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An Unusual Presentation of Mucocele of the Appendix: Case Report and Review of Literature

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Abstract: ***Introduction: Appendiceal Mucocele is a rare disease. Sometimes, it is discovered accidentally and sometimes mimic acute appendicitis. Correct diagnosis before surgery is very crucial for the selection of adequate surgical treatment to avoid intra-operative and postoperative complication. Ultrasonography and particularly, Computed Tomography should be used extensively for this purpose. If mucocele of the appendix is treated incorrectly Pseudomyxoma peritonei which is characterized by malignant process may develop. Presentation Of Case: A 53year old woman came to surgical opd with clinical features suggestive of Bilateral renal calculi, outside ultrasound abdomen and CT report showed - bilateral renal calculi and complex cyst in right iliac fossa, repeated CT abdomen showed- Features suggestive of Mucocele of appendix. Based on report, surgery right hemicolectomy was done with help of intraoperative squash cytology. Histopathologic diagnosis-low grade appendiceal mucinous neoplasm was reported. After 3months of surgery patient is doing well with no postoperative complications.***

Keywords: Mucocele, Appendix, Pseudomyxoma peritonei, renal calculi, hemicolectomy, squash cytology, low grade appendiceal mucinous neoplasm.

1. Introduction

Appendiceal mucocele is an obstructive dilatation of the appendiceal lumen caused by intra luminal accumulation of mucoid material. It is a rare disease. The incidence is 0.2% to 0.7% of all appendectomy specimens [1-3]. This transformation is caused by one of four pat-terns of epithelial proliferation: Retentioncyst, mucosal hyperplasia, mucinous cystadenoma and mucinous cystadenocarcinoma [4, 5].

This disease does not have a typical clinical picture. Sometimes the patient has pain in the lower right quadrant of the abdomen; therefore, a surgeon may mistake it for acute appendicitis. Sometimes it is discovered incidentally found while evaluating a patient for another condition as in this case. Acute appendicitis is one of the most common surgical diseases [3, 6, 7].

It is important to differentiate between these two pathologies before surgery and select adequate surgical tactics. If treated improperly, the mucocele may progress, epithelial cells may escape into the peritoneal cavity and Pseudomyxoma peritonei may develop which has a high mortality [7]. We present the case which was discovered intraoperative.

2. Case Report

A 53-year-old woman presented to the Surgical Outpatient Department with complaints of left loin pain. The pain has been on and off for the last 4years. Sudden in onset, colicky in nature, radiating to left groin associated with nausea and vomiting during episodes of pain. No pain in the right lower abdomen. No anorexia. Examination of the abdomen showed mild tenderness at left renal angle. For which she evaluated and diagnosed as bilateral renal calculi outside.

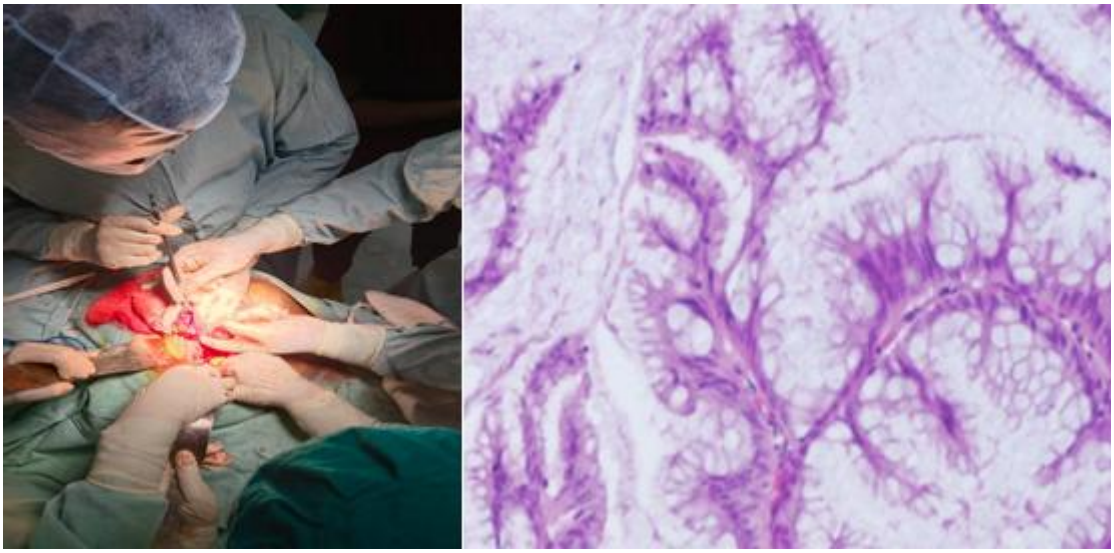

Figure 1 and Figure 2: Intraoperative finding of appendiceal mucocele with cytology

Then CT-KUB done it showed as bilateral multiple renal calculi, well defined soft tissue density lesion in RIF? complex cyst.

Then repeat CECT abdomen and pelvis done in our institute which showed-69x55x50mm [CCxAPxT] cystic lesion with wall calcifications in close proximity to caecum. Appendix not visualized. Features suggestive of mucocele of appendix. no e/o mural no dularity/ wall irregularity. Then patient planned for surgery after pre anesthetic checkup. Laparotomy was performed with vertical midline incision.6x4cm size of cyst [mucocele of appendix] with thick mucus as a content with surrounding adhesions. Base of appendix with inflammatory changes. Appendiceal specimen sent for squash cytology and reported as atypical cells. Extended right hemicolectomy with ileotransverse an astomosis was done. Figure 1 and Figure 2. Histopathologic diagnosis was low grade appendiceal mucinous neoplasm was reported. There was no complication in the post operative period. Three months after Surgery the patient is feeling well.

3. Discussion

Mucocele of the appendix was first described by Rokitansky [8]. It is a descriptive and unspecific term to define the cystic dilation of the appendix caused by the accumulation of a large amount of mucus secretion. This process is slow and gradual, with no signs of infection inside the organ. It results from lumen obstruction in the appendix, which is secondary to the inflammatory or neoplastic proliferation of the appendix mucosa or lesion in the caecum, adjacent to the appendiceal ostium. While some article, confirm its prevalence among women [9, 10] other demonstrate a higher incidence among men [11].

The appendix is lined by epithelium containing more goblet cells than the colon. As a result, most appendiceal epithelial tumors are mucinous and start as mucoceles.

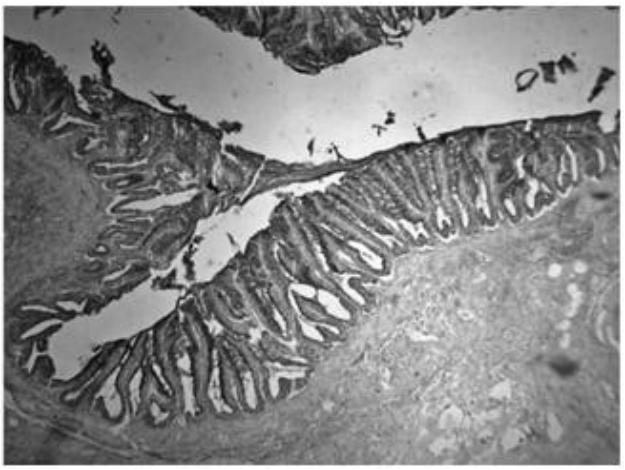

Figure 3

Mucocele of the appendix is classified into four pathological subgroups based on the epithelial characteristic [12, 13].

The first group consists of a simple retention cyst secondary to occlusion of the appendix by faceolith, scar tissue from previous inflammation or in rare cases dueto endometriosis [14]. It has a normal or flattened epithelium, moderate luminal dilation up to 2 cm and it constitutes about 15% of all appendiceal mucocele [14]. The case presented falls under this group. However, it was much larger than expected for this pathologic type. The second group with hyperplastic epithelium and moderate luminal dilatation: This constitutes about 25% of all mucocele of the appendix [14].

The third group is benign mucinous cystadenoma: This is characterized by tubular adenomatous epithelium with varying degree of epithelial atypia. It produces large amount of mucin with prominent luminal dilatation of up to 6 cm. it is the most common form, constituting about 48% cases and with associated 20% risk of perforation [13, 14].

The fourth group encompasses the malignant mucinous cystadenocarcinoma; characterized by glandular stromal invasion and/or tumour cells in peritoneal implants i.e., Pseudomyxoma peritonei. It sometimes resembles mucinous carcinoma of the colon. It constitutes about 11-20% of all cases with 6% risk of spontaneous rupture [13, 14]. Cystadenoma and cystadenocarcinoma are neoplastic appendiceal mucocele, constituting about 35% of all primary neoplasm of the appendix [11, 14].

The clinical presentation of the disease does not have a specific picture. It often flows asymptomatically as in this case. In about 50% of cases, it is discovered accidentally during radiologic and endoscopic examination or at surgery. However, a patient's clinical symptoms may include pain, palpable abdominal mass, nausea, vomiting, weight loss, gastrointestinal bleeding, and signs of intussusceptions of the intestine. [9, 13, 14].

Pre-operative diagnosis of appendiceal mucocele is very important for the selection of an adequate surgical method to prevent peritoneal dissemination to prevent intraoperative and postoperative complication and repeated surgery [13].

Sonographic examination, Computerized tomography and colonoscopy is used for diagnostics. USG is considered the first line diagnostic modality. An appendicular diameter of 15 mm or more has been determined the threshold for diagnosis of mucocele with a sensitivity of 83% and a specificity of 92%. (CT) scan is important to confirm the diagnosis and to evaluate the extent of the disease.

Fine needle aspiration cytology (FNAC) is not usually recommended as it increases the risk of perforation and dissemination in to peritoneal cavity [16]. Colonoscopy usually reveals an elevation of the appendicular orifice. In addition, a yellow mucous discharge would be visible as well. Colonoscopy is also important for the diagnosis of synchronous or meta chronous cancers when present.

In our patient outside USG and CT did not provide the correct information; so, repeated CECT abdomen and pelvis which confirmed diagnosis. As in our case presented, the patient had right hemicolectomy, done after pre-operative workup and intraoperatively squash cytology showed atypical cells - [mucus cells]. Cytology helps to guide the surgery as in our case. Right hemicolectomy is recommended when malignant mucocele is suspected by the

presence of a perforated mucocele, enlarged mesenteric lymphnodeora positive cytology. An accurate exploration of the abdomen is advised due to the well-known association between the appendiceal mucocele and other mucin-secreting cells such as colon and ovarian cancers [21].

4. Conclusion

Mucocele of the appendix is a rare disease with vague symptoms. Abdominal ultrasonographic scan important diagnostic tool, but histopathology is needed for definitivediagnosis.
Surgeryforbenignappendicealmucocelehasanexcellent long-term prognosis.

Consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her consent for images and other clinical in-formation to be reported in the journal. The patient understand that her name will not be published and due efforts will be mode to conceal her identity.

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Conflict of Interest
There are no conflicts of interest.

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