

Mucinous adenocarcinoma of the appendix presenting as intussusception in 27 year old female

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Submitted Feb 12, 2012. Accepted for publication May 15, 2012.

DOI: 10.3978/j.issn.2078-6891.2012.033

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Introduction

While intussusception is commonly reported in children, it is quite unusual in adults almost always reported secondary to a definable lesion (1). Incidence secondary to adult intussusception has been estimated to range from 0.003% to 0.02% (2). Rarely, as in our case, the appendix may be the lead point for intussusception (3). Appendiceal intussusception may occur secondary to a number of etiologies including: villous adenoma, mucinous cystadenoma, endometriosis, and adenocarcinoma of the appendix (4-6). The incidence of epithelial malignancies of the appendix has been estimated to be 0.12 per 1 million persons per year (7). 5% of the total cases of intussusception (adults and children) have been reported in adults. Intussusception is reported as the underlying cause of 1-5% of adult cases of bowel obstruction (8). We will discuss our case, a 27 year female presenting with abdominal pain and a palpable abdominal mass, as well as briefly review the topic of appendiceal carcinoma.

Case presentation

A 27 year-old Hispanic female G₁P₀₂₀₂ presented to the emergency room with severe abdominal pain. She described a 2 day history of worsening intermittent “crampy” pain located in the periumbilical region that was exacerbated with touch. She also reported 3 episode of non-bloody vomiting earlier that day. Review of systems was unremarkable. Past medical and surgical histories revealed a caesarean section of twins one month prior for premature rupture of membranes at 35 weeks. She denied any social or family histories. Vitals were within normal limits. The abdomen was soft with normal bowel sounds and focal tenderness elicited on palpation of the right lower quadrant. On deep palpation a 3 cm hardened mass was found in the right periumbilical area. Laboratory findings

revealed leukocytosis of 11.9 K (4.5-11.0 k/mm³). Complete blood count, coagulation profile, urine pregnancy, and basic metabolic panel were unremarkable. Liver related tests were mildly elevated: ALT-113 (0-31 U/L), AST-76 (0-32 U/L), alkaline phosphatase of 124 (39-117 U/L), total bilirubin of 1.2 (0.0-1.0 mg/dL). A CT of the abdomen/pelvis with oral and IV contrast revealed possible volvulus. A repeat CT of the abdomen/pelvis with rectal contrast was then ordered for further characterization demonstrating a suspicious rounded area of low attenuation with peripheral high density (appendix) as a lead point consistent with an intussusception (Figures 1,2). The patient was taken to the operating room where an exploratory laparotomy was performed urgently to relieve the intussusception. A midline excision was made extending from the xiphoid to the pubic symphysis. The uterus, fallopian tubes, and ovaries appeared grossly normal. On exploration of the abdomen intussusception was confirmed extending from the ileocecal region to the hepatic flexure. During reduction of the intussusception a 3.5 to 4.5 cm mass was uncovered in the

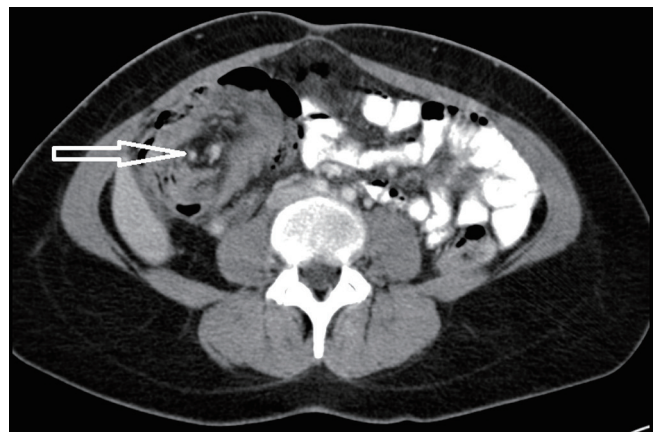


Figure 1 An axial CT Abdomen/Pelvis with rectal contrast showing the appendiceal-colonic intussusception with a suspicious rounded area of low attenuation (arrow), with peripheral high density. This served as the lead point for the intussusception

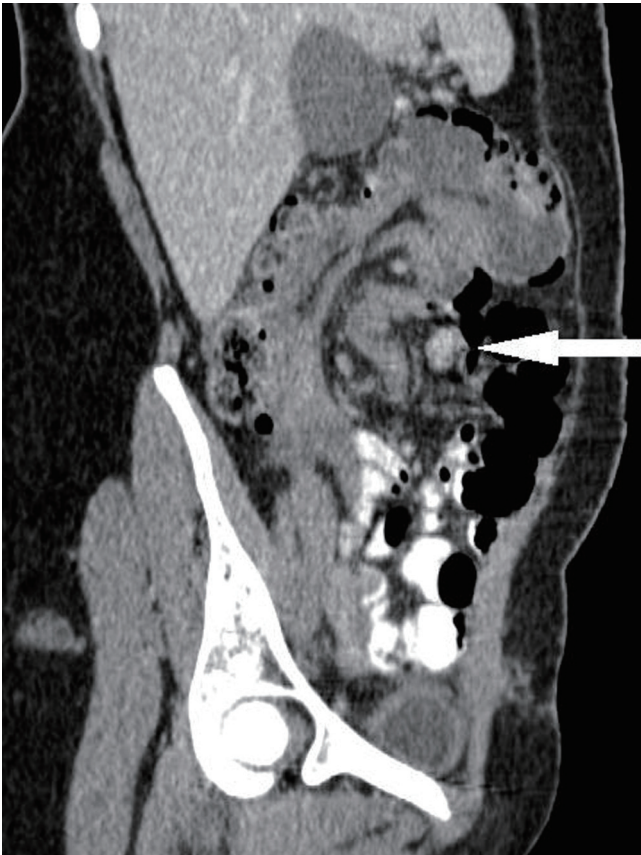


Figure 2 Axial and oblique coronal reformatted CT images of the appendiceal-colonic intussusception showing an area of high density (arrow), which was determined to be a mucinous adenocarcinoma

appendix (*Figure 3*). Subsequently a right hemicolectomy was performed, containing 15 cm of the right colon in continuity with 3.5 cm of the terminal ileum (*Figure 4*). An end-to-end ileocolonic anastomosis was performed prior to closure. Macroscopically the surgical specimen revealed a smooth tan bulging 4.5 cm × 4.5 cm × 3.7 cm mass located in the appendix. There were multiple pink tan lymph nodes dissected ranging from 0.3 cm to 1.7 cm. Microscopically, the mass was found to be a mucinous (colloid) adenocarcinoma (*Figures 5,6*), histologically grade 1 (well differentiated). No lymphovascular or perineural invasion was found, with all margins free of tumor: AJCC tumor stage pTispN0Mx.

On post-operative day 3 she was started on a clear liquid diet and advanced to a full diet on the day of discharge, post-operative day 5. The patient followed up 1 week later in the outpatient surgical clinic with no reported post-operative complications and was discharged from the clinic.

Discussion

There are two types of primary carcinoma of the appendix,



Figure 3 15 cm of the right colon in continuity with 3.5 cm of the terminal ileum. A smooth tan bulging 4.5 cm × 4.5 cm × 3.7 cm mass located in the appendix



Figure 4 Dissected appendiceal mass that later revealed mucinous adenocarcinoma of the appendix

adenocarcinoma (epithelial origin) and neuroendocrine tumor (neuroendocrine origin, formerly called “carcinoid”). The adenocarcinoma type can further be broken down into mucinous and non-mucinous (colonic), while the neuroendocrine tumors can be broken down into signet, malignant, and goblet subtype. Adenocarcinoma of the appendix is estimated at around 0.2/100,000 per year, whereas neuroendocrine tumors are estimated around 0.075/100,000 per year (9,10). Tumors of the appendix are found in approximately 1% of appendiceal specimens submitted for pathologic examination (11). The National Cancer Institute’s Surveillance, Epidemiology and End-Results (Seer) program analyzed 1,645 appendiceal malignancy cases from 1973-1998. 37.2% of the cases were identified as mucinous adenocarcinoma, 24.9%, “colonic type”, 19.6% “malignant carcinoid”, 13.7% “goblet

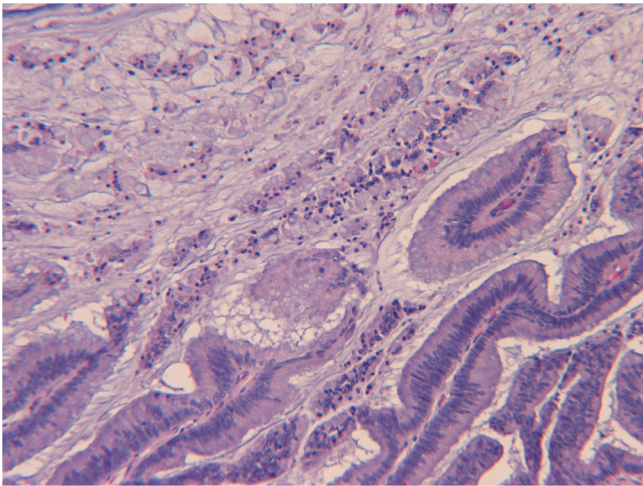


Figure 5 H&E stain 200× showing cystic mass occupying virtually the entire appendix containing pools of mucin with a focal complex epithelial structure without invasion of the appendiceal wall but with mucin extravasation into the wall. This can lead to pseudomyxoma peritonei

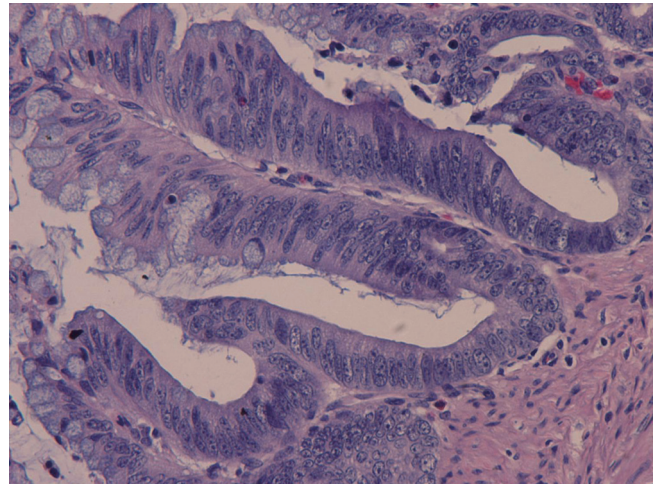


Figure 6 H&E stain 200× revealing a cystic mass occupying virtually the entire appendix containing pools of mucin with a focal complex epithelial structure without invasion of the appendiceal wall but with mucin extravasation into the wall

carcinoid”, and 4.3% “signet ring cell” carcinoma (12). Connor *et al.* reviewed a database of 7,970 appendectomies and found 74 patients with appendiceal tumors: 42 carcinoid, 12 benign, and 20 malignant (13).

Less than one third of mucinous appendiceal adenocarcinomas manifest as acute appendicitis, more commonly they are found incidentally on imaging studies as a cystic right lower quadrant mass or in a patient with increasing abdominal girth secondary to pseudomyxoma peritonei (11). CT is a sensitive technique for detecting the presence of an underlying appendiceal neoplasm. Changes such as the presence of cystic dilation of the appendix or a focal soft-tissue mass are present in the majority of cases (14). An appendiceal diameter greater than 15 mm is not specific, but this finding should be viewed with extreme suspicion of appendiceal malignancy. Although ultrasound (US) can be used to evaluate an abdominal mass CT is superior to US in regards to anatomical topography of an appendiceal adenocarcinoma with the ability to distinguish between cecum and mucocele, as well as the ability to detect mural calcifications within the neoplasm (15). The optimal treatment of any adenocarcinoma of the appendix is right hemicolectomy, either as a primary operation or as a secondary operation after adenocarcinoma of the appendix is noted on microscopic exam (11). When appendiceal mucocele is suspected controversy surrounds the topic of open versus laparoscopic appendectomy (16). Gonzalez *et al.* (17) reported dissemination of the mucocele after laparoscopic approach suggesting open appendectomy as the procedure of choice. Rupture of an appendiceal mucocele can result in dissemination of the epithelial

cells into the peritoneal cavity and incite pseudomyxoma peritonei a catastrophic complication (18). Care must be taken regardless of the approach when handling this neoplasm. Patients with appendiceal adenocarcinomas have a significant risk of synchronous and metachronous neoplasm, which often originate from the gastrointestinal tract (4).

Grading of appendiceal adenocarcinoma is the same as in the large intestines. Similar to the colon an adenoma-carcinoma sequence is assumed to occur in the appendix (19). In our patient there was no sign of adenoma and the adenocarcinoma was thought to be de novo. In comparison with colonic adenomas, adenomas of the appendix are more like to be serrated or villous (20). Staging for appendiceal adenocarcinoma uses the tumor-node-metastasis classification similar to colonic adenocarcinoma, however should be viewed differently secondary to the unique nature of appendiceal pseudomyxoma peritonei (M1/stage 4) in which malignant cells maybe noninvasive eluding to a worse prognosis than the same staged colonic induced pseudomyxoma peritonei that carries a worse prognosis (20). Misdraji *et al.* found patients with low-grade neoplasms confined to the appendix had a 100% survival at a median survival time of 6 years. Those with extra-appendiceal spread had a survival at 86% at 5 years. Those diagnosed with mucinous adenocarcinomas had a 5-year survival of 44% (21). SEER data 5-year survival for localized adenocarcinoma was 95%, and 80% in mucinous or cystadenocarcinomas (9). Right hemicolectomy with appropriate adjuvant therapy is recommended for both mucinous and nonmucinous carcinomas (21).

Conclusions

Mucinous adenocarcinoma of the appendix is a rare entity that in most cases is found incidentally. It is estimated that general surgeons may only see one or two cases of adult intussusception during their career (7). When intussusception is found in an adult a clinician should have malignancy high on their differential. We would like to impress the importance of pursuing the underlying etiology behind intussusception so appropriate treatment may be given prior to a malignancy becoming inoperable or untreatable if not diagnosed on initial presentation.

Acknowledgements

Disclosure: The authors declare no conflict of interest.

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Cite this article as: Siddiqui G, Lipka S, Freedman L, Takeshige U, Rizvon K, Mustacchia P. Mucinous adenocarcinoma of the appendix presenting as intussusception in 27 year old Female. *J Gastrointest Oncol* 2012;3(4):369-372. DOI: 10.3978/j.issn.2078-6891.2012.033