



## A Rare Case of Mucinous Cystadenoma of Appendix with Pseudomyxoma Peritonei Presenting as Umbilical Hernia : An Interesting Case Report

Arunvathani A, Balakrishnan S, Senthil Kumar M, Kudiyaarasu M, Prabhakar P.

Department of General Surgery, Madras Medical College, Chennai.

### Abstract

Mucinous cystadenoma of appendix is a rare condition that develops as a result of proliferation of mucin-secreting cells in an occluded appendix. Mucinous cystadenoma presenting as umbilical hernia is a rare clinical entity. Pseudomyxoma peritonei is a relatively uncommon presentation of appendiceal adenocarcinoma or ovarian cystadenoma. Presentation as hernia is even more rare and can include virtually any type of ventral hernia. We report a rare case of mucinous cystadenoma of appendix with pseudomyxoma peritonei presenting as an umbilical hernia in a 48-year old female patient. The patient had a one and a half year history of a reducible mass in the umbilical region and was diagnosed with umbilical hernia. Ultrasonography and Contrast enhanced computed tomography of abdomen and pelvis were performed and revealed ascites and the hernial defect. Ultimately, open appendectomy was performed and a low grade mucinous neoplasm of appendix was confirmed. In addition, the present report discussed the association between mucinous cystadenoma of the appendix and umbilical hernia, as well as the diagnostic process and treatment strategies.

**Keywords:** Mucinous cystadenoma, appendix, umbilical hernia, Pseudomyxoma peritonei

### Case Report

A 48-year old female patient presented to our OPD with complaints of swelling in the umbilical region for 1½ years, which was insidious in onset, progressive in nature, initially reducible, now irreducible for the past 6 months. No H/O of abdomen pain, distension, or vomiting. Patient didn't take any treatment for the same as the condition was painless without any symptoms. She was a known diabetic on irregular treatment with oral hypoglycemic

drugs for the past 5 years. There was no other significant past medical or surgical history.

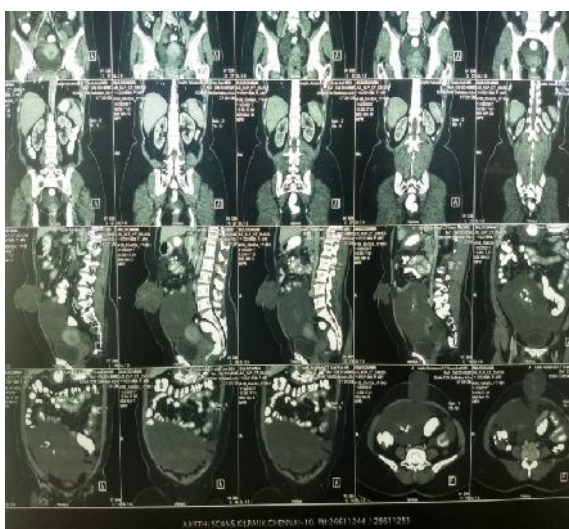
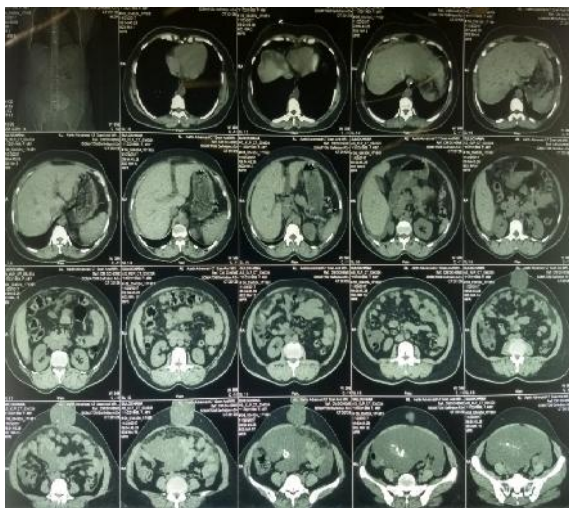
On examination a swelling of size 4 x 4 cm was present over the umbilical region with a visible cough impulse, which was spherical in shape, soft in consistency with a overlying skin stretched and shiny. Generalized abdominal distension was present with no fluid thrill or shifting dullness. No other palpable mass or organomegaly. Bowel sounds were normal. Per-Rectal examination was normal. Rest of the systemic examination were normal. Routine blood investigations were within normal limits. TFT showed thyronormalcy.

Ultrasound of the abdomen done showed 4 cm defect noted in umbilicus with omentum as content. Free fluid was present in the abdomen which was loculated and with septations. Final impression was umbilical hernia with moderate ascites. High frequency ultrasound showed evidence of 1.5 cm defect noted in umbilical region with omentum and bowel as its content. Evidence of peristalsis noted within the herniated bowel. Features suggestive of umbilical hernia with omentum and bowel as content.

CECT-abdomen and pelvis with IV contrast showed

- Free fluid seen in the right perihepatic space, perisplenic space, paracolic gutters and pelvis suggesting ascites.
- Anterior abdominal wall defect seen measuring 3.2 cm with herniation of the septated fluid through the defect.

- Omental nodularities and fat stranding suggesting caking noted with the involvement of the transverse mesocolon.
- Small nodes seen in the mesenteric region measuring 4 x 2 mm.
- Specks of calcification and soft tissue density seen (small bowel stricture ileal) involving the lower mesenteric region with soft tissue density lesion on the right side measuring 5.6 x 4 cm in a linear manner.
- Significant sized nodes in the central mesentery measuring 14 x 8 mm
- Both ovaries are normal



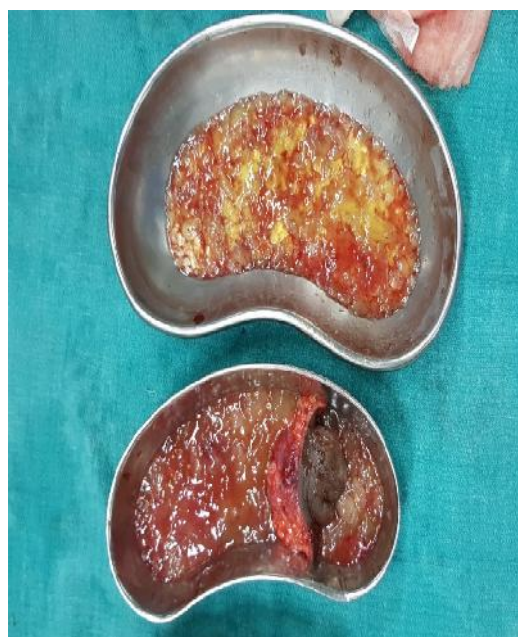
## Impression :

DIAGNOSTIC POSSIBILITIES ARE INFECTIVE CAUSE LIKE TUBERCULOSIS- SUGGESTED HISTOPATHOLOGICAL CORRELATION OF THE ASCITIC FLUID

Ultrasound guided aspiration of ascitic fluid attempted. Screening USG showed echogenic ascitic fluid with loculations-? Organized. Minimal thick fibrinous fluid aspirated and sent for cytology. The cytology report showed a moderately cellular smear composed of scattered lymphocytes and reactive mesothelial cells in a fluid background. Impression: Reactive effusion.

Thoracic physician opinion obtained. It suggested sputum AFB, digital chest X-ray and CECT chest. Sputum AFB came negative. Chest X-ray was normal. CECT chest showed no significant abnormality.

Since patient had persistent abdomen pain, distension and an irreducible umbilical hernia, planned to proceed with an elective umbilical hernia mesh repair. After getting diabetologist fitness and anaesthetic fitness, patient was taken up for surgery. Under general anaesthesia a transverse elliptical incision made around the umbilicus and deepened till rectus. Plane created all around the neck of the sac. Neck found to be wide and opened at the lower end. Sac content found to be a jelly like substance. Entire hernial sac excised above the rectus with umbilicectomy done. Rectus opened in midline, above and below. Ascitic fluid of mucinous content about 600 ml drained and sent for analysis and HPE.





Thorough laparotomy done. Liver and spleen surface found to be nodular with specks of mucin over it. Since the ascitic fluid was mucinous jelly like substance with a picture of pseudomyxoma peritonei, primary was searched for in the ovaries and the appendix. Appendix was found to be the primary with the mucinous cystadenoma (neoplasm) approximately of size 7 x 5x4 cm about 4 cm from the base of the appendix.



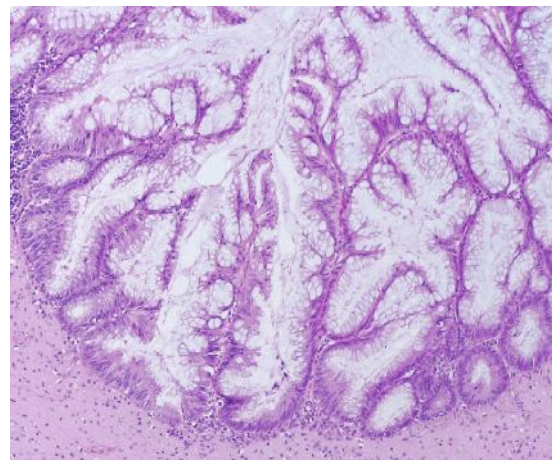
Appendicectomy was done in usual manner. Greater omentum excised in-toto. Thorough wash given with 3 litres of normal saline. Hemostasis secured. Two drainage tubes kept- left urobag DT kept in pelvis, right ICD tube kept in right paracolic gutter. Romovac DT kept in subcutaneous plane. Wound closed in layers. Rectus with 1-0 prolene, subcutaneous with 2-0 vicryl, skin with 1-0 ethilon.

Specimen sent for the histopathological examination are the following:

- Appendicectomy specimen with the mass
- Base of the appendix
- Two adjacent lymph nodes
- Umbilical hernial sac with the mucinous sac
- Entire omentum
- Ascitic fluid for analysis and HPE



Post operative period uneventful. Paracolic and subcutaneous drain were removed on day 3. Pelvic drain was removed on day 7. Sutures were removed on day 12. Postoperative HPE came as a low grade mucinous neoplasm, possibly from appendix. Medical oncology opinion obtained suggested palliative chemotherapy with 5-Fluorouracil and cisplatin for stage IV mucinous carcinoma appendix – post laparotomy status.



## Discussion

Umbilical hernia is a common surgical complication, with ~10% of all primary hernias comprised of umbilical and epigastric hernias. According to the European Hernia Society classification, abdominal wall hernias located 3 cm above to 3 cm below the umbilicus are defined as umbilical hernia. In adults, intra-abdominal hypertension is a major cause of umbilical hernia. Predisposing factors for intra-abdominal hypertension include obesity, pregnancy or ascites. There are two primary repair options for umbilical hernias: Suture and mesh. Currently, due to low recurrence rates, mesh reinforcement is recommended. Mucocoele of the appendix is a descriptive term that implies a dilated appendiceal lumen caused by abnormal accumulation of mucus. Mucinous cystadenoma is the most common cause of mucocoeles of the appendix (63–84%), and can generate a large amount of ascites. An appendiceal mucocoele is a rare condition that is observed in 0.2–0.6% of all appendectomy specimens. Mucinous cystadenoma of the appendix is a rare clinical entity with symptoms that can vary, which poses a diagnostic challenge. Appendectomy is advised for focal or diffuse mucosal hyperplasia and cystadenoma when the appendiceal base is intact. Cecal resection is performed for cystadenoma with a large base and right colectomy is recommended for cystadenocarcinoma. In the present case, the increased abdominal pressure followed by the accumulation of ascites contributed to the occurrence of an umbilical hernia.

In adults, intra-abdominal hypertension is a major cause of umbilical hernia. The possible causes of intra-abdominal hypertension include obesity, pregnancy or ascites. In the present case, it was believed that the main cause of the umbilical hernia was ascites. The diagnosis of mucinous cystadenoma of the appendix during umbilical hernia repair was unexpected.

An appendiceal mucocoele is a rare condition that is observed in 0.2–0.6% of all appendectomy specimens. Four histological types of appendiceal mucocoele exist: i) Retention cysts, ii) mucosal hyperplasia, iii) mucinous cystadenoma; and iv) mucinous cystadenocarcinoma. Mucinous cystadenoma is a rare cystic neoplasm of the vermiform appendix, which characteristic villous adenomatous changes of the appendiceal epithelium that are associated with marked distension of the appendiceal lumen with mucin. Among these four types, mucinous cystadenocarcinoma has the least developed etiology, and the clinical progression of the disease has not yet been determined. The clinical symptoms for mucinous cystadenocarcinoma may include right lower abdominal pain, palpable abdominal masses, weight loss, nausea, vomiting, gastrointestinal bleeding and signs of intestinal intussusception; however, in the present case, the disease presented as an umbilical hernia.

Peritoneal pseudomyxoma is a peritoneal or retroperitoneal accumulation of a gelatinous substance secondary to the rupture of a mucinous appendiceal lesion,

thus it occasionally combines mucinous ascites and peritoneal implants, in which case the prognosis is considerably poorer, synonymous with recurrent peritoneal involvement, which is known as a gelatinous disease of the peritoneum.

Pseudomyxoma peritonei, a term first used by Werth that actually means false tumour of the peritoneum, usually has its origins in an appendiceal adenocarcinoma or ovarian cystadenoma, although other sites have also been described. It is a progressive disease and mostly affects women in their fifth to sixth decades. The earlier suggestion of a common ovarian origin is now debated and it has been proposed that women may have synchronous involvement of the appendix and ovary, with ovaries being secondarily involved following appendix rupture due to an enlarging mucinous adenocarcinoma. Clinical presentation varies and usually starts with non-specific symptoms which may include unexplained weight loss quickly followed by abdominal distension and pain. The most common presentation which prompts patient to present to hospital is features of appendicitis (27%) followed by increased abdominal girth (23%). Presentation as an ovarian mass (20%) is the third most common followed by hernia (14%). Various other rare presentations have also been reported including recurrent incisional hernia.

In the present case, the intraoperative diagnosis was of a mucinous cystadenoma of the appendix, with suspected malignant potential. An appendectomy was the only surgical procedure performed, since it is sufficient for the treatment of such cases.

In conclusion, patients with appendiceal mucocoeles present with a range of clinical symptoms. In the present case, the patient presented with an umbilical hernia. An accurate diagnosis is essential for the selection of the appropriate surgical procedure. Although rare, appendiceal mucocoeles should be considered during the diagnosis of an umbilical hernia.

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