Mucinous cystadenoma of appendix presenting as mucocele of appendix: A rare finding report

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Abstract:

Mucocele of appendix although a rare clinical finding may be the presentation of mucinous cystadenoma of appendix. We present mucocele of appendix in a patient who had recurrent pain right iliac fossa. On ultrasonoghaphy a fluid filled tubular bowel loop and on CECT scan there was a hypodense oval shaped lesion in association with caecum. On exploration there was tense appendix with normal base, simple appendicectomy was done and patient was discharged after five days and is under follow up

Key words: mucocele of appendix, mucinous cystadenoma, CECT, CEA, CA 19-9

Introduction

Mucocele of appendix is a rare clinical presentation with varying symptoms mostly in the form of pelvic mass. The incidence is 0.2%-0.4% of all appendectomies specimens [1, 3]. With detailed preoperative investigations the diagnosis sometimes remains elusive and the findings are confirmed at the time of exploration only. Mucinous cystadenoma of the appendix may present in the form of appendiceal mucocele and is a quite rare condition that is incidentally discovered without any symptom despite dilatation of the appendix [4]. There are four histological types, which lead to individualized surgical treatment and course in each case [3]. Preoperative diagnosis is essential for the best surgical approach choice of surgical approach to prevent peritoneal dissemination.

Case history

A fifty year old women was admitted with recurrent pain right iliac fossa, there was no history of altered bowel habits, loss of appetite, weight loss or urinary symptoms. On examination there was mild tenderness in the right iliac fossa with palpable smooth mass in the same site. Her biochemical investigations, total leukocyte count, CEA, CA 19-9 and CA 125 were within the normal range. Ultrasonography was done which showed a persistant fluid filled tubular non peristaltic bowel loop in the right iliac fossa measuring 75 × 35 mm in size without any para-aortic lymphadenopathy or ascites 1], while contrast enhanced computed tomography (CECT) abdomen showed a hypodense oval shaped lesion of $63 \times 37 \times 34$ mm with thick gelatinous contents of volume of 50 cc in association

with caecum with anterior extension, and the tip was adherent to the anterior abdominal wall [fig 2]. The lesion was well defined showing thin enhancing wall with attenuation value of 18 U to 24 U and without any perilesional fat straining or ascites. On exploration appendix was tense with normal appearing base [fig 3]. Histology showed lining of columnar mucus epithelium with areas of papillary projections with mild nuclear atypia and focal areas of pseudo stratification. Wall showed fibrous tissue, thinned muscle wall without invasion of wall or evidence of malignancy [fig 4].

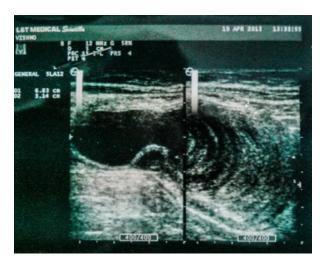


Figure 1: Ultrasonography abdomen showing persistant fluid filled tubular non peristaltic bowel loop





Figure 2 (a), (b) CECT abdomen showing a hypodense oval shaped lesion on axial and sagital sections



Figure 3: Showing mucocele of appendix

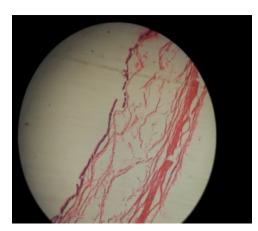


Figure 4: showing showed lining of columnar mucus epithelium with mild nuclear atypia and focal areas of pseudo stratification

Discussion

Appendiceal mucocele (AM) was first described by Rokitansky (1842) and was named by Feren (1876). Mucinous cystadenoma of appendix is very uncommon tumor presenting in the form of mucocele of appendix with an incidence of 0.2%-0.4% of all appendectomies specimens [1, 3]. The mucocele may be secondary to mucinous cystadenoma (63%), mucosal hyperplasia (25%), mucinous cystadenocarcinoma (11%) and retention cyst ^[5]. The clinical presentation ranges from right lower quadrant pain, change in bowel habits, per rectal bleeding to a palpable mass [6]. Approximately 23–50% of patients are asymptomatic, with the diagnosis made incidentally during surgery, radiological evaluations or 7] endoscopic procedures Preoperative investigations like CEA, CA 19-9 should be done so that a comprehensive surgical plan is made. Ultrasound is usually the first-line diagnostic modality and different sonographic findings of AM and acute appendicitis (AA) have been described [3, 8]. Appendix diameter 15 mm or more in USG examination has been determined as the threshold for AM diagnosis with a sensitivity of 83% and a specificity of 92% [3]. CT can also be done especially in elderly group patients to know the exact status of lesion and to rule out any other abdominal pathology [8]. In our case preoperative diagnosis was made with the help of contrast enhanced computerizes tomography but the exact anatomy of base of appendix was not confirmed and we went through right paramedian incision. Colonoscopy in such patients with abdominal pain is a useful test to rule out any intraluminal pathology and findings like 'volcano sign' [2, 9].

They are comprised of the following 3 distinctive clinicopathologic entities: focal or diffuse mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma. 5) Of the 46 cases with mucinous cystadenoma, 9 were associated with adenocarcinoma of the colon, and 4 with ovarian mucinous cystoma. Among the 18 cases with focal or diffuse mucosal hyperplasia, 5 were associated with adenocarcinoma of the colon, and 1 with ovarian mucinous cystoma [5]. Surgical resection is advised by laparoscopic approach as it has many advantages over open surgery but in such cases careful handling of appendix is advised to prevent the complication like pseudomyxoma peritonei. Cecal resection is advised for a cystadenoma with a large base, involvement of the caecum or adjacent organs is an indication for right hemi-colectomy and thorough exploration of the gastrointestinal tract and ovaries [10]

Conclusion

Although a rare disease, can have a varied presentation, once diagnosis of mucocele is made surgical treatment is mandatory to exclude malignant transformation and prevention fatal complications like pseudomyxoma peritonei

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