Ruptured Giant Low-grade Mucocele of the Appendix and its Management: A Case Report

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Key Clinical Message :

Preoperative prompt imaging is crucial for a definitive diagnosis of appendiceal mucocele, and double-J stenting of the ipsilateral ureter in a large retroperitoneal cyst is recommended for easy identification and prevention of complications.

Keywords :

Appendix; appendiceal neoplasms; mucocele; pseudomyxoma peritonei; neoplasms; laparotomy

Introduction :

Mucocele of the appendix is a well-described clinical condition of the appendix. It is a rare condition with an incidence of about 0.2-0.7 % of all appendectomies (1). The pathophysiology behind the appendiceal mucocele is an obstructive dilatation of the appendicular lumen by excess intra-luminal mucous secretions. Due to the presence of very vague presenting symptoms or even asymptomatic presentation, it is often diagnosed at an advanced clinical stage, or even misdiagnosed as acute appendicitis. It requires prompt imaging with preoperative planning and adequate surgical treatment, to avoid the rupture and spillage of the cystic content and to prevent the development of pseudomyxoma peritonei.

Case Presentation :

A 60-year-old hypertensive male came to the general surgery outpatient department with complaints of pain and distension in the abdomen for three years which was insidious in onset and progressive in nature. On examination, the abdomen was soft, non-tender, and distended with a vague lump felt in the right half of the abdomen (Fig. 1). There were no complaints of fever, weight loss, appetite loss, constipation, or vomiting. Laboratory investigations showed leucocytosis (13.20 x 10^3 /ul) with neutrophilia (9.09 x 10^3 /ul), and carcinoembryonic antigen (CEA) levels were elevated, 117.8 ng/ml.

Abdominal ultrasonography showed a large intra-abdominal lobulated hypoechoic, likely cystic mass with internal echogenic areas on the right side. Abdominal contrast-enhanced computed tomography showed a large well defined multilobulated lesion of 17x13x24 cm in the retroperitoneum on the right side along with areas of whorled calcification in the superior aspect of the lesion. This gigantic cyst was pushing the retroperitoneal organs towards the left side (Fig. 2).

Because of the large size of the mass, a decision to perform an exploratory laparotomy was made. A full-length midline incision was given and the mass was excised in toto with accidental spillage of content along

with the part of the cecum (Fig. 3, 4). Intraoperatively, a large retroperitoneal cystic mass of 15.5 x 12 x 20 centimetres was found in connection with the body of the appendix. The cyst was extending superiorly to the dome of the diaphragm, medially to the lateral border of the inferior vena cava and inferiorly to the pelvis. Histopathology samples of the mass and cell cytology samples of the spillage liquid were sent. Microscopic examination of the cyst wall showed flattened to undulating columnar lining epithelium which was getting morphed into papillary infoldings, exhibiting mild nuclear pleomorphism, round to oval coarsely chromatic nuclei, inconspicuous nucleoli and abundant mucinous cytoplasm. The underlying lamina propria showed moderate chronic inflammatory infiltrate and desmoplasia. Histopathological diagnosis of "Low grade appendiceal mucinous neoplasm, pT4a" was made. The appendix base and part of the cecum were not involved in the tumour. The patient was discharged on a postoperative day-7 and has been followed up in the outpatient department with no reported complications for 6 months.

Discussion :

Mucocele was initially described by Rokitansky in 1842 (2). Most often, benign or malignant epithelial proliferation results in an obstructive dilatation of the appendix. Less frequently, inflammation or obstruction by a fecalith is the cause of mucocele formation (3). Some studies state a higher prevalence in females (1), while others confirm a higher prevalence in males (4). At our tertiary care referral centre, we have found that males above 50 years of age are more commonly affected by appendiceal mucocele.

The clinical presentation of appendiceal mucocele is vague or sometimes asymptomatic. Due to this, patients often present late to the hospital. Usually delayed presentations of appendiceal mucocele are large abdominal lump, abdominal distension, acute appendicitis or acute abdomen or pseudomyxoma peritonei. Without proper in-time treatment, the whole abdomen may get involved due to perforation of the mucocele and dissemination of the mucin-producing epithelium in the peritoneal cavity, which may result in the development of pseudomyxoma peritonei. In our scenario, the patient presented to us with a large abdominal lump and abdominal distension. Ultrasound showed the classical onion skin sign suggestive of appendiceal mucocele (5). A contrast-enhanced computed tomography was performed for further characterization of the cystic mass and surgical planning. A correct diagnosis before surgery is of paramount importance for both planning the surgery and decreasing the risk of severe intraoperative and postoperative complications. The imaging techniques that can aid in the diagnosis of mucocele of the appendix are ultrasonography, computed tomography and colonoscopy.

On computed tomography, a mucocele will typically manifest as a homogenous hypoattenuating material that has Hounsfield values similar to water filling the lumen of the appendix (3). A mucocele is highly likely if there is curvilinear calcification in the appendix's wall (6). An elevated appendiceal mucocele can be seen during a colonoscopy and is called as "volcano sign" (7). A yellowish mucus discharge may be visible from this orifice (8). Histopathology is usually the standard investigation for establishing a definitive diagnosis of mucocele. Because of the risk of seeding neoplastic cells along the needle track or in the peritoneal cavity, which could produce pseudomyxoma peritonei, fine needle aspiration of an appendiceal mucocele is avoided (9).

The treatment modality of choice for appendiceal mucocele is surgery. The basic guiding principle behind the surgery for an appendiceal mucocele is to retrieve the cyst intact without spillage, as a perforated appendiceal mucocele carries a higher risk of development of pseudomyxoma peritonei. The surgical method preferred for operating on the mucocele of the appendix is open surgery because of the higher risk of rupture of the mucocele laparoscopically (10). In open surgery, it is possible to do a complete in-situ manual examination of the intestines including palpation of the inflamed structure and also direct inspection of the most probable spots of mucinous tumours in the abdomen. Laparoscopic exploration does help in the confirmation of diagnosis. But now, because of the availability of high-resolution computed tomography with good diagnostic accuracy, a laparoscopic examination is seldom required.

Our case showcases the importance of large size and location in the retroperitoneum of the appendiceal mucocele. Preoperative double-J stenting of the right ureter should be done in the retroperitoneal appendiceal

mucocele for easy identification of the right ureter. Meticulous intraoperative handling and dissection are required to prevent the rupture of the cyst and its complications. An appendectomy, ileocecectomy or right hemicolectomy should be the only procedure performed during the initial surgery for a ruptured appendiceal mucinous lesion at non-specialized centres. Additionally, careful inspection of the abdominal cavity with documentation and biopsy of any suspicious peritoneal lesions should be done. To minimize tumour cell implantation, the surgical and abdomen wounds should be cleaned thoroughly by irrigation (11).

Depending on the final detected pathology, a patient with gross peritoneal dissemination of mucin should thereafter be transferred to a specialized centre. A consensus classification developed by the Peritoneal Surface Oncology Group International has been instrumental in clearing up the ambiguity around diagnostic terminology (12).

Our case was reported as "T4a low-grade mucinous neoplasm", which should be managed with follow-up contrast-enhanced computed tomography or diagnostic laparoscopy at 6 to 12 months to look for recurrence or development of the tumour. Cytoreductive surgery/hyperthermic intraperitoneal chemotherapy can then be offered if evidence of disease is detected.

Conclusion :

Appendiceal mucocele, albeit a rare disease, should always be kept as a differential diagnosis when the presenting symptoms are like acute appendicitis. Clinicians should be aware that the underlying pathology behind a mucocele of the appendix can range from a mere retention cyst to malignant tumours. To correctly diagnose a mucocele pre-operatively, contrast-enhanced computed tomography abdomen should be used extensively. The importance of contrast-enhanced computed tomography and other imaging modalities increases in large-size cysts. For larger cysts located in the retroperitoneum, preoperative double-J stenting of the right ureter is recommended for easy identification. Meticulous dissection to prevent spillage and to avoid the development of pseudomyxoma peritonei is a must.

Author Contributions :

Hritvik Jain : Conceptualization; writing – original draft; writing – review & editing. **Dheerain Gupta** : Conceptualization; writing – original draft, writing – review & editing. **Jyoti Jain** : Writing – original draft, writing – review & editing. **Mahendra Lodha** : Methodology; project administration; supervision; writing – review & editing. **Binit Sureka** : Writing – original draft. **Prakriti Pokhrel** : Writing – original draft.

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Figure Legends :

Figure 1: Preoperative image of the patient's abdomen showing fullness in left hypochondrium and left lumbar region.

Figure 2: CECT abdomen figure showing the axial (left side) and coronal (right side) view. The white dashed arrow is showing the displaced colon and the black dashed arrow is pointing towards the displaced right kidney hence confirming it to be predominantly retroperitoneal in location.

Figure 3: Intraoperative image showing an in-situ cystic mass occupying the entire retroperitoneum.

Figure 4: Postoperative image showing an excised specimen with mucin.