

# Sonographic Findings of Malignant Appendix Tumors in Seven Cases

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## 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  $\pm$  standard deviation [SD],  $6.6 \pm 4.5$  mm; range, 2–15 mm) and prominent wall thickening (mean wall thickness  $\pm$  SD,  $6.2 \pm 2.3$  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.<sup>[1]</sup> Approximately 30%–50% of all appendiceal neoplasms are associated with signs and symptoms of acute appendicitis, and the rest of them are clinically silent.<sup>[2]</sup> 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.<sup>[2,3]</sup> However, preoperative detection of underlying appendiceal malignancy is important because it can lead to modification of the surgical approach and extent of resection.<sup>[2]</sup>

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.<sup>[2,4]</sup> A mucocele is a macroscopic morphological descriptive term representing intraluminal distension due to the accumulation of mucoid materials.<sup>[5–7]</sup> There have been several case reports about computed tomography (CT) and ultrasound (US) findings of each various appendiceal tumors.<sup>[1,2,8–10]</sup> 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.

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## 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.<sup>[11]</sup>

### 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 ( $\pm$  standard deviation,  $\pm 12.1$  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  $11.2 \text{ mm} \pm 9.4 \text{ mm}$  (range, 2–31 mm) and  $6.0 \text{ mm} \pm 2.0 \text{ mm}$  (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 ( $6.6 \text{ mm} \pm 4.5 \text{ mm}$ , 2–15 mm) and prominent thick walls ( $6.2 \text{ mm} \pm 2.3 \text{ mm}$ , 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 periappendiceal abscess, suggestive of perforation.

## DISCUSSION

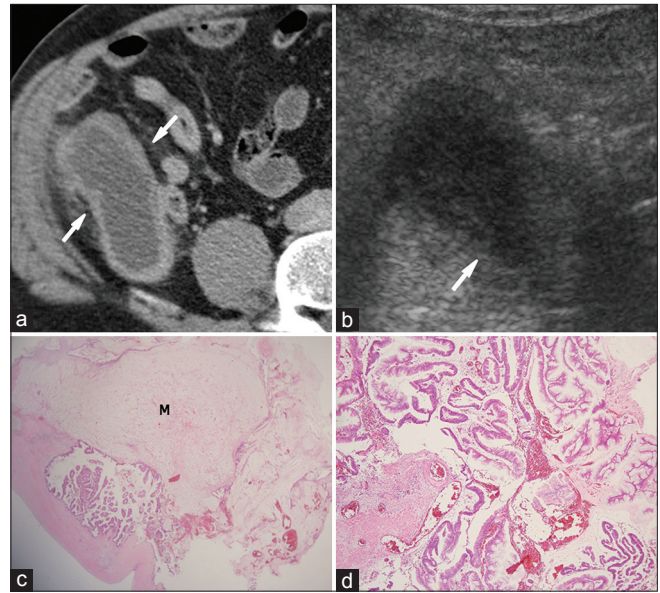
Primary carcinoma of the appendix is rare and constitutes <0.5% of all gastrointestinal neoplasms.<sup>[12]</sup> Even though primary appendiceal cancers are rare, the histology is diverse. Carcinoids are by far the most common, accounting for approximately 66% of appendiceal cancers, with cystadenocarcinomas accounting for 20% and adenocarcinomas accounting for 10%.<sup>[13]</sup> 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 performed 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.<sup>[3]</sup> 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.<sup>[12]</sup> 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

**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	Hyperechoic	No	Yes	Mucinous cystadenocarcinoma
2	6	44	31	Yes	No	No	No	Yes	Hyperechoic	No	Yes	Mucinous cystadenocarcinoma
3	5	13	4	Yes	Yes	Yes	No	No	Hypoechoic	No	Yes	Metastatic adenocarcinoma, poorly differentiated
4	7	17	5	Yes	No	Yes	No	Yes	Hypoechoic	Yes	Yes	Adenocarcinoma, poorly differentiated
5	10	24	7	Yes	No	No	No	No	Hypoechoic	Yes	Yes	Adenocarcinoma, moderately differentiated
6	3	18	15	Yes	Yes	Yes	No	No	Hyperechoic	No	No	Adenocarcinoma, well differentiated
7	6	15	2	No	No	No	No	No	Hypoechoic	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 periappendiceal echogenic fat encircling appendix and extending peripherally, ≥1 cm in short axis, round shape, or loss of hilar fat echo, ¶Discernable space-occupying lesion including segmental wall thickening, \*\*Evaluated at the measurable maximal wall thickening portion. LN: Lymph nodes

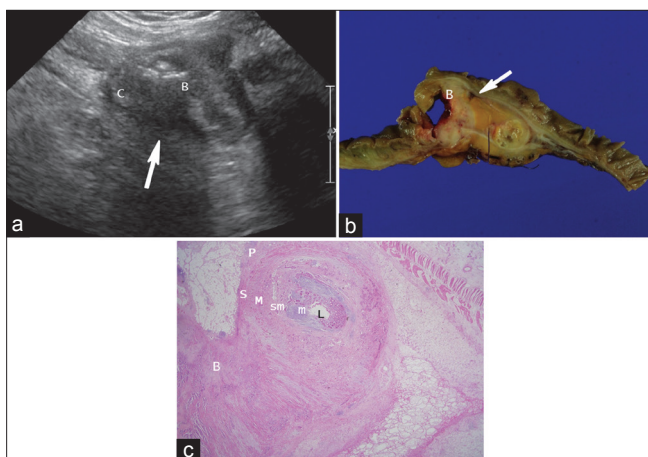


**Figure 1:** A 56-year-old man with a mucinous adenocarcinoma in the appendix (Case 2). (a) A coronal computed tomography scan of the appendix shows cystic dilatation of the appendix (arrow, A) and irregular wall thickening. (b) Axial sonography of the appendix also shows cystic dilatation of the appendix 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)

cases, preoperative detection of an underlying appendiceal malignancy is important because modification of the surgical approach and extent of resection may be required.

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 malignant appendiceal tumor, were present. In two mucinous cystadenocarcinomas, cystic dilatation of the appendix (mucocele) with irregular wall thickening was seen on sonography, which correlated with pathologic findings. These findings were consistent with those of a previous study,<sup>[12,14,15]</sup> 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,<sup>[16-20]</sup> 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, they can be also seen in malignant appendiceal tumors. In our five nonmucinous carcinomas of the appendix, four showed submucosal hypoechogenicity and two showed loss of a wall layer pattern.





**Figure 2:** A 79-year-old man with a nonmucinous adenocarcinoma in the appendix (Case 4). (a) An axial sonography shows an irregular, hypoechoic mass (arrow) at the appendiceal base and cecum. Loss of the wall layer pattern is also noted. (b) On gross examination, adenocarcinoma (arrow) was detected at the appendiceal base. (c) A low-power microphotograph shows tumor cells infiltrating the mucosa, submucosa, muscularis, and serosa layers and periappendiceal tissue (H and E stain,  $\times 100$ ). B, appendix base; C, cecum; L, lumen; M, muscle; m, mucosa; S, serosa; sm, submucosa; P, periappendiceal tissue

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,<sup>[2,21]</sup> but also in perforation in appendicitis.<sup>[22,23]</sup>

In addition, most malignant appendiceal tumors in our case series had sonographic findings suggestive of perforation, such as severe periappendiceal fat infiltration or periappendiceal 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.<sup>[24]</sup> As reported by Lim *et al.*,<sup>[8]</sup> there is an increased risk of perforation in malignant mucocèles.

## 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.

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## Conflicts of interest

There are no conflicts of interest.

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