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Intussusception of the Appendix and Endometriosis

Roberta E. Sonnino, MD,* and Mohammed R. Ansari, MD†

Intussusception of the appendix, though uncommon, is usually diagnosed during surgery. Endometriosis in an intussuscepted appendix is rarely described. A case of appendiceal intussusception of a rare morphologic type associated with endometriosis is presented, and the literature is briefly reviewed. Etiology, classification, and diagnosis are discussed. (Henry Ford Hosp Med J 1986;34:61-4)

Intussusception of the appendix is an uncommon clinical finding. Approximately 180 cases have been described thus far in the literature. An even less common occurrence is the finding of endometriosis within the intussuscepted appendix. A case where endometriosis was the cause of appendiceal intussusception is reported, and a brief review of the literature is presented herein.

Case Report

In February 1985, a 35-year-old woman presented to the gastroenterology service due to a sudden onset of midepigastric pain and melena. Her past medical history was noncontributory. A complete upper gastrointestinal workup showed negative results, but melena recurred, and her abdominal pain became lower and more crampy in nature. The patient's hemoglobin dropped from 14.5 g to 11 g over several weeks. A barium enema revealed a 2 cm pedunculated polypoid-filling defect near the ileocecal valve, which appeared to be bilobed and was described as consistent with lipomatosis of the ileocecal valve (Fig 1).

Colonoscopy with biopsy was performed in April 1985, which confirmed the presence of a multilobed mass at the ileocecal valve. The biopsy was interpreted as an adenomatous polyp, but no attempts at endoscopic removal were made. Physical examination was entirely unremarkable, and no abdominal masses could be felt. The patient elected surgery in May 1985, when an ileocecal resection was planned. Intraoperatively, however, the "polyp" was found to be an appendix approximately 50% intussuscepted into itself, with an edematous, dusky tip protruding in the cecum (Figs 2-4). The patient underwent appendectomy with excision of a small rim of cecum. A large corpus luteum cyst was found in the right ovary, which was drained. Pathology revealed endometriosis within the intussuscepted appendix (Figs 5,6). The remaining hospital course was uneventful, and the patient was discharged on the seventh postoperative day.

Discussion

Intussusception of the appendix is an uncommon entity. Approximately 180 cases have been reported since the first description by McKidd in 1858 from the autopsy findings in a seven-year-old child (1). In 1963 after a 40-year study of 71,000 human...
Eig 2—Intraoperative appearance of cecum. Short segment of appendix seen.

Fig 4—Resected specimen with tip as it appeared on colonoscopy.

Collins (2) described seven cases of intussuscepted appendix (incidence of 0.01%). The condition has occurred in patients ranging from ten months to 75 years of age (mean age 16 years) with a 5:1 male to female ratio. Compound ileocolic intussusception occurs in over 50% of the cases. Endometriosis was found to be present in 22 out of 50,000 appendices in another review by Collins (3). Only a dozen documented cases of endometriosis in an intussuscepted appendix have been described to date.

Appendiceal intussusception was classified into four types by McSwain in 1941 (4) and in 1964 was expanded into five types by Fink (5) (Fig 7). Type 3 is by far the most common. All types, with the possible exception of type 4, may proceed to complete ileocolic intussusception. Our patient seemed to represent an intermediated stage between type 1 and type 5, with the appendiceal tip completely free within the cecum (Fig 8) and a certain degree of isolated mucosal intussusception. Only two previous cases with a similar appearance have been reported (6). In our case, the presence of endometriosis in the tip of the appendix, acting as a leading point, may explain the unusual morphology with intussusception of the tip into the proximal lumen of the appendix, rather than the more common invagination of the base of the appendix into the cecum.

The conditions promoting intussusception of the appendix, described by Forshall in 1953 (7), include: 1) a fetal type of cecum with the appendix originating at the tip, 2) a wide appendiceal lumen with the proximal lumen larger than the distal, 3) a thin mesoappendix free of fat with a narrow base, 4) a healthy appendix capable of active peristalsis, and 5) an appendix not fixed by inflammatory adhesions. It is generally believed that an
Fig 6—Histology: low power magnification. Multiple islands of endometriosis present within serosa as well as muscular coat of appendix (X2).

Fig 7—Classification of appendiceal intussusception. Relative frequency of each type: type I = 2%, type II = 14%, type III = 53%, type IV = 2%, type V = 27%, unclassified = 2%.

attempt on the part of the appendix to extrude a foreign body with active peristalsis may be a significant leading cause of appendiceal intussusception. Several pathologic conditions have

Fig 8—Diagram of appendix in case described.
been associated with the disease, but in many cases no distinct cause, other than chronic inflammation, can be found (Table).

The symptoms with which the disease may present were classified by Fink (5). Group 1, the most common occurring group, includes acute cases "poorly mimicking acute appendicitis." Group 2 includes the symptoms associated with intestinal intussusception. Group 3 patients usually have a long history (months to years) of recurrent crampy pain. Group 4 includes cases where intussusception of the appendix is entirely asymptomatic. The clinical presentation in our case was similar to Fink's third group, where the patient experienced recurring attacks of lower abdominal pain, although her symptoms were initially most indicative of an upper intestinal/gastric problem.

As with our patient, in most reported cases the diagnosis of intussuscepted appendix is usually made intraoperatively. Reports of correct preoperative diagnosis are rare but have occurred since the first radiological diagnosis by Skarby in 1941 (8). Several authors describe endoscopic removal of the intussuscepted appendix, which was thought to be a polyp. The procedure clearly entails significant risks and, though advocated by some authors, would generally not be attempted if the correct diagnosis were known (9). The radiological findings, though quite characteristic, are rarely recognized preoperatively. A mass in the cecum on the same side as the ileocecal valve, which could have a "coiled-spring" appearance, together with non-visualization of the appendix should raise the suspicion of appendiceal intussusception (10). A correct preoperative diagnosis would allow the options of nonoperative management in selected cases (11). However, the treatment of choice is surgery. If a neoplasm has been excluded, either a simple appendectomy after reduction of the intussusception or, as in our case, excision of a small rim of cecum with the appendix should be performed.

While endometriosis of the intussuscepted appendix has been reported with increasing frequency in recent years, it remains a rare initiating cause of intussusception. The first case, based only on the gross findings, was reported by Ingersoll and Meigs in 1945 (12). Since then, just over a dozen cases have been reported in the literature (13). Endometriosis of the intussuscepted appendix should be considered when evaluating right lower quadrant pain in women with a known history of endometriosis.

Consideration of appendiceal intussusception in the differential diagnosis of cecal masses is important because an extensive surgical procedure for suspected malignancy may be avoided by prompt recognition of the problem. Although the condition is recognized more frequently, appendiceal intussusception is still not correctly diagnosed by many clinicians. It is possibly true, as Zeifer proclaimed in 1951 (14), that until clinical acumen leads to correct preoperative diagnosis, individual case reports described in the literature will be necessary.

### Table

Relative Incidence of Factors Causing Appendiceal Intussusception

<table>
<thead>
<tr>
<th>Causative Factor</th>
<th>Percentage of Cases Reported</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chronic inflammation</td>
<td>80%</td>
</tr>
<tr>
<td>Mucocele</td>
<td>6%</td>
</tr>
<tr>
<td>Endometriosis</td>
<td>6%</td>
</tr>
<tr>
<td>Carcinoid</td>
<td>3%</td>
</tr>
<tr>
<td>Polyps/tumors</td>
<td>3%</td>
</tr>
<tr>
<td>Foreign body</td>
<td>1%</td>
</tr>
<tr>
<td>Other (including inverted appendiceal stump)</td>
<td>1%</td>
</tr>
</tbody>
</table>

### References