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## Case Report

# Low-grade appendiceal mucinous neoplasm with puffer ball-like appearance: A case report <sup>☆,☆☆</sup>

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## ABSTRACT

We report a case of ruptured low-grade appendiceal mucinous neoplasm with an impressive toy puffer ball-like appearance on magnetic resonance imaging. A 79-year-old woman with lower abdominal pain underwent computed tomography scanning, revealing a 6-cm mass in the right lower abdomen. T2-weighted images showed a radial low-signal structure in the central area of the mass, which was presumed to be fibrotic. Pathology confirmed ruptured low-grade appendiceal mucinous neoplasm. The rupture point was at the tip of the appendix, coinciding with the center of radial fibrosis. The unique morphology of the puffer ball-like appearance in this case may be a characteristic of low-grade appendiceal mucinous neoplasms.

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## Introduction

The diagnosis of low-grade appendiceal mucinous neoplasm (LAMN) has been adopted from the 4th edition of the World Health Organization classification, 2010. LAMN was previously

known as mucinous cystadenoma or mucinous cystadenocarcinoma. LAMN rupture can lead to progressive peritoneal dissemination, resulting in a clinical syndrome referred to as pseudomyxoma peritonei (PMP). Herein, we report a case of ruptured LAMN with an impressive toy puffer ball-like appearance on magnetic resonance imaging (MRI).

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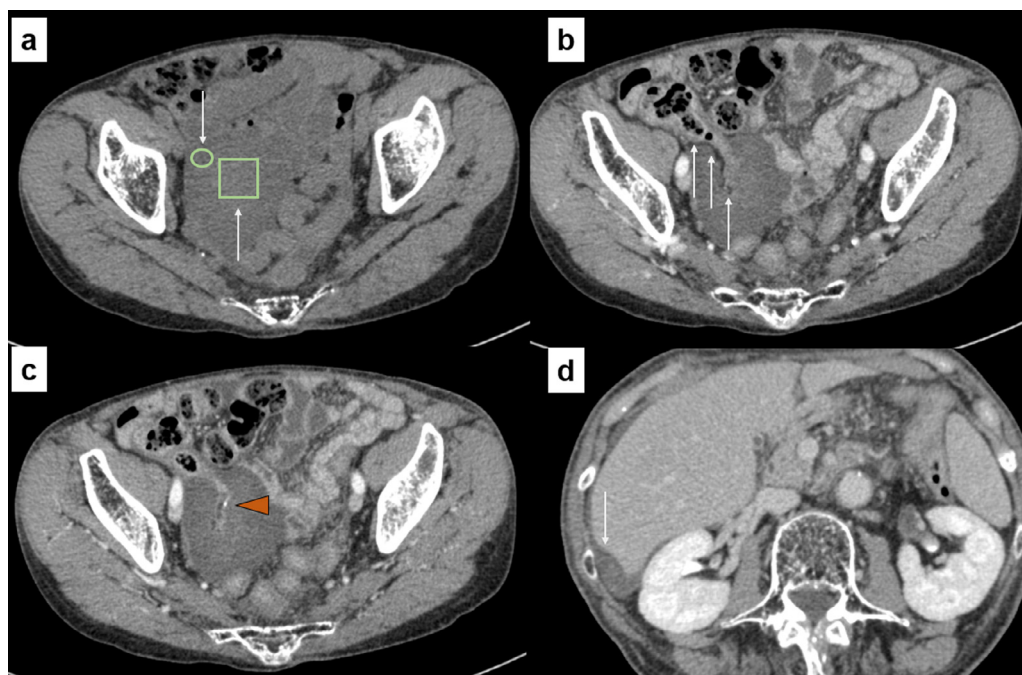
<sup>☆☆</sup> Competing Interests: We declare that we have no conflict of interest.

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**Fig. 1** – Computed tomography (CT) images. Plain CT image (A) (1-mm thickness). CT values were measured at the locations indicated in the figure. The CT values of the peripheral area (circled area 67 mm<sup>2</sup>) were 13 HU on pre-enhanced study, 14 HU in the early phase (not shown), and 15 HU in the delayed phase (not shown). The CT value of the central area (rectangle area 600 mm<sup>2</sup>) (appendiceal wall not included) was 27 HU on pre-enhanced study, 34 HU in the early phase (not shown), and 38 HU in the delayed phase (not shown). Early-phase images (B, C) (1-mm thickness). The central area of the mass, around the appendix, is faintly enhanced, but the peripheral areas are not. The appendix and mesenteric fat continuous with the cecum are identified in the tumor (B) (arrows). The appendiceal wall is smooth and nonthickened, and the adipose tissue of the appendiceal mesentery is preserved. Wall calcification is noted in the middle part of the appendix (C) (arrowhead). Delayed phase CT image (D) (1-mm thickness). Only 1 gelatinous mucinous implant is observed around the liver (arrow), except for the appendiceal mass.

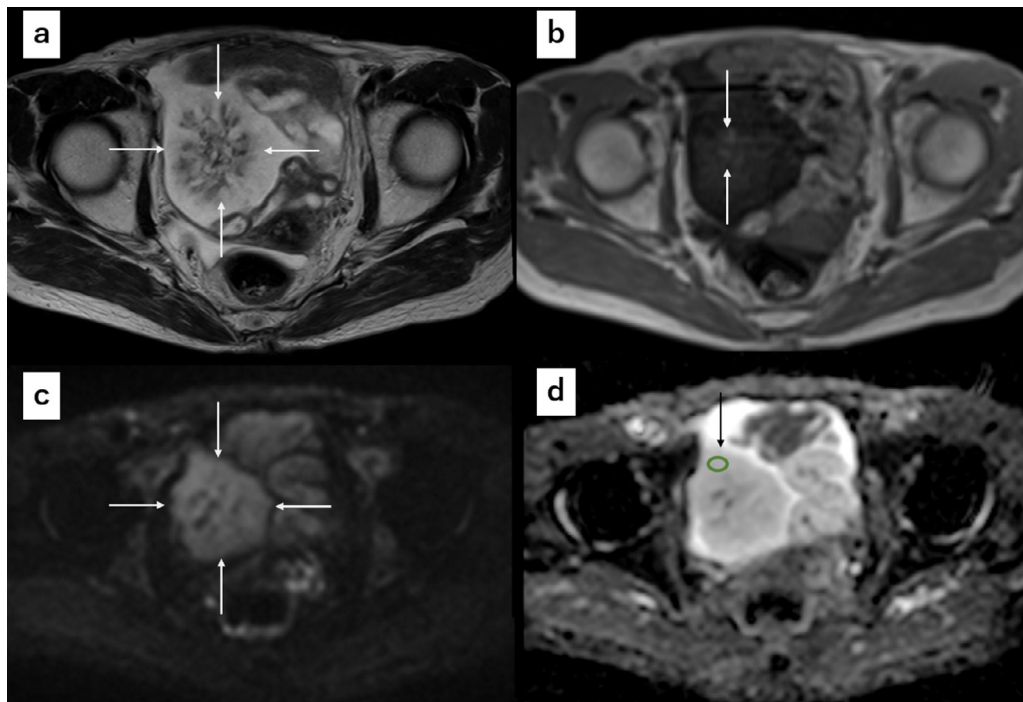
## Case report

A 79-year-old woman presented with a 3-month history of lower abdominal pain. Computed tomography (CT) and MRI revealed a 6-cm mass in the right lower abdomen. Serological examination revealed a carcinoembryonic antigen level of 56 ng/mL (normal range: 0.10–5.00 ng/mL) and carbohydrate antigen 19-9 level of 114 U/mL (normal range: 0–37.00 U/mL). The patient had diabetes (glycosylated hemoglobin, HbA1c: 7.6%; normal range: 4.6%–6.2%) and anemia (hemoglobin, Hb: 9.5 g/dL; normal range: 12–16 g/dL). She also had malignant lymphoma but had been in remission for the last 17 years.

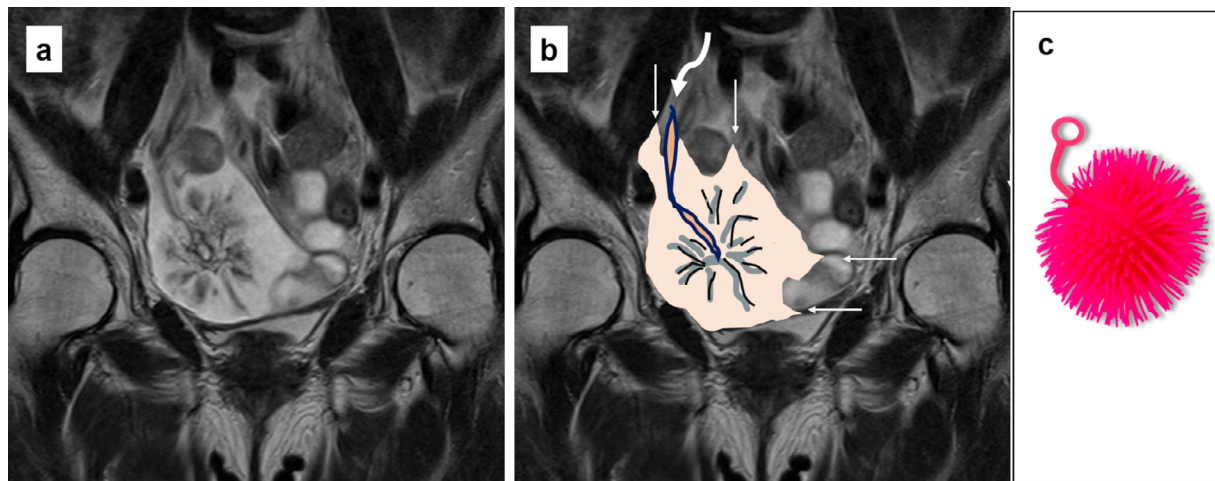
The mass was 6-cm in diameter with well-defined borders, resembling encapsulated ascites or peritoneal inclusion cysts. The mass showed low density and was almost isodense to urine on plain CT, and the peripheral area was not enhanced with contrast material (Fig. 1A, circled area). The appendiceal tip was located at the center of the mass, which was enhanced with the contrast material, and the surrounding area was faintly enhanced (Fig. 1A, rectangular area). CT with a slice-thickness of 1 mm showed that the continuity of the appendiceal wall was unclear at the apex but preserved in other parts of the appendix. The appendiceal wall was smooth and

nonthickened, and the adipose tissue of the appendix mesentery was included in the tumor (Fig. 1B, arrows). Wall calcification was observed in the middle part of the appendix (Fig. 1C, arrowhead). The appendiceal lumen was not dilated. Only 1 lesion resembling encapsulated ascites was observed around the liver (Fig. 1D, arrow).

MRI was performed for further qualitative evaluation of the tumor. On MRI examination, the mass showed low intensity on T1-weighted imaging and high intensity on T2-weighted imaging, and it was almost isointense to urine (Figs. 2A and B). T2-weighted images showed a radial low signal structure in the central area of the mass (Fig. 2A, arrows), which was presumed to be fibrosis. The center of the radial fibrosis was the tip of the appendix, showing a toy puffer ball-like appearance (Figs. 2A, 3A, B, C). On T1-weighted images, it was seen that the adipose tissue of the appendix mesentery was included in the tumor (Fig. 2B, arrows). On diffusion-weighted imaging (DWI) (b value = 800 mm/s<sup>2</sup>), the tumor showed slight hyperintensity (Fig. 2c, arrows), and the apparent diffusion coefficient value of the lesion was high ( $2.7 \times 10^{-3}$  mm<sup>2</sup>/s) in the circled area, as shown in Fig. 2D (arrow). The margin of the tumor was smooth, and several sharp apices were also observed (Fig. 3B, straight arrows), resembling the shape of encapsulated ascites or peritoneal inclusion cysts. The pre-

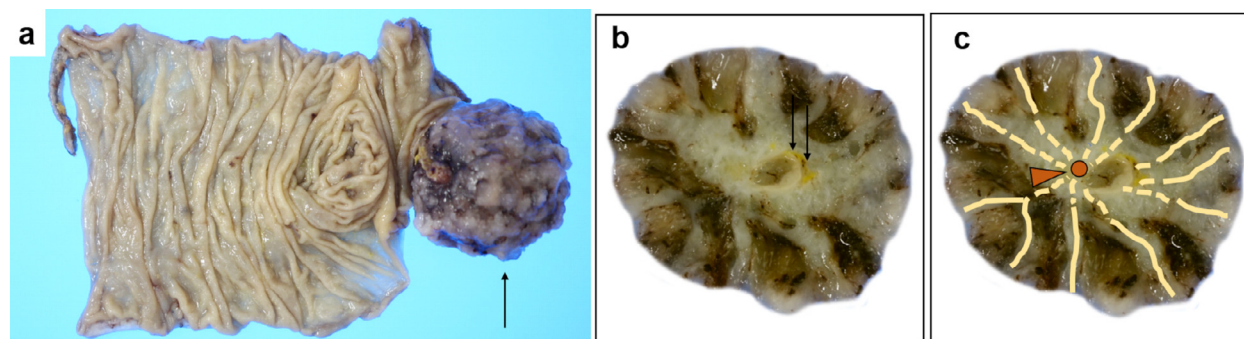


**Fig. 2** – Magnetic resonance imaging of the lower abdomen. T2-weighted image (A) revealing a radial low-signal structure (arrows) in the central mass area, presumed to be fibrosis. T1-weighted image (B) showing the adipose tissue (arrows) of the appendiceal mesentery to be present in the tumor. On diffusion-weighted imaging (C) ( $b$  value =  $800 \text{ s/mm}^2$ ), the tumor (arrows) has a faintly high intensity. The apparent diffusion coefficient value of the lesion is high ( $2.7 \times 10^{-3} \text{ mm}^2/\text{s}$ ,  $101 \text{ mm}^2$ ) at the circled area shown (D) (arrow).



**Fig. 3** – Puffer ball-like appearance. Coronal T2-weighted image (A), the schema (B), and a toy “puffer ball”-like appearance (C). There is a duct-like structure extending from the ileocecum, which is considered to be the appendix (B, curved arrow). Radial distinct low-signal linear structures are present, and the center of the radial structure is the tip of the appendix. The structure appears like a toy puffer ball. The margin of the tumor is smooth for the most part, and several sharp apices are also observed (B, straight arrows), which resemble the shape of encapsulated ascites or peritoneal inclusion cyst.





**Fig. 4 – Resected specimen of the appendiceal tumor. The peripheral tumor area, rich in mucus components, is removed during surgery. The central area of the tumor, rich in fibrous components, is shown (A) (arrow). Cross-section of the tumor perpendicular to the long axis of the appendiceal tip (B) and the schema (C). The appendiceal wall and adipose tissue (arrows) of the appendix mesentery were preserved. Radial fibrosis is shown using solid lines. The extension of the fibrosis towards the tumor center is shown using a dashed line. The center of radial fibrosis appears to be the point indicated by the arrowhead.**

operative differential diagnosis was LAMN, mucinous adenocarcinoma, or a seromucinous borderline tumor with ectopic endometriosis.

The tumor was surgically excised. Surgical findings revealed that the tumor's peripheral area was rich in mucus components, which were removed during surgery. Implants were noted around the liver. In addition, tumor cells were detected in the ascites fluid. The gross specimen of the central area of the tumor showed rich fibrous components (Fig. 4A, arrow). The appendix and appendiceal mesentery fat tissue were included in the tumor (Fig. 4B, arrows), which corresponded with the CT and MRI findings. The appendiceal wall was smooth and nonthickened, and the appendicular lumen was not dilated. Radial fibrosis was also confirmed in the tumor, and the center of the radial fibrosis was at the tip of the appendix (Fig. 4C, arrowhead; Fig. 5A, arrowhead). Histological examination revealed tumor cells with low atypia in the appendiceal mucosa inside the appendix, and no tumor cells were detected in the mucosa of the appendicular root. Mucinous and fibrous tissue was observed around the appendix with scattered tumor cells. The appendiceal wall was mostly preserved with no invasion or destruction beyond the mucosa, but there was a 2-3-mm area where tumor cells had penetrated the appendiceal wall near the tip of the appendix, which was inferred to be a ruptured point (Fig. 5B, arrowhead). This rupture site coincided with the center of radial fibrosis. The tumor did not contain any papillary or ductal adenocarcinoma components. Based on these pathological and radiographic findings, the tumor was diagnosed as ruptured LAMN, that is, localized PMP.

## Discussion

LAMNs are known to arise in the appendix and, by definition, do not infiltrate [1]. LAMNs usually consist of only low-grade mucinous epithelial cells, and do not include other histological types of colon cancer.

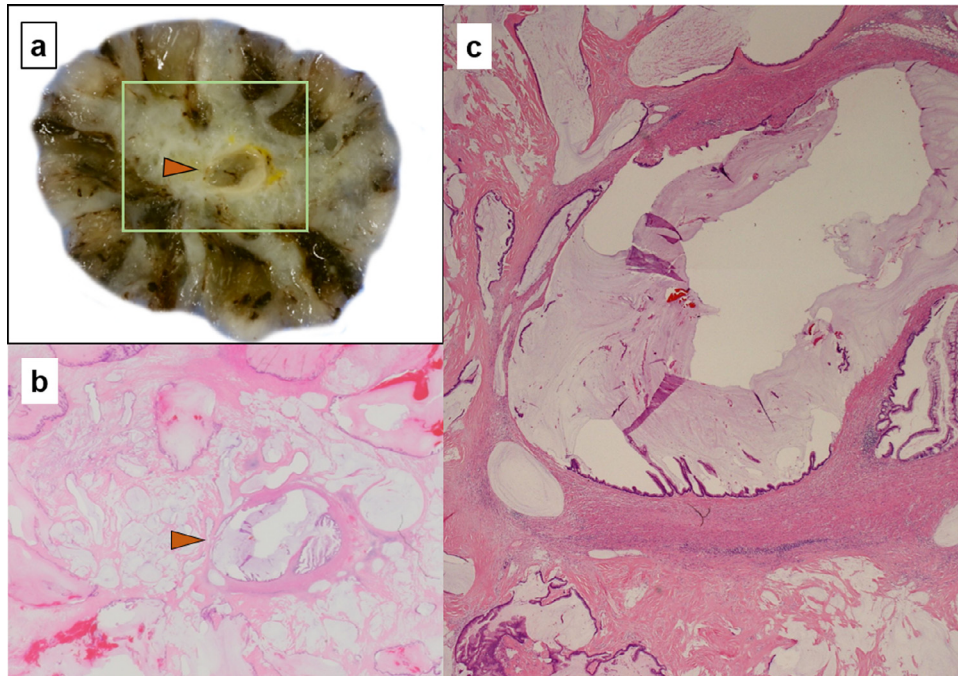
Mucinous tumors of the appendix are classified as LAMNs, high-grade appendiceal mucinous neoplasms (HAMNs), and mucinous adenocarcinomas [1]. HAMNs are distinguishable from LAMNs only by the degree of epithelial dysplasia. The World Health Organization classifies the majority of noninvasive epithelial appendiceal lesions as LAMNs [1].

Mucinous adenocarcinoma of the appendix, similar to other colorectal adenocarcinomas, is a subtype of adenocarcinoma that is usually mixed with other histologic types. They demonstrate frankly infiltrative invasion and are defined as invasive glands containing high-grade cytologic atypia and extracellular mucin, comprising >50% of the cross-sectional area of the lesion under a microscope [1]. In our case, the tumor cells penetrated the appendiceal wall at one site, which was 2-3 mm in area. If this site was evaluated as an invasion site, the diagnosis would be mucinous carcinoma; however, if it was evaluated as a ruptured site, the pathology would be consistent with LAMNs. Based on the radiological and pathological findings, the tumor was diagnosed as a localized PMP following LAMN rupture.

PMPs are rare, with an incidence of 1 per million, they occur in adults (mean age of occurrence: 53 years), and are 2-3 times more frequent in women [2]. On CT imaging, the masses show a low density (usually < 20 HU). A dominant cystic or solid mass is often present in the right lower abdomen (the expected location of the appendix). On MRI, implants usually show low signal intensity on T1-weighted images and hyperintensity on T2-weighted images. Areas of enhancement within the lesion are often more apparent on delayed phase images. DWI intensity depends on the cellularity. Moreover, mucus retention between adherent peritoneal membranes, called “scalloping,” is a characteristic feature [2].

The CT density and MR images in this case were consistent with previously reported features. The images in this case had the following 3 features, with the first not being generally described as a characteristic feature of ruptured LAMNs:

1. Radial fibrosis with a puffer ball-like appearance extending from the tip of the appendix (Figs. 3A and B).



**Fig. 5 – Gross and histological images of the rupture site. Cross-section of the tumor perpendicular to the long axis of the appendiceal tip (A) and the hematoxylin and eosin-stained image (B, C) (B: with a loupe) (C: x 20). The rectangular area of the resected specimen (A) coincides with the hematoxylin and eosin-stained image with a loupe (B). The center of radial fibrosis (A, arrowhead) coincides with the rupture site (B, arrowhead). The muscular layer is interrupted in one-quarter circumference, and collagen fibers and mucous lakes can be seen at the rupture site.**

2. A shape similar to that of encapsulated ascites or peritoneal inclusion cysts (Fig. 3A and B).
3. Dominant distribution near the appendix.

PMPs are thought to cause inflammation and fibrosis via the cytokines produced in the peritoneum [3]. The first feature, radial fibrosis, can be inferred to have been formed by the fibrosing process along with the slow expansion of the tumor. The second and third features may be related to the encapsulation of mucinous tumor cells by fibrosis.

To review the appendiceal MRI findings of ruptured LAMNs, we performed a literature search that included all literature published until October 19, 2022, using PubMed for studies published in English. The search terms were “rupture” AND (“low-grade appendiceal mucinous neoplasm” OR “mucinous cystadenocarcinoma” OR “mucinous cystadenoma”) AND “appendiceal” AND “MRI.” Eight cases were detected, of which 3 had MR images of the appendix reported [4–6]. Although not listed in PubMed, a reference case in a Japanese textbook on gynecologic MRI [7] also had MR images of the appendix. These 4 cases with images showed the typical MRI intensities that have already been reported. Of these 4 cases, 2 appeared to have a puffer ball-like appearance [4,7], but there was no mention of fibrosis, and the relationship between the rupture site and the center of the radial structure had not been described.

In summary, although the number of reported cases to date is small, a puffer ball-like appearance may be a characteristic feature of LAMNs, and this morphology may not be a rare finding. The reason for the scarcity of reported cases may be attributed to the appendiceal lesions being rarely investi-

gated by using an MRI examination; the described phenotype is difficult to recognize on CT, but is easily recognized on T2-weighted MR images. Our study suggests that the central area of the “puffer ball” is the ruptured point of the appendix, and that the radial spread of fibrosis from the rupture point is the cause of this unique morphology. To our knowledge, this is the first case report of a ruptured LAMN revealed through MRI and correlated with pathological findings. The limitations associated with this case is that this is just a rarely reported imaging feature, and that we have not been able to track the actual spread of fibrosis over time. The unique morphology of the puffer ball-like appearance observed in this case should be further investigated in future studies.

### Patient consent

Informed written consent was obtained from the patient for the publication of the case report and all imaging studies. Consent form is on record.

### REFERENCES

- [1] WHO Classification of Tumours Editorial Board. Digestive system tumors. World Health Organ Classif Tumours. 5th ed., 1. World Health Organization; 2019. p. 135–55.

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- [2] Siva P, Raman MD. Pseudomyxoma peritonei. In: *Diagnostic imaging: gastrointestinal*. Amsterdam: Elsevier; 2016. p. 148–51. p.
- [3] Lohani K, Shetty S, Sharma P, Govindarajan V, Thomas P, Loggie B. Pseudomyxoma peritonei: inflammatory responses in the peritoneal microenvironment. *Ann Surg Oncol* 2014;21:1441–7. doi:10.1245/s10434-013-3261-6.
- [4] Xu R, Yang ZL. Treatment of a giant low-grade appendiceal mucinous neoplasm: a case report. *World J Clin Cases* 2021;9:11056–60. doi:10.12998/wjcc.v9.i35.11056.
- [5] Suh DS, Song YJ, Kwon BS, Lee S, Lee NK, Choi KU, et al. An unusual case of pseudomyxoma peritonei associated with synchronous primary mucinous tumors of the ovary and appendix: a case report. *Oncol Lett* 2017;13:4813–17. doi:10.3892/ol.2017.6079.
- [6] Meer EM, Khazindar AR. Ruptured appendiceal mucocele presenting as a ventral hernia: a case report of a rare presentation of appendiceal mucocele. *Cureus* 2022;14:e23304. doi:10.7759/cureus.23304.
- [7] Imaoka I, Tanaka YO. A key to gynecologic MR imaging 2004:192.