

A Case of Appendiceal Mucocele Mimicking Adnexal Mass in a Young Woman with Chronic Abdominal Pain

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ABSTRACT

Background & Objective: Low-grade appendiceal mucinous neoplasm (LAMN) is a very rare condition, diagnosed in 0.2-0.7% of appendectomies. The pathophysiology of the neoplasm involves accumulation of large volume of mucin in the appendiceal lumen, resulting in its obstructive dilation. Most epithelial tumors affecting the appendix are of the mucinous type and are manifested by mucin accumulation within an inflamed appendix with fibrotic and hyalinized wall.

Case Presentation: A 29-year-old female presented with a history of chronic and intermittent episodes of abdominal pain and a palpable mass in the right lower abdominal quadrant. Sonography showed a 58 mm mass anterior to the uterus and adjacent to the ovary, probably a uterine myoma. Laparotomy revealed normal uterus, and ovaries with a translucent mass originating from the appendix and fixed to the cecum. Pathologic examination of the mass was reported as low grade appendiceal mucinous neoplasm.

Conclusion: This is a rare case of appendiceal mucocele in a young female, mimicking a pelvic mass, which was misinterpreted by sonography as a gynecologic solid mass near the uterus. Preoperative diagnosis of appendiceal mucocele is important due to the risk of concurrent GI and ovarian malignancy associated with the condition.

Keywords: Mucocele, Appendix, Pseudomyxoma peritonei, Mucinous neoplasm



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Introduction

Low-grade appendiceal mucinous neoplasm (LAMN) is a very rare condition, diagnosed in 0.2-0.7% of appendectomies (1-5). In this condition, accumulation of mucinous material results in obstructive dilation of the appendix. No typical presentation has been defined for the disease, rather it is often found incidentally during an operation, resulting in its late diagnosis in most instances. There is a wide range of presenting symptoms, mimicking other intra-abdominal pathologies (5-8). Distinguishing the condition from other diagnoses is essential, because if treated inappropriately, it can result in peritoneal seeding of mucin-producing cells.

This dreadful complication is called pseudomyxoma peritonei (PMP), and results in vast death (8). The World Health Organization (WHO) defined the cytoarchitectural categorization for LAMNs, in 2010, in order to aid the accurate diagnosis of the condition (1). Here we present a case of appendiceal mucocele in a patient presenting with chronic abdominal pain.

Case Presentation

A 29-year-old woman was referred to the subspecialty clinic of Imam Hossein medical center, with a one-year history of intermittent colicky

abdominal pain, which had intensified in the month preceding the presentation. The pain was initially epigastric, migrating to the right lower quadrant in previous days, and radiating to the lower back region. She denied fever, nausea, anorexia and fatigue and had no previous history of acute abdominal pain, gastrointestinal bleeding or weight loss; but experienced urinary symptoms such as dysuria and frequency in the previous week. Her past medical history included history of normal vaginal delivery, hypothyroidism and chronic constipation. Her regular medications included Levothyroxine and laxatives. On the physical examination she was afebrile, and had normal vital signs. Her abdomen was soft with a palpable mass in the right lower abdominal quadrant which was slightly tender on deep palpation, but with no rebound tenderness. Rectal examination was unremarkable.

Her blood tests showed mildly elevated leukocyte count ($13.6 \times 10^9/L$) and C-reactive protein (11 mg/L), with normal hematocrit, liver function tests and coagulation studies. Tumor markers, CA-125, CA19-9, alfa feto-protein, CEA and human chorionic gonadotropin (HCG) levels were normal. Transvaginal color Doppler ultrasound revealed an 86x57x51 mm solid heterogenous mass with regular borders, located in the right lower abdominal quadrant, anterior to the

endometrium and adjacent to the right ovary. The mass had no acoustic shadowing or vascular flow. The ovaries on both sides were reported to be slightly larger than normal. Liver, bile ducts, pancreas, and kidneys were normal on abdominal ultrasonography. On previous ultrasonography and transvaginal Doppler 5 months earlier, a large heterogenous solid mass measuring 80x56x57 mm with no abnormal flow was reported anterior to the uterus and adjacent to the right ovary, suggesting a probable uterine myoma. Both ovaries and uterus were reported to be normal in size.

The patient was transferred to the surgical unit for exploration. Laparotomy by Pfannenstiel incision revealed completely normal uterus, and ovaries. There was no evidence of adhesions. A translucent mass, measuring 5x8 cm, originating from appendix and fixed to cecum was observed which was resected and biopsy from mesenteric lymph nodes was obtained for frozen section (Figure 1). The frozen section reported a benign mucinous tumor of the appendix. No signs of mucinous implants or pseudomyxoma peritonei were detected in the abdominal cavity. The patient's pain completely resolved after the surgery. The final pathology report was low grade appendiceal mucinous neoplasm. Peritoneal cytology was reported negative for malignancy. The patient was discharged in a good general condition.

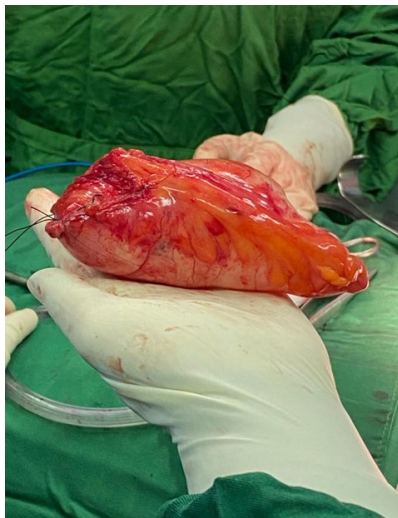


Figure 1. Macroscopic appearance of appendiceal mucocele in the presented patient

Discussion

LAMNs constitute a rare adenomatous malignancy of the appendix, manifested as obstructive appendiceal dilation from accumulation of large volumes of mucin. Most epithelial tumors affecting the appendix are of the mucinous type and are manifested by mucin accumulation within an inflamed appendix with fibrotic and hyalinized wall. The condition has male preponderance and affects patients mainly in their sixties. LAMNs are typically diagnosed incidentally, and occasionally their presentation can be of their complications, such as intussusception, volvulus, small

bowel obstruction (SBO), appendiceal rupture, pseudomyxoma peritonei and ureteral obstruction (1, 2, 9, 10). It is not uncommon for the disease to be mistaken for acute appendicitis, adnexal mass, or a retroperitoneal tumor in the right iliac fossa. Mucoceles smaller than 2 cm are considered as benign LAMNs (retention mucoceles). Those measuring more than 6 cm are associated with higher risk of complications such as malignancy, perforation, and PMP (2). Mild elevations of CEA, Ca 19-9, and Ca-125 is reported in 56.1-67.1% of LAMN cases (11). After surgery, these

markers can be used for investigating peritoneal malignancy. There is a 35% risk of coexisting gastrointestinal malignancy in LAMN patients (12). Although more than half of LAMNs are diagnosed incidentally during radiologic, endoscopic or surgical interventions, use of the preoperative diagnostic imaging is essential. Having the correct pre-operative diagnosis aids in appropriate surgical planning and preparations, which in turn minimizes the risk of intra/post-operative complications, such as peritoneal spreading. Ultrasonography, CT scan and colonoscopy are the most commonly used diagnostic tools for this purpose (5, 7, 8, 13, 14). In investigating patients with abdominal pain, ultrasound is considered to be the first line imaging modality. It has high sensitivity (83%) and specificity (92%) in differentiating between acute appendicitis and mucocele (15, 16). A more accurate and detailed description of mucocele characteristics including appendiceal lumen, its cystic dilation and wall calcifications can be obtained through CT scanning (5-7, 17). Colonoscopy aids in evaluation of the appendiceal opening and the presence of mucinous discharge through it, as well as detection and access to the possible concurrent colonic tumors (5, 7, 18, 19).

In the present case, ultrasound was not diagnostic and could not identify the appendiceal mucocele. Diagnosis of appendiceal mucocele should prompt its surgical resection in order to prevent complications such as unprompted or iatrogenic ruptures, intraperitoneal mucin leakage and subsequent malignant transformation (20, 21). Nonetheless, surgical treatment of mucocele has its potential problems, due to the high risk of peri-operative mucocele rupture resulting in mucin leakage into the peritoneal cavity with resultant pseudomyxoma peritonei (9, 14). Hence, correct choice of surgical method for treatment of mucocele is very important. Dhage-Ivatury and Sugarbaker proposed a system to help decision-making regarding the type of surgical intervention for appendiceal mucocele. Factors considered in this system include: presence of perforation, margins of resection involvement, and involvement of mesoappendiceal and ileocolic lymph nodes. A range of treatment options are defined for the condition which include: surgical resection procedures (ranging from appendectomy to right colectomy), debulking operation, heated intraperitoneal chemotherapy, and early postoperative intraperitoneal chemotherapy (14). If the mucocele is situated far from the appendicular base, is not very large in size and shows no signs of perforation, simple appendectomy is a satisfactory treatment option in the presence of clear

margins of resection and a normal caecum. Partial cecostomy is to be considered in the presence of wide mucocele base, in cases where mucocele projects into the cecal wall, or in the presence of positive resection margins with negative lymph nodes. Suspicion of malignant mucocele, presence of enlarged mesenteric lymph nodes or a positive cytology, should warrant performance of right hemicolectomy. Thus, the utilization of frozen section examination is a valuable tool in choosing the treatment method intra-operatively. A detailed peritoneal cavity exploration is mandatory to exclude the presence of co-existing mucin-secreting malignancies, such as colonic and ovarian cancers, given the high rate of their occurrence in the presence of an appendiceal mucocele (22). In our patient, mucocele was removed intact, with no leakage. The margins of resection were all negative, with negative regional lymph nodes on frozen sections. Since there was no pathologic development in the base of the appendix, appendectomy was performed. Exploration of the abdomen was negative for presence of co-existing malignancies. She is followed up by the general surgical service.

Conclusion

The present case is a rare occurrence of appendiceal mucocele in a young female, presenting with right lower abdominal pain and a pelvic mass which was initially misinterpreted by sonography as a gynecologic solid mass near the uterus. Nevertheless, gynecologic surgeons should be aware of the rare possibility of confronting appendiceal mucocele when adnexal and pelvic masses are suspected pre- and intra-operatively. Precise preoperative diagnosis of appendiceal mucocele is crucial, firstly due to its associated risk of concurrent GI and ovarian malignancy, and secondly because it necessitates appropriate surgical resection planning to prevent complications such as unprompted or iatrogenic ruptures, intraperitoneal mucin leakage, and subsequent malignant transformation.

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Conflict of Interest

There is no conflict of interest.

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