Appendiceal Intussusception due to Endometriosis: Radiologic and Surgical Correlation

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Objectives

1. Present a case of appendiceal intussusception.
2. Review causes of appendiceal intussusception.
3. Discuss presenting signs and symptoms.
4. Review imaging findings and differential diagnosis.
5. Discuss clinical implications and treatment.
Introduction

Intussusception of the appendix is a rare occurrence with reported rates of 0.01% in patients who have had an appendectomy.\(^{(2)}\) Patient presentation can range from asymptomatic to symptoms mimicking acute appendicitis. Multiple etiologies exist including inflammation, inflammation, carcinoid, and endometriosis, with endometriosis reported as the most common in adults.\(^{(3)}\) Although endometriosis is reported as the most common cause, it remains a rare condition with the majority of information obtained from case reports. Suggestion or diagnosis of this entity on imaging can help prevent unnecessary interventions. We present a case of appendiceal intussusception secondary to endometriosis in an asymptomatic 50 year-old female as diagnosed in a community hospital setting, with correlation of colonoscopy findings, CT findings, and final pathology.
A 50 year-old asymptomatic female was seen for routine screening colonoscopy with a local general surgeon. She denied any issues with bowel habits or family history of colon cancer. Past medical history was significant for endometriosis, for which patient had hysterectomy in 2004. Patient was found to have a submucosal mass at the cecum, which was subsequently biopsied, with pathology of colonic mucosa with no significant abnormality; however, no submucosal tissue was present for evaluation.
Colonoscopy

Submucosal mass found in the cecum, extending into the cecal lumen.
Case Presentation

Patient was subsequently sent for a CT scan which showed a 1.6 x 1.3 cm pedunculated intraluminal mass extending from the posterior aspect of the cecum.
Pedunculated intraluminal mass measuring 1.6 x 1.3 cm emanating from the posterior inferior cecum. Appendix not visualized. No lymphadenopathy
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Case Presentation

After discussion, patient opted for removal of cecal mass and underwent laparoscopic ileocecectomy. Specimen was sent to pathology. Final surgical pathology was positive for intussusception of the appendix with areas of endometriosis.
Gross Specimen

Right Colon: 6.2 cm of right colon with a 3.0 x 1.3 cm cylindrical structure protruding into the cecum, which appears to emanate from the appendiceal os
Pathology Slides

Low power of the appendiceal tip extending into the cecum
Low power view of endometriosis within appendix
High power view of endometriosis within appendix
Final Pathology

Terminal ileum and right colon resection with intussusception of the appendix with focal areas of endometriosis. Negative for adenomatous epithelium. Negative for malignancy.
First reported in 1858, primary intussusception of the appendix is a rare entity, with estimated occurrence of 0.01%\(^{(2)}\) and with only case reports and small case series found in surgical and radiology literature. A comprehensive review performed by Chaar et al in 2009 found that appendiceal intussusception is more common in adults than children, with women affected more than men. Although rare, intussusception of the appendix is important to consider in the differential diagnosis of a cecal mass, as preoperative diagnosis can help prevent unnecessary interventions.\(^{(8)}\)
Discussion: General Information

In 1910 Moschowitz initially classified appendiceal intussusception, which was modified in 1941 by McSwain, with 5 types described. All types are based on which part of the appendix is the intussusceptum and where the intussuscipiens is located.\(^{(3)}\)
McSwain Classification\(^{(3)}\)

**Type I:** Appendix tip (intussusceptum) invaginated into the proximal appendix (intussuscipiens)

**Type II:** Invagination of the middle portion of the appendix (intussusceptum) into the proximal appendix (intussuscipiens)

**Type III:** Base of appendix (intussusceptum) invaginates into the cecum (intussuscipiens)

**Type IV:** Proximal appendix (intussusceptum) invaginates into distal appendix (intussuscipiens)

**Type V:** Complete invagination of appendix (intussusceptum) into cecum (intussuscipiens)
Discussion: Causes

Intussusception is defined as an invagination of a loop of bowel into the lumen of an adjacent portion of bowel secondary to peristalsis. Many causes of intussusception of the appendix have been reported in the literature. In their comprehensive literature review, Chaar et al found that inflammation was the main cause in pediatric patients, accounting for 76% of cases. In adults, endometriosis was found to be the most common cause, causing 33% of cases, follow by mucocele and inflammation (both 19%), adenoma (11%). Carcinoid (7%) and adenocarcinoma (6%) rounded out the list of the more common causes.
Discussion: Signs and Symptoms

Patient with intussusception of the appendix can present with a range of symptoms. Patients may be asymptomatic with diagnosis made incidentally on colonoscopy or imaging studies such as CT or barium enema.\(^{(8)}\) Other patients may present with acute symptoms mimicking appendicitis\(^{(8)}\) with abdominal pain, nausea, and vomiting. Most patients present with a more chronic course, with waxing and waning symptoms occurring over a period of weeks to months. The most common symptoms reported were abdominal pain, vomiting, and blood per rectum. Additionally, some patients may present with palpable RLQ mass on examination, although this is more common in pediatric patients.\(^{(3)}\)
Discussion: Imaging

Although a relative rare entity, appendiceal intussusception can be diagnosed on imaging, with preoperative diagnosis possible in 58% of cases.\(^{(3)}\) Barium enema may be the first imaging modality in a suspected case of appendiceal intussusception. The intussuscepted appendix can appear as rounded intraluminal filling defect, usually in the medial wall of the cecum, with the classic findings being a finger-like filling defect within the cecal lumen.\(^{(1)}\) Levine described a coiled spring appearance of the cecum with non-visualization of the appendix as an additional findings for intussusception of the appendix.\(^{(6)}\)
Discussion: Imaging

Ultrasound can also play a role in diagnosing intussusception of the appendix, especially in the pediatric population. Findings on ultrasound include a donut-like or target-like mass identified within the cecum.\(^4\) Additional findings that may suggest an intussuscepted appendix include an outer diameter measuring 6 mm of a concentric mass within the cecum.\(^4\)
Discussion: Imaging

Patients may also undergo CT scan as a preoperative workup, especially if neoplasm is a working diagnosis. With modern CT scanners, there is better anatomic resolution, as well as the ability for 3-D reconstruction, both which can aid in the diagnosis. On CT, an intraluminal mass originating from the appendiceal orifice can be seen. The filling defect may have a target, layered, or sausage/reniform appearance.\(^7\) Multiplanar reconstructions as well as 3-D applications may aid in determining the appendix to be the source of the intraluminal mass.
Discussion: Differential Diagnosis

In patients with abdominal pain and vomiting, appendicitis or ileocolic intussusception should be considered as diagnoses. In patients who are asymptomatic with findings seen on imaging obtained for other reasons, intestinal polyp or malignancy should be considered. If intussusception of the appendix is suspected, this can help your referring physicians determine appropriate management. If patient is to undergo colonoscopy, it may also prevent attempted removal of the “mass” at colonoscopy, which if attempted can lead to perforation.\(^3\)
Discussion: Treatment

The optimal treatment for appendiceal intussusception is appendectomy with cecal cuff resection.\(^{(3)}\) Attempts at reducing the intussuscepted appendix can be attempted with barium enema; however, patients may be at high risk for recurrence depending on underlying cause. Other possible treatments include wider surgical excision including ileocecectomy, right colectomy, and subtotal colectomy. Since a majority of causes are benign, surgical resection is the definitive treatment.\(^{(3)}\)
Conclusion

Although a relatively rare diagnostic entity, knowledge of the presenting symptoms and imaging findings of appendiceal intussusception can help aid in its diagnosis. Most patients present with chronic, intermittent pain over a period of weeks to months. Also, women with history of endometriosis should raise suspicion for this entity with the appropriate imaging and clinical findings. Imaging with barium enema, ultrasound, or CT can suggest the diagnosis and help aid in patient management and possibly help prevent aggressive surgery or complications from attempted endoscopic removal. Definitive treatment is appendectomy with resection of the cecal cuff.
References