

Appendicealmucocele: A case report**Dr. Dharmveer Jajra¹, Dr. Pukhraj², Dr. Mahendra Kumar³, Dr. Ashok Kumar⁴, Dr. Dilip Chaudhary⁵,
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Abstract: The appendicealmucocele[AM] is a rare disorder. Preoperative diagnosis is difficult due to nonspecific clinical manifestations. Ultrasonography, computed tomography, and colonoscopy can suggest the diagnosis. Treatment is always surgical and depends on the integrity, size of base and on histological type of original lesion of appendix. Preoperative diagnosis is important to avoid intra-operative and post-operative complications. If mucocele is treated incorrectly pseudomyxomaperitonei, which is characterized by malignant process and has very poor prognosis, may develop. We report a case of a 42-years-old man who was admitted to the surgery department with abdominal lump in right lower quadrant. Only appendectomy was done. Hystopathological diagnosis was mucinous cystadenoma.**Keywords:** appendix, mucocele, mucinous cystadenoma, surgical treatment

INTRODUCTION

The term appendicealmucocele [AM] was coined by Von Rokitansky in 1842[1]. AM is a rare disorder and is characterized by a distended, mucus filled appendix. It results from the lumen obstruction in the appendix, which is secondary to the inflammatory or neoplastic proliferation of the appendix mucosa, or of lesion in the caecum, adjacent to the appendicealostium and rarely by endometriosis. The incidence is 0.2%-0.4% of all appendectomies[2]. There are four histologic types of AM: retention cyst(1%), mucosal hyperplasia(25%), mucinous cystadenoma (63%), and mucinous cystadenocarcinoma(11%). Mucocele of size less than 2 cm are usually associated with retention cyst and mucosal hyperplasia. Larger mucocele are more likely to be neoplastic [3]. Mucinous cystadenoma is a benign condition and adequately treated by a simple appendectomy. Mucinous cystadenocarcinoma is a malignant condition that requires more radical surgery and its rupture can lead to a grievous outcome in form of pseudomyxomaperitonei [4].

CASE REPORT

A 42-years-old man was presented to our surgery department with chief complain of pain in right

lower quadrant of abdomen for last six months. Previously, two times he was admitted at district level hospital. Conservative treatment for appendicular lump was given and discharged with advice of interval appendectomy. On examination, the lump was palpated at right iliac fossa. Lump was firm, painful, non-mobile, with smooth surface and well defined margins. Temperature was 37⁰c. TLC showed mild leucocytosis. Ultrasound showed cystic mass arising from the caecum base with non-visualization of appendix. Computed tomography of abdomen demonstrated a cystic, oval shaped, thin walled structure in right lower quadrant in continuity with caecum and there was no surrounding inflammation or abscess formation [figure-1]. Based on radiological findings, an initial diagnosis of a mucocele was made. An elective appendectomy was performed. . At the time of surgery, a cystic mass of the appendix (6×4×3 cm) was discovered [figure-2]. No discharge was found in the peritoneal cavity. Only appendectomy was performed because base of appendix and caecum were free of disease and the lymph nodes were not increased in size. Cut section of specimen showed gelatinous material. Hystopathologic diagnosis was mucinous cystadenoma. The patient was discharged on the fourth postoperative day without any complication. One year after the surgery the patient is feeling well.

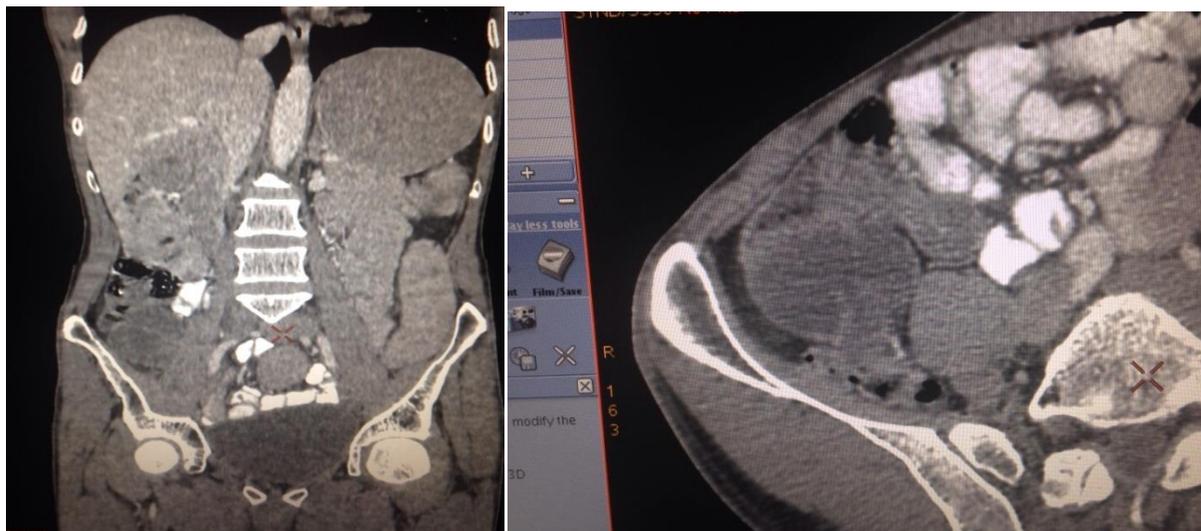


Fig. 1: CT Scan Image of AM



Fig. 2: Intraoperative Image of AM

DISCUSSION

AM is a descriptive term referring to a dilated appendix with abnormal intraluminal accumulation of mucus. It is a rare disease. Its incidence ranges between 0.2% - 0.4% of all appendectomies. The appendix is lined by epithelium containing more goblet cells than the colon. As a result, most appendiceal epithelial tumors are mucinous and start as mucoceles. According to modern classification, there are four histologic types: retention cyst, mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma.

Clinically, AM does not have a specific picture. Patients are often asymptomatic. In about 50% of cases it is discovered accidentally during physical, radiological, and endoscopic examinations or at surgery. Patients may present with complain of pain in

the right lower quadrant of the abdomen, palpable abdominal mass, nausea, vomiting, weight loss, gastrointestinal bleeding, and features of intestinal obstruction due to intussusception or volvulus. Abdominal pain is the most common symptom followed by abdominal lump [5].

Diagnosis of AM can be established by USG, computed tomography (CT), and colonoscopy. USG is the first-line diagnostic method which shows a cystic, encapsulated lesion, in relation to the caecum with variable internal echogenicity and calcification of the cyst wall. USG can be used to differentiate between mucocele and acute appendicitis. 6 mm outer diameter of appendix is suggestive of acute appendicitis while 15 mm and more indicate the presence of a mucocele, with 83% sensitivity and 92% specificity. The 'onion

skin sign' is considered to be specific for AM [6, 7]. CT is regarded as the most accurate method of diagnosis. Appendix lumen more than 1.3 cm, its cystic dilatation, and wall calcification is specific for AM. On barium enema, there is usually non filling or partial filling of the appendix with contrast and lateral displacement of caecum and terminal ileum. Colonoscopy can demonstrate 'volcano sign' i.e. soft erythematous mass, with a central crater due to the protrusion of appendicealostium, which can increase and decrease according to the respiratory movement. Furthermore, synchronous and metachronous tumors of colon can be identified. The carcinoembryonic antigen (CEA) level may suggest malignancy in the appendix or in the colon. FNAC can distinguish between benign and malignant form of AM, but it should not be used due to the risk of cell dissemination and evolution to pseudomyxomaperitonei [8, 9, 10].

The differential diagnosis should include carcinoid tumor, adenocarcinoma, lymphoma, hydrosalpinx, ovarian cystic lesions, secondary duplication cyst, mesenteric and omental cysts, mesenteric and retroperitoneal hematoma or tumors and abdominal or retroperitoneal abscesses. Radiological findings i.e. calcifications, imaging of the mucin content and relationship with the cecum, combined together, can differentiate AM from mimicking disease [11].

The treatment of AM is surgery. An algorithm for the selection of the type of surgery has been furnished by Dhage-Ivatury and Sugarbaker [12]. It includes several factors such as: (1) mucocele is perforated or not; (2) the base of the appendix (margins of resection) is involved in the process or not; and (3) whether the lymph nodes of mesoappendix and ileocolic region are positive or not. So for patients with benign mucocele that has negative margins of resection without perforation simple appendectomy is sufficient. No long term follow-up is needed for these patients. For patients with perforated mucocele, with positive margins of resection, positive cytology and positive appendiceal lymph nodes, right colectomy/cytoreductive surgery (CRS)/heated intraperitoneal chemotherapy (HIIC) and early postoperative intraperitoneal chemotherapy (EPIC) should be performed. Long term follow-up is obligatory for these patients. In our patient the mucocele was not perforated, one cm of base of the appendix was free from disease and the regional lymph nodes were negative. Therefore, only appendectomy was performed, which is an adequate surgery in such a case. Also, according to the algorithm, no long-term follow-up is advised for our patient.

The spontaneous and surgery induced complications of AM include intestinal obstruction,

intussusceptions, intestinal bleeding, fistula formation, and volvulus [13, 14, 15]. The worst complication is pseudomyxomaperitonei, characterized by peritoneal dissemination caused by iatrogenic or spontaneous rupture of the mucocele. The tissues should be handled carefully during surgery in order to avoid rupture of the mucocele. Therefore, some surgeons consider that laparotomy should be preferred against laparoscopy [16]. In case AM is diagnosed during laparoscopy, the surgery should be converted into a laparotomy. So that the cyst is not ruptured and the filling is not scattered into the peritoneal cavity and with laparotomy compared to the laparoscopic method, it is possible to have a fuller inspection, palpation, and direct inspection of the spots in the abdomen where mucinous tumors are most common. Some surgeons consider that the operation can be performed using a laparoscopic method by atraumatic handling of the appendix and use of impermeable bag for removal of the specimen [17].

CONCLUSION

In conclusion AM is a descriptive term of a distended, mucus filled appendix caused by various conditions, both benign and malignant. Preoperative diagnosis is difficult due to nonspecific clinical manifestations. Correct preoperative diagnosis and careful resection is necessary to prevent pseudomyxomaperitonei like complications.

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