Abstract: Mucocele of appendix is a very rare entity present in 0.2-0.3 percent of appendicectomies. Here we present a 40 years old female admitted with complaints of right lower abdominal pain for the past one month. Initially diagnosed as having chronic appendicitis. USG abdomen revealed tubular structure in right iliac fossa suggestive of Mucocele of appendix. CT abdomen confirmed the diagnosis. Laparotomy revealed a distended appendix of about 8 x 3 cm, which was not adherent to the adjacent structures. The patient underwent appendicectomy with wide resection of meso appendix. She recovered well and the histopathological report came as mucocele of appendix probably as retention cyst. The patient is now free of symptoms. Although mucocele of appendix is uncommon, preoperative diagnosis to rule out underlying malignancy is important for patient management.

Keyword: Mucocele of Appendix, cystadenoma, cystadenocarcinoma, pseudomyxomaperitonei.

Introduction: Mucocele of the Appendix is an obstructive dilatation of the appendix caused by either a benign or a malignant process. It is a rare lesion found in only 0.2% to 0.3% prevalence among appendectomies. (1) Approximately 25-50% of Mucoceles of appendix are asymptomatic and are found incidentally during abdominal imaging or surgery. Mucocele of the appendix was first described by Rokitansky in 1842. (2) The common causes of Mucocele of appendix include, mucinous cystadenoma, mucinous cystadenocarcinoma, diffuse or focal villous hyperplasia and retention cyst. (3) The most dreaded complication of benign or malignant mucocele is pseudomyxoma peritonei, which is difficult to treat surgically or medically. It has an uncertain prognosis, with a 5-year survival rate reported to be around 53%. A consensus on the optimal management of the appendicular mucocele has not yet been reached. Some recommends appendicectomy alone while other supports the undertaking of right hemicolectomy. (4)
Here we report a case of 40 years old female who underwent an laparotomy with appendectomy and wide resection of the mesoappendix.

CASE REPORT:
A forty year old woman admitted with right sided lower abdominal pain for the past one month. There were no associated bowel or bladder symptoms. Patient had no other systemic illness. She had no personal or family history of trauma, inflammatory bowel disease or malignancy. Patient was awake, alert with stable general condition. Her abdomen was soft with tenderness in the right iliac fossa at the time of admission. She had no guarding or rigidity. Her laboratory test values were all within normal limits. Ultra sonogram abdomen revealed tubular anechoic fluid containing lesion of size 67 x 23mm seen in right iliac fossa, suggestive of mucocele of appendix. CT scan abdomen was taken which revealed Tubular hypo dense lesion extending for a length of 6.5 x 2.3 cm seen in the right iliac fossa with attachment to the caecum with no evidence of lymphadenopathy. On laparotomy the appendix was found to be distended measuring 8 x 3 cm with no free fluid, not adherent to the surrounding structures. Entire peritoneal cavity was inspected and found to be normal. Liver, ovaries, uterus, colon, stomach, small bowel and large bowel were normal. As the base of the caecum was not involved and there was no local invasion, appendicectomy with wide resection of mesoappendix was done. Histopathology report came as Mucocele of appendix probably retention cysts. Post operatively patient passed flatus on 2nd POD. Oral fluids started on 4th POD. Patient discharged on 8th POD without any postoperative complications and advised follow up.
Discussion:
Mucocele of the Appendix is a rare entity often present as an incidental surgical or radiological finding. It is found in only 0.2% to 0.3% prevalence among appendectomies. Approximately 25-50% of Mucoceles of appendix are asymptomatic and are found incidentally during abdominal imaging or surgery. Mucocele of the appendix was first described by Rokitansky in 1842. The term mucocele is widely used in diagnosing both benign and malignant lesions. Mucocele of appendix are sub divided according to the histopathological morphology into 4 sub groups – Retention cyst (18%), mucosal hyperplasia (20%), mucinous cystadenoma (52%), and mucinous cystadenocarcinoma (10%). Microscopically mucinous cystadenoma is characterised by cellular atypia, glandular and papillary proliferation. Mucinous cystadenocarcinoma is characterised by invasion, local spread or peritoneal spilling. Mucosal hyperplasia is characterised by additional hyperplasic epithelium and retention cyst are lined by flat epithelium with dystrophic mineralisation, fibrosis and mucous in the lumen of the cyst.. The clinical presentation is often nonspecific and it is often an incidental finding while doing radiological investigation or at doing surgery for appendicitis (23-50%). (5) Signs and symptoms occur in fewer than 50% of cases and are generally associated with malignancy. These include pain in the right lower abdominal quadrant, an abdominal mass, weight loss, nausea, vomiting, and change in bowel habits, anaemia, and hematochezia. (5) The initial detection of the lesion may be facilitated by radiological, sonographic or endoscopic means. In our patient initially the diagnosis was made with the help of Ultrasound later confirmed by CT abdomen. On barium enema, there is usually non filling or partial filling of the appendix with contrast. The lesion may be seen as a sharply outlined sub mucosal or extrinsic mass indenting the caecum and laterally displacing it.
Ultrasound findings in mucocele of appendix can vary from purely cystic lesions with anechoic fluid, hypo echoic masses with fine internal echoes as well as complex hyper echoic masses depending on the contents. Internal septations, polypoid lesions extending into the lumen and irregular shapes seem to be associated with the malignant variety. The onion skin sign in a cystic mass lesion in the right lower quadrant is useful in assisting the diagnosis of mucocele appendix. (6,7) In our patient the Ultrasound abdomen revealed tubular anechoic fluid containing lesion of size 67 x 23mm seen in right iliac fossa suggestive of mucocele of appendix CT of the abdomen usually shows a cystic, well-encapsulated round or ovoid mass centred in the right iliac fossa with Mural calcification and absence of periappendiceal inflammation or abscess. CT scanning has the advantage of
allowing precise observation of the relationship between the lesion and adjacent organs and any other abnormalities associated with mucocele. The dreaded complications of mucocele of appendix pseudomyxoma peritonei is characterised on CT by the presence of low attenuation ascities with scalloping of liver contour due to peritoneal implants. Loculation of ascitic fluid with associated mass effect should lead to a consideration of pseudomyxoma peritonei. (8,9) In our patient the CT-scan abdomen revealed Tubular hypo dense lesion extending for a length of 6.5 cm seen in the right iliac fossa with attachment to the caecum with no evidence of lymphadenopathy.

In suspected cases of appendiceal mucocele, fine needle aspiration should be avoided to preserve integrity of the appendix and prevent tumour inoculation. Colonoscopy findings include the 'volcano sign', the appendiceal orifice seen in the centre of a firm mound covered by normal mucosa or a yellowish, lipoma-like submucosal mass. (10) Although complications from appendiceal mucocele are minimal, there is evidence that complications are associated with concomitant neoplasm. Therefore surveillance colonoscopy and removal of polyps in any patient with an appendiceal mucocele should be done. Most investigators agree that the adenoma-adenocarcinoma sequence is similar to the colonic polyp-adenocarcinoma sequence. Complication of Mucocele of appendix include intussusceptions, bleeding, perforation, peritonitis, rupture and pseudomyxoma peritonei. (11) The most dreaded complication of benign or malignant mucocele is pseudomyxoma peritonei, which is difficult to treat surgically or medically. All patients, whether they have benign or malignant appendiceal mucocele, should be evaluated for pseudomyxoma peritonei. Pseudomyxoma peritonei more commonly occur in malignant appendiceal mucocele of about 95% as opposed to 13% in patients with non-malignant appendiceal mucocele.

It has an uncertain prognosis, with a 5-year survival rate reported to be 53%. (13) The differential diagnosis includes ovarian cysts and tumours, duplication cyst, mesenteric cyst and omental cysts, mesenteric haematoma or tumour, abdominal abscess, retroperitoneal haematoma, renal cyst and pseudo cyst of pancreas. (14) In our case, laparotomy and appendicectomy with wide resection of mesoappendix was made as a result of diagnostic certainty. An intact mucocele presents no future risk for the patient; however the opposite is true if the mucocele had ruptured into the peritoneal cavity.

Laparoscopic surgery provides the advantages of good exposure and evaluation of entire abdominal cavity for pseudo myxoma peritonei, as well as more rapid recovery with avoidance of a large incision and a better cosmetic outcome. Careful handling of the specimen is recommended as spillage of the contents can lead to pseudomyxoma peritonei. This can be achieved by atraumatic handling of the appendix and use of impermeable bag for removal of the specimen. Conversion to laparotomy should be considered if the lesion is traumatically grasped or if the tumour clearly extends beyond the appendix or if there is evidence of malignancy such as peritoneal deposits. Involvement of the caecum or adjacent organs is an indication for right hemicolectomy and thorough exploration of the gastrointestinal tract and ovaries. (15)
Conclusion:
Mucocele of appendix is a very rare entity present in 0.2-0.3% of appendicectomies. Approximately 25-50% of Mucocele of appendix are asymptomatic and are found incidentally during abdominal imaging or surgery. In our case preoperative evaluation, including sonography and CT scan, resulted in diagnosis of mucocele of appendix confirmed by surgery. Appendicectomy is curative for this condition, as neither regional nor distant lymph node metastases nor haematogenous spread have been reported. It is concluded that preoperative diagnosis by imaging modality is extremely helpful to the operating surgeon for careful mobilisation and resection of the appendix without peritoneal contamination which is essential for optimal results. Since the occurrence of cystadenoma and villous hyperplasia is more when compared to cystadenocarcinoma in mucocele of appendix, appendicectomy with wide resection of appendix rather than right hemicolectomy is justifiable, however if the diagnosis of cystadenocarcinoma is confirmed by histopathological examination right hemicolectomy has to be done. If symptoms due to pseudomyxoma peritonei occur, a continued aggressive surgical approach is justified.

Reference:


