"Appendicular mucocele" – Case report and review of literature

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IN BRIEF

Introduction: Mucocele of appendix is a rare condition which majority of time is diagnosed incidentally or during working up for a pelvic cause. Clinical presentation is non specific, Majority of cases presents to gynaecologists with suspicion of adnexal pathology. Accurate preoperative diagnosis is difficult to arrive at, given the findings of USG and CT may be confusing. As it can be associated with malignancy, operative handling of specimen should be optimum. Case Presentation: Two cases of Am were diagnose preoperatively by USG and CECT . Patient underwent surgery with exicision of the appendix. Conclusion: A female patient presenting with complaints of lower abdominal pain with no findings in pelvis should be considered for differential diagnosis of Appendiceal mucocele.

Introduction

Appendiceal mucocele is a rare clinical entity presenting variably, With an incidence of 0.2%-0.4%, occurs due to occlusion of the lumen with/or without increased mucus glands in the wall. majority of cases are asymptomatic (upto 50%) and those with symptoms are nonspecific. Mass may be felt on abdominal or pelvic examination. AM can also present as an incidental finding on imaging studies or surgery for other reason.

Ultrasound of abdomen is the primary imaging modality, though non specific can be a useful tool to provide a suspicion of AM. CT abdomen has high Sensitivity to detect AM compared to sonography with addition of MRI, diagnostic accuracy increases. Barium enema and Colonoscopy are also a part of diagnostic kit with acceptable sensitivity. No specific laboratory indices yet found which relate to AM.

Surgery is mandatory with various procedure like simple appendicectomy, right hemicolectomy , excision of the appendix etc. Extended resection should be considered if malignancy cannot be ruled out. Accidental spillage of content of mucocele harbouring malignancy leads to pseudomyxoma peritoneii.

Case Presentation:

1. A sixty year old lady presented to surgery OPD with complaints of "left iliac fossa" pain of 6 months duration, pain was colicky in character with no bowel or bladder disturbances. She had been referred from gynaecology department with suspicion of pelvic pathology, whom she presented

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primarily to. Clinical examination was essentially normal with normal pelvic examination. Routine laboratory investigations were inconclusive and CA 125 and CEA were under normal limit.

Sonography revealed the presence of mass in the right side of pelvis with mixed echogenicity extending from abdomen suggesting right adnexal pahtology. Patient underwent computerised tomography which showed presence of blind ending distended mass in the right lower abdomen with peripheral enhancement measuring 6 x 5 x 4 cms in the region of caecum. She underwent laprotomy and appendicectomy through local approach. Surgical exploration revealed a mass in appendix withish grey in colour. Pathological examination of the specimen revealed mucinous cystadenoma with no evidence of malignancy. Postoperative course was uneventful and patient was discharged on 3rd postoperative day and she is under regular follow up.

2. A forty five year old non-smoker male presented with lump in RIF for 10 years and pain for eight months. Patient gave history of taking ATT for 6-8 months USG revealed AM with mild ascitis. Other history was not significant. On examination a mobile lump 5 x 6 cms was present in Rif with right bubonocele. CECT showed periappendicular fluid with? metastasis in liver with? hemangioma with bilateral multiple renal cortical cyst with pericardial effusion. Patient underwent laparotomy and appendectomy through local approach. Postoperative course was uneventful and patient was discharged on 3rd postoperative day and he is under regular follow up.

DISCUSSION

Appendicular mucocele is the term used to describe a pathological entity of appendix which is distended by mucus either due to increased mucus secretion or obstruction of lumen or both. Incidence of appendicular mucocele is estimated to be 0.29% to 0.4%, with no data available for gender preponderance. Etiology of the mucocele is either due to Mucinous cystadenoma which is the major cause (63%) followed by Mucosal hyperplasia (25%), Mucinous adenocarcinoma (11%) and retention cyst. Thus mucocele of appendix can be either due to benign or malignant cause. Other causes like endometriosis of appendix, caprolith obstruction, carcinoid tumor of appendix are rare. Based on architectural and cytologic features appendicular mucocele has been classified as 1) Low grade appendiceal mucinous neoplasm, 2) Mucinous adenocarcinoma and 3) Discordant variety. Such classification is of prognostic value. Mucoceles with a diameter of < 2cms are often benign.

Clinically around 23% to 50% of patients are asymptomatic in whom diagnosis is made incidentally during surgery, radiological evaluation and endoscopic procedure for other reasons. Other patients may present with Right lower abdominal pain, change in bowel habits, complains of palpable abdominal mass, lower gastrointestinal bleeding. Majority of patients also present to gynaecology department first and surgeon called in gynaecology operation theatre. Examination of the patient usually reveals right lower abdominal mass, sometimes pelvic examination is clearer but physical examination may be entirely normal as in case of our patient.

Radiological investigations are indispensible for diagnosis as majority of patients present with non specific symptoms and signs. Ultrasonography, computerised tomography, barium enema and colonoscopic examination are recognised modality of investigations. Role of MRI is yet to be established.

Sonological findings of cystic lesion in right iliac fossa with variable echogenicity and outer appendiceal diameter of >15mm is 83% sensitive and 92% specific for mucocele of appendix. Onion skin sign of appendix is also pathognomonic for appendicular mucocele.

Computerised tomography in majority of cases is the confirmatory investigation, a cystic soft tissue mass with enhancing wall nodularity, occational mural calcification

occupying the region of caecum, pushing it is suggestive of appendicular mucocele. Through use of USG and CT scan possible differential diagnoses can be ruled out.

Barium enema also adds to the diagnostic tool – partially filled mass indenting the caecum displacing it laterally is the finding on x-ray.

Colonoscopy apart from diagnosing appendicular mucocele also rules out synchronous or metachronous colonic tumors which can be present in around 29% of cases. Elevation of the orifice of appendix known as 'Volcano sign' with yellowish mucus discharge seen from the appendiceal orifice points towards mucocele.

*In the present cases ultrasound and CT scan of the abdomen confirmed the diagnosis of appendicular mucocele.

Surgical treatment is the primary modality and complete excision of mucocele of appendix is done either by laparoscopy or laparotomy. The advantages of laparoscopy being good exposure and visualisation of entire abdomen, also shorter hospital stay. In case of suspected malignancy laparotomy and proceed is a better option as risk of spillage and subsequent peritoneal carcinomatosis is present. Simple appendicectomy would suffice in case of mucocele of appendix confined to it. If suspicious of malignancy with involvement of caecum and adjacent organ right hemicolectomy has to be done with lymph node clearance.

CONCLUSION

Appendicular mucocele is the diagnosis of exclusion and high index of suspicion is the key for its diagnosis. Female presenting to gynaecology with right iliac fossa pain and with no clinical features suggesting gynaecologic pathology should be evaluated for appendicular mucocele.

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