complete luminal occlusion by the endometrioma [14]. Another mechanism suggested is that of endometrium hemorrhage within the seromuscular layer of appendix, which is followed by edema, obstruction and inflammation. Pain in the right lower abdominal quadrant is one of the most common symptoms of appendiceal endometriosis. Although laboratory results are not specific, leukocytosis along with subfebrile fever is mostly present. Leukocytosis with the predominance of polymorph nuclear leukocytes accompanies acute appendicitis in most cases, along with elevated C-reactive protein [15]. Because endometriosis of the appendix can manifest in many ways without any specific indications, it is difficult to make an accurate preoperative diagnosis. Laboratory tests are of limited value. CT of the abdomen and pelvis may show evidence of acute appendicitis, or appendiceal abnormality. Laparoscopy is considered the gold standard for the diagnosis of endometriosis. Pain is the most common indication for surgical management. The correct diagnosis of appendiceal endometriosis is only established by the histological presence of endometrial tissue in the specimen [14]. About half of endometriosis of the appendix involves the body and half involves the tip of the appendix. Glandular tissue, endometrial stroma and hemorrhage are typical examinations conducted in patients with endometriosis [16].

Muscular and seromuscular involvement occurs in two-thirds of patients, while the serosal surface is involved in only one-third of patients. The mucosa is not involved, but Langman et al. [17] found that the submucosa was involved in one-third of patients with endometriosis of the appendix. In our patient, endometrial glands and stroma are found in muscular propria of appendix, too. Appendiceal intussusception secondary to endometriosis is extremely rare with fewer than 30 cases reported in the literature during the last fifty years. Endometrial involvement of the appendix is usually accompanied by chronic fibrosis, inflammation and hyperplasia or hypertrophy of the muscularis propria. This hypertrophic segment serves as a lead point for hyperperistalsis hence making it prone to intussusception particularly when combined with a fully mobile appendix that has a wide proximal lumen and a fat free mesoappendix. CT abdominal scans may demonstrate a soft tissue mass in the region of the caecum, although in this particular case the CT scan did not point towards the diagnosis [18]. The diagnosis of appendiceal intussusception is often difficult. Most appendiceal intussusceptions are founded at the time of operation of the patients suspected to have appendicitis [19]. Ultrasound is diagnostic imaging modality of choice. A few cases in asymptomatic patients have been incidentally diagnosed by barium enema, colonoscopy, CT scan of the colon and endoscopic sonography. In multiple cases appendiceal intussusceptions was mistaken for cecal polyp in colonoscopy. Sonographic examination demonstrated a lead point within a characteristic multiconcentric ring sign, and longitudinal

sonograms showed the inverted appendix protruding into the cecal lumen.[20, 21]. CT scan is also a modality for diagnosis of appendiceal intussusceptions [22, 23]. Appendectomy is choice treatment of appendiceal intussusception but it may relapse in spite of previous appendectomy. In these cases partial cecotomy also may be curative. Although surgical removal of appendix is treatment of choice, but in this case hydrostatic reduction of appendiceal intussusception occur.[24]. The treatment consists mainly of surgery and hormone therapy. The treatment tends to be determined by the age of the patient and the degree of the patient's symptoms. A gynecological assessment should be performed to determine the extent of endometriosis. At laparotomy or laparoscopy, a careful examination of the abdominal cavity is carried out in order to fully evaluate the extent of disease. Postoperative follow-up is mandatory for appendiceal endometriosis. Medical treatments for endometriosis are secondary. The postoperative gynecological examination did not reveal any other endometriotic lesion in our patient and she had normal serum CA125 (cancer antigen 125) level, so it is not necessary for her to receive additionally treatment.[25].

IV. Conclusion

Gastrointestinal endometriosis should be considered as a cause of appendiceal intussusception in postmenopausal women with periodic symptoms and confirmed disease. As in all cases of intussusception, the index of suspicion must be high since 90% of all intussusceptions in adults are due to an underlying neoplastic process. Intestinal endometriosis should be considered as a differential diagnosis in post-menarchal women who present with occasional gastrointestinal symptoms particularly in conjunction with gynaecological symptoms. The best way in the exploration of similar cases would appear to be laparoscopy or laparotomy followed by surgical resection in order to obtain histological confirmation of the main point which is of crucial importance in cases of appendiceal intussusception, in order to exclude an underlying neoplastic process. Subsequently, surgical resection is necessary either by an open or a laparoscopic approach.

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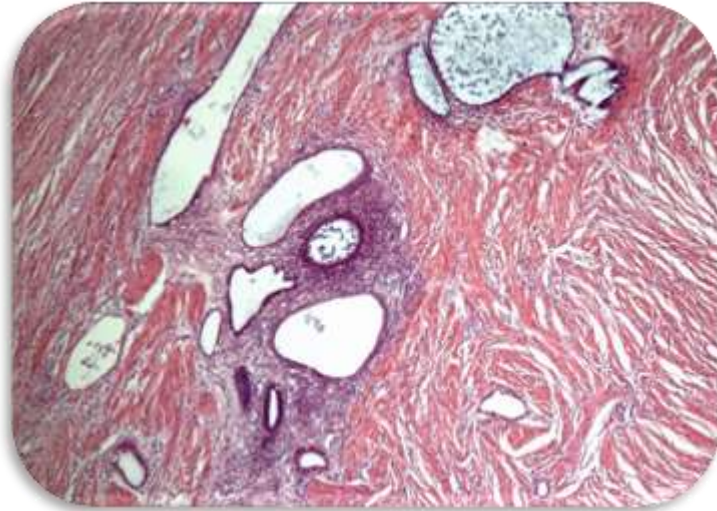


Photo 1: histopathological study showing appendix wall with multiple foci of endometriosis within the muscle layer.

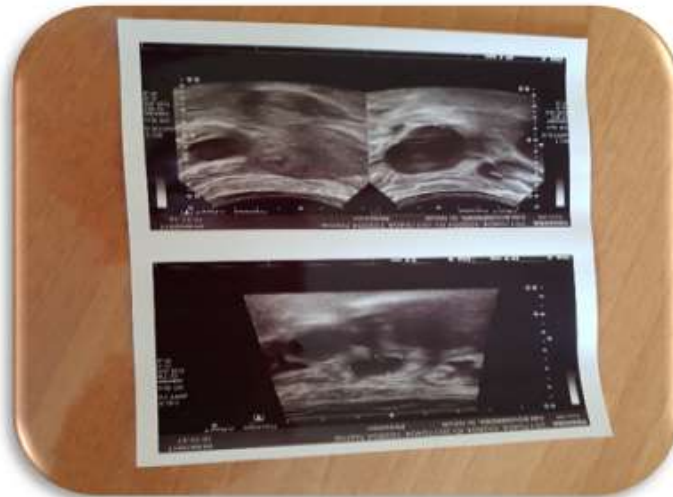


Photo 2 USE showing agglutination of the digestive loops with presence of a picture in cockade

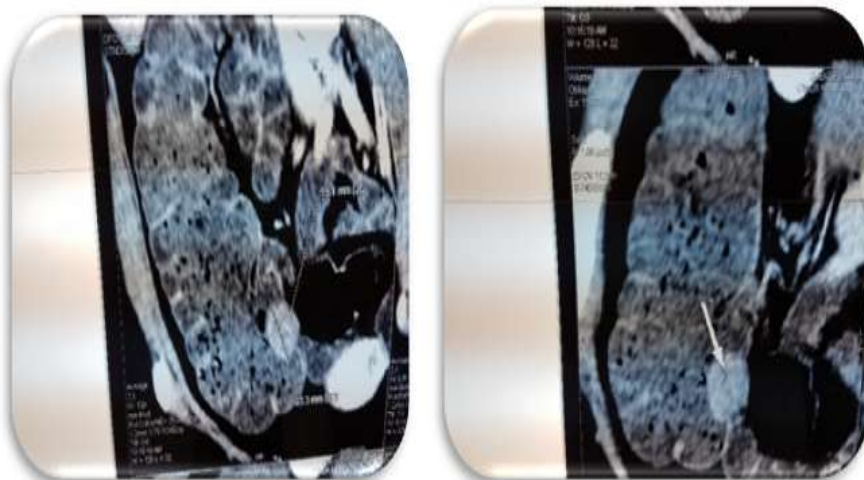


Photo 3, 4 & 5 CT scans showing Nodular formation of the internal wall of the lower caecal base



Photo 6 showing intussuscepted appendix covered by the caecal mucosa



Photo 7 showing intussuscepted appendix well individualized from the caecal wall