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Case Report

Incidental finding of LAMN during hysterectomy for adnexal tumor

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Abstract

Low-grade appendiceal mucinous neoplasm (LAMN) is a rare malignancy originating from the epithelium of the appendix. The diagnosis is difficult to make, and it is often found during surgery.

In the present case, we prepared our patient for a total abdominal hysterectomy with bilateral salpingo-oophorectomy, but during laparotomy, a cystic dilation of the appendix was found. We did an appendectomy and the pathological diagnosis was LAMN. Gynecologists may encounter this disease incidentally. Whenever there is a right abdominal mass, cystic or not, found in the preoperative imaging, gynecologists should always consider appendiceal origin.

Keywords

adnexal tumors, appendiceal neoplasm, case report, low-grade appendiceal mucinous neoplasm

Introduction

Low-grade appendiceal mucinous neoplasm (LAMN) is a rare malignancy originating from the epithelium of the appendix with a reported incidence between 0.4% and 1.0% of all appendiceal specimens.[1] It usually presents with atypical clinical signs and symptoms. It can be seen during examination with CT imaging or ultrasound, more commonly mimicking adnexal masses.[2] It is not uncommon for a cyst, an abscess, a cancer of the gastrointestinal tract, or another pelvic organ to be identified through imaging as an adnexal mass, so they should not always be assumed to be of ovarian origin.[3] Sometimes LAMNs can be identified as suspicious appendiceal tumors during surgery, but, in most cases, they constitute an incidental finding on the following pathologists' report. The appendix tumor itself does not invade lymph nodes or demonstrate distant hematogenous metastases nor infiltrate surrounding tissues. However, it produces mucin that causes luminal obstruction and patients usually complain of abdominal pain.[4] The examination can indicate appendicitis or non-specific abdominal pain. Rarely, the expanding mucin may cause thinning and rupture of the appendiceal wall leading to spillage in the abdomen and creating a serious clinical syndrome, pseudomyxoma peritonei (PMP).[5] We present the case of a 59-year-old woman who was initially diagnosed with right and left ovarian cystic lesions and possibly fallopian tube formations using imaging but was later diagnosed with an appendiceal mass near a right ovarian cyst during exploratory laparotomy. This case highlights the need for a good preoperative imaging diagnosis and the importance of having LAMN in mind during differential diagnosis of cystic formations of the right lower quadrant of the abdomen.

Case presentation

A 59-year-old female patient was referred to our department because of incidental findings at her yearly scheduled gynecological examination. A transvaginal ultrasound described a longitudinal cystic formation 4.3 cm behind the uterus extending to the right without internal vessels and with low echogenicity content. Physical examination showed no findings and the patient described no other symptoms.

The past medical history of the patient included hypertension, hyperthyroidism, and transaminasemia under investigation. She denied any family history. She had no prior surgeries, no family history, and she was not a smoker. From her gynecologic history, she had one miscarriage, one abortion, and two pregnancies both with natural childbirth, and she entered the menopause period five years ago. Her last smear test of the same year was normal with a negative HPV test.

Her physician was concerned about her transaminasemia, so an MRCP and MRI were performed, but no abnormal findings were discovered. Liver elastography showed fatty liver disease of 2nd degree (F0-F1).

A follow-up abdominal ultrasound showed an oval cystic formation 7.5×5 cm in the anatomical position of the left ovary and another allantoid cystic formation 10.3×3.3 cm along the dilated right fallopian tube, with echogenic elements inside it. MRI showed a formation with smooth borders in the right lower pelvis, possibly coming from the right adnexal, with high intensity on T2-weighted imaging 12×3.5 cm and another cystic formation 4.9×5.5 cm.

Preoperative laboratory evaluations were within normal limits, as well as the tumor markers, except for the already-known transaminasemia.

Based on the findings, the patient's age, and due to the patient's wish, she was prepared for a routine total hysterectomy with salpingo-oophorectomy. During laparotomy, a left adnexal mass with a cystic formation was found. It was removed and sent for a frozen section that came back negative for malignancy. Afterward, we proceeded to a total hysterectomy with right adnexal removal, which also appeared to be normal.

*During the exploratory laparotomy, a largely dilated appendix was found. The surgical oncologist was consulted. The possibility of a malignant nature of the appendiceal cyst was not very high according to the surgeon, and therefore he proposed a single appendectomy, which was subsequently performed using a GIA stapler **(Fig. 1)**. The post-surgery period was unremarkable and the patient was sent home awaiting the biopsy results.*

The pathologist review described a dilated appendix 12×5×4 cm in size. The appendiceal wall was intact on gross examination, with smooth serosa and luminal mucin component on dissection.

The microscopic examination revealed a mucinous neoplasm composed of epithelial cells with abundant apical cell mucin, flat or micropapillary histological pattern, and low-grade nuclear atypia. Infiltrative growth features were absent.

The neoplasm was confined mostly in the lumen. Focally acellular mucin invaded into the muscularis propria or extended to the serosa surface, or into the adipose tissue of the mesoappendix, with no infiltration to the uterus and ovary. In the surgical margin of resection, acellular mucin was also found in the serosal surface of the appendix.

*Serosal reaction was recognized, with chronic inflammatory reaction and neovascularization. The diagnosis was of a low-grade mucinous appendiceal neoplasm (LAMN) grade I who involved the appendix serosa (pT4a) **(Fig. 2)**.*

The case was discussed in our MDT. According to the PSOGI/EURACAN clinical practice guidelines, a pT4M0 patient with no peritoneal disease, such as ours, is recommended to undergo further surgical interventions. Following the guidelines, our patient was submitted to a right hemicolectomy and HIPEC. The biopsy from the right hemicolectomy came back negative for evidence of residual

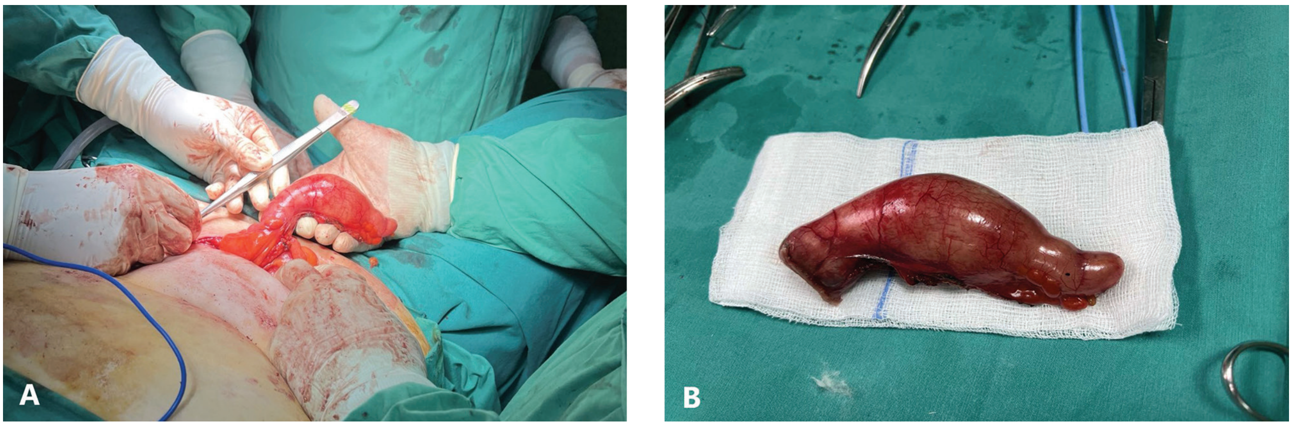

Figure 1. **A)** Intraoperative look during exploratory laparotomy of dilated appendix; **B)** Dilated appendix resected with GIA; The biopsy results showed a serous cystadenoma of the left ovary. The uterus included a leiomyoma, an adenomyoma, and superficial spots of adenomyosis.

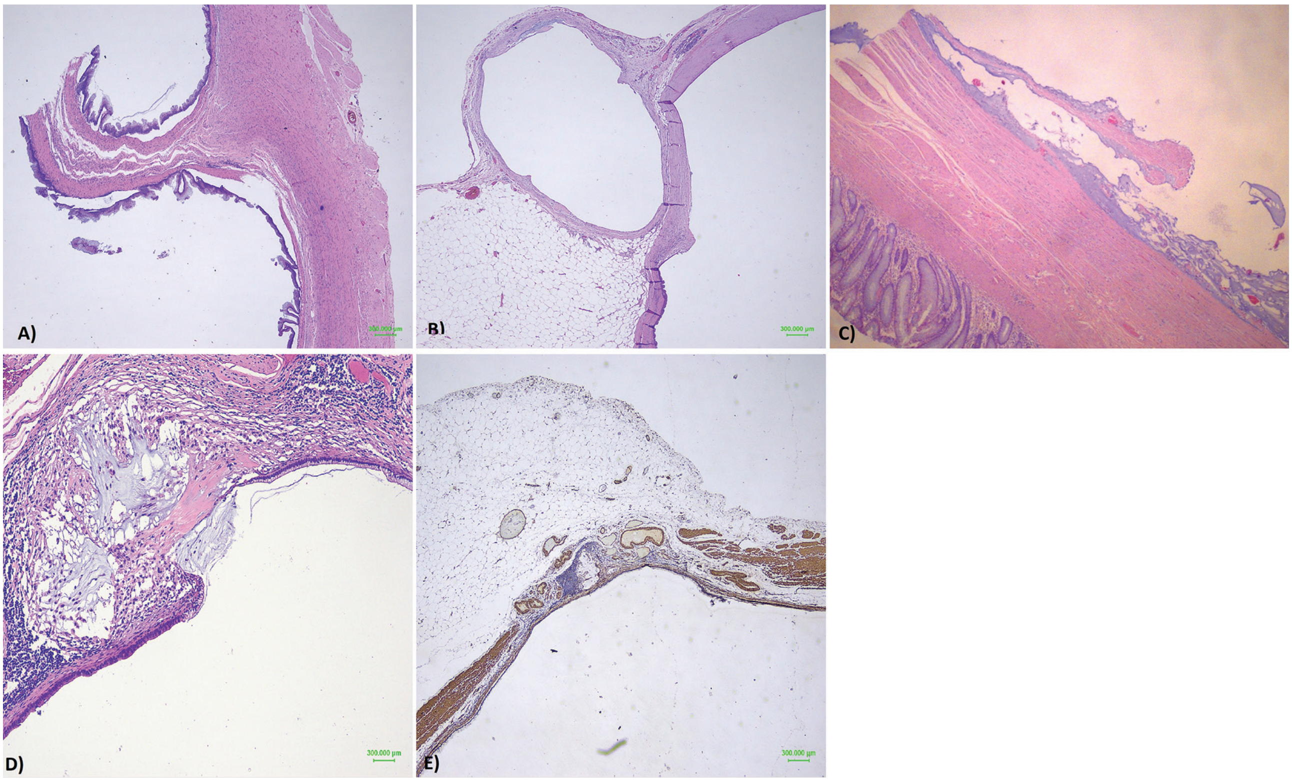

Figure 2. **A**) Flat histological pattern of mucinous epithelial cells originating from appendiceal lumen with elongated nuclei and low-grade nuclear atypia, HE ×20; **B**) Tumor invading through muscularis propria into subserosa or mesoappendix, HE ×20; **C**) Acellular mucin invading visceral peritoneum (serosa), HE ×40; **D**) Pools of acellular mucin invading muscularis propria, HE ×100; **E)** Pools of acellular mucin invading muscularis propria, SMA ×20.

disease, except for acellular mucin on the site of appendicectomy. In light of the new biopsy, a follow-up plan was made for the patient. The plan consisted of tests for tumor markers and abdominal MRI every 3 months for two years and then every year for 3 more years for the early detection of recurrence. At her one-year follow-up, the patient was found to be disease-free.[6]

Discussion

LAMN is a very rare disease. Approximately 0.4%–1% of specimens of appendicectomy are consistent with LAMN.[2] The true number of LAMN cases in the population, including asymptomatic patients, is unknown. It is more common in women.

Patients can present with mostly non-specific symptoms, including nausea, vomiting, fever, and pain in the right iliac fossa. A lot of patients are asymptomatic and the disease is either found during routine checkups or incidentally during laparotomy or laparoscopy for various diseases. Diagnosis is difficult, especially given the disease's rarity, which causes radiologists and clinical doctors to frequently overlook it in their differential diagnosis.

Moreover, due to the anatomical proximity of the appendix to the right adnexa, a LAMN can often – similarly to our case – be mislabeled in diagnostic imaging as an adnexal mass.

Abdomino-pelvic CT scans remain the gold standard diagnostic tool. In CT, an appendiceal lump >1.3 cm is often identified, with cystic dilation of the lumen and irregular thickening of the wall. Calcifications can be observed in approximately 50% of such cases. MRI is an equally good alternative imaging modality, though it is more difficult to interpret by less experienced observers. Ultrasound may be of some use, in experienced hands. In ultrasound, an appendix with cystic formation, close to the cecum could be found. An 'onion skin' or 'whipped-cream' sign has been described as a characteristic diagnostic sign, where a unilocular mass with layered hyperechoic content is present due to mural calcification.[7] Colonoscopy may sometimes be useful and show a mass protruding from the appendix in the cecum.[4,8] In case of appendiceal PMP patients eligible for CRS and HIPEC, colonoscopy should be performed preoperatively, to exclude second primaries, as it is not uncommon, due to the fact that the appendix is embryologically derived from the colon and they share similar histological structure.[6] Lastly, the high-grade appendiceal mucinous neoplasm (HAMN) should also be included in the differential diagnosis. LAMN diagnosis is based on histopathological examination in which the lesion shows a villous or flat proliferative mucinous epithelium with low-grade cytologic features, whereas HAMN has micropapillary and cribriform patterns with high-grade cytological features.[9]

Due to the rarity of this entity, treatment and follow-up guidelines have not yet been established. The decision for the proper surgical treatment depends on the dimensions and the histology of the appendiceal tumor. The treatment of LAMN with an intact base, without perforation or lymph node involvement or free surrounding tissues, or if the diameter of the appendix is less than 2 centimeters, is a simple appendicectomy with mesenteric removal. In the case of a large cystadenoma with a large base, a cecal resection is the proposed treatment and a right hemicolectomy for adenocarcinoma.[7] A cytoreductive surgery (CRS) with hyperthermic intraperitoneal chemotherapy (HIPEC) is advised when perforation of the appendix is identified. Early postoperative intraperitoneal chemotherapy (EPIC) is also another option. Some studies suggest that when the appendicectomy specimen has positive margins, a new surgery for re-excision is not necessary, as it is shown at their studies that a recurrence rate is rare to non-existent.[11] Disease progression called pseudomyxoma peritonei (PMP) is rare but very aggressive. It is possible when the wall of the appendix is perforated from the disease and mucin containing tumor cells is spread in the peritoneal cavity. It may also be perforated through surgical maneuvers.[5,10] Mortality rate of PMP is close to 40% within 10 years. If any of these conditions is present, treatment with HIPEC and CRS is recommended.[12]

As this disease entity is extremely rare, not enough research has been done. A completely resected LAMN mass without mucin outside of the appendix specimen in theory has no recurrence risk, therefore some authors suggest no post-op surveillance. Usually the proposed postoperative surveillance consists of radiographic imaging every six months for two years following appendicectomy. The aim is the early detection of any potential tumor recurrence or PMP-related complications.[9] Follow-up for a T4a LAMN, where mucin is located outside of the appendix, or a rupture has been identified, includes tumor marker and imaging follow-up for at least 5 years for early detection of recurrence. Tumor markers are not universally accepted as a helping tool for LAMN diagnosis or follow-up. The 5-year recurrence-free survival rate is 78%–95%.[1,13]

In 2020, the PSOGI/EURACAN Clinical Practice Guidelines were published, based on expert opinions and Delphi consensus. The new experts' recommendations suggest right-sided hemicolectomy in case of no perforation. In case of perforation, with peritoneal disease or not, CRS and HIPEC are recommended. These recommendations, though, are not strong.[6]

Conclusion

LAMN is a rare disease. It is more common in women. For that reason, gynecologists and those who perform gynecological ultrasound should be aware of this entity and stay diagnostically vigilant when faced with an adnexal mass of the right lower quadrant. More research should be done to conclude the best treatment and follow-up plans.

References

1. Köhler F, Reese L, Hendricks A, et al. Low-grade mucinous neoplasms (LAMN) of the appendix in Germany between 2011 and 2018: a nationwide analysis based on data provided by the German Center for Cancer Registry Data (ZfKD) at the Robert Koch Institute (RKI). Langenbeck's Archives of Surgery 2022; 407(8):3615–22. doi: 10.1007/s00423-022-02639-w
2. Gorgas EJ, Dowling SC. Low-grade appendiceal mucinous neoplasm causing equivocal appendicitis: a case report. Cureus [Internet] 2024; 16(3):e56286. doi: 10.7759/cureus.56286
3. Toffaha A, El Ansari W, Aleter A. What you see might not be what you get: discrepancies between intraoperative findings and preoperative diagnosis of ovarian tumors. Appendicular mucocele presenting as an adnexal mass - Case report and review of literature. Int J Surg Case Rep 2020; 75:543–9. doi: 10.1016/j.ijscr.2020.09.112
4. Chang HC, Kang JC, Pu TW, et al. Mucinous neoplasm of the appendix: A case report and review of literature. World J Gastrointest Surg [Internet] 2024; 16(3):944–54. doi: 10.4240/wjgs.v16.i3.944
5. Katsuyuki K, Fujiwara H, Kitagawa M, et al. Low-grade appendiceal mucinous neoplasm encountered during risk-reducing salpingo-oophorectomy: A case of laparoscopic surgery. J Obstet Gynaecol Res 2023; 49(12):2975–8. doi: 10.1111/jog.15802
6. Govaerts K, Lurvink RJ, De Hingh IHJT, et al. Appendiceal tumors and pseudomyxoma peritonei: Literature review with PSOGI/EURACAN clinical practice guidelines for diagnosis and treatment. Eur J Surg Oncol 2021; 47(1):11–35.
7. Fourie H, Al Memar M, Tuomey M, et al. The "onion skin" sign of a low-grade appendiceal mucinous neoplasm: An incidental finding during early pregnancy assessment. Australasian J Ultrasound Med [Internet] 2024; 27(2):131–5. doi: 10.1002/ajum.12377
8. Qian W, Curtain BMM, Deshwal A, et al. Massive appendiceal mucinous neoplasm diagnosed after abdominal trauma: Pitfalls in imaging and diagnosis. Radiol Case Rep [Internet] 2024; 19(5):1956–9. doi: 10.1016/j.radcr.2024.02.010
9. Priya Dharshini R, Agarwal N, Chander RV. An incidental finding of low-grade appendiceal mucinous neoplasm in a case of borderline Brenner tumor of the ovary. Cureus 2024; 16(5):e61151. doi: 10.7759/cureus.61151
10. Yao MQ, Jiang YP, Wang YY, et al. Asymptomatic low-grade appendiceal mucinous neoplasm: A case report. World J Clin Cases [Internet] 2024; 12(2):361–6. doi: 10.12998/wjcc.v12.i2.361
11. Guner M, Aydın C, Güner M. Low-grade appendiceal mucinous neoplasm: what is the best treatment? Cureus 2023;15(10). doi: 10.7759/cureus.46591
12. Mouawad C, Bardier A, Wagner M, et al. Active surveillance for low-grade appendiceal mucinous neoplasm (LAMN). Pleura and peritoneum [Internet] 2023; 9(1):31–7. doi: 10.1515/pp-2023-0032
13. Fatima K, Mushtaq A, Hasan M. Rare presentation of low-grade appendiceal mucinous neoplasms (LAMN) as an appendicular lump: A case report. Ann Med Surg 2022; 84:104848. doi: 10.1016/j.amsu.2022.104848