Sonographic Findings of Malignant Appendix Tumors in Seven Cases

Kyung Su Kwag1, Hyuk Jung Kim1*, Suk Ki Jang1, Jae Woo Yeon1, Soya Paik2, Byeong Geon Jeon3, Ki Ho Kim4, Ji Hoon Park4, Eun Shin5

Departments of 1Radiology, 2Pathology and 3Surgery, Daejin Medical Center Bundang Jesaeng General Hospital, Departments of 4Radiology and 5Pathology, Seoul National University Bundang Hospital, Seongnam-Dong, Gyeonggi-Do, South Korea

Abstract

We report the sonographic features of confirmed malignant appendiceal tumors in seven cases. The histologic diagnoses of these tumors were mucinous cystadenocarcinoma (n = 2), colonic type adenocarcinoma (n = 4), and signet-ring cell carcinoma (n = 1). The 2 mucinous cystadenocarcinomas showed mucocele type, which had markedly enlarged inner luminal diameters (mean, 23 mm; range, 15–31 mm) and thick, irregular walls (mean wall thickness, 5.5 mm; range, 5–6 mm). In contrast, the 5 nonmucinous carcinomas (4 adenocarcinomas and 1 signet-ring cell carcinoma) showed nonmucocele type, which had relatively small inner luminal diameters (mean ± standard deviation [SD], 6.6 ± 4.5 mm; range, 2–15 mm) and prominent wall thickening (mean wall thickness ± SD, 6.2 ± 2.3 mm; range, 3–10 mm). Of the 5 nonmucinous tumors, only one had a discernible mass, three had thick irregular walls, two had loss of the wall layer pattern, and four had submucosal hypoechogenicity. Regardless of the histologic type, five of the seven malignant appendiceal tumors showed a severe periappendiceal fat infiltration or periappendiceal abscess, suggestive of perforation. Although the sonographic findings of the malignant appendiceal tumors were nonspecific, some of the sonographic features seen in these seven cases may help radiologists consider the possibility of underlying malignant appendiceal tumors.

Keywords: Appendix, malignant tumor, sonography

Introduction

Appendiceal tumors are very rare. They are found in approximately 1% of appendectomies, and malignant appendiceal tumors account for 27% of all appendiceal neoplasms.[1] Approximately 30%–50% of all appendiceal neoplasms are associated with signs and symptoms of acute appendicitis, and the rest of them are clinically silent.[2] The radiologic findings of these tumors are also usually nonspecific. Therefore, the possibility of an underlying appendiceal neoplasm is seldom suspected before surgery. The correct diagnosis is usually made by evaluating the frozen section at the time of surgery or later during the pathologic evaluation of the surgical specimen.[2,3] However, preoperative detection of underlying appendiceal malignancy is important because it can lead to modification of the surgical approach and extent of resection.[2]

Appendiceal neoplasms can lead to morphologic changes of the appendix, which can be divided into the two following subgroups according to their morphologic features: mucocele type and nonmucocele type.[2,4] A mucocele is a macroscopic morphological descriptive term representing intraluminal distension due to the accumulation of mucoid materials.[3,7] There have been several case reports about computed tomography (CT) and ultrasound (US) findings of each various appendiceal tumors.[1,2,8-10] However, to our knowledge, there has been no case report about US findings including both mucocele and nonmucocele type appendiceal tumor.

Therefore, the purpose of this case report was to show characteristic US features of malignant appendiceal tumor classified into mucocele and nonmucocele type.

Address for correspondence: Dr. Hyuk Jung Kim, Department of Radiology, Daejin Medical Center Bundang Jesaeng General Hospital, 255-2, Seohyung-Dong, Bundang-Gu, Sungnam-Si, Gyeonggi-Do 463-774, South Korea. E-mail: hyukjungkim@naver.com

Received: 13-03-2017    Accepted: 26-10-2017    Available Online: 28-03-2018

How to cite this article: Kwag KS, Kim HJ, Jang SK, Yeon JW, Paik S, Jeon BG, et al. Sonographic findings of malignant appendix tumors in seven cases. J Med Ultrasound 2018;26:52-5.
Case Report

This retrospective case series study was approved by the institutional review board, and the requirement for informed consent was waived.

Case samples

By conducting a computer search of the pathology database at our institution, we identified 86 patients who had underlying appendiceal tumors from a total of 9585 registered appendix specimens that were collected during a simple appendectomy, cecectomy, ileocecectomy, and right hemicolectomy over a 16-year from January 2000 to December 2015. Patients with a benign tumor (n = 30) and borderline malignancy (n = 37), which cannot be clearly classified as benign or malignant from the histopathological results, were excluded. Of the remaining 19 patients with malignant tumors, only seven underwent preoperative US, and they were included in the final case samples.

The histologic diagnoses of these tumors were mucinous cystadenocarcinoma (n = 2), colonic type adenocarcinoma (n = 4), and signet-ring cell carcinoma (n = 1). Among the current case samples, one case has been reported in a case report that described the sonographic findings of signet-ring cell carcinoma (Case 7) in the appendix.[11]

Ultrasound techniques

All US examinations were performed with Acuson Aspen and Sequoia (Siemens, Forchheim, Germany) and an iU22 US system (Philips Healthcare, Eindhoven, Netherlands) using 5–8 MHz curved or 5–12 MHz linear probes. In two patients, color Doppler US was performed at the end of the gray-scale US examination to evaluate blood flow.

Patient characteristics

The mean age at presentation was 62.6 years (± standard deviation, ± 12.1 years; age range, 51–79 years). Two patients were males and five were females. All patients had the clinical symptom of the right lower quadrant pain. On physical examination, there was no palpable mass in the right lower quadrant of the abdomen. Two patients (Cases 4 and 6) initially underwent ileocecectomy and right hemicolectomy because the malignancy was confirmed by frozen section examination performed during the operation. Four patients (Cases 2, 3, 5, and 7) underwent a radical second operation after initial appendectomy and cecectomy because the malignant appendiceal tumors were confirmed in the final pathological report. The remaining patient (Case 1) underwent cecectomy and did not undergo a radical secondary operation based on the surgeon’s decision because the tumor showed only focal destructive invasion of the muscularis propria. Two patients (Cases 2 and 4) had elevated levels of carcinoembryonic antigen.

Pathologic findings

Two mucinous cystadenocarcinomas (Cases 1 and 2) were enlarged, cystic, and porcelain-like—this appearance is called a mucocele—and had an irregular, thick wall. In contrast, among the 5 nonmucinous carcinoma (4 adenocarcinomas [Cases 3–6] and 1 signet-ring cell carcinoma [Case 7]), two had a mass at the base of the appendix, and the other three cases did not have a mass, but diffuse transmural and mucosal tumor infiltration was seen microscopically. Regardless of the histologic types, five of the seven malignant appendiceal tumors had perforation in the pathologic or surgical reports.

Sonographic findings

Table 1 shows the sonographic findings of each case. The mean inner luminal diameter and wall thickness of the seven cases were 11.2 mm ± 9.4 mm (range, 2–31 mm) and 6.0 mm ± 2.0 mm (3–10 mm), respectively. Two mucinous cystadenocarcinomas had markedly enlarged inner luminal diameters (mean, 23 mm; range, 15–31 mm) and irregular, thick walls (5.5 mm; 5–6 mm) [Figure 1]. On the other hand, five nonmucinous carcinomas (4 adenocarcinomas and a signet-ring cell carcinoma) had relatively small inner luminal diameters (6.6 mm ± 4.5 mm, 2–15 mm) and prominent thick walls (6.2 mm ± 2.3 mm, 3–10 mm) [Figure 2].

Of the five nonmucinous tumors, only one had a discernable mass on sonographic images, and the other four had wall thickening with or without irregularity. In addition, most of them (4 of 5) had submucosal hypoechogenicity and loss of the wall layer pattern was noted in two cases.

Regardless of the histologic types, five of the seven malignant appendiceal tumors showed severe periappendiceal fat infiltration or periaappendiceal abscess, suggestive of perforation.

Discussion

Primary carcinoma of the appendix is rare and constitutes <0.5% of all gastrointestinal neoplasms.[12] Even though primary appendiceal cancers are rare, the histology is diverse. Carcinoids are by far the most common, accounting for approximately 66% of appendiceal cancers, with cystadenocarcinomas accounting for 20% and adenocarcinomas accounting for 10%.[13] Moreover, there are rare forms of appendiceal cancers that include adenocarcinoid tumor, signet-ring cell carcinoma, nonHodgkin’s lymphoma, ganglioneuroma, and pheochromocytoma. In our database, four malignant appendiceal neuroendocrine tumors were found but excluded because they did not perform preoperative sonography. Finally, we included cystadenocarcinoma, adenocarcinoma, and signet-ring cell carcinoma.

Most commonly, patients with malignant appendiceal tumors present with symptoms and signs suggestive of acute appendicitis, as was seen in the current case series.[1] According to the current recommendations, all noncarcinoid, malignant, appendiceal tumors should be removed by a right hemicolectomy. Therefore, if an underlying appendiceal neoplasm is not suspected before surgery, a secondary radical operation, including a right hemicolectomy, is needed after the primary appendectomy.[12] Most of our patients underwent a radical operation, including four cases, underwent a secondary radical operation because malignancy had not been suspected at the time of preoperative diagnosis or surgery. As seen in these
A 56-year-old man with a mucinous adenocarcinoma in the appendix (Case 2). (a) A coronal computed tomography scan of the appendix shows cystic dilatation of the appendix (arrow, A) and irregular wall thickening. (b) Axial sonography of the appendix also shows cystic dilatation of the appendix and irregular wall thickening (arrow, B). (c) A low-power microphotograph shows a cystic, dilated appendix with abundant intraluminal mucin (H and E, ×10). (d) The appendiceal lumen is lined by a mixture of high-grade, pseudostratified, columnar, neoplastic epithelium and low-grade, mucinous epithelium (H and E, ×40)

Kwag, et al.: Sonographic findings of malignant appendix tumors

On sonography, all of our cases showed nonspecific appendiceal wall thickening and luminal dilatation, which are suggestive of acute appendicitis. Interestingly, some atypical findings, which may be suggestive of malignant appendiceal tumor, were present. In two mucinous cystadenocarcinomas, cystic dilatation of theappendix (mucocele) with irregular wall thickening was seen on sonography, which correlated with pathologic findings. These findings were consistent with those of a previous report,[12,14,15] which reported that when cystic dilatation of the appendix is present, wall irregularity and internal soft-tissue density with nodular thickening are associated with malignancy on CT. On the other hand, five nonmucinous carcinomas (four adenocarcinomas and one signet-ring cell carcinoma) had relatively small inner luminal diameters and thickened appendiceal walls ($n$ = 4) or mass ($n$ = 1) combined with submucosal hypoechogenicity. According to a previous report,[16-20] malignant tumors of the colon demonstrated the following wall characteristics on a high-resolution sonographic examination: heterogeneous hypoechoic mass, irregular wall thickening, and absence of a layered appearance of the wall. As the submucosal hypoechogenicity and loss of wall layer pattern are associated with tumor cell infiltration and loss of wall pattern thickening and absence of a layered appearance of the wall, these findings were consistent with those of a previous report.[12,14,15] Malignant tumors of the appendix also showed the following wall characteristics: heterogeneous hypoechoic mass, irregular wall thickening, and absence of a layered appearance of the wall. As the submucosal hypoechogenicity and loss of wall layer pattern are associated with tumor cell infiltration and loss of wall pattern thickening and absence of a layered appearance of the wall, these findings were consistent with those of a previous report.[12,14,15]

### Table 1: Summary of sonographic features of patients

| Case | Wall thickening* (mm) | Outer diameter† (mm) | Inner diameter‡ (mm) | Severe fat infiltration§ | Abscess | Cecal wall thickening | Enlarged LNs¶ | Mass** | Submucosal echo** | Loss of wall layer pattern** | Wall irregularity** | Pathologic results |
|------|-----------------------|---------------------|---------------------|-------------------------|---------|----------------------|---------------|---------|------------------|--------------------------|------------------|-------------------|
| 1    | 5                     | 26                  | 15                  | No                      | No      | No                   | No            | No      | Hyperchoic       | No                       | Yes              | Mucinous cystadenocarcinoma |
| 2    | 6                     | 44                  | 31                  | Yes                     | No      | No                   | No            | No      | Hyperchoic       | No                       | Yes              | Mucinous cystadenocarcinoma |
| 3    | 5                     | 13                  | 4                   | Yes                     | Yes     | Yes                  | Yes           | No      | Hyperchoic       | No                       | Yes              | Metastatic adenocarcinoma, poorly differentiated |
| 4    | 7                     | 17                  | 5                   | Yes                     | Yes     | Yes                  | Yes           | No      | Hyperchoic       | Yes                      | Yes              | Adenocarcinoma, poorly differentiated |
| 5    | 10                    | 24                  | 7                   | Yes                     | No      | No                   | No            | Yes     | Hyperchoic       | Yes                      | Yes              | Adenocarcinoma, moderately differentiated |
| 6    | 3                     | 18                  | 15                  | Yes                     | Yes     | Yes                  | Yes           | No      | Hyperchoic       | No                       | Yes              | Adenocarcinoma, well differentiated |
| 7    | 6                     | 15                  | 2                   | No                      | No      | No                   | No            | No      | Hyperchoic       | No                       | No               | Signet-ring cell carcinoma |

*Maximal wall thickening of appendix, †Outer to outer wall diameter at the maximal wall thickening portion, ‡Luminal diameter at the maximal wall thickening portion, §Large amount of perappendiceal echogenic fat encircling appendix and extending peripherally, ¶≥1 cm in short axis, round shape, or loss of hilar fat echo, **Evaluated at the measurable maximal wall thickening portion.
However, these sonographic findings are nonspecific, so they can be seen not only in infiltrative appendiceal tumors, such as lymphoma and goblet cell carcinoid tumor,[22,23] but also in perforation in appendicitis.[22,23]

In addition, most malignant appendiceal tumors in our case series had sonographic findings suggestive of perforation, such as severe peripapillary fat infiltration or peripapillary abscess, except for two cases. Nitecki et al. reviewed 94 consecutive patients with primary adenocarcinomas of the appendix and reported that 46% of the patients had an appendiceal perforation.[24] As reported by Lim et al.,[8] there is an increased risk of perforation in malignant mucocles.

**Conclusion**

Malignant appendiceal tumors are rare, and their sonographic findings do not suggest a definitive diagnosis, but some features may prompt the radiologist to consider the possibility of a malignant appendiceal tumor.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

**References**

1. Ruoff C, Hanna L, Zhi W, Shahzad G, Gotlieb V, Saif MW, et al. Cancers of the appendix: Review of the literatures. ISRN Oncol 2011;2011:728579.
2. Pickhardt PJ, Levy AD, Rohrman CA Jr., Kende AI. Primary neoplasms of the appendix: Radiologic spectrum of disease with pathologic correlation. Radiographics 2003;23:645-62.

3. Benedix F, Reimer A, Gastinger I, Mroczkowski P, Lippeit H, Kube R, et al. Primary appendiceal carcinoma — Epidemiology, surgery and survival: Results of a German multi-center study. Eur J Surg Oncol 2010;36:763-71.
4. Pickhardt PJ, Levy AD, Rohrmann CA Jr., Kende AI. Primary neoplasms of the appendix manifesting as acute appendicitis: CT findings with pathologic comparison. Radiology 2002;224:775-81.
5. Zhou ML, Yan FH, Xu PJ, Zhang LJ, Li QH, Ji Y, et al. Mucinous cystadenoma of the appendix: CT findings. Chin Med J (Engl) 2006;119:1300-3.
6. Chiou YY, Pitman MB, Hahn PF, Kim YH, Rhea JT, Mueller PR, et al. Rare benign and malignant appendiceal lesions: Spectrum of computed tomography findings with pathologic correlation. J Comput Assist Tomogr 2003;27:297-306.
7. Madwed D, Mindelzun R, Jeffrey RB Jr. Mucocoele of the appendix: Imaging findings. AJR Am J Roentgenol 1992;159:69-72.
8. Lim HK, Lee WJ, Kim SH, Kim B, Cho JM, Byun JY, et al. Primary mucinous cystadenocarcinoma of the appendix: CT findings. AJR Am J Roentgenol 1999;173:1071-4.
9. Fusari M, Sorrentino N, Botazzi EC, Del Vecchio W, Cozzolino I, Maurea S, et al. Primary signet ring cell carcinoma of the appendix mimicking acute appendicitis. Acta Radiol Short Rep 2012;1:p1:ar12.200107.
10. Tirumani SH, Fraser-Hill M, Auer R, Shabana W, Walsh C, Lee F, et al. Mucinous neoplasms of the appendix: A current comprehensive clinicopathologic and imaging review. Cancer Imaging 2013;13:14-25.
11. Cho YJ, Kim HJ, Jang SY, Yeon JW, Kim KH, Paik SY, et al. Signet-ring cell carcinoma of the appendix: A case report with an emphasis on sonographic findings. Ultrasoundonography 2014;35:164-7.
12. Wang H, Chen YQ, Wei R, Wang QB, Song B, Wang CY, et al. Appendiceal mucocoele: A diagnostic dilemma in differentiating malignant from benign lesions with CT. AJR Am J Roentgenol 2013;200:W590-5.
13. McCusker ME, Coté TR, Clegg LX, Sobin LH. Primary malignant neoplasms of the appendix: A population-based study from the surveillance, epidemiology and end-results program, 1973-1998. Cancer 2002;94:3307-12.
14. Waku N, Fujita M, Yamauchi Y, Takeda Y, Ueki N, Otsuka T, et al. Mucinous cystadenocarcinoma of the appendix in which contrast-enhanced ultrasonography was useful for assessing blood flow in a focal nodular lesion in the tumor cavity: A case report. Exp Ther Med 2013;6:3-8.
15. Kim SH, Lim HK, Lee WJ, Lim JH, Byun JY. Mucocele of the appendix: Ultrasonographic and CT findings. Abdom Imaging 1998;23:292-6.
16. Shirahama M, Koga T, Ishibashi H, Uchida S, Ohta Y. Sonographic features of colon carcinoma seen with high-frequency transabdominal ultrasound. J Clin Ultrasound 1994;22:359-65.
17. Richardson NG, Heriot AG, Kumar D, Joseph AE. Abdominal ultrasonography in the diagnosis of colonic cancer. Br J Surg 1998;85:530-3.
18. Lim JH. Colorectal cancer: Sonographic findings. AJR Am J Roentgenol 1996;167:45-7.
19. Price J, Metreweli C. Ultrasonographic diagnosis of clinically non-palpable primary colonic neoplasms. Br J Radiol 1988;61:190-5.
20. Chaubal N, Dighe M, Shah M, Chaubal J. Sonography of the gastrointestinal tract. J Ultrasound Med 2006;25:87-97.
21. Kim HJ, Ha HK, Cho KS, Yu E, Kim JC, Yoo CS, et al. CT features of primary colorectal signet-ring cell carcinoma. J Comput Assist Tomogr 2001;25:225-30.
22. Blurnf eld E, Nayak G, Srinivasan R, Muranaka MT, Blitman NM, Blurnf eld A, et al. Ultrasonography for differentiation between perforated and nonperforated appendicitis in pediatric patients. AJR Am J Roentgenol 2013;200:957-62.
23. Quillin SP, Siegel MJ, Coffin CM. Acute appendicitis in children: Value of sonography in detecting perforation. AJR Am J Roentgenol 1992;159:1265-8.
24. Nitecki SS, Wolff BG, Schlinkert R, Sarr MG. The natural history of surgically treated primary adenocarcinoma of the appendix. Ann Surg 1994;219:51-7.