



Contrast Enhanced Ultrasonography, As An Additional Imaging Feature, Provides an Accurate Identification of Appendiceal Mucinous Cystadenoma: A Case Report

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Abstract

Appendiceal mucocele is a rather rare entity mimicking acute appendicitis, ranging from simple cystadenoma to malignant adenocarcinoma. Depending on radiological diagnosis, treatment varies from simple appendectomy to right hemicolectomy. Moreover complications in case of malignancy such as pseudomyxoma peritonei can be avoided if early diagnosis is obtained. Typically US appearance includes onion skin sign and on CT lack of uptake is considered in favour of a benign case. Contrast Enhanced Ultrasound is applied in order to investigate nodule uptake indicative of malignancy.

Keywords: Contrast enhanced ultrasound; Appendix; Mucinous cystadenoma

Introduction

Right lower quadrant pain is a common presenting complaint in the Emergency Department usually attributed to acute appendicitis. However, various uncommon entities can mimic acute appendicitis. One such rare entity is appendiceal mucocele [1]. Mucocele presentation varies from a simple retention cyst to malignant adenocarcinoma [2]. Rupture of the mucocele can be complicated with pseudomyxoma peritonei [3].

We present a case of mucocele of appendix in a 42 year old male subject complaining of lower quadrant discomfort, that was diagnosed sonographically and confirmed by CT. Contrast enhanced ultrasonography (CEUS) was conducted to exclude potential focal areas with wash out, which indicate malignancy [4]. Simple appendectomy was performed and the histopathological analysis showed a mucinous cystadenoma with no evidence of malignancy.

Case Presentation

A 42 year old man presented to the emergency department at 251 Hellenic Air Force and VA Athens General Hospital complaining of a 5 day duration right lower quadrant pain. The patient was a febrile and did not mention vomiting or diarrhea. Physical examination showed mild tenderness, localized in the right lower quadrant pain. Laboratory exams were normal.

Ultrasound examination was performed with an Aixplorer ultrasound system (SuperSonic Imagine, Aix-en-Provence, France), equipped with a multi frequency sector transducer (SL10-2) having a bandwidth of 2-10 MHz and a Single Crystal Curved Array XC6-1, having a bandwidth of 1-6 MHz. An enlarged vermiform appendix was identified in the right iliac fossa close to the iliac vessels. A hypoechoic structure was recognized inside the appendix with fine internal echoes, as the onion skin sign [5], which represents lamellated mucin and is considered pathognomonic for mucocele of the appendix [6] (Figure 1).

The patient was examined by the surgeons, who suggested further imaging studies in order to rule out a secondary acute inflammatory process and malignancy. A contrast enhanced ultrasonography examination was performed by a sole Radiologist with 10 years'

experience on CEUS, with an Aixplorer ultrasound system (SuperSonic Imagine, Aix-en-Provence, France), equipped with a Single Crystal Curved Array XC6-1, having a bandwidth of 1-6 MHz. Imaging was performed with a mechanical index of 0.09 and the focus was adjusted to the depth of the mass. According to the non-liver CEUS guideline, the contrast agent SonoVue (Bracco, Milan, Italy) was injected as a bolus 2.4 mL SonoVue (Bracco) through a 20 gauge catheter into an antecubital vein, and followed by injection of 5 mL of normal saline solution (0.9% NaCl). The CEUS examination was recorded from the end of the bolus and for a 120 seconds period. The appendiceal wall started to enhance at 10th second and the enhancement was equally present in the entire wall of the appendix. Previous studies suggested that the presence of a focal nodular lesion in the tumor cavity is an important predictor of malignancy [4,7]. In our case there was no evidence of appendiceal abscess or malignancy in the scanning area because of the absence of defect sites and the absence of focal areas with wash-out (Figures 2-4).

A CT-abdomen examination was also performed and revealed an enlarged fluid-filled appendix with axial diameter of 3 cm and without peri appendicular fat stranding. A curvilinear calcification was identified in the wall of the appendix, which is highly predictor of a mucocele [8]. There was no evidence of pseudomyxoma peritonei and no presence of intra-abdominal lymph node enlargement (Figure 5).

Based on the normal thickened and normal contrast-enhanced wall of the appendix in the abdominal CT and CEUS, mucocele of appendix of the appendix was diagnosed without radiological evidence of malignancy. Simple appendectomy was performed and surrounding connective tissue was excised. The histopathological findings confirmed the diagnosis of mucinous cystadenoma with no evidence of malignancy.

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Figure 1: In B mode ultrasound, the onion skin sign of the appendix presented as hypoechoic structure with fine internal echoes (white arrows).

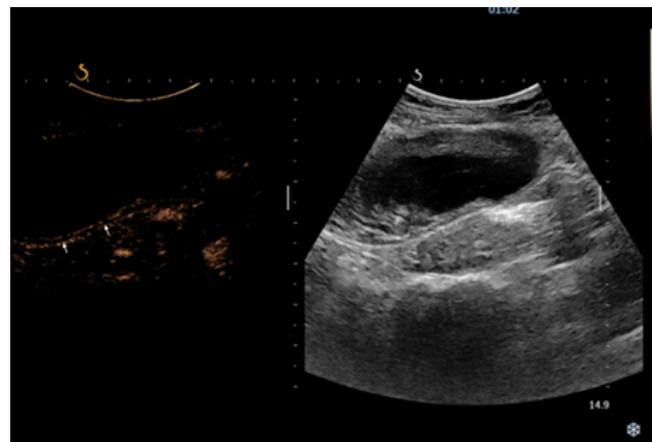


Figure 4: Contrast-enhanced ultrasound at Late phase 62 sec after bolus injection: normal contrast-enhanced wall of the appendix (white arrows). No presence of wash.



Figure 2: Contrast-enhanced ultrasound at arterial phase (13 sec after bolus injection): normal thickened and normal contrast-enhanced wall (white arrows) of the appendix.



Figure 3: Contrast-enhanced ultrasound at arterial phase venous phase (39 sec after bolus injection): normal contrast-enhanced wall (white arrows) of the appendix.



Figure 5: CT revealed an enlarged fluid-filled appendix without peri-appendicular fat stranding. A curvilinear calcification was identified in the wall of the appendix (white).

Discussion

Mucocele is a rare tumour of the appendix. It was first described by Rokitsky in 1842 as hydrops processus vermiformis [9]. Approximately 50% of the cases are asymptomatic and detected

incidentally [10], during surgery, radiological evaluations or endoscopic procedures [5]. Mucocele is identified in about 0.07-0.3% of all appendectomy cases [11]. Mucocele of the appendix affects women 4 times more often as men with peak age of incidence after fifth decade [11]. However, adenocarcinomas of the appendix are more often than men with peak age at 60 or 70 years old decades and there is an association with other colonic neoplasms and chronic ulcerative colitis [12]. Primary appendiceal adenocarcinomas are very rare malignant neoplasm accounting only 6% of all malignant tumors of appendix [13].

Various classifications have been proposed. A commonly used pathological classification system was described by Higa et al. [7], and includes three types: (i) focal or diffuse mucosal hyperplasia (ii) mucinous cystadenoma and (iii) mucinous cystadenocarcinoma. Simple mucoceles may mimic cystic lymphangioma, mesenteric cyst, enteric duplication cyst, Meckel diverticulum, retroperitoneal tumor and ovarian cystic lesion [14]. The presence of a solid enhancing cecal mass could indicate a cecal tumor [2]. Mucoceles are treated surgically [2]. Benign mucinous tumors require simple appendectomy [8], whereas bulky adenomas with large base may need cecal resection [15]. All appendectomies should include a wide mesoappendix resection in

order to exclude lymph node involvement [16]. Right hemicolectomy is reserved for malignant lesions [8], when peritoneal disease is absent [10]. In the present case, an enlarged appendix was successfully visualized by B-mode US and the mucocele was diagnosed by the pathognomonic onion skin sign. CEUS examination and additionally to CT showed no evidence of malignancy and guided the surgeons to proceed to a simple appendectomy instead of right hemicolectomy.

CEUS is being increasingly used as a first-line tool for detecting and characterizing hepatic and non-hepatic lesions [17]. CEUS is a new technique providing information about blood perfusion of the lesion in real-time through intravenous injection of ultrasound contrast agent [4]. In comparison with other imaging techniques CEUS provides a higher level of spatial resolution and more detailed information on blood perfusion of the lesion in real-time [4,7]. It can differentiate the microvasculature in bowel wall between healthy and diseased intestines [18]. In our case the use of CEUS helped to visualize precisely the solidified mucus and exclude a solid malignancy tumor. These findings suggest the utility of B-mode US combined with CT for diagnostic imaging of mucocele. CEUS is particularly useful for the assessment of blood flow in projections to exclude potentially malignant sites.

Conclusion

In current literature there are only a few studies examining the mucocele of the appendix by CEUS [4,7]. It has been suggested that CEUS is an important tool for determining the treatment strategy [7]. Future studies may show that this new modality is an important preoperative diagnostic tool for this condition.

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