ORIGINAL RESEARCH PAPER

Volume-7 | Issue-6 | June-2018 | PRINT ISSN No 2277 - 8179

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

MUCINOUS ADENOCARCINOMA OF APPENDIX : A RARE CASE

General Surgery

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ABSTRACT

The mucocoele of the appendix was first described in 1842 by Rokitansky. It is a rare entity that can present with variety of clinical symptoms or occur as an incidental surgical finding. Appendiceal mucinous neoplasms represent an exceptionally rare form of pathology. Mucinous cystadenocarcinoma, causes a mucocele by the neoplasm occluding the narrow lumen which allows the mucin to build up and distend the appendix. The treatment of choice is often surgical resection combined with adjuvant chemotherapy. Here we report a case of 55 years old lady who presented with a swelling in the right iliac fossa of a short duration. Upon evaluation she was diagnosed to have an appendiceal mucocele which was identified as mucinious cystadenocarcinoma on histopathology

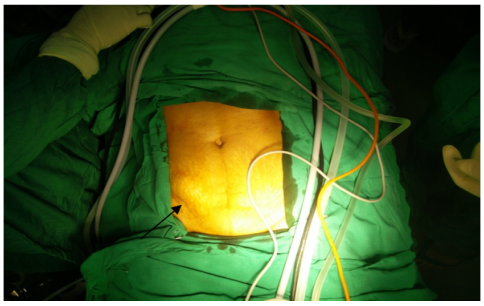
KEYWORDS

INTRODUCTION

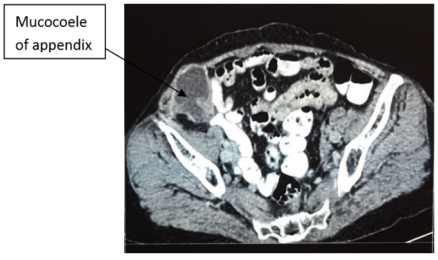
The mucocoele of the appendix was first described in 1842 by Rokitansky. It is a rare entity that can present with variety of clinical symptoms or occur as an incidental surgical finding. The incidence is 0.2%-0.4% of all appendectomy specimens. Mucocoele of the appendix denotes an obstructive dilatation of the appendiceal lumen due to abnormal accumulation of mucus resulting in a cystic dilatation of the lumen. There are 4 histologic types of appendiceal mucocele: retention cyst, mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma.[1-4] Appendiceal mucinous neoplasms represent an exceptionally rare form of pathology.

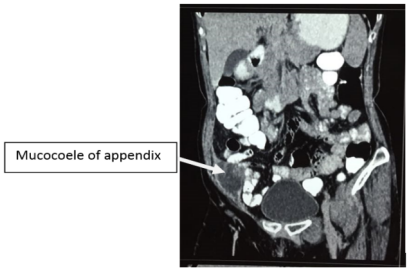
CASE REPORT

A 55years old thin built lady presented with complaint of swelling in the right iliac fossa since 2 months which progressed in size gradually, which was associated with pain in right lower quadrant of abdomen. On examination vital parameters were normal. On per abdomen examination, lump of size 5 x 6 cm present in right iliac fossa, which was hard in consistency, non mobile, non tender, intraperitoneal, with other systemic examinations being normal.

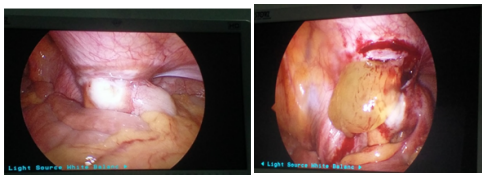


An ultrasonography of abdomen and pelvis was performed which showed 4.0 x 4.7 x 4.5 cm well defined collection in the right iliac fossa reaching upto abdominal wall anteriorly and ileocecal junction posteriorly. The appendix was not separately visualised and more likely represented as an appendicular abscess. Her CT report stated mildly peripherally enhancing cystic lesion of size 4.8x3.9x4.7 cm noted in right iliac fossa. The lesion is extending up to base of caecum. Appendix not visualised separately. Anteriorly the lesion is abutting the anterior abdominal wall in the inguinal region with mild intramuscular extension.There is mild wall thickening with heterogenous enhancement noted at its junction with base of cecum likely suggestive of mucocoele of appendix. Reports of blood investigation were unremarkable.





She underwent laproscopic appendicectomy. During the surgery, only the base was visualised, while the tip had pierced through the rectus sheath .The rectus sheath was opened and the remaining part of appendix was pulled into the abdomen, whose tip was dilated like a cystic mass with mucus (yellowish in colour) filled in it .



Appendicectomy was done and the specimen sent for frozen section which was suggestive of benign pathology. Post operative recovery was uneventful. However, Histopathological examination of the specimen revealed mucinous adenocarcinoma of appendix. Following histopathology report ,patient was started on chemotherapy regimen-mFOLFOX and will be reassessed after 3 months for the need of surgery.

DISCUSSION

Appendiceal mucinous cystadenomas are characterized by hyperplasia of glandular epithelium along with hypersecretion of

mucous resulting in a grossly dilated appendix. It is typically found in adults of 50 to 60 years old, they often presents with symptoms of acute appendicitis. Mucinous cyst-adenocarcinoma, causes a mucocele by the neoplasm occluding the narrow lumen which allows the mucin to build up and distend the appendix. Perforation may occur allowing the spill of cancerous cells out into the peritoneum, which creates the condition of pseudomyxoma peritonei. [6] These cells then seed the organs of the peritoneum and continue to produce the mucin. As the mucin accumulates, the abdomen becomes distended which is referred to as "jelly belly" .[6]

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.[10] Preoperative detection of appendiceal adenocarcinoma is rarely feasible. The imaging diagnosis of mucinous neoplasms hinges primarily on detection of the resulting mucocele. Abdominal radiography may suggest a soft-tissue mass in the right lower quadrant, but specificity is increased when calcification is identified. Curvilinear mural calcification is highly suggestive of the diagnosis.[8]

Mucinous adenocarcinoma of appendix tumors are usually well differentiated and do not undergo metastatic spread until the late stages of the disease. [9]After an initial urgent operation if the histological diagnosis reveals positive lymph nodes, adenocarcinoma of the intestine, mucinous adenocarcinoma, carcinoid or adenocarcinoid tumors larger than 2.0 cm, or high mitotic rate, a right hemicolectomy should be performed. The treatment of choice is often surgical resection combined with adjuvant chemotherapy.In case of an appendiceal specimen with perforation and adenomucinosis, follow-up with CT scans every six months for five years and also CEA and CA19-9 tumor marker surveillance is recommended.[7]

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