



MUCINOUS ADENOCARCINOMA OF APPENDIX PRESENTING AS INTUSSUSCEPTION IN ADULT PATIENT

General Surgery

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ABSTRACT

Intussusception is found to be a cause of bowel obstructions in 1% adults cases diagnosed with intestinal obstruction. Unlike in children, where intussusception is most often idiopathic in nature, malignancy is the most common etiology identified in adults. Symptoms are usually non-specific, but timely identification and management is crucial in reducing the morbidity and mortality as there are high rates of carcinomatous lesion to be present as the lead point of intussusception. Here we present a rare case of mucinous adenocarcinoma of the appendix that presented as ileocolic intussusception.

KEYWORDS

Acute Appendicitis, Mucinous Adenocarcinoma, Ileocolic intussusception

INTRODUCTION

Intussusception and acute appendicitis are common surgical conditions with similar clinical presentation. Both require urgent attention and careful examination because more the delay in diagnosis, greater the chances of increased in morbidity and mortality¹. Intussusception is the invagination of one segment of bowel, the intussusceptum, into an adjacent segment of bowel, the intussusciptens, which can lead to obstruction and subsequent ischemia. Clinical presentation of intussusception can vary significantly between children and adults². Appendiceal intussusception may occur because of a number of etiologies that include villous adenoma, mucinous cystadenoma, endometriosis, and adenocarcinoma of the appendix³. Malignancy of the appendix is a rare condition, making up approximately 0.5% of all gastrointestinal tumors. These entities are usually carcinoid tumors, mucinous cystadenocarcinomas, colonic adenocarcinomas, and adenocarcinoid tumors⁴. This is a report of an elderly female with ileo-colic Intussusception secondary to mucinous adenocarcinoma of appendix.

CASE REPORT:

62 years female presented with right lower abdominal pain since for 10 days, constipation and abdominal distension for 5 days. O/E- vitals were normal with tenderness over right iliac fossa. Rectal examination was unremarkable. Laboratory investigations were within normal limits. X-ray abdomen showed dilated bowel loops. Computed Tomography abdomen was suggestive of Ileocolic intussusception with non-enhancing hypodense lesion showing peripheral calcification represent the lead point (Figure 1). Exploratory laparotomy was performed, which revealed Ileocolic intussusception with dilated tip of appendix (>2 cm) with appendix as lead point and a cecal mass. Right hemicolectomy was performed with ileo-transverse anastomosis (Figure 2).

Macroscopically, cecum showed blackish discoloured area of 3x2x2 cm with mucinous material. Cut section of appendix showed lumen filled with mucinous material. Microscopically, the mass was found to be a mucinous (colloid) adenocarcinoma, histologically grade 1 (well differentiated). No lymphovascular or perineural invasion was found, with all margins free of tumor: AJCC tumor stage pTispN0Mx. Postoperative course was uneventful.

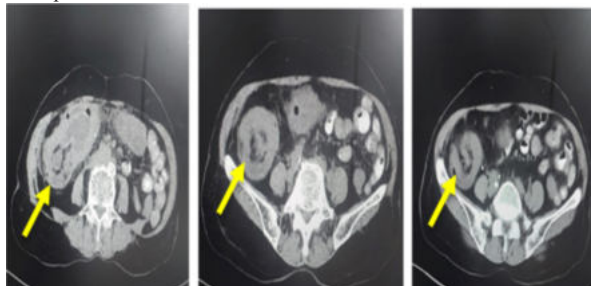


Figure 1: An axial CT Abdomen with contrast showing the

appendiceal-colonic intussusception with a suspicious rounded area of low attenuation, with peripheral high density. This served as the lead point for the intussusception

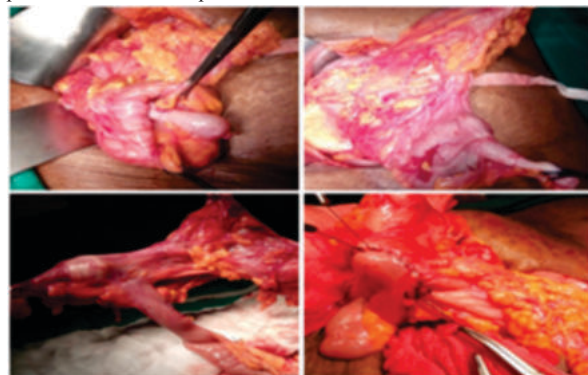


Figure 2: Ileocolic intussusception with dilated tip of appendix with appendix as lead point with right hemicolectomy and end-to-end Ileo-colic anastomosis.

DISCUSSION:

Adult intussusception is a rare surgical condition representing the cause of approximately 0.003% of hospital admissions. The diagnosis of adult intussusception is difficult due to the varied, nonspecific clinical presentations like intermittent abdominal pain, vomiting, nausea and hematochezia⁵. Clinical manifestations of an appendiceal neoplasm is often presents as acute appendicitis. Our patient presented with right lower abdominal pain, constipation and abdominal distension. The preoperative diagnosis of appendiceal neoplasms is often difficult. Therefore, they are usually identified during the pathological examination of surgical specimens⁶.

In adults, less than 20% of intussusceptions are primary or idiopathic, with the majority being secondary to a pathologic lesion or irritant in the bowel lumen that leads to alterations in normal peristaltic activity. Risk factors in adult typically include endometriosis, intestinal malignancies, Meckel's diverticulum or even appendiceal pathology as seen in this case. Unfortunately in adult, malignancies are the most common cause, with 30% of small bowel intussusceptions being attributed to malignant lesions and over 60% in the large bowel. Metastasis is more common lead point in the small intestine and adenocarcinomas are the most common malignant lead point in the colon. In view of malignancies being associated with intussusception in adults, a close follow period is essential⁷. Honjo *et al.* published a case series of intussusception in adults and reported one myxoma and two cystomyxomas of the appendix was the lead point of ileocecal intussusception Similarly, Kang *et al.* reported a case of appendicitis in an elderly patient resulting in colocolic intussusception⁷. The cause of intussusception in our case is secondary to malignant lesion involving the appendix.

Contrast studies are useful diagnostic tool for adult intussusception which can help to identify the site and cause of the intussusception. "Stacked coin" or "coiled spring" appearance is visible on upper gastrointestinal series. Barium enema is useful in patients with colonic or ileocolic intussusception in which a cup-shaped filling defect is a characteristic finding. "Target lesion" or "doughnut sign" with the presence of several concentric rings is classic findings on transverse scanning. Diagnostic yield of CT abdomen is around 78%, and is useful for identification the lesion and the underlying cause. The CT appearance is often a complex sausage-shaped soft tissue mass with an eccentric area of fat density contained within, which represents the mesenteric fat. The mesenteric vessels may be visible⁷. The CT abdomen in our case was suggestive of Ileocolic intussusception with non-enhancing hypodense lesion showing peripheral calcification may represent lead point.

There is some controversy regarding appropriate methods for management of adult intussusception. Several methods could be utilized depending on the location, etiology, suspicion of malignancy and clinical condition of the patient. Surgical resection of the lesion is most often indicated in adult, especially in colonic intussusception given the high likelihood of malignancy⁵. Uncomplicated, small, lesions of the appendix are usually treated by simple appendectomy, whereas larger lesions are treated by right hemicolectomy due to the higher potential for malignancy⁸. In our case, intussusception was secondary to malignant lesion involving the appendix and cecum so right hemicolectomy was performed.

Neuroendocrine tumors of appendix are estimated around 0.075/100,00 per year, whereas adenocarcinoma is estimated at around 0.2/100,000 per year. Tumors of the appendix are found in approximately 1% of specimens submitted for pathologic examination. The National Cancer Institute's Surveillance, Epidemiology and End Results (SEER) program analyzed 1,645 appendiceal malignancy cases from 1973-1998. Mucinous adenocarcinoma was found in 37.2% of the cases, "colonic type" in 24.9% of the cases, "malignant carcinoid" in 19.6%, "goblet carcinoid" in 13.7% and "signet ring cell" carcinoma was found in 4.3% of the cases. Connor *et al.* reviewed a database of 7,970 appendectomies and found 74 patients with appendiceal tumors: 42 carcinoid, 12 benign, and 20 malignant³.

In general, prognosis is good in adult patients with intussusception due to nonmalignant causes. Unfortunately, since a vast number of patients develop intussusceptions secondary to malignant lesions, the overall survival rate in this population is adversely affected. Mortality is less than 10% in adult intussusception cases where the lead point is a benign lesion, whereas mortality in cases where malignancy is the causative factor is over 50%⁵.

CONCLUSION:

Mucinous adenocarcinoma of the appendix is quite rare, often detected incidentally. Adult intussusception should raise a strong index of suspicion for a malignant etiology. CT scan abdomen is a valuable tool for diagnosis. Operative resection without reduction is the preferred treatment in adult intussusception.

CONFLICTS OF INTEREST:

Authors declare that they have no conflicts of interest.

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