



A MUCINOUS NEOPLASM OF THE APPENDIX SHORT LITERATURE REVIEW AND CASE REPORT

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A MUCINOUS NEOPLASM OF THE APPENDIX. SHORT LITERATURE REVIEW AND CASE REPORT. [ENGLISH]

SUMMARY

Mucinous neoplasms of the appendix are a rare pathology with nonspecific or missing clinical appearance and typical CT findings. Occasional finding of such a process during laparotomy/laparoscopy is even rarer nowadays. The vast majority of surgeons know little about the various cases and the appropriate management for each.

We present the case of H.H, a 64-year-old female, admitted to the General Surgery Ward of MPHAT – Burgasmed after a query appendicular neoplasm, which was confirmed by the Imaging Department Team and surgically treated in our clinic. Appendectomy and atypical ablasic resection of the cecum were performed. After an uneventful postoperative period, the patient was discharged home with a recommendation for follow-up at the Complex Oncology Centre – Burgas.

KEYWORDS

Mucinous neoplasms of the appendix, appendectomy, LAMN

LITERATURE REVIEW

Mucinous neoplasms of the appendix (MNA) are rare tumours, found in approximately 0,4–1% of all the neoplasms in the GIT. According to the literature, in 0,2–0,3% of all the appendectomies, a mucinous neoplasm can be found [1].

In the USA, approximately 1500 cases per year have been reported, with a consistent rise in recent years, according to the vast majority of surgeons. Unlike CRC, this process is slower and rarely spreads outside the peritoneal cavity.

In reality, the vast majority of cases are missed during pathology testing. Because appendectomy is an effective treatment for early MNA, statistics and case descriptions are difficult to obtain. It is difficult to conduct randomized trials, which would lead to management algorithms and unified treatment strategies. As the treatment of LAMN is resection of the organ, adenocarcinoma requires more complex strategies, including surgery, HIPEC, and systemic chemotherapy.

Mucinous neoplasms of the appendix are a heterogeneous group of diseases whose therapeutic strategy remains a challenge. The malignant potential is quite different. The process may be limited to the organ or, in advanced cases, may spread to involve large mucinous masses throughout the abdominal cavity, a condition often described as Pseudomyxoma peritonei (PMP). There are many classifications, often conflicting across different aspects, based on the clinical picture, molecular criteria, or management strategies. As a result, discrepant reports and difficulties in the interpretation of various management strategies appear.

A consensus on the matter was published in the American Journal of Surgical Pathology, 2016; 40;14; however, significant contradictions in nomenclature remain.

By definition, there must NOT be an infiltrative component. In the presence of the latter, the correct term should be mucinous adenocarcinoma.

Basic characteristics:

LAMN is a low-grade, noninvasive epithelial proliferation that may cause PMP in cases of appendicular rupture.

Lesions containing high-grade nuclear dysplasia are classified as High-grade Appendiceal Mucinous Neoplasm (HAMN).

Terms:

LAMN is a lesion, developing from the appendix, containing only low-grade epithelial characteristics, in the absence of infiltrative growth.

HAMN is a lesion, developing from the appendix, containing high-grade epithelial characteristics, in the absence of infiltrative changes.

PMP is a clinical term used for obviously mucinous ascites or peritoneal mucinous masses.

Mucocele is a clinical term for a dilated appendix filled with mucus.

Cystadenoma is an outdated diagnostic term that should no longer be used.

Molecular/cytogenetic characteristics:

KRAS mutations and 5q deletions are frequently reported. [8] Various GNAS alterations are reported in 50% of cases. [9] HAMN can contain p53 or ATM mutations. [10] Microsatellite instability and BRAF mutations are not observed.

Clinical presentation: Mucinous neoplasms of the appendix can have a clinical presentation quite similar to acute appendicitis – right lower quarter abdominal pain, nausea, vomiting, and elevated inflammatory markers. A dilated appendix with a width > 15 mm is usually identified preoperatively and may have diagnostic value. A small proportion of patients report a palpable abdominal mass as their first symptom and the reason for the appointment.

Part of the cases present with typical urogenital symptoms – haematuria, pain, dysuria, and hydronephrosis on US examination. Occasionally, this type of neoplasm may be asymptomatic and detected incidentally during a prophylactic or screening examination for CRC.

In 1995, Carr et al. investigated a series of 184 patients with noncarcinoid and “tumor-like” findings in the appendix. They found that 32 % of the cases were operated for acute appendicitis and 23% were accidental findings.

Currently, we observe even fewer cases in the theatre due to improved preoperative imaging. Advanced cases of HAMN often present with chronic abdominal pain, bloating, anaemia, infertility, and inguinal or umbilical hernia as a result of dilation of the abdominal cavity, often with sudden onset.

Imaging: Abdominal ultrasound can detect cystic transformation, a dilated appendix with a porcelain wall, calcifications, and lamellated mucin, resembling onion skin. If ruptured, a wall fracture could be seen, as well as a mucous leak. Pseudomyxoma peritonei (PMP) typically presents as thickening of the peritoneum or omentum, with anechogenic areas containing echogenic spots and septa. [2]

CT can find an enlarged appendix with wall thickening, septa, calcification, and liver margin scalloping in pseudomyxoma [2,3].

MRI may find hyperintense dilation of the appendix with glittering mucous in T2, and nodular appearance in contrast series.

A biopsy is not recommended due to the risk of disease dissemination. In cases with confirmed peritoneal spread, biopsy may be helpful for diagnostic purposes and treatment planning.

Endoscopy can sometimes accidentally find mucinous lesions while screening for colorectal cancer is performed. The mucinous lesions are submucosely or externally situated, so they can be seen as a smooth impression of the lumen of the cecum due to external compression.

Differential diagnosis: LAMN must be differentiated from Neuroendocrine neoplasms (NEN) of the appendix, which are significantly more frequent (3-5 /1000 appendectomies). [4] NEN

usually affects the apex of the appendix, and because of that, it rarely gives the symptoms of acute appendicitis. Their metastatic potential is low, with distant metastases reported in 0.7% of the cases. It's possible that they spontaneously involute, because paediatric incidence is much higher. Cases with distant metastatic disease have less favourable outcomes – less than 25% 5-year survival.

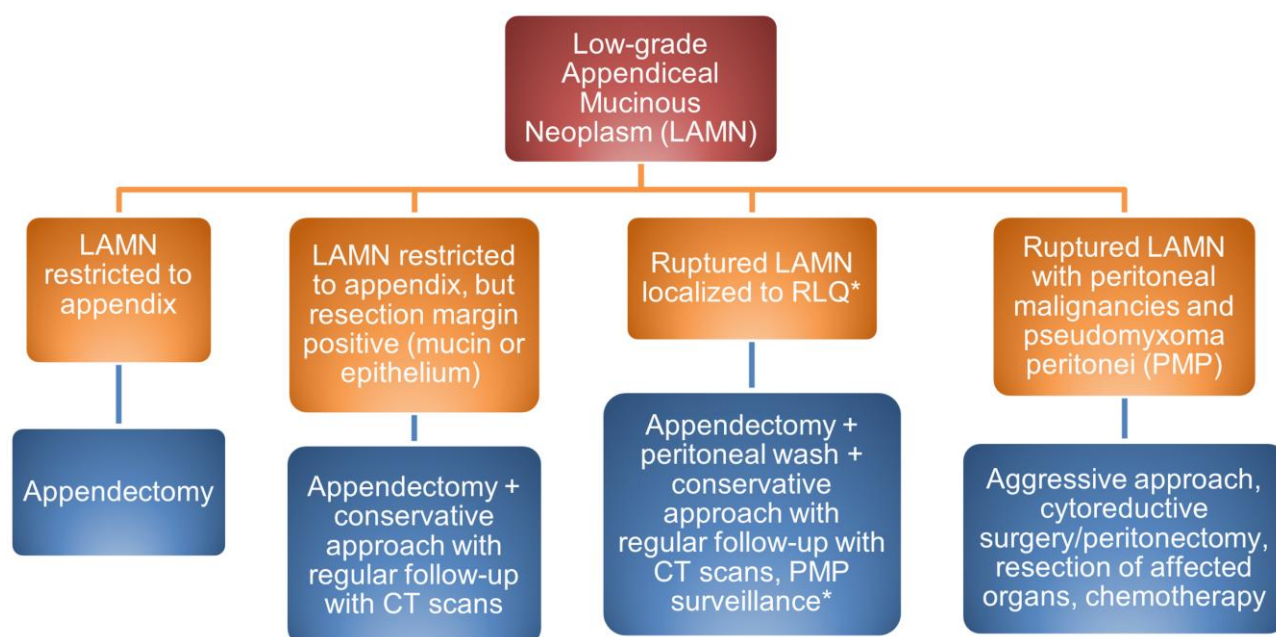
Adenocarcinoma of the appendix presents more often with clinical signs of acute appendicitis. They can provoke GI bleeding, bowel obstruction, palpable mass, and symptomatic distant metastases. Management strategies include a mandatory right hemicolectomy, as this approach yields a higher 5-year survival rate than appendectomy alone. In general, the outcome for adenocarcinoma of the appendix is worse than that of colorectal carcinoma.

Management: Appendectomy, performed conventionally or laparoscopically, is considered an effective treatment for LMNA pT3 or lower. It has been thought that open surgery carries a lower risk of perforation and of mucinous masses spreading into the peritoneal cavity. Perforation has been reported even in open-surgery cases. There is no single survey comparing different techniques. Nowadays, laparoscopic appendectomy is the treatment of choice. In Bulgaria, patients still must pay for laparoscopic surgical consumables, which limits the method's implementation nationwide. Some authors recommend a small portion of the cecum to be resected as well to perform a radical surgery. [5]

Management of cases with positive resection lines, rupture of LAMN, and spreading into the peritoneal cavity: In cases of positive resection lines with acellular mucous or the presence of atypical cells, some surgeons prefer conservative management, including annual CT for 5–10 years. The same approach can be used for localized perforation in the right lower quadrant of the abdomen. A significant point during surgery is the peritoneal lavage to destroy mucous masses. In cases of PMP and peritoneal malignant masses, a more aggressive approach is recommended, including cytoreductive surgery, excision of the peritoneum, resection of the organs affected, and chemotherapy.

Right hemicolectomy is not routinely recommended, as this approach requires retroperitoneal dissection and increases the risk of spreading the process in this particular zone. [6,7] However, some surgeons still recommend right hemicolectomy in cases of a positive resection line.

Pathology: LAMN is characterized by atypical glands, neoplastic mucinous epithelium, densely situated crypts, loose lamina propria, and widely spread villi. The process of growth is externally orientated, producing diverticula or tongue-like sprouts of the epithelium. The mucous and neoplastic epithelium can penetrate the appendix wall, allowing mucin to spread into the peritoneal cavity and PMP to develop. However, there is no data on invasive spreading and desmoplastic stromal reaction.



*No consensus for ruptured LAMN with malignant cells in RLQ – most take a conservative approach

Figure 1: Management Algorithm of LAMN

CASE PRESENTATION

We present a case of LAMN operated by our team.

History: Female patient, 64 years old, under treatment for breast cancer, operated and followed up in an Oncology department. Comorbidities include arterial hypertension, ICD – currently well managed. PET/CT shows a cyst-like mass in the right lower quarter, which does not retain isotope. The images have not changed significantly between 2 consecutive PET/CT scans performed 1 year apart. The patient was seen by a gastroenterologist and referred to our clinic for operative management.

On examination: Overall condition is fine. No significant pathology found in head and neck examination. Chest movement is equal bilaterally. Good air entry on both sides. Abdomen soft and non-tender. Palpable resilient mass in the right lower quarter. Other areas NAD. Good peripheral pulses in all four limbs; muscle tone and strength preserved.

Laboratory findings:

WCC 4.02×10^9 , Hb 120 g/l, Er 3.68×10^9 , Hct 0.361, Ca 2.5 mmol/l, Na 132 mmol/l, K 4.01 mmol/l, Cl 105 mmol/l, CRP 9, 29 mmol/l. Others were unremarkable.

CT abdo-pelvis with contrast:

Liver – normal dimensions, preserved phasecontrast characteristics. Calcified formation in the VII-th segment, 0.85 x 0.63 cm.

Pancreas –reduction of parenchyma in all three parts.

Kidneys – normal dimensions and localization. Preserved phasecontrast characteristics. No drainage alterations bilaterally. Three cysts, class I, dominant in the left kidney sinus 3.65/4.09 cm.

Retroartiac left renal vein – anatomic variation.

Appendix vermiformis – significantly dilated – 4,39 cm wide, pelvic localization.

Markedly prominent degenerative changes in the lumbar and thoracic segments of the vertebral column – osteochondrosis and spondylarthrosis. L3-L4 – protrusion of IVD, forming a median disc hernia.

Impression: The images presented here demonstrate the characteristics of low-grade appendicular mucinous neoplasm.

After appropriate preoperative management, including standard blood tests (FBC, U&E, LFTs), coagulation screen, CXR, cardiology, and anesthesiology input, the patient underwent surgery as an elective case. We proposed a laparoscopic approach, but the patient declined because of financial reasons.

We performed open surgery under general anesthesia with endotracheal intubation and muscle relaxation. After Roux laparotomy, we explored the abdomen and found an extremely dilated appendix, whitish in colour, markedly different from the normal tissue. (Figure 2 A and B)

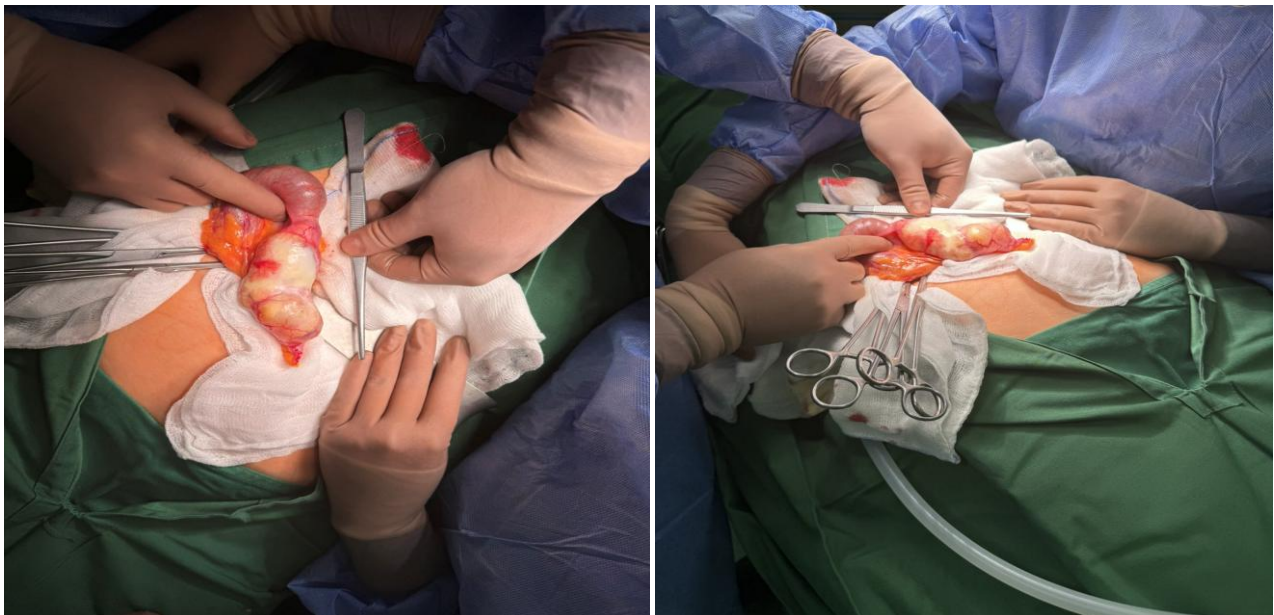


Figure 2 A and B: Intraoperative finding.

We performed an appendectomy and an atypical coecal resection. After haemostatic control of the mesoappendix and cutting the coecum 1 cm from the visible line of the process, we closed the coecal wall with double-layer running sutures. (Figure 3) We left a drain in the abdomen and closed the operative wound layer by layer. Skin closed by running suture. After an uneventful postoperative period, the patient was discharged home.

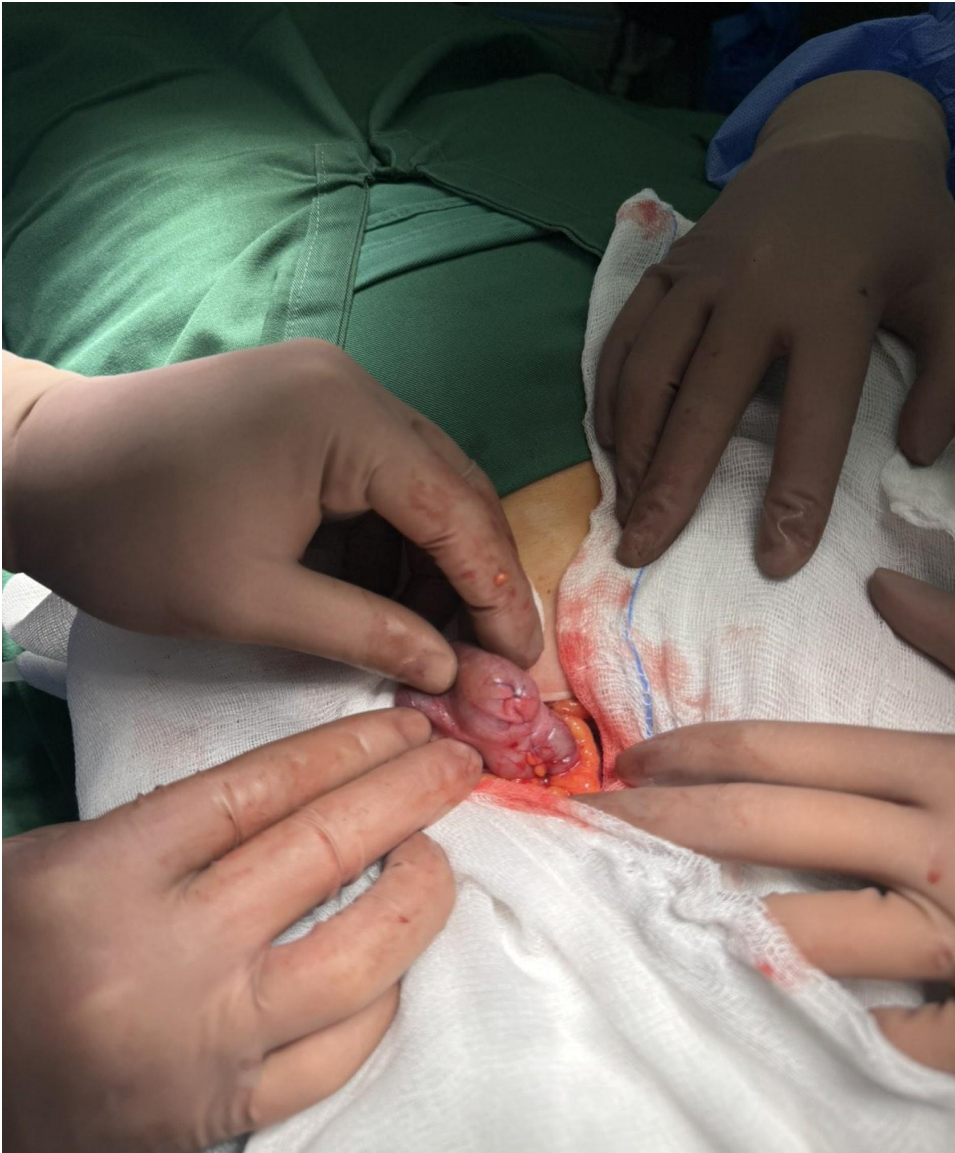


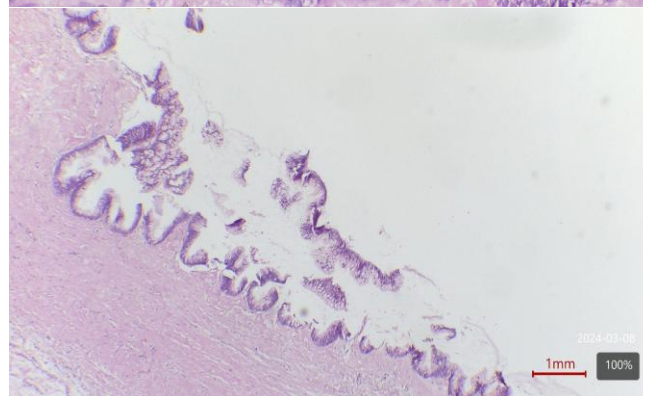
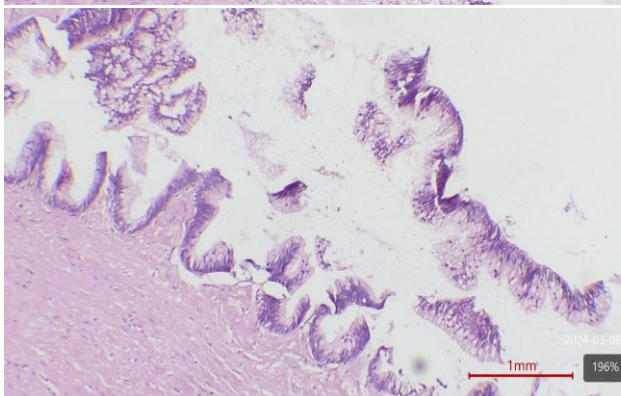
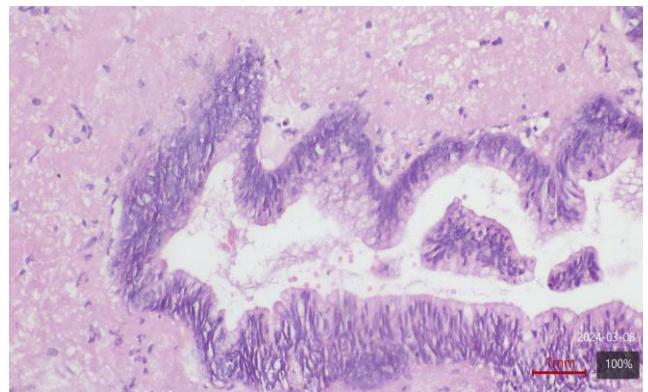
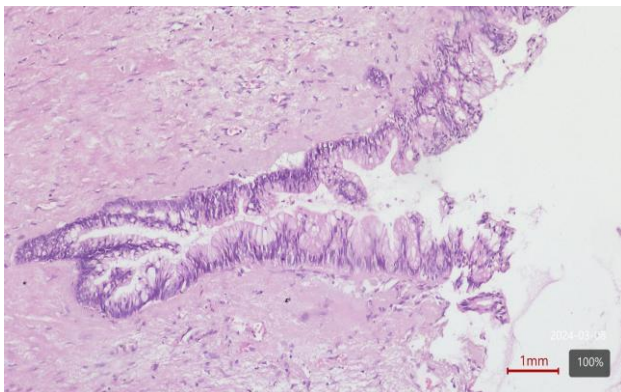
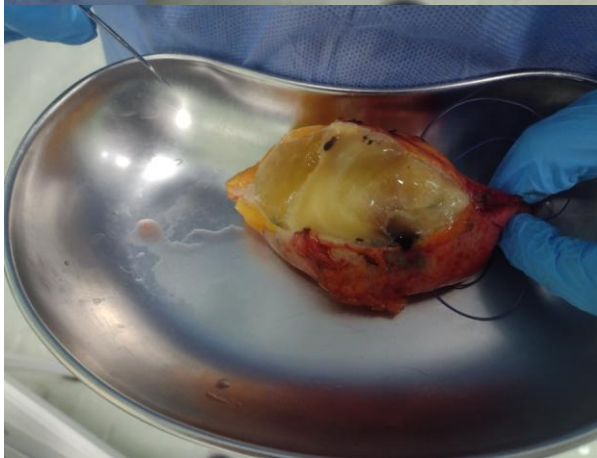
Figure 3 Closure of the appendicular stump

Pathology report: (See pictures below)

Macroscopic: Mucocelle – mucinous cyst with pappilarity.

Hystology: Lumen filled with mucous, tapered by neoplastic epithelium, ulcerated, withoutmarked stromal reaction. Finding correlates with the LAMN of the appendix.

Free resection lines.



CONCLUSION

Mucinous neoplasms of the appendix should come into consideration every time an extremely dilated appendix vermiformis is present. Special attention should be paid to cases without a typical clinical presentation of acute appendicitis. Contemporary imaging offers numerous opportunities for accurate diagnosis and management.

In cases presenting with the typical clinical appearance of acute appendicitis, contemporary pathology, genetics, and cytologic analyses provide opportunities for accurate diagnosis.

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Conflict of interest: We report no conflict of interest.

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