A Rare Case of Appendiceal Intussusception Secondary to Low-Grade Mucinous Neoplasm

ABSTRACT

Appendiceal intussusception is an unusual clinical entity, and when it is caused by a low-grade mucinous neoplasm, it becomes exceptionally rare. A 27-year-old female sought medical attention with a 3-day history of abdominal pain and recurrent vomiting. Clinical examination revealed right iliac fossa tenderness. Emergency imaging, ultrasound of abdomen, revealed a blind-ending, thick-walled, non-compressible tubular structure with a target appearance, suggesting appendiceal intussusception. Laboratory findings showed lymphocytosis, while other parameters were within the normal range. Confirmation of the diagnosis and assessment of disease extent were achieved through contrast-enhanced computed tomography (CECT) of the abdomen and pelvis. CECT revealed a soft tissue mass at the ileocecal junction with peripheral enhancing walls, indicative of an inflammatory process. A dilated and inflamed appendix, with post-contrast enhancement, was observed to be contributing to this mass. The patient underwent laparotomy, where approximately 100-150ml of inflammatory fluid was drained, and the inflamed appendix was identified intussuscepted into the cecum with associated adhesions. Appendicitis intussusception is a rare condition that requires appendectomy for treatment. Based on a study of 71,000 human appendix surgical specimens, the incidence of appendix intussusception has been estimated to be.01%.

Keywords: Appendiceal Intussusception, Low-Grade Mucinous Neoplasm, hemicolectomy, appendectomy, appendicitis

INTRODUCTION

Intussusception is the invagination of one bowel segment into another, resulting in blockage and ischemia. [1, 2] Although this condition primarily affects children, adults account for 5% of cases and are caused by benign or malignant intestinal lesions in 70% of cases. [3] The presence of lesions in the intestine disrupts normal peristalsis, promoting sick segment invagination. [1, 3] Because of its etiology, intussusception must be considered as a differential diagnosis in cases of intestinal obstruction. It is categorised based on the segment of the bowel that is affected: 80% are enteroenteric, including just the small intestine, 10% are ileocolonic, involving the terminal ileum invaginating into the ascending colon, 7% are colocolonic, compromising the large bowel, and 1% are gastroenteric. [1,3]The difficult diagnosis is determined through clinical observations in 32% of instances, whereas computed tomography (CT) scans have the ability to diagnose 78% of cases. [1,3]

Appendiceal neoplasms are detected in approximately 0.2-0.3% of histopathological samples obtained from appendectomies. Among gastrointestinal tumors, appendiceal mucinous neoplasms (AMN) account for about 0.4-1% of cases. [4,5] The early detection of an AMN may be coincidental because its signs and symptoms resemble those of acute appendicitis. However, in the later stages, apart from abdominal pain, there is also abdominal distension caused by the buildup of mucin in the peritoneal cavity. [5] The management of local appendiceal mucinous neoplasms involves removing the tumor through surgery. Hemicolectomy may be necessary depending on factors such as the extent of the lesion, the level of cell differentiation, involvement of the base of the appendix, and the presence of lymph node metastasis. [5,6]

For many years, the standard treatment for appendicealtumors was appendectomy (for tumors less than 2 cm) and right hemicolectomy (for tumors bigger than 2 cm). In the last two decades, laparoscopic surgery has surpassed open surgery in terms of reliability, yet open surgery may be more trustworthy in large and giant tumors due to the possibility of iatrogenic rupture and spread of mucin in the peritoneal cavity. [7-11]
We present a case of a 27-year-old female with appendicular intussusception resulting from mucinous adenocarcinoma, emphasizing the clinical, diagnostic, and therapeutic aspects of this condition.

CASE PRESENTATION

A 27-year-old female presented to the outpatient department with a chief complaint of abdominal pain and three episodes of vomiting over the past three days. No relevant past history or family history.Upon examination, the patient was found to be vitally stable, with focal tenderness in the right iliac fossa on abdominal palpation.
An ultrasound of the abdomen with pelvis were performed. The imaging study revealed a tubular, non-compressible, non-peristaltic, blind-ending pouch with a diameter of 13mm, characterized by a thickened wall, indicative of an inflamed appendix. Interestingly, the appendix appeared to be intussuscepted into the base of the cecum, displaying a target appearance with a diameter of 2.4x2.5cm and a length of approximately 3cm. A few tiny calcific foci were noted along the wall of the appendix, which were likely to be appendicoliths, with minimal peri-appendiceal fluid collection suggesting appendiceal intussusception.Laboratory investigations revealed lymphocytosis with a white blood cell count of 16,000 cells/mm^3, while other lab parameters were within the normal range.
To confirm the diagnosis and assess the extent of the condition, a contrast-enhanced computed tomography of the abdomen and pelvis was conducted. The imaging findings showed a soft tissue density mass at the ileocecal junction measuring 27x29x35mm, with peripheral enhancing walls on post-contrast images and surrounding inflammatory changes. Additionally, a dilated (9mm diameter) and inflamed appendix with post-contrast enhancing walls was observed, conglomerating with the previously mentioned mass.
Subsequently, the patient was taken for laparotomy with a midline infra umbilical incision. Intraoperatively, approximately 100-150ml of inflammatory fluid was identified in the pelvic region and the right hypochondriac region, which was drained. An inflamed, enlarged tip of the appendix was observed, invaginating into the cecum with surrounding adhesions. The appendiculocecal intussusception extended approximately 3cm below the ileocecal junction, with no evidence of intestinal obstruction (Figure 1). Careful adhesiolysis was performed, and the appendix was retracted from the cecum. At the base of the appendix, thickening was identified, which served as the lead point. A frozen section was sent for pathological evaluation, which was inconclusive for malignancy. Subsequently, an appendectomy was performed at the base of the appendix with a 1mm margin from the thickened area, and mesenteric lymph node biopsy was obtained.
The final histopathological report revealed a low-grade mucinous neoplasm at the base of the appendix, with no involvement of lymph nodes. Given these findings, the patient was scheduled for hemicolectomy one month later, as the initial financial constraints limited immediate intervention. The final hemicolectomy specimen displayed no evidence of malignancy, extracellular mucin pools, tuberculosis, or lymph node metastasis.
The patient is currently under regular follow-up on a three-month basis and has not reported any fresh complaints, signifying a positive outcome following the surgical intervention and resection of the neoplasm.

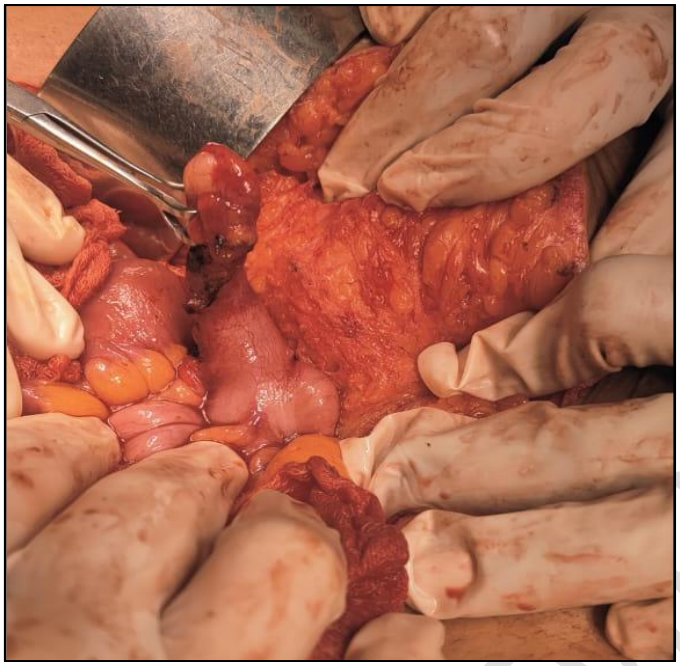

Figure 1. The appendiculocecal intussusception

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DISCUSSION

In adults, intussusception is a very rare occurrence that frequently mimics other abdominal pathologies. [12] About 90% of intussusception lead points in adult patients are pathogenic, with 65% being neoplastic. [13] The annual incidence of intussusception in adults is roughly 2 to 3 cases per 1,000,000 in the general population. [14,15] The age range of 30 to 50 years old has the highest occurrence rate. [15]

When the cecal appendix is the primary site of intussusception, the clinical picture may resemble acute appendicitis. [16] However, in certain cases, patients may be asymptomatic or exhibit symptoms consistent with intussusception, such as abdominal discomfort and vomiting, [17] as in the present case.McSwain has given five types of appendiceal intussusception. In

Type I there is mild invagination of the appendiceal tip, Type II there is moderate invagination of the appendiceal tip within the proximal appendix, in Type III, intussusception of the appendiceal base only while in Type IV a retrograde intussusception occurs, and Type V there is complete appendiceal intussusception into the cecum.[18] In our case, it was Type III.

The primary diagnostic technique for intussusception has been abdominal CT. [14-16] However, it has been suggested that in some situations, due to poor discrimination of the edematous intestinal wall, discriminating between the different anatomical aspects of the main mass is difficult. [15] Despite this, the abdominal CT report in the present case finds that there were indications of soft tissue density mass at the ileocecal junction. If the histology results reveal malignancy, an interval right hemicolectomy may be considered subsequently. [16]

In our case, the definitive diagnosis was made through histopathological examination, which revealed a low-grade mucinous neoplasm of the appendix at the base. These neoplasms are characterized by the production of mucin-secreting cells and are known for their indolent clinical course. Here, the neoplasm was the lead point for the intussusception. Notably, the presence of lymphocytosis in the laboratory findings can be considered an atypical manifestation in the context of this condition and warrants further investigation into its clinical significance.

The treatment of choice for appendiceal intussusception is surgical intervention, which is aimed at addressing the intussusception and ensuring the removal of the involved appendix. In this case, laparotomy was performed, and the surgical team successfully addressed the intussusception by retracting the inflamed appendix from the cecum. Given the inconclusive results of the frozen section, an appendectomy was performed at the base of the appendix with a 1mm margin from the thickened area. A mesenteric lymph node biopsy was also undertaken to assess the potential spread of malignancy. Subsequent histopathological reports confirmed the presence of a low-grade mucinous neoplasm at the base of the appendix, without lymph node involvement.

CONCLUSION

Appendiceal intussusception, particularly when attributed to a low-grade mucinous neoplasm, is an exceptional clinical entity. This case report highlights the significance of considering rare conditions in patients presenting with abdominal symptoms and the necessity of a multidisciplinary approach for their effective management. Early diagnosis, comprehensive surgical intervention, and diligent follow-up contribute to a successful outcome in such cases, underscoring the importance of continued research and documentation of rare medical phenomena.

Consent

We obtained informed written consent from the patient to authorize publication of her case and her photographs.

Ethical Approval:

As per international standard or university standards written ethical approval has been collected and preserved by the author(s).

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