Abdominal Radiology

Published online: 26 October 2018

Abdom Radiol (2019) 44:3246–3251
https://doi.org/10.1007/s00261-018-1813-8

Appendiceal endosalpingiosis: clinical presentation and imaging appearance of a rare condition of the appendix

James Tudor,[1] Todd R. Williams,[1] Daniel T. Myers,[1] and Beena Umar[2]

[1]Department of Radiology, Henry Ford Hospital, 2799 West Grand Blvd, Detroit, MI 48202, USA
[2]Department of Pathology, Henry Ford Hospital, 2799 West Grand Blvd, Detroit, MI 48202, USA

Abstract

Endosalpingiosis rarely affects the appendix but can be mistaken for acute appendicitis or appendiceal tumors. The medical literature regarding appendiceal endosalpingiosis is sparse; consisting of only four case reports which are primarily focused on the histopathology but provide little radiologic correlation. Endosalpingiosis is a rare condition characterized by the presence of benign fallopian tubal-like glandular epithelium derived from Mullerian ducts, usually affecting the serosal surfaces of the pelvis and peritoneum. It is histologically differentiated from endometriosis as endosalpingiosis lacks endometrial stroma. Endosalpingiosis tends to affect older women and has been associated with ovarian serous tumors of low malignant potential. After a retrospective review of a pathology database, we present pathologically proven cases of appendiceal endosalpingiosis with correlative imaging. We discuss the clinical presentation, illustrate the CT and MRI appearance, histologic characteristics, and review the current medical literature of appendiceal endosalpingiosis.

Key words: Appendix—Endosalpingiosis—Endometriosis—Appendicitis—Tumors

Endosalpingiosis refers to the condition of ectopic glandular fallopian tube-like epithelium implanted on serosal surfaces of the pelvis and adnexa. Grossly, these implants resemble endometriosis, however, histologically these implants consist of fallopian tube-like ciliated epithelium that lack the endometrial stroma seen in endometriosis [1–3]. Endosalpingiosis is most commonly seen along serosal surfaces in the pelvis, in an older age group than endometriosis, and has been associated with an increased rate of developing gynecological malignancies, specifically ovarian serous tumors of low malignant potential [1]. The medical literature is scant regarding appendiceal endosalpingiosis, consisting of 5 case reports, four of which are in the pathology literature. We report two cases of pathologically proven cystic appendiceal endosalpingiosis that presented with right lower quadrant pain, initially clinically thought to represent acute appendicitis or appendiceal neoplasm. We illustrate the imaging findings of appendiceal endosalpingiosis on CT, endosalpingiosis on MRI and summarize the current literature.

Materials and methods

After approval by the Institutional Review Board (IRB), a 4-year retrospective review of both the pathology and radiology archive was performed at our institution for the prospective diagnosis of appendiceal endosalpingiosis. A data search utilizing a medical record search engine (Illuminate Softek, Overland Park, Kansas, USA) of the pathology database searched for pathology reports containing both the words "appendix" and "endosalpingiosis" to identify potential cases. Twenty-four reports were identified having both words. Eleven of these cases identified the presence of endosalpingiosis involving the appendix. Six cases had confounding, coexisting appendiceal pathologic diagnoses including endometriosis, neuroendocrine tumor, Brenner tumor, and three cases with concomitant acute appendicitis and were excluded. Of the remaining five cases, three had no correlative imaging within 30 days prior to the date of the pathologic specimen report, leaving the two cases presented here, both of which had contrast enhanced multidetector computed tomography (CEMDCT) imaging performed prior to appendectomy. Single phase CEMDCT images provided were performed on 64-slice multidetector scanner (GE Medical Systems, Milwaukee WI, USA) after IV contrast administration of 100 mL isovue-370, performed during the ve-

Correspondence to: Todd R. Williams; *email:* Toddw@rad.hfh.edu

nous phase utilizing a 70 s time delay after the onset of injection and a 3.75 mm reconstructed axial slice thickness.

Results

The two identified cases of pathologically proven appendiceal endosalpingiosis with pre-operative CT imaging both presented to the emergency department with right lower quadrant pain and were initially suspected of having acute appendicitis. In both cases the patients underwent abdominal/pelvic CEMDCT scans as part of the initial work-up. Similar to the previously reported case of cystic endosalpingiosis of the appendix by Pollheimer [2], our two cases also presented with a cystic mass abutting the appendiceal tip, although the masses in our cases are considerably larger measuring 18 mm in diameter in Case #1 and 23 mm in Case #2 vs. the 8 mm mass originally reported. Both patients had subsequent appendectomies shortly after the CEMDCT scans. Case #1 underwent a laparoscopic appendectomy. Case #2 required an open laparotomy as the patient had a previous laparoscopic appendectomy 5 years earlier and represented with a cystic mass along the suture margin at the tip of the appendiceal stump.

Case #1

Patient is a 41-year-old female who presented to the emergency department with a two-day history of increasing right lower quadrant pain, reported as 8/10 on the pain scale at the time of presentation. Pertinent past medical history included tubal ligation. The patient's white blood cell (WBC) count was not elevated at 8 K/microliter. Patient underwent contrast enhanced MDCT scan revealing an 18 mm eccentric cystic mass at the tip of the appendix, with little or no inflammatory response (Fig. 1). Pre-operative differential diagnosis included early tip appendicitis and appendiceal mucinous neoplasms. The patient went on to have simple uncomplicated laparoscopic appendectomy. Pathology was reported as cystic appendiceal endosalpingiosis.

Case #2

The second case is more complicated. The patient is a 36-year-old female who presented to the emergency department with increasing right lower quadrant pain over a 4-day period, similar to the prior RLQ pain which lead to her original appendectomy, 5 years earlier. Because her surgical history included prior appendectomy, appendicitis was not initially considered at the time of presentation. She had nausea, but no fever or chills, and her WBC count was not elevated at 8.9 K/microliter. Pertinent past surgical history included a prior partial hysterectomy and laparoscopic appendectomy as mentioned above. Her past medical history also included the diagnosis of endometriosis. The patient underwent a

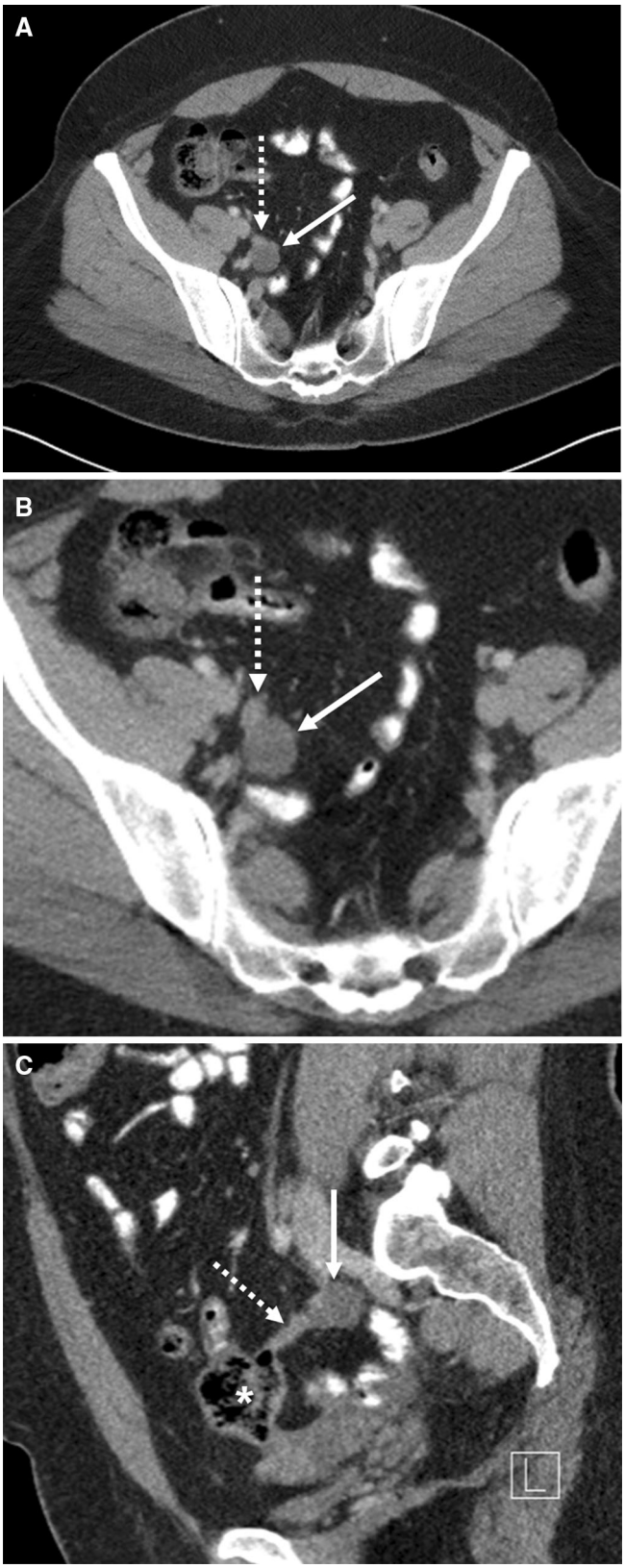


Fig. 1. Axial (**A**, **B**) and sagittal (**C**) CEMDCT images demonstrating a thin walled cystic mass (white arrow) eccentrically abutting the tip of the appendix (dashed arrow), * denotes cecum.

single phase CECT revealing a 19 × 23 mm rim enhancing eccentric cystic mass at the tip of the appendiceal stump, along the suture margin. Differential diagnosis pre-operatively included stump appendicitis and appendiceal mucinous neoplasm (Fig. 2).

Discussion

Background

Pelvic endosalpingiosis was first described in 1930 by Sampson in women who had prior surgery including tubal ligation or salpingectomies [4]. Endosalpingiosis is a rare condition where fallopian tube-like epithelium is deposited along serosal surfaces in the pelvis. Although the mechanism of peritoneal seeding is unclear, it has been postulated that chronic tubal inflammation or injury may lead to the proliferation of these cells, which are then deposited along serosal surfaces similar to endometriosis [1, 5]. This was initially described as occurring through transplantation of cells during surgery or as part of a reactive reparative process in the setting of tubal damage/injury. Of note, both of our patients had past surgical histories of tubal surgery, which supports the above proposed mechanism of seeding.

The other postulated theory for the development of endosalpingiosis includes a multi-focal metaplastic process (celomic metaplasia) involving the mesothelium called Mullerianosis and may be subject to hormonal influence. This Mullerianosis triad also includes endometriosis and endocervicosis [1, 6]. Both endosalpingiosis and endometriosis are thought to originate from the Mullerian ducts, however, endosalpingiosis differs histologically from endometriosis as it lacks endometrial stroma and tends not to elicit the inflammatory changes and cyclical pain associated with endometriosis due to this different histology [1, 2].

Endosalpingiosis is usually an incidental pathologic diagnosis made histologically after pelvic surgery or biopsy. It has been associated with benign and malignant gynecologic conditions, including endometriosis. Pelvic endosalpingiosis has been found histologically in lymph nodes and ovaries, and serosal implants have been reported along the colon, bladder, and uterus, and as in our cases, rarely the appendix [7, 8].

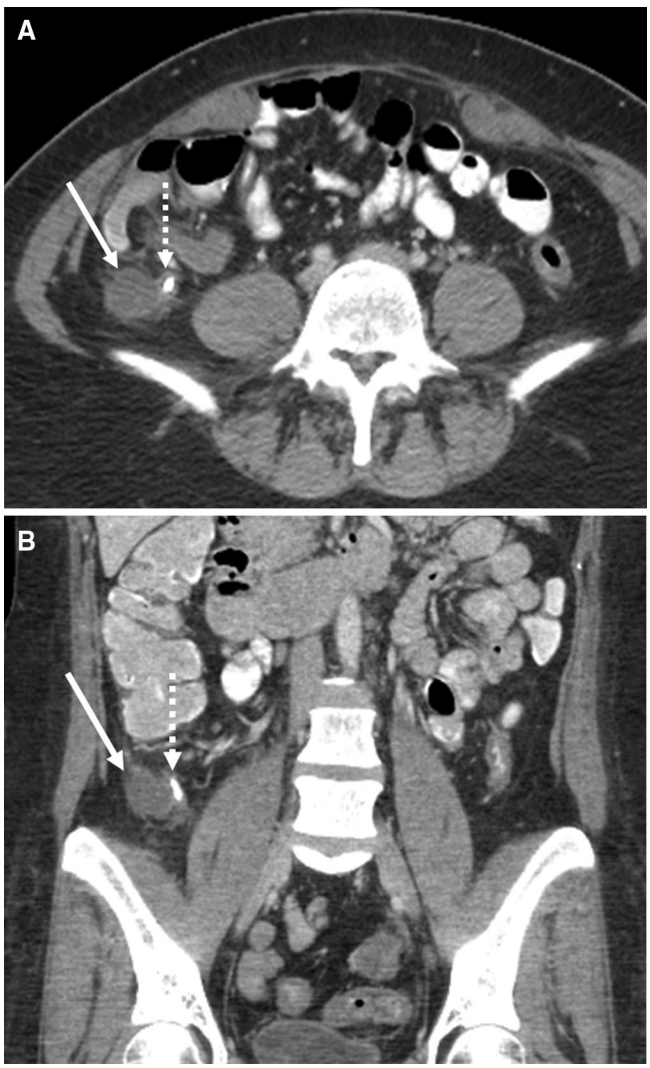


Fig. 2. Axial (**A**) and coronal (**B**) CECT images of the abdomen demonstrate a low density cystic mass adjacent to the tip of the appendiceal stump with high density appendectomy clips seen abutting the endosalpingiosis deposit.

Histology of endosalpingiosis

Microscopic examination shows multiple cystically dilated glands lined by single layer of fallopian tube type epithelium. The three cell types of normal fallopian tube epithelium (pale ciliated columnar cells, non-ciliated secretory (mucous) cells and dark intercalated/peg cells) are seen in varying numbers. Glands are surrounded by stroma which is composed of dense connective tissue and may contain mononuclear inflammatory cells. Psammoma bodies are frequently present within the gland lumina or in the adjacent stroma. In contrast, endometriosis is characterized by the presence of endometrial type glands and stroma along with hemosiderin laden macrophages (Fig. 3) which are absent in endosalpingiosis [9].

Imaging findings of endosalpingiosis

Pelvic endosalpingiosis is rarely diagnosed by imaging but has been described as either ill or well defined predominantly cystic lesion with mural and septal enhancement on post contrast imaging. The MRI appearance of endosalpingiosis has been described at dark T1, bright T2 signal with septal and mural

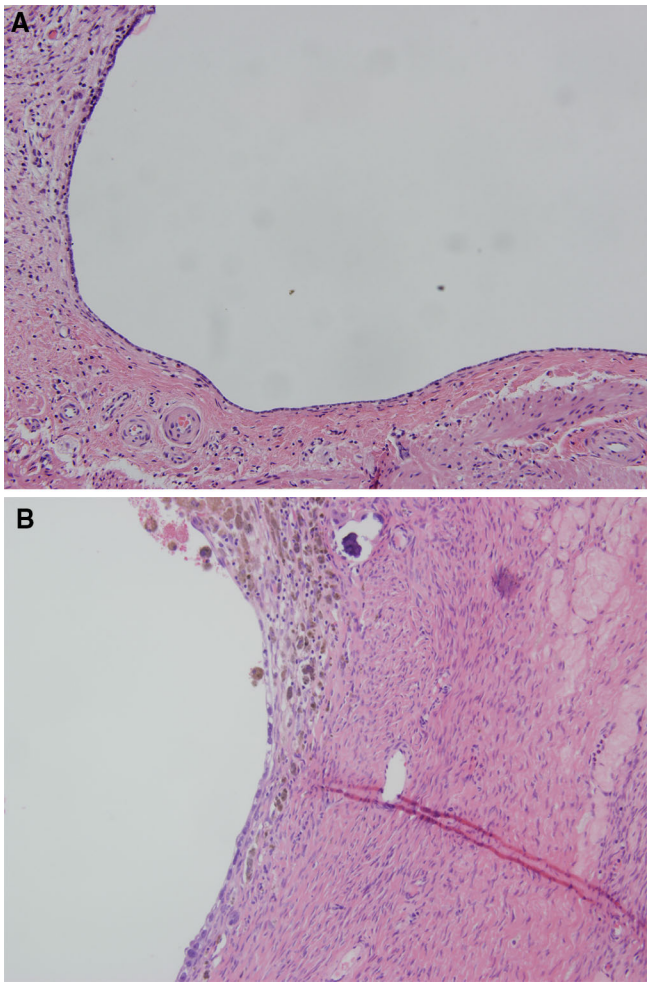


Fig. 3. Histopathology of endosalpingiosis with haematoxylin and eosin stain (20×) demonstrating a cystic space lined by ciliated columnar epithelial cells without atypia (**A**). Histopathology of endometriosis with haematoxylin and eosin stain (20×) demonstrating cystic space showing endometrial lining, endometrial stroma, and hemosiderin laden macrophages, with psammoma bodies (**B**).

enhancement (Fig. 4), which is much different that the T1 bright appearance of endometriosis. This cystic appearance of endosalpingiosis on MRI is non-specific and therefore can be mistaken for neoplasm, especially ovarian cystadenomas or metastatic disease when affecting the ovaries [10]. In both examples presented here, appendiceal endosalpingiosis presented as a thin walled cystic lesion abutting the appendiceal tip on CEMDCT. In both cases, the appendix or appendiceal stump itself was of normal caliber without any adjacent inflammatory change, and the lesion abutted the appendix without distending the lumen.

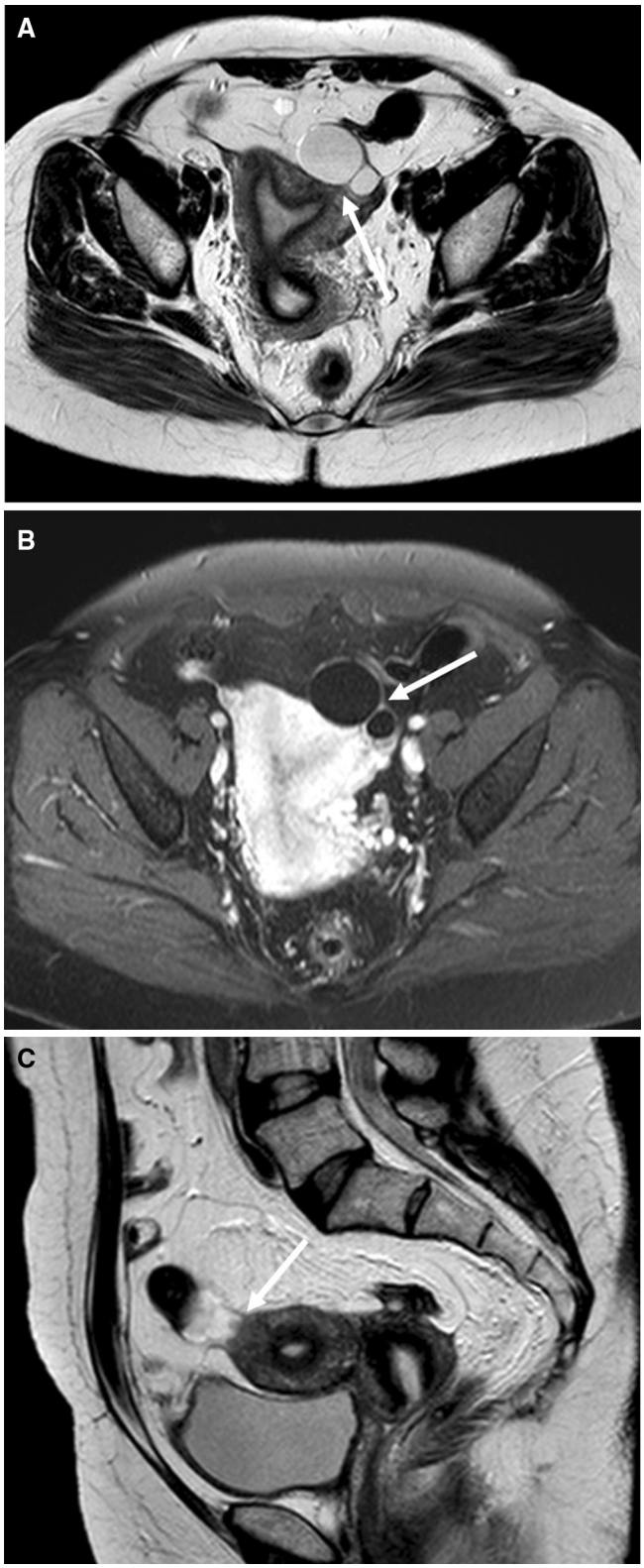
Appendiceal endosalpingiosis

Appendiceal endosalpingiosis is a rare entity. Cajigas reported the first case of appendiceal endosalpingiosis in 1991. Since then, there have only been five total reported cases of appendiceal endosalpingiosis described in four pathology case reports and one review article [3, 5, 11]. Based on our limited experience, the literature may under represent the true incidence of this finding.

Endometriosis of the appendix is much more common, affecting up to 2.6% of appendectomy histology specimens in one pathology study [2]. In general, endometriosis is a disease of pre-menopausal women, often presenting with chronic pelvic pain, whereas pelvic endosalpingiosis is seen in older females, with up to 40% being post-menopausal. There are currently conflicting study results, linking pelvic endosalpingiosis to chronic pelvic pain, however, both of our patients presented with a brief history of increasing right lower quadrant pain which resolved after appendectomy, and neither had pathologic evidence of appendicitis. Histologically, pelvic endosalpingiosis has been reported to occur concurrently with endometriosis in 34–66% of specimens but is often clinically occult and incidentally detected histologically [1]. Interestingly, our second case had a past medical history of endometriosis; however, endometriosis was not present in the appendiceal surgical specimen. One other important reported association of pelvic endosalpingiosis is the increased risk for the development of gynecological malignancies, specifically ovarian serous tumors of low malignant potential or the so called serous borderline ovarian tumors (SBTO) and to a lesser degree, ovarian cystadenocarcinomas [1, 6, 12]. To date neither of our reported cases of appendiceal endosalpingiosis has developed an ovarian malignancy.

Conclusion

Pelvic endosalpingiosis is a disorder consisting of tubal epithelial deposits occurring in the pelvis usually after tubal injury or from multi-focal mesothelial metaplasia. Histologically, endosalpingiosis is similar to endometriosis, but lacks endometrial stoma. Endosalpingiosis tends occurs an older age group and is less frequently associated with chronic pelvic pain. However, pelvic endosalpingiosis has a reported association with the increased risk of developing ovarian malignancies, specifically serous tumors of borderline malignant potential. Appendiceal endosalpingiosis is reportedly very rare. Both our cases presented with increasing right lower quadrant pain, no elevated WBC and a thin walled cystic mass at the tip of the appendix or appendiceal stump on CEMDCT without adjacent inflammatory



◀Fig. 4. Axial FSE T2 (**A**) and axial T1 fat saturated post contrast (**B**) MRI images demonstrating two thin walled cystic uterine serosal implants along the uterine fundus, pathologically proven after hysterectomy. The cystic component is dark on T1 and bright on T2 imaging with only thin mural/septal enhancement (white arrows). Sagittal T2 FSE (**C**) showing the interface with the serosal surface (white arrow).

changes. The findings of a peri-appendiceal cystic mass near the tip is non-specific with a range of diagnostic possibilities, including tip appendicitis, mucinous neoplasms of the appendix, peri-appendiceal implants from non-appendiceal origin mucinous neoplasm such as ovarian, and rarely cystic appendiceal endosalpingiosis. Both patients went on to appendectomy with resolution of their symptoms.

Compliance with ethical standards

Funding No grants or funding was involved in this project.

Conflict of interest All authors (Drs. Tudor, Williams, Myers and Umar) declare no conflicts of interest.

Ethical approval The study is a retrospective review of the medical record and is compliant with Henry Ford. Hospital's Institutional Review Board guidelines (IRB# 12225) and was performed in accordance with the ethical standards as laid down in the 1964 Declaration of Helsinki and its later amendments or comparable ethical standards. For this type of study (retrospective review) formal consent is not required.

Informal consent This article does not contain studies with human or animal participants performed by any of the authors.

References

1. Prentice L, Stewart A, Mohiuddin S, Johnson NP (2012) What is endosalpingiosis? Fertil Steril 98(4):942–947. https://doi.org/10.1016/j.fertnstert.2012.06.039
2. Pollheimer MJ, Leibl S, Pollheimer VS, Ratschek M, Langner C (2007) Cystic endosalpingiosis of the appendix. Virchows Arch 450(2):239–241. https://doi.org/10.1007/s00428-006-0328-9
3. Demir MK, Savas Y, Furuncuoglu Y, et al. (2017) Imaging findings of the unusual presentations, associations and clinical mimics of acute appendicitis. Eurasian J Med 49(3):198–203. https://doi.org/10.5152/eurasianjmed.2017.17218
4. Sampson JA (1930) Post salpingectomy endometriosis (endosalpingiosis). Am J Obstet Gynecol 20:443–480
5. Tallón-Aguilar L, Olano-Acosta C, López-Porras M, Flores-Cortés M, Pareja-Ciuró F (2009) Endosalpingiosis of the appendix. Cir Esp 85(6):383. https://doi.org/10.1016/j.ciresp.2008.09.017
6. Djordjevic B, Clement-Kruzel S, Atkinson NE, Malpica A (2010) Nodal endosalpingiosis in ovarian serous tumors of low malignant potential with lymph node involvement: a case for a precursor lesion. Am J Surg Pathol 34(10):1442–1448. https://doi.org/10.1097/PAS.0b013e3181f17d33
7. Katre R, Morani AK, Prasad SR, et al. (2010) Tumors and pseudotumors of the secondary Mullerian system: review with emphasis on cross-sectional imaging findings. AJR 195(6):1452–1459. https://doi.org/10.2214/AJR.10.4302
8. Esselen KM, Terry KL, Samuel A, et al. (2016) Endosalpingiosis: more than just an incidental finding at the time of gynecologic

surgery? Gynecol Oncol 142(2):255–260. https://doi.org/10.1016/j.ygyno.2016.05.036
9. Irving JA, Young RH, Clement PB (2015) *Sternberg's diagnostic surgical pathology*, 6th edn. The Netherlands: Wolters Kluwer Health
10. Kaneda S, Fujii S, Nosaka K, et al. (2015) MR imaging findings of mass-forming endosalpingiosis in both ovaries: a case report. Abdom Imaging 40(3):471–474. https://doi.org/10.1007/s00261-014-0327-2
11. Cajigas A, Axiotis CA (1990) Endosalpingiosis of the vermiform appendix. Int J Gynecol Pathol 9(3):291–295
12. Ryuko K, Miura H, Abu-Musa A, Iwanari O, Kitao M (1992) Endosalpingiosis in association with ovarian surface papillary tumor of borderline malignancy. Gynecol Oncol 46(1):107–110