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Appendicular mucocele: a case report and review of literature

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ABSTRACT: Appendicealmucocele is a rare disease. Sometimes it is discovered accidentally and sometimes it resembles acute appendicitis. Correct diagnosis before surgery is very important for the selection of adequate surgical treatment to avoid intraoperative and postoperative complications. Ultrasonography, and particularly computed tomography, should be used extensively for this purpose. If mucocele is treated incorrectly pseudomyxomaperitonei, which is characterized by malignant process, may develop. We present a case of 69 year old man presented with pain lower abdomen for few days. Imaging showed cystic mass in right iliac fossa . Appendicular mucocele was suspected. Laparotomy was performed. At the time of surgery, a cystic mass of the appendix with dimensions approx. 10 × 5 ×3 cm, with thin walls, but without perforation was discovered in the right iliac fossa. No discharge was found in the peritoneal cavity. Only appendectomy was performed because no pathologic process was found in the base of the appendix and lymph nodes were not palpable. Histopathologic diagnosis was mucinous cystadenoma.

Keywords:appendicular mucocele, mucinous cystadenoma , mucinous tumour , appendectomy

I. INTRODUCTION

Appendicularmucocele is a rare disease where there is obstructive dilatation of the appendix caused by intraluminal accumulation of mucoid material.The incidence is 0.2% to 0.7%.[1-5]There are 4 histologic types of appendicularmucocele: retention cyst, mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma.[6-7]Appendicular mucocele may present the same way as acute appendicitis with pain in right lower quadrant of abdomen, fever, leukocytosis etc. It can present as nonspecific lower abdominal pain or even asymptomatic. If treated improperly, the mucocele may progress, epithelial cells may escape into the peritoneal cavity, and pseudomyxomaperitonei may develop, which has a high mortality.[8-9]

II. CASE REPORT

A 69 years old male patient presented to General Surgery OPDwith complaints of pain in right lower quadrant of abdomen for last three weeks. Pain was mainly present in right iliac fossa, which was gradually progressive in nature, dull aching type of pain and non-radiating. There were no aggravating or relieving factors.When palpating the lower right quadrant of the abdomen, mild tenderness elicitated in right iliac fossa. An ill-defined lump palpated. Patient was afebrile. Leucocyte count was normal.Patient came with reports ofUltrasound abdomen and CT scan abdomen which suggested of 11.1 x 5.7 cm mass in Right iliac fossa & bilateral renal calculusand well capsulated thin walled mesenteric lymphangiomaor cyst & bilateral renal cortical cyst respectively. Patient underwent repeat ultrasound abdomen in our hospital which showed “approximately 10.4 x4.8 cm sized elongated tubular hypoechoic structure with peripheral calcification & internal echogenic contents seen in right iliac fossa. No internal vascularity seen. Suggestive of appendicular mucocele more likely than a mesenteric cyst”.

He was planned for exploratory laparotomy and found a large cystic mass of appendix (10 x 5 x 3)cm.No discharge was found in the peritoneal cavity. Base of the appendix was healthy and free from macroscopic disease. There was no lymphnodes palpable. Appendectomy performed and specimen sent for histopathological examination which showed mucinous cystadenoma without any evidence of malignancy. Post operative recovery was uneventful.

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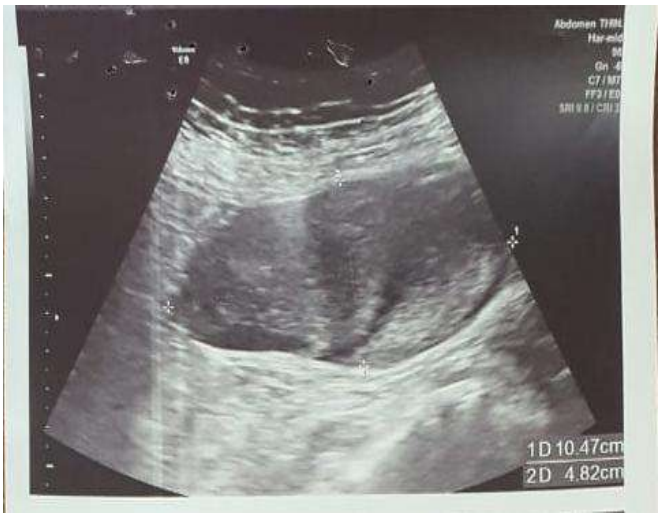

Fig 1 – ultrasonography suggestive of appendicular mucocele

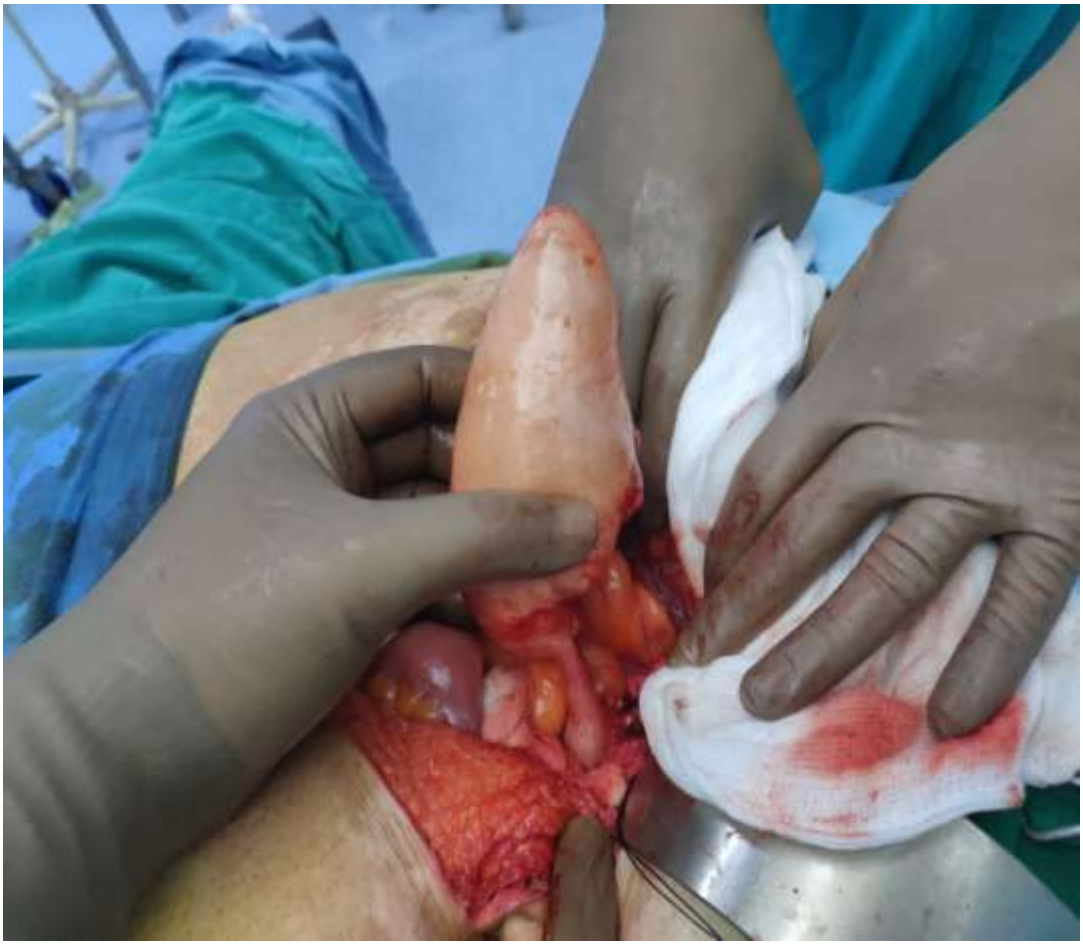

Fig 2 – mucocele of appendix seen at laparotomy. The base is healthy and free from macroscopic disease.

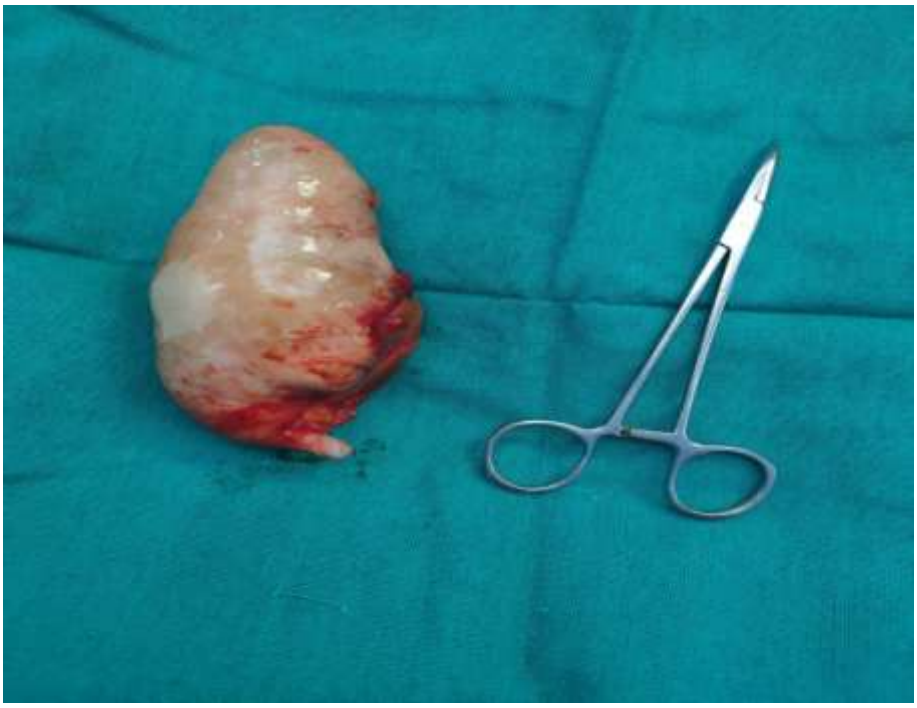

Fig 3 – resected specimen

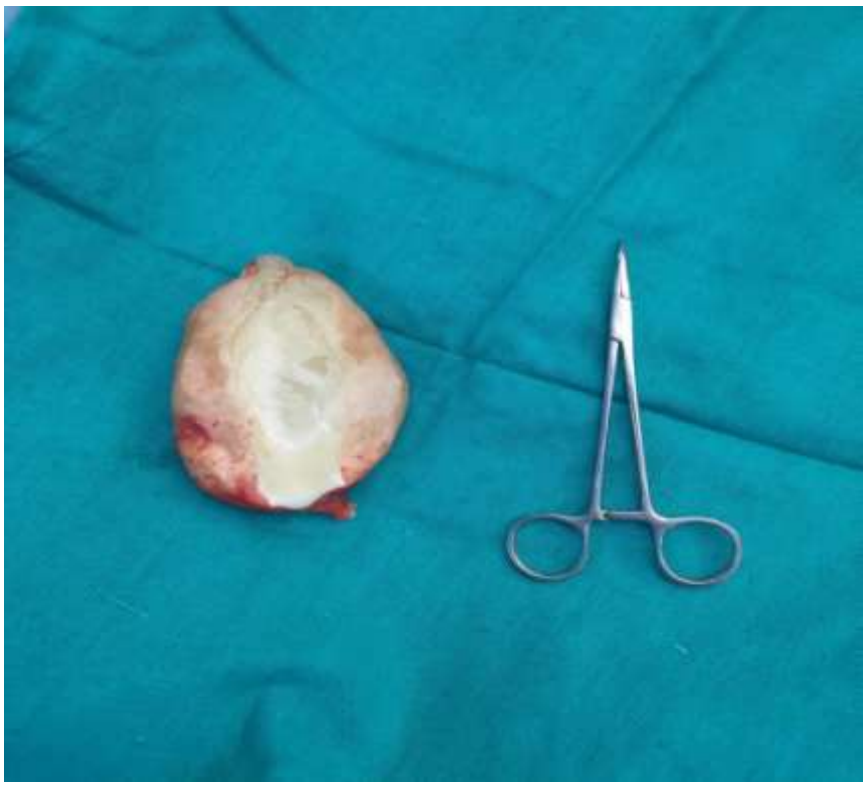

Fig 4 – specimen showing mucoid material upon incising

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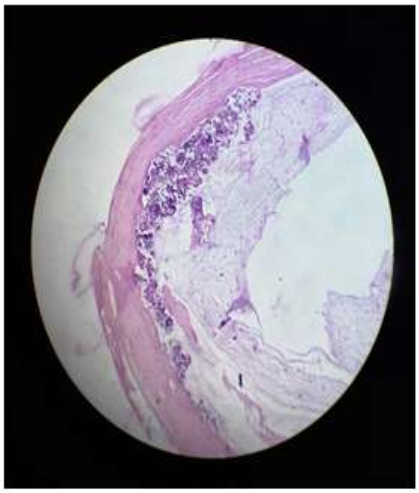

Fig 5

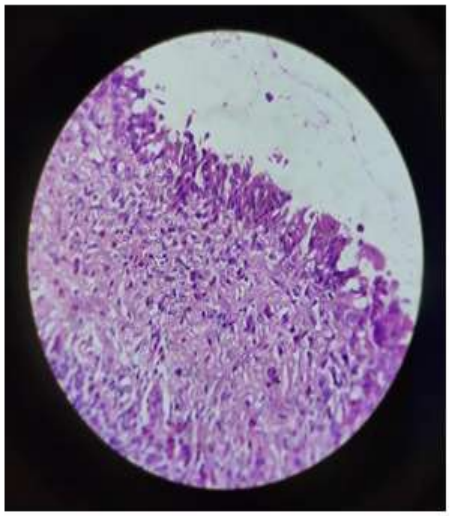

Fig 6

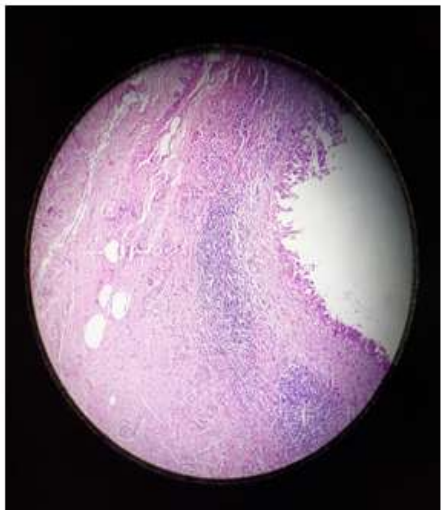

Fig 7

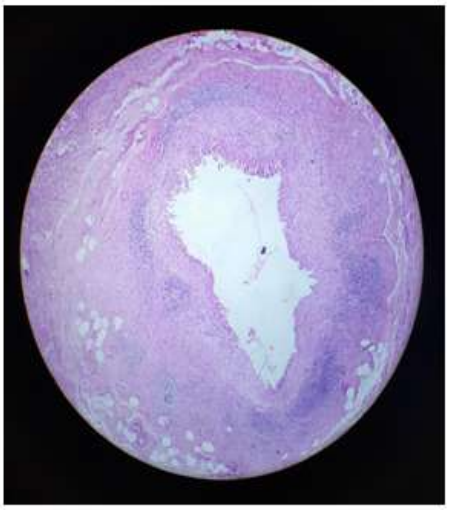

Fig 8

Fig [5-8] – histological sections show mucinous cystadenomaof appendix. There is no evidence of malignancy

III. DISCUSSION

Mucocele of the appendix was first described by Rokitansky.[11] This disease is characterized by dilatation of a lumen as a result of an accumulation of a large amount of mucus. The appendix is lined by epithelium containing more goblet cells than the colon. As a result, most appendicular epithelial tumors are mucinous and start as mucoceles.[12] It falls under the category of rare diseases. Its incidence ranges between 0.2% and 0.7% of allexcised appendixes. This condition can have benign as well as malignant processes. According to modern classification, there are 4 histologic types: retention cyst, mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma.[1-7,10,13]The clinical flow of the disease does not have a specific picture. It often flows asymptomatically. In about 50% of cases it is discovered accidentally during radiologic and endoscopic examinations or at surgery. A patient's clinical symptoms may include pain in the right lower quadrant of the abdomen, palpable abdominal mass, nausea, vomiting, weight loss, gastrointestinal bleeding, and signs of intussusception of the intestines.[14-19]

Mucocele of the appendix is no longer considered a benign tumour but is regarded as a low-grade borderline malignant lesion or, more appropriately, mucinous tumour of uncertain malignant potential (UMP) to distinguish it from the overtly more aggressive mucinous adenocarcinoma. In contrast to the latter, tumours

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in the UMP category appear benign on histology, rarely metastasize to lymph nodes and, although they may give rise to Psedomyxomaperitonei (PMP), they rarely invade parenchymal organs except for the ovary or spread to regional lymph nodes. Thus PMP caused by UMP tumours (usually following rupture) carries a much better prognosis than in cases caused by mucinous adenocarcinomas.

One classification of appendiceal mucinous tumours proposed by Pai and Longacre qualifies the designation UMP for mucinous tumours of uncertain malignant potential with M-UMP for mucinous tumours which have spread to the pertinoneum but are clearly not invasive. The details of this classification are as follows.

1 Adenoma: simple or focally stratified columnar epithelium with goblet cells; mild to moderate atypia; no atypical mitosis, no stromal invasion, no extra-appendiceal epithelium – its oncological significance is that when completely excised without rupture it does not recur; but rupture may cause acellular mucinous ascites.

2 Mucinous tumour of UMP: as in 1 but (1) with involvement of proximal margin, (2) mucin with epithelium though present within the wall is not overtly invasive, (3) there is uncertainty
on the presence of epithelium within extra-appendicealmucin – its oncological significance relates to the risk of PMP and need for close follow-up.

3 Mucinous tumour of M-LMP: as in 1, but neoplastic cells are present in peritoneal implants. May require hyperthermicintraperitoneal chemotherapy.

4 Mucinous adenocarcinoma: a frankly invasive tumour which may cause aggressive PMP with visceral invasion and nodal metastases.

However, there are other classifications and there seems to be a lack of agreement amongst expert pathologists in defining a generally accepted classification and some have suggested a simpler histological grouping into (1)appendicealmucinous neoplasm with low dysplasia, (2) appendiceal mucinous neoplasm with high-grade dysplasia and (3) invasive mucinous adenocarcinoma.[AlfredCuschieri, George B. Hanna ; Essential surgical practice , 5th Ed , p 932-933]

Preoperative diagnosis is important for the selection of the appropriate surgical procedure in order to prevent intra-operative complications especially particular peritoneal dissemination, to prevent intraoperative and postoperative complication, and repeated surgery.[1]USG, computed tomography (CT), and colonoscopy is used for diagnostics. USG is the first-line diagnostic method for patients with acute abdominal pain. USG can be used to differentiate between mucocele and acute appendicitis. In case of acute appendicitis, the outer diameter threshold of the appendix is 6 mm, and 15 mm and more indicates the presence of a mucocele, with 83% sensitivity and 92% specificity.CT is regarded as the most accurate method of diagnostics. CT can be used to discover the signs specific to mucocele with high accuracy: appendix lumen more than 1.3 cm, its cystic dilatation, and wall calcification.[20,23]By colonoscopy an elevation of the appendicular orifice is seen and a yellowish mucous discharge would be visible from this orifice. Furthermore, synchronous and metachronous tumors of colon can be identified.[24-25]

One of the cardinal principles of surgical treatment of this disease is that intact mucoceledo not pose a threat for the patient. If it is perforated and the filling turns up in the peritoneal cavity, there is a high probability that pseudomyxomaperitonei will develop, for which treatment is very problematic and long-term results are quite unsatisfactory. [26]Therefore, the selection of an adequate surgical method is very important. Most surgeons think that open surgery should be favored against laparoscopy. If the surgery was launched using a laparoscopicmethod and it appears that there is an appendicularmucocele, it must be converted into open surgery. This has 2 objectives: (1) to perform surgery carefully so the cyst is not ruptured and the filling is not scattered into the peritoneal cavity and (2) with an open surgery 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.[27]Some surgeons consider that the operation can be performed using a laparoscopic method by adhering to safety rules, especially when removing the mucocele from the abdomen and an endobag must be used. [28-29]

An algorithm for the selection of the type of surgery has been furnished by Dhage-Ivatury and Sugarbaker. It envisages several factors: (1) whether or not a mucocele is perforated; (2) whether the base of the appendix (margins of resection) is involved in the process; and (3) whether there are positive lymph nodes of mesoappendix and ileocolic. As a result patients

may require different operations: appendectomy to the right colectomy, including cyst reductive surgery, heated intraoperative intraperitoneal chemotherapy, early postoperative intraperitoneal chemotherapy.[14]For mucinous cystadenoma, appendectomy is sufficient if the lesion doesnotinvolve the appendiceal base. Occasionally the mass will rupture prior to or at the time of removal, but this rupture is typically contained to the right lower quadrant and is considered localized pseudomyxomaperitonei. If the mass is benign, appendectomy and removal of any residual mucin is curartive.[7]The recommended treatment of mucinous adenocarcinoma consists of right hemicolectomy with debulking of any gross spread of disease and removal of all mucin. It is not uncommon, however, for the diagnosis to be unknown until the time of pathologic evaluation of the appendectomy specimen. In such cases, re-operation with right hemicolectomy is recommended.[30]In our patient the mucocele was not perforated (no discharge into the peritoneum cavity), there was no pathologic process in the base of the appendix (negative margins of resection), 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.Histopathologic examination confirmed the diagnosis of mucinous cystadenoma without any evidence of malignancy.

IV. CONCLUSION

Appendicularmucocele is a rare disease and has a clinical picture thatmay resemble acute appendicitis. A correct diagnosis before surgery is very important for the selection of surgical technique to avoid severe intraoperative and postoperative complications. USG, particularly CT, should be used extensively for this purpose.

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