International Journal of Biomedical and Advance Research
ISSN: 2229-3809 (Online); 2455-0558 (Print)
Journal DOI: 10.7439/ijbar
CODEN: IJBABN

Case Report

Appendicular Mucocele: High Frequency Sonography Imaging – A case report and short review of literature

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Abstract

Appendicular mucocele, a cystic mass resulting from a dilated appendiceal lumen caused by abnormal accumulation of mucus that often is not considered when problems of the right quadrant lower abdominal pain are assessed. It is rare entity; occurring in 0.25% of 43,000 appendectomy specimens in one series. A correct preoperative diagnosis is rarely made on the basis of clinical examination. Accurate diagnosis is essential in order to prevent rupture at surgery with development of pseudomyxoma peritonei and predict malignant transformation. We describe high frequency sonography findings that led to the correct preoperative diagnosis confirmed with surgery and histopathological confirmation.

Keywords: Abdominal Pain, Appendix, Imaging, Mucocele, Ultrasonography.

1. Introduction

Appendiceal mucocele(AM) is a descriptive term applied for mucinous distension of the appendiceal lumen regardless of the underlying pathology, is a rare clinical entity; the reported prevalence in appendectomy specimens at surgery is 0.2-0.3%[1]. It may be an outcome of various processes. Most important from the surgical point of view is the mucocele caused by mucinous cystadenoma and cystadenocarcinoma. Less common causes are retention cyst, carcinoid, appendicolith, endometriosis, adhesions and volvulus. Diagnosis is however, important, as some of these lesions are malignant. An early detection should also reduce the incidence of pseudomyxoma peritonei and its associated morbidity[2,3]. It is therefore important to identify the disease process preoperatively and to plan a careful resection. AM can present in a variety of clinical syndromes or can occur as an incidental surgical finding in 50% of cases. The anatomic location of AM in the right lower quadrant of the abdomen includes it in the differential diagnosis of masses in this region. The early preoperative clinical diagnosis can therefore be difficult because of this lack of specific clinical symptomatology. The advent of advanced imaging techniques, particularly CT and sonography, has been invaluable in the diagnosis of mucocele of the appendix. Currently, the clinicians rely heavily on high resolution USS as the primary diagnostic tool in emergency hours being quick, sensitive and cheap.

2. Report of the case

A 60-year-old male presented with pain in the right lower quadrant of the abdomen since 4 days. It was of moderate intensity, intermittent in nature. The patient had a febrile illness at the onset of his symptoms. There was no change in bowel habits. On examination, the clinician felt a vague mass in the right iliac fossa, which was also mildly tender and mobile. No guarding and rigidity present. Laboratory data was normal (sedimentation ESR): 110 mm/h, leucocytes: 3.64, haemoglobin: 115 g/L, platelets: 222 g/L). Carcino Embryonic Antigen 4.43Transabdominal high frequency sonography was done using a Philips with a high frequency 8.2 mhz linear probe. The appendix measured. The

appendiceal surface appeared smooth and thin in outline. The appendix appeared as a cystic anechoic mass with posterior acoustic enhancement. There were few mobile echo of inflammatory debris inside the lumen.

A radiological diagnosis of mucocele of the appendix was made. At surgery, the diagnosis was confirmed with subhepatic caecal mucocele present. The appendix was carefully resected(Figure 2). The cecum, terminal ileum and mesentery were noted to be normal, and no evidence of infection was noted. The histopathological diagnosis was mucocele of the appendix with microscopy showing atypical proliferating mucous cells and papillary formation with some areas showing mucosal atrophy with fibrosis and inflammation. The mucosa of the appendix appeared intact, hyperplastic with minimal inflammation.

Figure 1: Appendicular Mucocele on gray scale

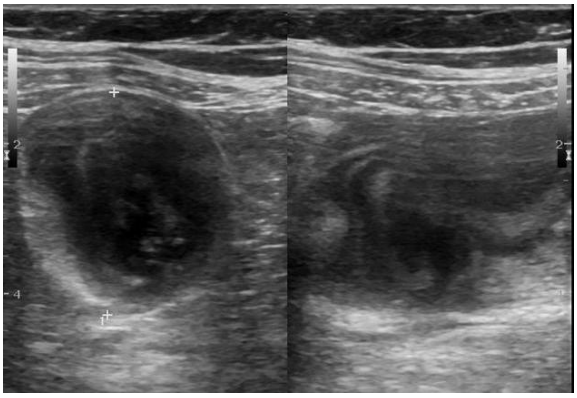
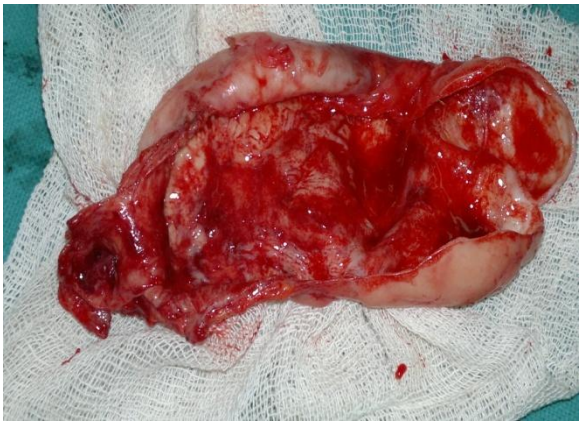


Figure 2: Surgical resected speciemen of appendix



3. Discussion

Mucocele of the appendix is a appendix distended by mucous secretions (31), secondary to mucinous cystadenoma (63%), mucosal hyperplasia (25%), mucinous cystadenocarcinoma (11%) and retention cyst[4]. A very rare type is secondary to occlusion of the lumen from post-inflammatory scarring, agerelated atrophy, congenital obstruction of Gerlach's valve or extramural compression. This type leads to an atrophic mucosa. All other types are classified into a spectrum, from mucous hyperplasia to mucinous cystadenoma to mucinous cystadenocarcinoma, as discussed above. Mucocele can also occur due to occlusion of the lumen by endometriosis or carcinoid tumor. Clinical manifestations include palpable abdominal mass, gastrointestinal bleeding and lower right abdominal pain, as in our case[5,6]. Leukocytosis is usually absent. Malignant to benign disease Symptomatic patients were reported, more likely, to have a malignant disease. Other tumors have been associated with AM, including gastrointestinal tract, ovary, breast and kidney tumors; the most common association being colorectal cancer. Whether AM represents an increased risk for colorectal cancer is unknown. A very important fact to be stressed here is the need for more mucoceles of the appendix to be diagnosed preoperatively. This makes the surgeon aware of the need for more careful surgery and consequently reduces the chances of iatrogenic damage to a mucocele with resultant leakage of the contents in the abdominal cavity with serious repercussions, especially pseudomyxoma peritonei[7].

The typical imaging finding of an appendiceal mucocele is a cystic mass in the expected region of the appendix. Plain abdominal radiographs may show mass effect on the adjacent bowel or bladder. Sonography shows an oblong, heterogeneous cystic mass containing mucin that is both liquid and gelatinous and viscous. The degree of internal echogenicity is related to the number of acoustic interfaces provided by the mucin[8]. Excellent through-transmission and posterior enhancement are usually present. The mass is well encapsulated unless rupture has occurred. The primary sonographic differentiation from uncomplicated acute appendicitis is the lack of appendiceal wall thickening of more than 6 mm. This distinguishing feature produces a characteristic target lesion, with an echogenic submucosal layer sandwiched by an edematous inner hypoechoic lamina propria,muscularis mucosa and outer hypoechoic muscular layer. Fine echo spots and/or concentric, echogenic layers within the cystic mass ("onion skin", Figure 1) are thought to be specific alteration[1]. The reason for the layered appearance is unclear; one may suggest that fluctuations of the concentration of mucin are responsible for this phenomenon either by fluctuation in the secretion of mucin into the cavity along with gradual absorption of water or by fluctuation of the degree of excretion

blockage from the cavity. Most cases did not show posterior echo enhancement.

Internal septations, polypoid lesions extending into the lumen and irregular outline seem to be associated with the malignant variety, although some papillary processes may be seen in mucinous cystadenomas[1,5,6]. The differential diagnosis on ultrasound include fluid filled small bowel,fluid in a small or large bowel diverticulum, appendiceal / diverticular abscess, mesenteric cyst seroma and particularly in females of reproductive age group, salpingitis and ectopic pregnancy masses[5,7].

On CT, a low-attenuation, well-encapsulated mass with smooth regular walls is seen in the right lower quadrant.

4. Conclusion

In conclusion, high-resolution sonography provides a sensitive, rapid, and noninvasive method for localizing the mucocele of the appendix. Thus, although it is nonspecific, when a mass is palpable or detected incidentally by imaging studies in the lower right abdomen in a patient without a history of appendectomy, we must consider the possibility of AM.

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