Acute intestinal obstruction: an unusual complication of mucocele of appendix

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Abstract. – A rare case of intestinal obstruction due to compression of ileocecal junction by mucocele of appendix is presented. Imaging with ultrasound or computed tomography (CT) is useful in establishing preoperative diagnosis. Avoidance of rupture of during appendicectomy is critical in preventing potential complication of pseudomyxoma peritonei.

Key Words: Mucocele, Appendix, Intestinal obstruction.

Introduction

Mucocele of appendix (MA) is characterized by aseptic dilatation of lumen of appendix as a result of distal obstruction. It is an infrequent condition, reported in only 0.2-0.3% of 43,000 appendicectomies. Acute presentation of MA as intestinal obstruction is still rarer. MA is usually asymptomatic and is found incidentally during radiological or endoscopic/surgical intervention undertaken for another reason. Appendicectomy is the treatment in view of possibility of malignant transformation of benign lesions and potential for development of pseudomyxoma peritonei. Gentle handling of MA during the exploration is of utmost importance as the inadvertent rupture of MA may lead to dissemination of mucoid material into the peritoneal cavity causing pseudomyxoma peritonei.

Cas Report

A seventy years old man presented to Surgery Emergency with complaints of pain, distension of abdomen, bilious vomiting, and absolute constipation of one week duration. On examination, tachycardia and dehydration were noted. Abdomen was distended and bowel loops were palpable. No abdominal mass was detected. Bowel sounds were exaggerated. X-ray abdomen showed grossly dilated jejunal and ileal loops, and absence of gas in colon (Figure 1). Laparotomy was undertaken with diagnosis of acute intestinal obstruction. A cystic mass, measuring about 8 cm in size, was found in the right iliac fossa. It was arising from caecum at the site of confluence of tenia coli. The mass was causing compression of ileocecal junction leading to obstruction of the small bowel (Figure 2). The caecum was mobilized and examined; no other appendix-like structure was found. Mesenteric lymph nodes were not enlarged and there was no mucoid material in the peritoneal cavity. The mass was carefully mobilized after releasing adhesions, and ligating vessels and caecal end. The compression on bowel was released and caecum and ascending colon became distended. Patient recovered well after operation and was discharged on 4th postoperative day.

Gross pathologic examination of the resected specimen revealed a 7.5 cm globular mass with an outer glistening whitish surface and a 2 mm long tubular structure at one end. The mass was filled with gelatinous material and there was no solid area. Microscopic examination showed mucinous epithelium with large areas of ulceration. Sections of tubular structure were suggestive of remnant of appendix. Histopathological features were consistent with diagnosis of mucinous cystadenoma of appendix (Figure 3).

Discussion

MA denotes dilated, mucus filled appendiceal lumen which includes four histopathological
groups: simple or retention cyst with normal appendiceal mucosa, mucosal hyperplasia, cystadenoma with neoplastic epithelium similar to that seen in adenomatous polyps of the colon, cystadenocarcinoma with neoplastic epithelium similar to that seen in adenocarcinoma of the colon. Rare causes of MA include carcinoid and endometriosis. MA is usually asymptomatic or presents with nonspecific clinical features. It presents as incidental finding in 50% of patients at the time of surgery. Pain in right iliac fossa is the most common clinical feature followed by abdominal mass, weight loss, nausea, vomiting. It may also present as acute appendicitis. This presentation is usually associated with simple or retention cyst occurring as result of proximal obstruction due to faecolith causing distension of lumen with mucus.

Preoperative radiological investigations [Ultrasoundography/Computed Tomography (CT) scan] may be of diagnostic help. Purely cystic lesions with anechoic fluid, hypoechoic masses with fine internal echoes as well as complex hyperechoic masses are the sonological features, depending on the contents. Multiple echogenic layers along the dilated appendix producing the appearance of “onion skin-like” circles are pathognomonic features of MA. Cystic well-encapsulated mass sometimes with mural calcification, in the expected location of the appendix which may be causing extrinsic pressure on the caecal wall without any surrounding inflammatory reaction are the CT scan findings in a suspected patient.

Other diagnostic modalities include colonoscopy, as first reported by Ponsky in 1976. Colonoscopy may demonstrate mucin emanating through the appendiceal orifice while evaluating the remainder of the colon.

The complications of MA are intussusception, torsion, intestinal bleeding, ureteral obstruction, hematuria, fistula formation, and volvulus. The most serious complication is the pseudomyxoma peritonei, described as the macroscopic presence of mucinous material in the peritoneal cavity.

Appendicectomy is the treatment, considering potential of benign lesions to progress to malignant and risk of rupture leading to pseudomyxoma peritonei. As a rule, MA should be taken out in toto without rupture to avoid potential complications. Misdaji et al., in his review of 107 patients of appendiceal mucinous neoplasm, found that an intact represents a benign process and does not lead to progression of disease. Gonzalez-Moreno et al. studied 501 patients of append-
diceal epithelial neoplasms and concluded that right hemicolecctomy does not have any survival advantage. These studies raised doubts about the conventional treatment of right colectomy with appendiceal epithelial tumors. Dhage-Ivatury and Sugarbaker\(^\text{13}\) emphasized the importance of assessing for appendiceal lymph nodes and appendiceal stump. If lymph nodes are positive, a right hemicolecctomy should be done while caecectomy should suffice if margin is positive in absence of lymph nodes.

If MA is encountered during laparoscopy, conversion to laparotomy should be done so as to prevent any rupture and to have a thorough examination of the peritoneal cavity. If the laparoscopy is to be done for the excision, should not be grasped and endobag must be used for the extraction\(^\text{13}\).

Intestinal obstruction resulting from the compression of the ileocaecal junction by a large MA is a highly unusual phenomenon. Due to its rarity, the surgeon may fail to consider MA as a possibility when a globular cystic mass in the vicinity of caecum in a case of intestinal obstruction is encountered. Transformation of the whole appendix into a mucocele may mask the appendicular origin of the lesion. The tiny connection to the caecum may be divided along with other adhesions without taking note of it. In a worse scenario, the MA may get ruptured due to inapt handling of not so alert surgeon, endangering the patient to the potential complication of pseudomyxoma peritonei. Once possibility of MA is considered, a deliberate search should be made for appendix, absence of which around the caecum would reinforce the operative diagnosis of MA. The lesion should be assessed for solid area and the bowel mesentery for enlarged lymph nodes, both of which indicate a malignant lesion.

**Conclusion**

MA is a rare condition. Intestinal obstruction as a complication of MA is still rarer. Presence of a cystic mass in relation to caecum should raise the possibility of MA. While excising the lesion, all efforts should be made to avoid rupture with a potential complication of pseudomyxoma peritonei.
References


