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Case Report

Appendiceal Mucocele: A Masquerade

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ABSTRACT

Appendiceal mucocele is a rare disease entity that has been infrequently diagnosed before surgery because of its lack of specific diagnostic features. Most often it is asymptomatic but sometimes mimics acute appendicitis like symptoms , adnexal mass especially in females. We herein, report and discuss such a case in a 58 years old female patient to highlight this appendiceal pathology.

Keywords: Appendix, Mucocele, Appendicitis.

INTRODUCTION

Mucocele of the appendix is a descriptive term for an abnormal mucous accumulation distending the appendiceal lumen, regardless of the underlying cause. This process is slow and gradual, with no signs of infection inside the organ. It was first described by Rokitansky in 1842. [1] They are found in 0.2%-0.3% of appendicectomies.[2,3,4]

CASE REPORT

A 58 years old female patient presented with history of right sided abdominal pain, dull and intermittent in nature since two months. Her past history is insignificant.

On physical examination, her abdomen was soft. A tender palpable mass measuring approximately 12x8cm was noted in right iliac fossa without rigidity, muscle guarding and rebound tenderness. No signs of peritoneal irritation were elicited. No associated vomiting or fever was observed. There were no associated urinary or bowel symptoms.

The laboratory findings include total leucocyte count of 11,000 cells/mm. [3] CRP was within normal limits. Serum CA-125, CEA were within normal limits.

Ultra sonography revealed mixed echogenic mass measuring 14x9cm in right lower abdomen with an echogenic rim.

CT scan demonstrated a well demarcated cystic mass measuring 14x9cm in right lower quadrant of abdomen.

Surgical exploration revealed grossly dilated appendix, excision of portion of terminal ileum and ascending colon with

appendix was performed and the same was subjected for histopathological evaluation.

On gross examination, specimen consisted of portion of terminal ileum and ascending colon with appendix measuring altogether 14x9x4cm. The appendix was markedly dilated, already opened with smooth inner surface showing large amounts of mucin. The appendiceal wall was markedly thickened and parchment like. The terminal ileum and ascending colon were unremarkable.

Histology of H&E stained sections of appendix revealed hypertrophic appendiceal wall. Mucinous material noted in mucosa. No lining epithelium was identified. Muscularis propria was thickened. No evidence of any malignancy. Terminal ileum and ascending colon were unremarkable morphologically. With the above features, we could arrive at the diagnosis of Retention cyst ('Mucocele') of appendix.

Figure 1: Mucocele: cut opened gross specimen showing dilated appendix with mucin on the inner surface.

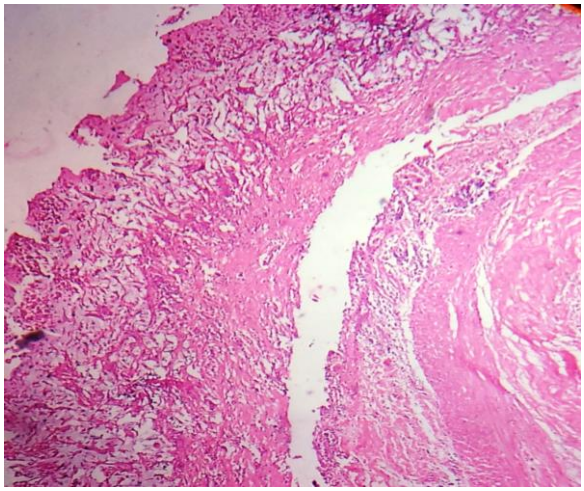

Figure 2: Mucocele: Photomicrograph showing mucin coated thickened appendiceal wall. No evidence of lining epithelium. H&E stain, 10X.

DISCUSSION

In 1940, Woodruff and Mc Donald classified appendiceal mucoceles into a benign type, representing a mucocele caused by obstruction of the appendiceal lumen, and a malignant type, representing a mucin secreting adenocarcinoma. Higa et al preferred to consider all mucoceles as mucinous neoplasms within a clinicopathologic spectrum comprising mucinous hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma. The term mucocele has been retained by popular usage; those without an apparent pathogenesis are called primary, and those with an evident pathogenesis are termed secondary.[1]

Of all mucoceles, 23% - 50% are incidental findings at surgery.[1,4]

Mucoceles of appendix has been described to occur more often in females with a female predominance of 4:1. [5] The average age at time of diagnosis is 54 years for benign mucoceles and 64 years for malignant disease.[3]

Various etiologies have been reported. [5] Appendiceal mucocele may come as a consequence of obstructive or inflammatory processes. Causes for obstruction of the appendiceal lumen may include inflammatory stricture, carcinoid, carcinoma, villous adenoma, appendicolith, mucosal web, endometriosis, and extrinsic compression.[1] Besides these cause, other

tumour lesions in the appendix or caecum may present as mucocele. There has been one report of a mucocele secondary to diverticulitis .[5]

An association between appendiceal mucocele and synchronous colon neoplasms has been previously noted. The most common synchronous neoplasms occur in the large bowel (19.5%-21%).[3]

The external appearance is gross enlargement of the appendix, the lumen is distended by mucin .[6]

The neoplastic diagnosis is only determined by pathology.[5]

Histopathologic classification of appendiceal mucoceles is dependent on the characteristics of their lining epithelium, these include retention cysts (18%), mucoceles with mucosal hyperplasia (20%), mucinous cystadenomas (32%), and mucinous cystadenocarcinomas (10%). Classification is important, because the course of the disease and prognosis are related to these subtypes.[3]

Simple mucoceles (retention cysts) are characterized by degenerative epithelial changes and may result from appendiceal obstruction and distension. There is no evidence of hyperplasia or neoplasia of the mucosa.[3]

Hyperplastic mucoceles are sessile or pedunculated lesions that represent hyperplastic polyps of the colon and are not known to have any malignant potential.[3]

Mucinous cystadenomas also have been referred to as low-grade appendiceal mucinous neoplasms. They typically are circumferential cystic lesions composed of mucin rich epithelium.They can be considered as the equivalent of adenomatous colon polyps.[3]

The mucinous cystadenocarcinoma presents with high grade cellular dysplasia and stromal invasion.[3]

Myxoglobulosis is a variant of mucocele seen in 0.35%-8% of cases, in which appendix is filled with ‘many solid translucent globules’. [1]

The preoperative clinical diagnosis of appendiceal mucoceles can be difficult because of lack of clinical symptomatology.[4]

Occasionaly, it may present with palpable abdominal mass, lower right abdominal pain, gastrointestinal bleeding or non specific signs including weight loss, nausea, vomiting, changes in bowel habits, hematuria, unexplained anemia and acute appendicitis .[2,6,7]

In women presenting with right iliac fossa mass and clinical features not indicative of gynaecological pathology, an appendiceal origin should be considered in the differential diagnosis.[3]

However, most of them are found incidentally by imaging studies or during surgery.[7] Symptomatic patients were reported, more likely, to have a malignant disease . [6]

Both Benign and malignant mucoceles may spontaneously rupture, secondary presumably to hypersecretion of mucus. These patients may present with a peritoneal cavity filled with mucus, informally described as ‘jelly belly’.[5]

Cheng considered ‘jelly belly’ or pseudomyxoma peritonei a type of foreign body peritonitis where all the gelatinous material originates from the mucocele and induces a fibrotic response in the peritoneum leading to encapsulation.[1]

The ultrasound shows an encapsulated cystic lesion in the lower quadrant of the abdomen with a liquid content of variable echogenicity, according to the density of the mucus .[5]

On ultrasound, they have been known to mimic ovarian cyst torsion .[5]

On CT, the typical feature of mucocele of the appendix is a cystic mass with a thin wall and of low density, which communicates directly with the caecum.[5]

The porcelain appendix and the so called volcano sign are two non specific diagnostic clues, but CT remains the most suggestive diagnostic tool.[5]

However, careful handling of the specimen is recommended as spillage of the contents can lead to pseudomyxoma peritonei.[4] So, the mucocele should be removed intact. [7]

Prognosis is related to the histopathological subtypes.[5]

The 5 year survival rate for simple or benign neoplastic mucocele ranges from 91% to 100%. [3]

Simple and benign mucoceles have an insidious evolution and are rarely perforated; on the other hand malignant mucocele evolution is faster, like in acute appendicitis, usually presenting as an organ perforation .[1]

CONCLUSION

In conclusion, mucocele of appendix is very rare. It is very difficult to diagnose pre-operatively as it lacks specific symptomatology. A variety of abnormalities (benign and malignant) of the appendix emphasizes the need for routine pathologic examination of the appendix.

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