

CONTINUING MEDICAL EDUCATION ΣΥΝΕΧΙΖΟΜΕΝΗ ΙΑΤΡΙΚΗ ΕΚΠΑΙΔΕΥΣΗ

Surgery Quiz – Case 36

An otherwise-healthy 43-year-old female patient referred to our surgical department for investigation of an unspecified lower abdominal pain over the last 6 weeks. Upon admission, body temperature was normal and physical examination revealed no signs of peritoneal irritation. Complete blood count test, CRP and tumor markers CEA, CA19-9, CA125 were within normal range. Abdominal computed tomography (CT) revealed abnormal wall thickening of the appendiceal base and body with a diameter of 9 mm (fig. 1), abnormal dilatation of the appendiceal tip due to accumulation of fluid with a diameter of 13 mm and an intraluminal air bubble (fig. 2), absence of regional and distal lymphadenopathy, and absence of peritoneal involvement. Colonoscopy revealed a mass like protrusion at the appendiceal base without any synchronous colonic lesion. The patient considered to have a suspected appendiceal neoplasm and scheduled for surgery.

Which is the appropriate surgical procedure?

- (a) Simple appendectomy
- (b) Radical appendectomy
- (c) Right colectomy with complete mesocolic excision
- (d) Cytoreductive surgery (CRS) and hyperthermic intraperitoneal chemotherapy (HIPEC)

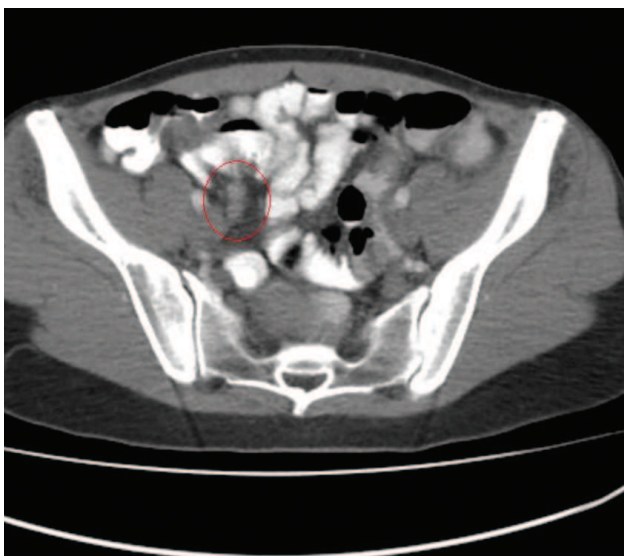


Figure 1

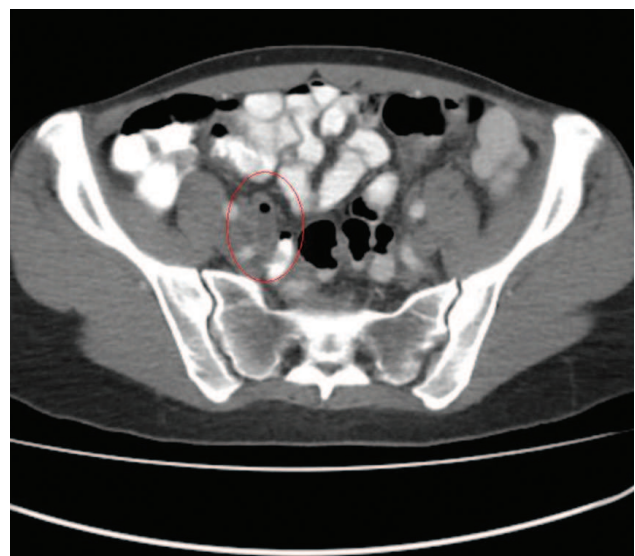


Figure 2

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ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2021, 38(6):857–859

A. Xenou,¹
K. Boulas,²
M. Nathanailidou,²
E. Kyriakidou,²
K. Sitaridis,²
I. Filippidis,²
A. Paraskeva,²
A. Triantafyllidis,²
A. Hatzigeorgiadis²

¹Department of Radiology, General Hospital of Drama, Drama

²Department of General Surgery, General Hospital of Drama, Drama, Greece

Comments

Based on preoperative assessment, an inflammatory appendiceal process ruled out. The most prominent diagnosis was a low-grade appendiceal mucinous neoplasm due to the mucocele-like formation of the appendiceal tip. As histology was not known at the time of initial surgery, the patient submitted to radical appendectomy with partial cecectomy instead of simple appendectomy or right colectomy, because: (a) dissection along tissue planes provided avoidance of appendiceal disruption and spillage of cancer cells; (b) resection of periappendiceal peritoneum and mesoappendix

lymph nodes provided valuable information for pathologic staging; (c) partial cecectomy provided negative resection margins at the appendiceal base.

Histology revealed a low-grade appendiceal mucinous neoplasm resected in negative margins. Consequently, in our patient's case radical appendectomy provided definitive treatment. In fact, radical appendectomy would have provided definitive treatment in most of the suspected appendiceal neoplasms, such as low- and high-grade appendiceal mucinous neoplasm, neuroendocrine tumors (NET) <2 cm, lymphoma, mesenchymal tumor, except from mucinous or colonic-type adenocarcinoma which would have required right colectomy. Moreover, radical appendectomy would have been the initial procedure of choice in patients with pseudomyxoma peritonei referred for interval CRS and HIPEC as initial right colectomy would not have provided any survival benefit.

Although primary and secondary appendiceal neoplasms are rare, surgeons should be familiar with appendiceal pathology as a primary neoplasm (tab. 1) is found in approximately 1% to 2% of appendectomy specimens. The most frequent initial manifestation of appendiceal neoplasms is acute appendicitis seen in 30–50% of patients. Acute appendicitis occurs more commonly in patients with NETs than epithelial neoplasms, as 25% of NETs develop at the appendiceal base. Epithelial neoplasms occur most frequently in middle-aged and older adults, while NETs occur more common in younger patients.

According to the World Health Organization (WHO), among primary appendiceal neoplasms, epithelial neoplasms are the most common. Epithelial mucinous neoplasms represent 37% of appendiceal neoplasms, including (a) adenoma; (b) low-grade appendiceal mucinous neoplasm (LAMN), corresponding to the former used terms mucocele and mucinous cystadenoma; (c) high-grade appendiceal mucinous neoplasm (HANM); (d) mucinous adenocarcinoma (low, moderate, poorly differentiated); (e) low- and

high-grade pseudomyxoma peritonei (PSP), corresponding to the former used terms disseminated peritoneal adenomucinosis (DPAM) and peritoneal mucinous carcinomatosis (PMCA), respectively. Epithelial nonmucinous (colonic-type) adenocarcinoma, goblet cell adenocarcinoma and malignant NET represent 27%, 11% and 19% of appendiceal neoplasms, respectively.

Epithelial neoplasms are more often detected at imaging than NETs due to their larger size and propensity for peritoneal spread and metastatic disease. Characteristic imaging features of mucinous epithelial neoplasms include appendiceal mucocele, periappendiceal or localized mucinous deposits, peritoneal dissemination resulting in PSP. Characteristic imaging features of nonmucinous epithelial neoplasms include focal soft-tissue mass or soft-tissue infiltration of the entire appendix, absence of mucocele formation, periappendiceal fat stranding, regional and distant lymphadenopathy, and metastatic disease. Appendiceal NETs are often difficult to visualize due to their small size, with a mean diameter of <1 cm. The lesions typically manifest as an avidly enhancing nodular wall thickening with microcalcifications mimicking appendicoliths.

According to 2019 American Society of Colon and Rectal Surgeons (ASCRS) clinical practice guidelines, treatment of epithelial appendiceal neoplasms depend on histologic type and extent of disease. Patients with LAMN and HAMN without peritoneal spread are optimally treated with appendectomy alone when resected in negative margins without perforation. Patients with mucinous and nonmucinous appendiceal adenocarcinoma without peritoneal spread are optimally treated with right colectomy, which allows complete staging and has therapeutic benefit due to the high risk of nodal metastasis which ranges from 20% to 67%, especially in the nonmucinous adenocarcinoma. In patients with mucinous appendiceal adenocarcinoma and peritoneal spread, routine right colectomy is not recommended, as several

Table 1. Primary appendiceal neoplasms.

| Primary appendiceal neoplasms | Cells types | Subtypes | Neoplasms |
|-------------------------------|------------------------|-------------|--|
| Epithelial neoplasm | Enterocyte | Mucinous | Low-grade appendiceal mucinous neoplasm |
| | | | High-grade appendiceal mucinous neoplasm |
| | | | Mucinous adenocarcinoma |
| | | Nonmucinous | Adenoma |
| | | | Colonic-type adenocarcinoma |
| | | | Goble cell carcinoma |
| Neuroendocrine tumor (NET) | Enterochromaffin cells | | NET |
| Lymphoma | Lymphoid tissue | | Lymphoma |
| Mesenchymal tumor | Mesenchymal cells | | GIST |
| | | | Leiomyoma |
| | | | Leiomyosarcoma |

GIST: Gastrointestinal stromal tumor

studies failed to demonstrate survival benefit of right colectomy over appendectomy alone in patients subsequently undergoing CRS and HIPEC.

For nonmetastatic appendiceal NETs, treatment is based on size and histological characteristics. Tumors <1 cm and without unfavorable histological characteristics, are optimally treated with appendectomy. Tumors >2 cm are optimally treated with right colectomy because the reported risk of nodal metastasis is as high as 40%. Tumors between 1 and 2 cm have an intermediate risk of nodal metastasis. However, the largest clinical series found no nodal disease in tumors <2 cm and under this threshold some authors propose appendectomy alone. In addition to size, histological characteristics influence surgical strategy. Findings on histology that may be unfavorable include mesoappendiceal invasion >3 mm, advanced grade consisting of mitotic index >2 mitoses per high-power field, Ki-67 index >3% and lymphatic or vascular invasion.

References

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Corresponding author:

K. Boulas, Department of General Surgery, General Hospital of Drama, 661 00 Drama, Greece
e-mail: boulaskonstantinos@gmail.com