Case Report

DOI: 10.7860/JCDR/2024/68272.18937

Oncology Section

Mucin-secreting Adenocarcinoma of the Appendix Presented as Abdominal Lump with Distension: A Case with Diagnostic Dilemma

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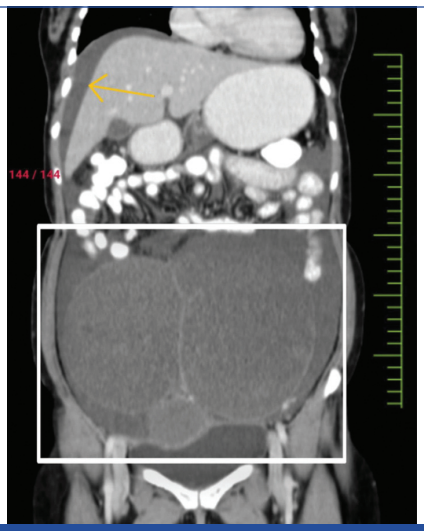
ABSTRACT

Appendicular adenocarcinoma is a rare tumour, and the mucin variety is common. It is usually presented with symptoms such as abdominal pain, abdominal tenderness, nausea, vomiting, change in bowel habits, and unexplained weight loss. These symptoms can resemble those of other clinical conditions, making the diagnosis of appendicular adenocarcinoma challenging. Screening modalities such as Magnetic Resonance Imaging (MRI), Computed Tomography (CT) imaging, Contrast-Enhanced Computed Tomography (CECT), blood tests, and histopathological analysis can help confirm the diagnosis. This case is reported to highlight the unusual presentation and diagnostic difficulties associated with appendiceal adenocarcinoma. A 48 years old female patient presented with complaints of dull, continuous abdominal pain for the past six months, along with an abdominal lump and distension, without any other associated symptoms. Laboratory investigations for Cancer Antigen 125 (CA 125), β-Human Chorionic Gonadotropin (HCG), and α-fetoprotein were found to be within normal limits. Further, CECT imaging was suggestive of mucin-secreting adenocarcinoma of ovarian origin. The final diagnosis was confirmed by histopathological analysis as mucin-secreting adenocarcinoma of the appendix with pseudomyxoma peritonei and was managed through cytoreduction surgery.

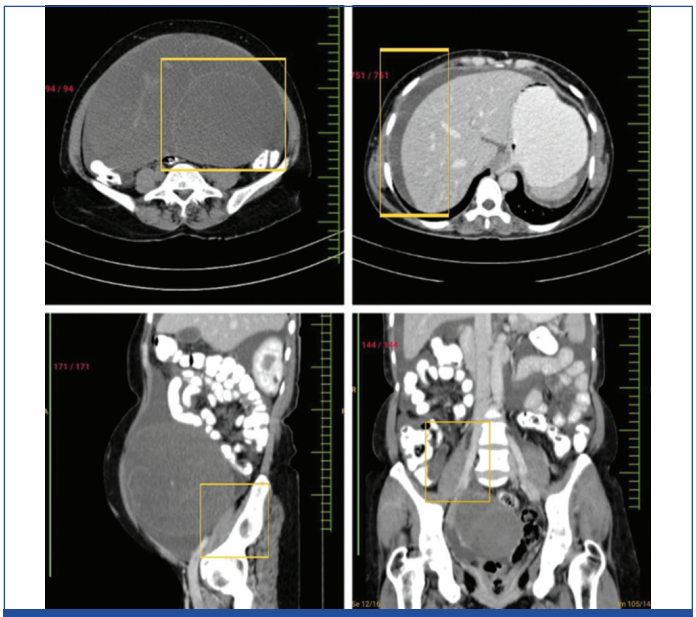
Keywords: Cancer antigen, Mucin, Ovarian cyst, Screening modalities

CASE REPORT

A 48-year-old female (G2P0L2A0) with a past history of hysterectomy for vaginal prolapse six years ago presented with a chief complaint of continuous, dull aching pain in the lower abdominal region and abdominal distension for the past six months. The patient did not have any other related complaints such as changes in micturition, bowel habits, or passing of stools. During abdominal examination, an abdominal lump was palpated in the lower abdomen, along with ascites. Clinical findings suggested that the lump originated from both ovaries. Laboratory investigations for CA 125, β-HCG, and α-fetoprotein were performed and found to be within the normal range. To confirm the diagnosis, a CECT scan of the abdomen and pelvis was conducted, revealing a 15×10 cm lump, likely of ovarian origin, along with the presence of ascites [Table/Fig-1,2].



[Table/Fig-1]: Computed Tomography(CT) image suggestive of enlarge ovary with ascites.

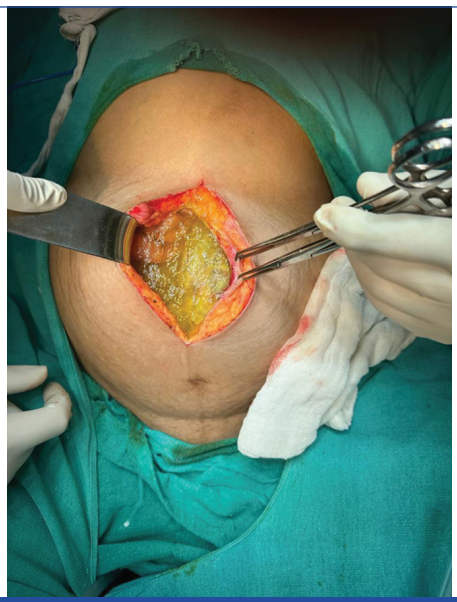


[Table/Fig-2]: Enlarge ovarian cysts on CECT findings, arrow focuses on ascitic fluid.

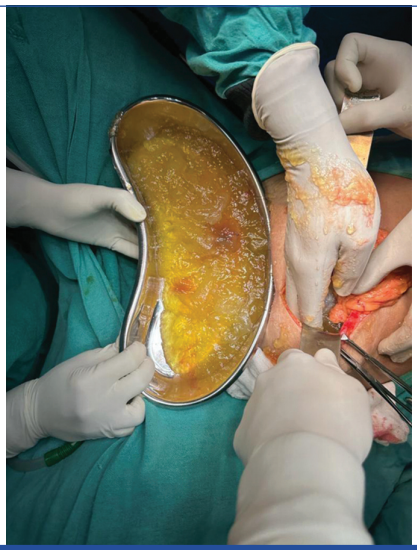
These findings indicated the presence of ascitic fluid and enlarged ovaries.

Based on the CECT findings, cytoreduction surgery of the ovaries was planned using an open abdominal approach. Intraoperative observations revealed the presence of jelly-like fluid throughout the abdomen [Table/Fig-3]. After removing the right ovarian mass, an abnormal morphology of the appendix was discovered, with jelly-like secretion coming out from its tip [Table/Fig-4,5]. The appendix was then removed from its base. The entire appendix, along with both ovaries and the extracted ascitic fluid, underwent histopathological and cytological examinations [Table/Fig-6]. The histopathological report indicated mucin-secreting adenocarcinoma of the appendix

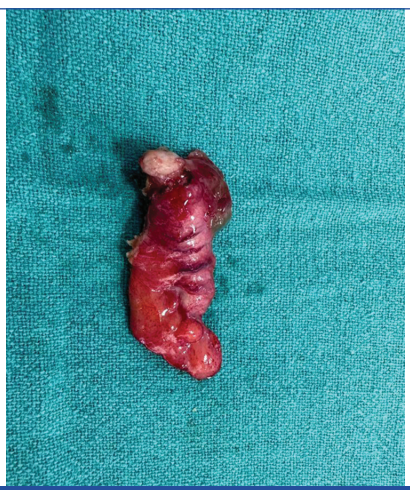
(low-grade), characterised by irregular and jagged glands infiltrating the appendiceal wall and floating in mucin arranged in stripes. The base of the appendix showed no infiltration by malignant epithelial cells, but direct tumour invasion was observed in both ovaries. Cytological examination of the extracted ascitic fluid suggested pseudomyxoma peritonei [Table/Fig-7]. The patient visited the outpatient department for follow-up every three months. At the six-month follow-up, there were no new complaints, the scar site was healthy, and the patient was recovering well.



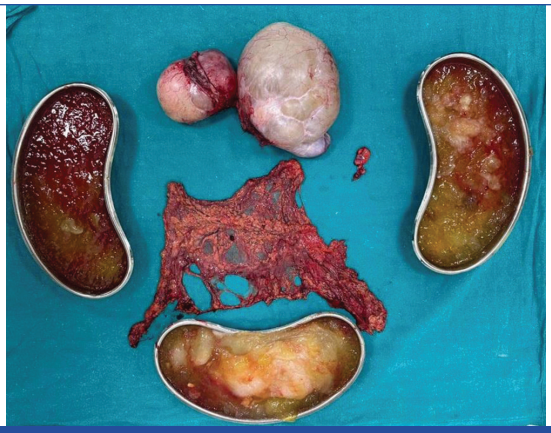
[Table/Fig-3]: Intraoperative mucinous ascitis fluid collected in abdomen.



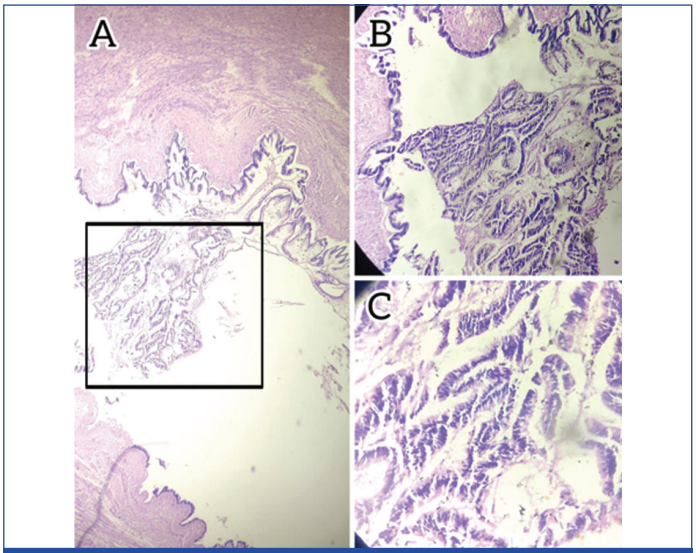
[Table/Fig-4]: Intraoperative mucinous ascitis fluid extraction.



[Table/Fig-5]: Mucin secreting perforated appendix.



[Table/Fig-6]: Excised specimen from abdomen, ovary and extracted mucin.



[Table/Fig-7]: Histopathology images of the extracted sample: A) Section of mucin secreting adenocarcinoma; B) 20X view, H&E, irregular and jagged glands infiltrating the appendiceal wall, floating in the mucin arranged in stripes; C) high power showing the arrangement of glands in stroma 40X view, H&E.

DISCUSSION

Appendiceal adenocarcinoma, also known as mucin-secreting adenocarcinoma of the appendix, is a rare type of cancer originating from the appendix, with reported incidence rates of 0.05% to 0.2% [1,2]. Case presentations of this condition have been reported as early as 1980. Mucin-secreting adenocarcinoma of the appendix primarily affects elderly individuals, both males and females, aged 55 years and older, with some instances of relapse following surgery [3,4].

Mucin-secreting adenocarcinoma is characterised by the production and release of excessive amounts of mucus by cancerous cells in the appendix. The accumulation of mucus can cause enlargement and distension of the appendix, leading to various symptoms and complications [4]. A 10-year study on malignant neoplasms of the appendix reported five cases in individuals aged 14 to 22, four of which were initially diagnosed as appendicitis. Common symptoms of mucin-secreting adenocarcinoma of the appendix may include abdominal pain, changes in bowel habits, bloating, loss of appetite, weight loss, and the presence of mucoceles [1]. Mucoceles are commonly associated with the appendix and can have either a neoplastic or non-neoplastic origin. Non-neoplastic mucoceles are small and rare compared to mucinous neoplasms characterised by mucus secretion. These neoplasms are differentiated into benign and malignant types based on their histological features [5,6].

In this case study, the diagnosis may be unclear based on the clinical symptoms presented. Common symptoms of mucin-secreting adenocarcinoma of the appendix were lacking, leading to the discovery of the disease during surgical management. Treatment approaches are based on tumour staging and histopathology.

Low-grade tumours are often treated surgically with resection of the primary site in the early stages of the disease, while peritoneal debulking and HIPEC (hyperthermic intraperitoneal chemotherapy) are options for advanced-stage disease. Treatment of high-grade tumours requires further prospective trials, and options may include debulking surgery and HIPEC, with or without pre-operative chemotherapy [8]. Therefore, a detailed evaluation-based diagnosis is recommended, combining results from different diagnostic methods such as physical examination, CT scan, biopsy, and, if necessary, MRI scan, to accurately diagnose and manage mucin-secreting adenocarcinoma of the appendix, as in this case.

CONCLUSION(S)

This case presentation highlights a diagnostic dilemma, as the initial complaints in this case were lower abdominal pain and distension. Further evaluation through CT imaging revealed a heterogeneously enhancing lesion in the bilateral ovaries, along with ascites in the abdomen. The presence of mucin-secreting adenocarcinoma of the appendix, with secondary involvement of the ovaries and pseudomyxoma peritonei, was only discovered during surgery. Given the presentation and findings of this case, it emphasises the importance of surgical management in susceptible elderly populations, rather than relying solely on medical management.

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PLAGIARISM CHECKING METHODS: [Jain H et al.]

- Plagiarism X-checker: Oct 26, 2023
- Manual Googling: Nov 22, 2023
- iThenticate Software: Dec 09, 2023 (9%)

ETYMOLOGY: Author Origin

EMENDATIONS: 6

AUTHOR DECLARATION:

- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Oct 25, 2023**
Date of Peer Review: **Nov 15, 2023**
Date of Acceptance: **Dec 12, 2023**
Date of Publishing: **Jan 01, 2024**