

LAMN/HAMN: Behavior and Histology

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Outline

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TURNING CANCER DATA
INTO DISCOVERY

- LAMN/HAMN Terminology
- LAMN Definition, background and behavior coding
- HAMN Definition, background, and behavior coding
- Solid Tumor Rules

LAMN/HAMN Terminology



- Appendiceal mucocele: A distended, mucus-filled appendix.
 This term is ambiguous and best utilized to describe an imaging appearance rather than pathologic entity.
- Acellular mucin: Mucin *without* tumor cells. Low risk for recurrence or progression which occurs in about 4% of cases.
- Cellular mucin: Mucin with tumor cells. High risk for recurrence or dissemination which occurs in 33-75% of cases.

LAMN versus HAMN



- High-grade appendiceal mucinous neoplasm (HAMN) are rare while low-grade appendiceal mucinous neoplasms are somewhat more common
- LAMN is often an incidental finding during appendectomy and not readily apparent in the organ
- 5th Ed WHO GI book and C.A.P agree these neoplasms are staged as Tis when there is no evidence of invasion through the muscularis propria into submucosa
- Grade (low versus high) is based on KRAS and GNAS mutations

Low Grade Appendiceal Mucinous Neoplasm: LAMN



- LAMN has a has a behavior of /2. They are slow-growing neoplasms which have the potential for peritoneal spread and can result in patient death. LAMNs demonstrate an interesting biology in that they do not have hematogenous dissemination risk, but risk for **appendiceal perforation**, which can result in peritoneal dissemination, repeated recurrences after surgery and even death.
- /2 = Tis(LAMN) confined by muscularis propria (T1-T2 are not used for LAMN), and such lesions are designated as Tis
- /3 = T3-T4 extending into subserosa or serosa

High Grade Appendiceal Mucinous Neoplasm: HAMN



- Rare, complex type of epithelial appendiceal cancer. It is a mucinous tumor that shows high-grade cytologic atypia. It does not have infiltrative growth, but rather a pushing-type invasion.
- Shows histological features like those of LAMN
- Neoplasms with high-grade nuclear dysplasia are termed HAMN
- Histological features like those of LAMN, with the addition of micropapillary features, cribriforming, piling up of epithelial cells with high-grade features
- Because of limited data, HAMN is staged like mucinous adenocarcinoma

Behavior in a Nutshell



- Both LAMN and HAMN can have a behavior of /2 based on staging and pathologic description of invasion
- Both LAMN and HAMN can have a behavior of /3 based on staging and pathologic description of invasion
- A diagnosis of LAMN or HAMN does not require the tumor be comprised of greater than 50% mucinous to be coded 8480.
- While LAMN is a slow growing neoplasm, it can be a stage T3 or T4 at diagnosis
- LAMN may also present with distant disease at time of diagnosis or later date.
- LAMN and HAMN are treated with surgical reduction with addition of hyperthermic intraperitoneal chemotherapy (HIPEC)

Solid Tumor Colon Rule H5: 1/1/2022 forward



H5 Code low grade appendiceal mucinous neoplasm (LAMN) and high grade appendiceal mucinous neoplasm (HAMN) 8480/2 when:

- Diagnosis date is 1/1/2022 forward AND
- Behavior is stated to be in situ/non-invasive OR
- Behavior is not indicated

Note 1: ICD-O-3.2 lists LAMN with behavior of /1. WHO 5th Ed Digestive Systems Tumors indicates this neoplasm is considered in situ. After consulting with WHO Digestive System editors, College of American pathologists, and AJCC GI chapter experts, the standard setting organizations have agreed LAMN should be collected and should be assigned a behavior code of /2 beginning with cases diagnosed 1/1/2022 forward.

Note 2: A diagnosis of LAMN or HAMN does not require the tumor be comprised of greater than 50% mucinous in order to be coded 8480.

Note 3: If the pathologist indicates LAMN or HAMN is invasive or has a malignant behavior, continue through the rules.

Solid Tumor Colon Rule H6: 1/1/2022 Forward

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• Rule H6 Code invasive mucinous adenocarcinoma 8480 when the diagnosis is any of the following:

Note 1:

- Exactly "mucinous adenocarcinoma" (no modifiers)
- High grade appendiceal mucinous neoplasm (HAMN) stated to be invasive (DX 1/1/2022 forward)
- High-grade pseudomyxoma peritonei
- **Invasive** pseudomyxoma peritonei
- Low grade appendiceal mucinous neoplasm (LAMN) stated to be invasive (DX 1/1/2022 forward)
- Malignant pseudomyxoma peritonei
- Two histologies and mucinous is documented to be **greater than 50%** of the tumor o Mucinous carcinoma must meet a percentage requirement to be coded. Do not use majority of tumor, predominantly, or predominant part of the tumor to code mucinous 8480.
- **Note 2:** Report the appendiceal mucinous neoplasm as malignant /3 using the ICD-O matrix principle when the **pathology** from the appendix is **low-grade mucinous** neoplasm (not reportable prior to 1/1/2022) **AND**
 - The pseudomyxoma peritonei are **high-grade**/invasive/malignant **OR**
 - Patient is treated for malignant pseudomyxoma peritonei OR
 - The diagnosis is LAMN, and the physician states it is malignant **OR**
 - The diagnosis is HAMN, and the physician states it is malignant OR
 - The pathologist staged the tumor as T3 or T4

Questions





Thank You



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