IF : 4.547 | IC Value 80.26 VOLUME-6, ISSUE-5, MAY-2017 • ISSN No 2277 - 8160

Original Research Paper Pathology

PORCELAIN APPENDIX – A RARE BENINGN ENTITY

MANIKA KHARE	ASSISTANT PROFESSOR, JNUIMSRC JAIPUR
MANISH KUMAR GUPTA	ASSISTANT PROFESSOR, JNUIMSRC JAIPUR
K M GARG	PROFESSOR, JNUIMSRC JAIPUR
U B SHARMA	PROFESSOR, JNUIMSRC JAIPUR

ABSTRACT Appendiceal mucocele was first described by Rokitanski in 1842.[1] Mucocele of appendix is the rare entity. Secondary changes in mucinous cyst adenoma include thinning of the wall, extensive ulceration, calcification and ossification ("porcelain appendix")

KEYWORDS :

INTRODUCTION -

Appendiceal mucocele was first described by Rokitanski in 1842.[1] Mucocele of appendix is the rare entity. When it is associated with the marked calcification it is known as porcelain appendix.

CASE REPORT –

A 60 years female presented with the recurrent pain in the right iliac fossa since 6 months. It was associated with malaise and fever. The vitals of the patient were within normal limits. USG abdomen of the patient was done with showed multiple stones in the gall bladder along with thickened and dilated appendix (15mm), which was non-compressible. The impression of acute appendicitis with cholelithiasis was given on USG. The surgery was planned for appendicectomy. On per op examination, appendix was thickened, dilated and inflamed. Appendicectomy was performed and the specimen was send to the histopathology lab for examination.

On gross examination the appendix was enlarged and dilated. External surface was congested. On cut section lumen showed many mucus plugs within the lumen. On microscopic examination, the wall of the appendix was thickened and showed large areas of calcification. Whole of the lining mucosa was denuded. Mild chronic inflammatory infiltrate was also noted. Based on above histological findings the diagnosis of calcified mucocele was given.[fig1,2]

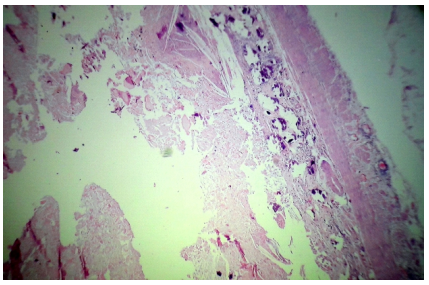

Fig 1. Photomicrograph showing scanner view of the wall of the appendix with areas of calcification. Lumen is filled with mucin

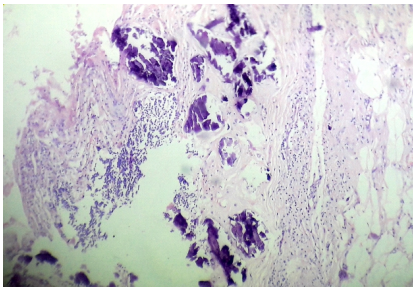

Fig 2 photomicrograph showing high power view with complete denudation of the mucosa and large areas of calcification in the wall of the appendix.

DISCUSSION –

Appendiceal mucocele is a rare entity. The reported prevalence in appendectomy specimens at surgery is 0.2-0.3%.[2] It is a descriptive term for the accumulation of the mucous in the appendiceal lumen, which causes distention of the appendix, irrespective of the underlying cause.[3] Previously, it was thought that most mucoceles occurred as a result of postinflammatory obstruction or due to obstructive faecaelith with distal accumulation of mucus.

Appendicial mucocele is usually a incidental finding in 50% of the cases.[4,5] Aho et al also reported in his study that almost a quarter of cases were asymptomatic and discovered incidentally.[6] The symptoms associated with the mucocele includes mild abdominal distress and pain, chronic intermittent pain(as was seen in this case). The complications includes intussusception, right iliac fossa mass and urinary symptoms.[7,8]

Histologically mucocele are divided into four types.[9,10]

Simple/non neoplastic mucocele or retention mucocele is a cystic dilatation of the distal appendix with accumulation of abnormal mucoid material. Secondary changes in mucinous cyst adenoma include thinning of the wall, extensive ulceration, calcification and ossification ("porcelain appendix")[11]

Benign neoplastic mucocele-mucinous cystadenoma[12] is a dilated mucus /mucin filled appendix containing adenomatous mucosa lined by atypical mucinous epithelium containing basal nuclei and showing only minimal dysplastic features.

Malignant mucocele[11]-mucinous cystadenocarcinoma is defined as adenocarcinoma associated with mucus-filled cystic dilatation of the appendix presenting as mucocele. A malignancy is suspected at surgery in about 30%.[9,11] In the others the diagnosis is made during pathologic examination.

CT scan demonstrates a low-attenuated, well encapsulated mass in the right lower quadrant, with a smooth regular wall. The presence of curvilinear or punctuate wall calcification in the right lower quadrant strongly suggests the diagnosis of appendiceal mucocele. Calcifications are thought to be a dystrophic response to a chronic inflammatory process incited by mucus in the appendiceal wall (porcelain appendix).[2] The presence of wall calcification in a right lower quadrant cystic mass with intraluminal gas allows one to

differentiate an infected mucocele from an abdominal abscess cavity.

The clinical and radiological differential diagnosis of mucocele of appendix includes mesenteric cyst, colonic duplication cyst, intussusception, right ovarian cyst and hydrosalphinx.[12,13] Enteric duplication is rare in adults and calcification is rare in mesenteric cysts. A right ovarian cyst or hydrosalpinx should also be considered in the differential diagnosis in women. In a patient who has not had an appendectomy, CT findings of a cystic mass containing mural calcification in the anatomic location of the appendix and adherent to the adjacent caecum are characteristic of an appendiceal mucocele.[14]

We present a rare case of porcelain appendix which showed marked calcificationwith complete denudation of the mocosa.

REFERENCES –

1. Gonzalez JE, Hann SE, Trujillo YP. Myxoglobulosis of the appendix. Am J Surg Pathol 1988; 12: 962-96.
2. Dachman A, Lichtenstein J, Friedman A. Mucocele of the appendix and pseudomyxoma peritonei. AJR Am J Roentgenol 1985;144:923-929.
3. Lo RH, Kan PS. Appendiceal mucinous cystadenoma presenting as "porcelain" appendix with myxoglobulosis—a rare cause of a right lower quadrant mass. Singapore Med J 1998;39: 174-176.
4. Wolff M, Ahmed N. Epithelial neoplasms of the vermiform appendix. Cancer 1976; 37:2511-2521.
5. Isaacs KL, Warshauer DM. Mucocele of the appendix: Computed tomographic, endoscopic and pathologic correlation. Am J Gastroenterol 1992;87:787-789.
6. Aho AJ, Heinonen R, Lauren P. Benign and malignant mucocele of the appendix. Histological types and prognosis. Acta Chir Scand 1973;139:392-400.
7. Dachman AH, Lichtenstein JE, Friedman AC. Mucocele of the appendix and pseudomyxoma peritonei. AJR Am J Roentgenol. 1985;144:923–929.
8. Huang HC, Liu TP, Jeng KS. Intussusception of mucocele of the appendix: a case report. Zhonghua Yi Xue Za Zhi (Taipei) 1994;53:120–123.
9. Appelman HD. Epithelial neoplasia of the appendix. In: Norris HT, ed. Pathology of the colon, small intestine, and anus 2nd ed. New York: Churchill Livingstone pub, 1991: 263-303
10. Stocchi L, Wolff BG, Larson DR, Harrington JR. Surgical treatment of appendiceal mucocele. Arch Surg 2003; 138: 585-59; discussion 585-59
11. Parakrama Chandrasoma. Gastrointestinal Pathology. 1st ed. Appleton& Lange pub, 1999: 239-241
12. Huang HC, Liu TP, Jeng KS. Intussusception of mucocele of the appendix: a case report. Zhonghua Yi Xue Za Zhi (Taipei) 1994; 53: 120-123
13. Viswanath YK, Griffiths CD, Shipsey D, Oriolowo A, Johnson SJ. Myxoglobulosis, a rare variant of appendiceal mucocele, occurring secondary to an occlusive membrane. J R Coll Surg Edinb. 1998;43:204–206.
14. Zissin R, Gayer G, Kots E, Apter S, Peri M, Shapiro-Feinberg M. Imaging of mucocele of the appendix with emphasis on the CT findings: a report of 10 cases. Clin Radiol 1999;54: 826-832