Included in this Additional Handout are:

• LAMN/HAMN Scenarios for reference during the presentation

Please have the Presentation Slides (PDF) open during the presentation. We have learned from prior years that the display of the online slides may be blurry depending on connectivity and bandwidth issues. Displaying the pdf of the slides concurrently gives you better visual accessibility.

LAMN/HAMN 01 Scenario

2023 SEER Workshop Cases

Abstracted Text

Physical Exam

06/01/2023 - HPI: Pt w/ hx of sigmoid ca recently found to have a dilated, fluid-filled appendix on follow-up imaging. Abd is non-distended and non-tender. Remainder of PE negative for any pertinent findings. IMP: Appendiceal swelling on CT, non-PET avid. Plan: Resection (hemicolectomy vs. appendectomy).

Scans

04/01/2023 - CT Chest/Abd/Pelvis: IMP: Fluid-filled, dilated, probable mucocele vs. mass. Adenocarcinoma cannot be excluded. Remainder of abd and pelvis negative. Chest unremarkable.

04/04/2023 - MRI Pelvis: IMP: Redemonstrated dilated fluid-filled distal appendix, may be mucocele, but underlying associated neoplasm is high on the differential.

04/15/2023 - PET-CT (Skull base to mid-thigh): IMP: No hypermetabolism associated w/ tubular, fluid-filled structure in the Rt abd. No enlarged or hypermetabolic LNs.

Labs

04/15/2023 - CEA 21.9 ng/mL (0.0 - 3.0 ng/mL normal).

Operative Reports

06/15/2023 - Laparoscopic Rt hemicolectomy: Findings: On lap a suspicious dilated appendix w/ obvious large mass found. Decision made to proceed w/ hemicolectomy given suspicious mass. No evidence of enlarged LNs or disseminated disease.

Treatment Plan

07/12/2023 - GI Tumor Board Note: Pt w/ LAMN s/p complete resection w/ Rt hemicolectomy. Case discussed and there was no recommendation for chemotherapy for this LAMN. Recommend observation only.

Stage

07/12/2023 - GI Tumor Board Note: pT4a pN0 cM0, Stg 2B LAMN.

06/15/2023 - Path Report #1

FINAL DIAGNOSIS

A. Right colon, terminal ileum and appendix, right colectomy:

Low-grade appendiceal mucinous neoplasm (LAMN), see Synoptic Staging Summary below.

Uninvolved colonic, small intestinal and omental tissue with no significant histopathologic abnormality.

Thirty-one pericolic lymph nodes, negative for metastatic tumor (0/31).

SYNOPTIC STAGING SUMMARY

Specimen: Procedure: Appendectomy and right colectomy
Tumor Site: Distal half of appendix
Histologic Type: Low-grade appendiceal mucinous neoplasm (LAMN)
Histologic Grade: G1, well differentiated
Tumor Size: 5 x 2.4 x 2.4 Centimeters (cm)
Tumor Deposits: Not identified
Tumor Extent: Acellular mucin invades visceral peritoneum (serosa)
Lymphovascular Invasion: Not identified
Perineural Invasion: Not identified
Margins: All margins negative for tumor and mucin
Regional Lymph Node Status: All regional lymph nodes negative for tumor
Number of Lymph Nodes Examined: 31

Pathologic Stage Classification (pTNM, AJCC) pT Category: pT4a pN Category: pN0

Additional Findings: Uninvolved colonic, small intestinal and omental tissue with no significant histopathologic abnormality. Special Studies:

Biomarkers (including Mismatch Repair Proteins and Microsatellite Instability): Not performed

CLINICAL HISTORY

Appendiceal mass

GROSS DESCRIPTION

A. Received in formalin, labeled with patient's name and "A. Right colon and appendix", is a hemicolectomy specimen, consisting of a 4 cm long by 4 cm circumference terminal ileum, contiguous with a 22 cm long by 9-15 cm range in circumference cecum and colon with an attached 7.5 cm long by 0.5-2.8 cm range in diameter intact vermiform appendix. The appendiceal serosa is covered in a marked amount of dusky fibrous adhesions (inked green). The distal 5 cm of the appendix has a markedly dilated lumen up to 2.4 cm diameter and is filled with opaque mucinous material. It is 1.5 cm from the radial margin (inked orange). The remainder of the segment has tan mucosa with a normal folding pattern and a uniform wall thickness of 0.2 cm. The remainder of the serosa is smooth with scant dusky fibrous adhesions and a moderate amount of attached pericolic adipose tissue. A 12 x 6.5 x 2.5 cm portion of yellow-tan lobulated omentum is also attached to the specimen. Also received is a container is a 2.5 cm diameter by 1.2 cm long annular portion of colon and attached soft tissue.

Representative sections are submitted as follows:

- A1: Margins, perpendicular; radial margin, en face.
- A2: Appendiceal orifice, entirely submitted.

A3-A13: Appendix, entirely submitted from proximal to distal (A13-distal tip longitudinally trisected).

- A14: Omentum.
- A15: Six possible lymph nodes.
- A16: Six possible lymph nodes.
- A17: Six possible lymph nodes.
- A18: Six possible lymph nodes.
- A19: Six possible lymph nodes.
- A20: Three possible lymph nodes.

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LAMN/HAMN 02 Scenario

2023 SEER Workshop Cases

Abstracted Text

Physical Exam

03/01/2023 - cc: RLQ pain x 1 wk. Pt seen at outside ED for suspected appendicitis, but imaging showed appendiceal mucocele vs. mass and pt was referred here. PE: Abd tender to palpation, but otherwise unremarkable PE. IMP: Appendix mucocele vs. neoplasm. Plan: Surgical rsxn.

Scans

02/17/2023 - PTA CT Abd/Pelvis: IMP: Dilated, fluid-filled appendix measuring approx. 2 cm without inflammation, extending towards base of cecum. Appendix concerning for mucocele or possible low-grade neoplasm. No enlarged mesenteric LNs.

Labs

03/28/2023 - CEA: 1.8 ng/mL (0.0 - 3.0 ng/mL normal).

Operative Reports

03/04/2023 - Laparoscopic appendectomy: Findings: Appendix was quite dilated, firm, appeared slightly inflamed and filled w/ mucin. No other suspicious findings noted and an uncomplicated appendectomy performed.

Treatment Plan

03/28/2023 - Surg Note: Lap appendectomy proved HAMN with clear margins. Reviewed pathology and outcomes for HAMN w/ pt and spouse. Pt requires no additional surgery or systemic therapy for this appendiceal neoplasm.

03/04/2023 - Path Report #1

FINAL DIAGNOSIS

Appendix (laparoscopic appendectomy):

High-grade appendiceal mucinous neoplasm (HAMN), with no evidence of invasive carcinoma and negative margin (see comment and synoptic staging summary below).

COMMENT

Given the negative margin and lack of mucin extravasation, this pTis neoplasm is considered adequately treated by appendectomy.

SYNOPTIC STAGING SUMMARY

Procedure: Appendectomy Tumor Site: Diffusely involving appendix Histologic Type: High-grade appendiceal mucinous neoplasm Histologic Grade: G2, moderately differentiated Tumor Size: At least 8 cm. Tumor Deposits: Not identified 11/8/23, 7:21 AM

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Tumor Extension: Mucin extends into but not through muscularis propria Lymphovascular Invasion: Not identified Margin Status for Invasive Carcinoma: Not applicable Margin Status for Non-Invasive Tumor and Mucin: All margins negative for non-invasive tumor and mucin Margin Comment: HAMN is estimated to be > 0.5 cm from the resection margin. Regional Lymph Node Status: Not applicable (no regional lymph nodes submitted or found)

Pathologic Stage Classification (pTNM, AJCC) pT Category: pTis pN Category: pN not assigned (no nodes submitted or found) Comments: This represents pT category: pTis (HAMN).

CLINICAL HISTORY

Mucocele of appendix

GROSS DESCRIPTION

A. Received in formalin labeled with patient's name and "A. Appendix". Size/Integrity: Intact and 9 cm long by 1-1.3 cm in diameter. Appearance: Serosa-pink gray and smooth; lumen-focally dilated, 0.2-0.7 cm, filled with a light opaque mucoid-like fluid and fecal material; wall-tan, 0.3-0.5 cm and slightly fibrotic. Perforation: Absent. Additional findings: None.

Summary of sections:

A1-A2. Representative sections with inked margin en face and bisected tip. Remainder of appendix is submitted in A3-A10.

MICROSCOPIC DESCRIPTION

Sections examined. The initial slides, A1-A2, shows at least a low-grade appendiceal mucinous neoplasm, which is not identified at the resection margin (slide A1). For complete histologic assessment and to exclude an invasive carcinoma, the remainder of the appendix is submitted for histologic evaluation.

These additional slides, A3 - A10, are reviewed and show no evidence of invasive carcinoma. In some areas the mucosa shows features reminiscent of sessile serrated polyp, while other areas (slides A2 and A10) show higher grade nuclear features including micropapillary features. No definitive cribriform architectural pattern is identified. The overall size of this noninvasive neoplasm is difficult to determine but likely measures up to 8 cm as it appears to involve the majority of the appendix. The resection margin is uninvolved, and the neoplasm is estimated to measure greater than 5mm from this margin.

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LAMN/HAMN 03 Scenario

2023 SEER Workshop Cases

Abstracted Text

Physical Exam

03/01/2023 - HPI: Pt had a witnessed fall and taken to outside urgent care. Pt complained of Rt sided pain and PTA U/S incidentally found a distended appendix w/ cystic structure (images not available for review). Referred here due to PTA concern for appendicitis. PE: Rt lower abd tender to palpation. No hepatomegaly. Rest of PE negative. IMP: Appendicitis vs. mucocele. Plan: Imaging and surgery.

Scans

03/01/2023 - CT Chest/Abd/Pelvis: IMP: 5.4 cm rim enhancing cystic lesion in region of appendix and cecum likely represents a ruptured appendix mucocele. Diffuse ascites which could be pseudomyxoma peritonei w/ possible peritoneal carcinomatosis. No evidence of adenopathy. Remainder of imaging negative.

Labs

03/04/2023 - CEA: 9.26 ng/mL (0.0 - 5.0 ng/mL normal).

Operative Reports

03/12/2023 - Exploratory lap, Rt hemicolectomy and distal ileectomy, extensive cytoreduction of peritoneal implants, omentectomy, cholecystectomy, splenectomy, partial peritonectomy: Findings: Presumed ruptured appendix w/ ascites and omental tumors. Large mucinous ascites and extensive peritoneal studding throughout abd and pelvis, including in Lt lower quadrant, perihepatic, and subdiaphragmatic areas. Large appendiceal tumor replacing appendix and possibly involving cecum. Serosal involvement of spleen requiring splenectomy. Pt cytoreduced and at completion of cytoreduction, score estimated as 1 with small residual disease in pelvis and very minimal abdominal disease remained, predominantly on small bowel.

Treatment Plan

04/14/2023 - Surg Note: Pt s/p cytoreduction for pseudomyxoma peritonei. Discussed potential for recurrence w/ pt and recommend close follow-up. Plan to see pt back in 3 months for imaging.

Stage

04/01/2023 - Staging Form: pT4a pN0 pM1b, Stg IVA

03/12/2023 - Path Report #1

FINAL DIAGNOSIS

A. Abdominal peritoneum, excision:

- Mucin with inflammatory cells ("acellular") involving the peritoneum
- Negative for neoplastic epithelium.

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- B. Abdominal peritoneum nodules, excisions:
- Mucin with fibrosis and inflammatory cells ("acellular").
- Negative for neoplastic epithelium.
- C. Abdominal peritoneum falciform, excision:
- Mucin with inflammatory cells ("acellular") involving the peritoneum.
- Negative for neoplastic epithelium.
- D. Abdominal peritoneum appendices epiploicae, excision:
- Mucin with inflammatory cells ("acellular") involving the peritoneum.
- Negative for neoplastic epithelium.

E. Right colon and ileum, right hemicolectomy and distal ileectomy:

Low-grade appendiceal mucinous neoplasm (LAMN) with the following features:

- Transmural extension of mucin with fibrosis and inflammatory cells ("acellular") onto the appendiceal serosa.
- 10 ileocolic lymph nodes negative for neoplasia (0/10).
- See synoptic summary below.

F. Omentum, omentectomy:

- Metastatic low-grade appendiceal mucinous neoplasm (LAMN), with neoplastic epithelium within pools of mucin.

G. Lesser omentum, excision:

- Metastatic low-grade appendiceal mucinous neoplasm (LAMN), with neoplastic epithelium within pools of mucin.
- H. Pelvic peritoneum, excision:
- Mucin with fibrosis and inflammatory cells ("acellular").
- Negative for neoplastic epithelium.
- I. Right upper quadrant peritoneum, excision:
- Mucin with fibrosis and inflammatory cells ("acellular").
- Negative for neoplastic epithelium.
- J. Gallbladder, cholecystectomy:
- Serosal involvement by mucin with fibrosis and inflammatory cells ("acellular").
- Negative for neoplastic epithelium.
- Gallbladder with no other pathologic alterations.

K. Liver, cyst wall, excision:

- Cyst wall serosa involved by mucin with fibrosis and inflammatory cells ("acellular").
- Negative for neoplastic epithelium.
- L. Spleen, splenectomy:

- Metastatic low-grade appendiceal mucinous neoplasm (LAMN), with neoplastic epithelium within pools of mucin, involving splenic serosa without splenic parenchymal involvement.

SYNOPTIC STAGING SUMMARY

Specimen: Right colectomy and distal ileectomy Tumor Tumor Site: Diffusely involving appendix Histologic Type: Low-grade appendiceal mucinous neoplasm Histologic Grade: G1, well differentiated Tumor Size: 5.5 Centimeters (cm) Tumor Extent: Acellular mucin invades visceral peritoneum (serosa) Lymphovascular Invasion: Not identified Margins Margin Status for Invasive Carcinoma: Not applicable 11/8/23, 7:21 AM about.blank
Margin Status for Non-Invasive Tumor: All margins negative for non-invasive tumor
Regional Lymph Nodes
Regional Lymph Node Status: All regional lymph nodes negative for tumor
Number of Lymph Nodes Examined: 10
Distant Metastasis
Distant Site(s) Involved: Intraperitoneal acellular mucin without identifiable tumor cells in the disseminated peritoneal
mucinous deposits; Intraperitoneal metastasis (including peritoneal mucinous deposits containing tumor cells): Omentum,
lesser omentum, spleen
Pathologic Stage Classification (pTNM, AJCC)
pT Category: pT4a
pN Category: pN0
pM Category: pM1b

CLINICAL HISTORY

New diagnosis of appendiceal mucinous neoplasm.

GROSS DESCRIPTION

A. Received in formalin, labeled "abdominal peritoneum", is a 6.5 x 4.5 x 1.5 cm portion yellow lobulated adipose tissue with a red, focally hemorrhagic external surface covering approximately 50% of specimen. Specimen is serially sectioned to reveal a homogenous yellow lobulated cut surface. Multiple 0.1-0.3 cm white ill-defined nodules are grossly identified. Representative sections submitted in A1.

B. Received in formalin, labeled "abdominal peritoneum nodules", are multiple, 0.7 x 0.5 x 0.5-3.9 x 2.5 x 2.5 cm pink-brown to focally dusky gelatinous, soft nodular tissue fragments with focal hemorrhagic areas on the external surfaces. Sectioning reveals brown-yellow, mucinous cut surfaces intermixed with 0.1 and 0.3 cm tan-white, nodules. Representative sections submitted in B1-B2.

C. Received in formalin, labeled "abdominal peritoneum falciform", are three, $3.5 \times 2.5 \times 2.5$ and $13.\times 9 \times 2$ cm portions of yellow lobulated adipose tissue with finely encapsulated surfaces remarkable for multiple petechial hemorrhagic areas. Sectioning reveals homogenous yellow lobulated cut surfaces with a singular $1.5 \times 1.0 \times 0.6$ cm white, firm nodule. Representative sections submitted in C1-C2.

D. Received in formalin, labeled "abdominal peritoneum appendices epiploicae", is a 16 x 10.5 x 6 cm aggregate of yellow lobulated adipose tissue with a single yellow lobulated to red hemorrhagic and lobulated external surface. It is serially sectioned to reveal a uniformly yellow lobulated cut surface with multiple tan-brown soft, mucinous nodules in the external surface measuring from 0.1-0.6 cm. Representative sections submitted in D1-D2.

E. Received in formalin, labeled "right colon and ileum", is a right hemicolectomy specimen consisting of: terminal ileum (85 x 3.5-4.5 cm in internal circumference), colon spanning from cecum to hepatic flexure (32 cm long by 7-10 cm internal circumference), appendix (7.5 x $3.2 \times 2.5 \text{ cm}$), scant omentum (5 x 5.5 cm), extensive attached pericolonic fat and mesentery. The serosal surface is tan-brown hemorrhagic and roughened with numerous adhesions and multiple mucinous tan-white gelatinous nodules on the small and large bowel serosa ranging in size from $0.2 \times 0.2 \times 0.2-9.0 \times 6.0 \times 3.5 \text{ cm}$ which grossly come within 0.1 cm of the colonic wall that do not grossly invade. Opening the specimen reveals a $5.5 \times 3.5 \times 2.5 \text{ cm}$ red-brown, mucinous friable mass within the appendix invading through the appendiceal wall at the tip extending to the serosal surface (inked blue), 1.0 cm from the nearest mesenteric resection margin (green), 34.5 cm from the distal resection margin (inked black and 86.5 cm from the proximal resection margin (inked blue). The remaining appendix is filled with a mucinous and tan yellow purulent material. The remaining colon has a uniformly 0.3 cm thick colonic wall and a rugose, tan-brown mucosa with focal attenuated areas and is remarkable for multiple intact diverticula and a single bosselated superficial polyp, 10.0 cm from the distal resection margin. The ileal mucosa is tan-green and mostly unremarkable with no additional masses or lesions identified. A regional lymph node search is within 15 cm of the appendix to reveal thirteen possible lymph nodes ranging from 0.3-1.0 cm.

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Representative sections are submitted as follows:

E1: Proximal, distal, mesenteric margins, perpendicular.

E2-E5: Appendix to include deepest depth of invasion in E4-E5 and appendiceal orifice (inked yellow) in E2.

E6-E7: Largest serosal nodule attachment site.

E8: Representative largest ileal nodule.

E9: Representative additional colonic and ileal nodules.

E10: Colonic polyp, entirely submitted and representative diverticula.

E11: Unremarkable ileal mucosa to include ileocecal valve.

E12: Unremarkable colonic mucosa.

E13-E14: Five possible lymph nodes in each cassette.

E15: Three possible lymph nodes.

F. Received in formalin, labeled "omentum", is a 55 x 10.5 x 4.5 cm portion of omentum. The external surface is yellow and lobulated with multiple $0.7 \times 0.7 \times 0.7 \times 0.7 = 3.8 \times 1.5$ cm brown-tan mucinous nodules. Representative sections submitted in F1.

G. Received in formalin, labeled "lesser omentum", is a $4.5 \times 4.5 \times 1.5$ cm portion of yellow lobulated adipose tissue with a single, $1.5 \times 1.5 \times 1.5$ cm white-tan, mucinous nodule. The cut surface is yellow lobulated and homogenous. Representative sections submitted in G1.

H. Received in formalin, labeled "pelvic peritoneum", is a $19 \times 11 \times 4$ cm portion of yellow lobulated adipose tissue with a finely encapsulated dusky surface. Specimen is serially sectioned to reveal multiple $1 \times 1 \times 1-5 \times 2 \times 1.7$ cm tan-brown, mucinous nodules. Representative sections submitted in H1.

I. Received in formalin, labeled "right upper quadrant peritoneum", is a 26 x 19 x 1.5 cm portion of peritonealized tissue with attached yellow lobulated adipose tissue. Peritoneal surface is hemorrhagic with loosely adherent, clotted blood. Sectioning reveals an unremarkable yellow lobulated cut surface. No masses or lesions are identified. Representative sections are submitted in I1.

J. Received in formalin labeled "gallbladder", Size/Integrity: 9.8 x 3.5 x 3.5 cm, intact. Serosa, Wall thickness, Mucosa: Serosal surface is purple-green, ragged with no lesions or masses identified. The wall thickness ranges from 0.3-0.2 cm and the mucosal surface is green-velvety to black-velvety. No calculi. No additional findings. Summary of sections: J1. Inked cystic duct and representative gallbladder wall.

K. Received in formalin, labeled "liver cyst wall", is a 2.0 x 1.2 x 0.9 cm portion of unoriented, white-red membranous tissue. Specimen is serially sectioned and entirely submitted in K1.

L. Received fresh, labeled "L: Spleen", is a 110 g, 9 x 7.5 x 4 cm spleen, with a moderate amount of attached hilar adipose tissue. The splenic capsule is mottled pale-tan-red, with dense fibrous adhesions, minimal focal possible hemorrhage, and areas of ill-defined mucinous nodular tissue. A 2.5 cm capsular mucinous nodule is identified. The remaining cut surface of the spleen is unremarkable.

Representative sections are submitted as follows:

- L1: Suspected vascular margins, en face, and longitudinal section of hilum.
- L2: Capsular mucinous nodular lesion.
- L3: Additional capsular tumor implants.
- L4: Representative spleen.

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LAMN/HAMN 04 Scenario

2023 SEER Workshop Cases

Abstracted Text

Physical Exam

05/15/2023 - Surg Consult: Pt referred to review and discuss symptomatic RLQ abdominal pain/mass. Pt endorses episodic abd pain since 01/2023. PTA pelvic U/S noted a tubular mass concerning for appendiceal mass. PE: Negative. IMP: Appendiceal mucocele vs. mass. Plan: Surgery.

Scans

04/01/2023 - PTA Pelvic U/S: IMP: Rt ovary was 2.0 cm and appeared unremarkable. There is an 8.7 cm tubular avascular mass w/ debris that is superior and lateral to ovary concerning for appendicitis vs. mass.

05/18/2023 - CT Abd/Pelvis: Findings: Large, dilated, blind-ended tubular structure arising from cecum measuring 2 cm in thickness. No gross periappendiceal inflammatory stranding. Findings could represent appendicitis vs. appendiceal mucocele. No adenopathy in abd/pelvis. Remainder of imaging negative.

Labs

05/20/2023 - CEA: 1.9 ng/mL (0.0 - 4.5 ng/mL normal)

Operative Reports

05/28/2023 - Laparoscopic appendectomy converted to Rt hemicolectomy: Findings: Appendix dilated and inflamed w/ large firm mass extending towards cecum and terminal ileum preventing simple appendectomy. No adenopathy or other disease identified.

Treatment Plan

06/12/2023 - Surg Note: Pt s/p Rt hemicolectomy for LAMN. No adjuvant therapy is required. Recommend follow-up w/ GI in 6 months.

Stage

06/12/2023 - Surg Note: pT3 pN0 cM0, Stg IIA

05/28/2023 - Path Report #1

FINAL DIAGNOSIS

A. Appendix, colon, and small intestine, right hemicolectomy (colon 17 cm, terminal ileum 10 cm):

- Low-grade appendiceal mucinous neoplasm (see Staging Summary below).
- Seventeen (17) lymph nodes negative for tumor.
- B. Additional segment of colon (segmental colectomy):
- Colon with no significant pathologic changes.
- Three (3) lymph nodes negative for tumor.

- There is no neoplastic mucinous epithelium, mucin, or malignancy.

SYNOPTIC STAGING SUMMARY

Procedure: Right hemicolectomy. Tumor site: Appendix, diffuse (full length) involvement. Tumor size: Approximately 15 cm, based on gross examination. Histologic type: Low-grade appendiceal mucinous neoplasm. Histologic grade: G1, well differentiated. Tumor extension: pT3: Tumor diffusely involves the appendix, with focal extension through the muscularis propria and into the subserosa; the serosal surface is uninvolved. The appendix is focally adherent to the colon, but neither neoplastic mucinous epithelium or mucin involves the colon (elastic stain and an immunostain for pan-keratin examined). Lymph-vascular invasion: Not identified. Perineural invasion: Not identified. Margins: Proximal small intestinal margin: Uninvolved. Distal colonic margin: Uninvolved. Radial mesenteric margin: Uninvolved. Regional lymph nodes: Number of lymph nodes identified: Twenty. Number of lymph nodes involved by metastasis: Zero (0/20). Pathologic stage: pT3 N0 (AJCC). Distant metastasis: Cannot be assessed.

CLINICAL HISTORY

ICD code(s): R19.03.

GROSS DESCRIPTION

A. Received fresh, labeled "A. terminal ileum, appendiceal mass, and ascending colon", is a right hemicolectomy specimen composed of terminal ileum (10 cm in length by 3.8 cm in circumference), colon (17 cm in length by 9.5-14 cm in circumference), appendix (15.0 x 6.5 x 5.2 cm), and a moderate amount of attached mesenteric fat. The proximal and distal margins are stapled. The specimen is inked as follows: blue-proximal ileal margin, black-distal colonic margin, orange-radial mesenteric margin, and green-appendiceal serosal surface. The appendix is intact, smooth, dilated, and focally adherent to the colon. The appendiceal orifice is not grossly identified. Within the entire appendix is tan yellow, gelatinous fluid lined by a tan-white, flattened, smooth mucosa with adherent tan-yellow patches of exudative material. The appendiceal wall thickness averages 0.2-0.3 cm. No discrete solid tumor is identified. Two possible polyps measuring 0.6 x 0.6 x 0.4 cm and 0.6 x 0.4 x 0.3 cm are identified in the colon. The small intestinal mucosa shows unremarkable folds with no lesions. The ileocecal valve is grossly unremarkable. Thirteen possible ileocolic lymph nodes ranging from 0.2-1.2 cm are identified. A representative section of the appendiceal wall is submitted for frozen section analysis and resubmitted in A1. Additional representative sections, including the entire appendix, are submitted as follows:

A1: Frozen section remnant of the appendiceal wall.

A2: Proximal ileal and distal colonic resection margins, perpendicular sections.

A3: lleocecal valve.

A4-A5: Appendiceal wall adherent to colon.

A6-A13: Representative sections of appendiceal wall.

A14-A16: Tip of appendix, entirely submitted.

A17: Largest possible lymph node, trisected.

- A18: Six possible whole lymph nodes.
- A19: Six possible whole lymph nodes.
- A20: Mesenteric radial soft tissue margin.
- A21: single bisected possible lymph node (adherent to appendiceal wall)

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A22-A55: remainder of appendix, submitted roughly from proximal to distal.

A56: representative sections of terminal ileum

A57: representative sections of colon, including two possible polyps.

B. Received in formalin, labeled "B. additional colon", is a 4.5 cm in length by 9.5 cm in circumference unoriented segment of colon with closed and stapled end margins, which are differentially inked blue and black. The mucosa is tan with grossly unremarkable folds with no lesions grossly identified. The small amount of attached mesenteric fat yields two possible lymph nodes measuring 0.3 and 0.7 cm. Representative sections are submitted as follows:

B1: Resection margins, contiguous perpendicular sections.

B2: Two possible whole lymph nodes.

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LAMN/HAMN 05 Scenario

2023 SEER Workshop Cases

Abstracted Text

Physical Exam

06/01/2023 - cc: Pt incidentally found to have mucinous deposits throughout peritoneum on PTA hiatal hernia repair. Cytologic assessment during PTA 05/02/2023 surgery proved atypical cells only. Pt referred for further workup. PE: Limited due to pt discomfort w/ no apparent organomegaly or LAD. IMP: Pseudomyxoma peritonei. Plan: Imaging, followed by surgery.

Scans

06/10/2023 - CT Abd/Pelvis: IMP: Moderately distended appendix supportive of presumptive dx of mucinous ca. Small lesion in liver most c/w cyst. Multiple pockets of fluid or mucin present throughout abd. Rest of imaging negative.

06/13/2023 - CT Chest: IMP: Several tiny pulmonary nodules are unchanged since prior CT dated 09/21/2021, presumed benign. No clear evidence of thoracic mets or LAD.

Labs

06/01/2023 - CEA: 1.1 ng/mL (0-5 ng/mL WNL)

Operative Reports

06/25/2023 - Exploratory laparotomy w/ cytoreduction of intra-abdominal tumor, partial peritonectomy, greater omentectomy, splenectomy, Rt hemicolectomy w/ distal small bowel rsxn, BSO: Findings: Extensive mucinous peritoneal disease along diaphragm, spleen, colon, bilateral ovaries, pelvic peritoneum, distal small bowel, and diffuse mucinous disease throughout omentum. Stringy mucinous disease along proximal small bowel as well as the porta hepatis. Liver negative. Appendix diffusely dilated w/ obvious perforation at base and mucinous extravasation. Small ascites present. At end of procedure, felt we had obtained adequate cytoreduction and HIPEC w/ Mitomycin-C completed.

Treatment Plan

07/19/2023 - GI Tumor Board Note: LAMN w/ acellular mucin in peritoneum, but no neoplasm in mucinous deposits. Pt s/p extensive cytoreduction and HIPEC. No further systemic therapy is recommended at this time and recommendation is for surveillance.

Chemo Text

06/25/2023 - Heated intraperitoneal chemotherapy w/ Mitomycin-C (intraoperative).

Stage

07/19/2023 - GI Tumor Board: pT4a pN0 pM1a, Stg IVA LAMN

06/25/2023 - Path Report #1

FINAL DIAGNOSIS

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A. Anterior partial peritoneum, falciform ligament, epigastric fad pad, right pericolic gutter peritoneum, median umbilical ligament, excision:

- Peritoneal tissue with reactive mesothelial hyperplasia, negative for neoplasm.

B. Mesenteric nodules, excision:

- Peritoneal tissue with reactive papillary mesothelial hyperplasia, negative for neoplasm.

C. Intra-abdominal mucin, excision:

- Mucin and inflammatory cells, no definite neoplasm.

D. Greater omentum and spleen, omentectomy and splenectomy:

- Spleen with serosal mucin and reactive changes, negative for neoplasm.

- Adipose tissue with reactive changes, negative for neoplasm.

E. Distal small bowel, appendix, cecum, and right colon, hemi-colectomy:

- Low-grade appendiceal mucinous neoplasm (LAMN) of appendix with mucin extravasation through serosa, see Synoptic Report.

F. Diaphragm and Morrison's pouch, excision:

- Peritoneal tissue with inflammation and reactive changes, negative for neoplasm.

G. Glisson's capsule, excision:

- Fibroadipose tissue with mucin, inflammation, and reactive changes, negative for neoplasm.

H. Porta hepatis tumor, excision:

- Fibroadipose tissue and mucin with fat necrosis, multi-nucleated giant cells, and reactive changes; negative for neoplasm.

I,J. Bilateral ovaries and fallopian tubes, bilateral salpingo-oophorectomy:

- Ovaries with follicular cysts, negative for neoplasm.

- Fallopian tubes with mild chronic serositis and focal mucin, negative for neoplasm.

K. Pelvic peritoneum, excision:

- Fibroadipose tissue with inflammation, reactive changes, and calcification, negative for neoplasm.

L. Periduodenal tumor, excision:

- Fibroadipose tissue with focal mucin and reactive changes, negative for neoplasm.

M. Left diaphragm peritoneum, excision:

- Fibroadipose tissue with mucin, inflammation, and reactive changes, negative for neoplasm.

N. Epiploic appendages, excision:

- Fibroadipose tissue with focal mucin and reactive changes, negative for neoplasm.

SYNOPTIC STAGING SUMMARY

APPENDIX: Resection SPECIMEN: Appendectomy and right colectomy TUMOR Tumor Site: Diffusely involving appendix Histologic Type: Low-grade appendiceal mucinous neoplasm Histologic Grade: G1, well differentiated Tumor Size: Greatest Dimension (Centimeters): 2.2 cm Tumor Deposits: Not identified Tumor Extent: Acellular mucin invades visceral peritoneum (serosa) Lymphovascular Invasion: Not identified MARGINS Margin Status for Invasive Carcinoma: All margins negative for invasive carcinoma Margin Status for Non-Invasive Tumor and Mucin: All margins negative for non-invasive tumor and mucin

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11/8/23. 7:22 AM **REGIONAL LYMPH NODES** Regional Lymph Node Status: All regional lymph nodes negative for tumor Number of Lymph Nodes Examined: 8 **DISTANT METASTASIS** Distant Site(s) Involved: Intraperitoneal acellular mucin without identifiable tumor cells in the disseminated peritoneal mucinous deposits

PATHOLOGIC STAGE CLASSIFICATION (pTNM, AJCC) pT Category: pT4a pN Category: pN0 pM Category: pM1a

CLINICAL HISTORY

Pre-op diagnosis: Pseudomyxoma peritonei

GROSS DESCRIPTION

A. Peritoneum. Received in formalin labeled, "1. ANTERIOR PARITAL PERITONEUM, FALCIFORM LIGAMENT, EPIGASTRIC FAT PAD, RIGHT PERICOLIC GUTTER PERITONEUM, MEDIAN UMBILICAL LIGAMENT" is a 28.0 x 13.0 x 2 cm portion of fibrofatty membranous tissue. Is partially covered by a glistening hemorrhagic serosa which is focally retracted and has numerous attached minute nodules averaging 0.1 cm. The nodules are sectioned and appear to be confined to the serosa. There are fibrofatty cut surfaces and no intramural masses are appreciated. Representative sections of the nodular serosa and retracted serosa are submitted in cassette A1-A2.

B. Lymph Node, Mesenteric. Received in formalin labeled, "2. MESENTERIC NODULES" is a 0.8 x 0.8 x 0.2 cm aggregate of tan-pink soft tissue fragments and translucent mucoid material. Entirely submitted in B1.

C. Peritoneum. Received fresh labeled, "3. INTRA-ABDOMINAL MUCIN" is a 5.0 x 5.0 x 2.0 cm aggregate of mucoid material. 2.5 cm of the material are arranged in grapelike clusters. Representative sections are submitted in C1.

D. Spleen. Received in formalin labeled, "4. GREATER OMENTUM AND SPLEEN" is a 410 g, 28.0 x 21.0 x 2 cm in greatest dimension portion of greater omentum. Adheres to the omentum is a 255 g, 14.0 x 10.0 x 5 cm spleen. The omentum serosa is tan-yellow smooth and glistening with numerous attached minute tan-pink nodules averaging less than 0.1 cm greatest dimension. The nodules are sectioned and are grossly confined to the serosa. The remaining omentum is sectioned and has unremarkable cut surfaces.

The splenic capsule contains a 11 x 4 x 1 cm area of adherent fibrous adhesions and adherent mucinous neoplasm. Spleen is sectioned and there is dark red splenic parenchyma on cut surfaces with normal-appearing red and white pulp. There is mucinous neoplasm is confined to the capsule of the spleen.

Representative sections are submitted as follows:

D1-D3: Random serosal nodules omentum

D4: Apparent hilar lymph node spleen

D5: Mucinous neoplasm attached to splenic capsule

E. Small Bowel, resection, for tumor. Received in formalin labeled, "5. DISTAL SMALL BOWEL, APPENDIX, CECUM AND RIGHT COLON" is a right colectomy specimen that is stapled at both ends (proximal inked blue, distal inked black). It consists of 49.0 cm long by 3.5 cm circumference of small bowel is contiguous with a 14.0 cm long by 6.0 cm circumference cecum and ascending colon. The cecum has an attached 5.5 cm long by 1.2 cm diameter vermiform appendix. At the appendix tip, the wall is firm white and thickened, up to 0.4 cm spanning 2.2 cm. The firm area is 2.3 cm from the appendiceal orifice and 4.0 cm from the closest radial margin (inked orange). The remaining wall is up to 0.2 cm thick. The lumen at the appendiceal orifice is a 0.6 cm diameter and gradually narrows to 0.3 cm diameter toward the tip and is filled with extensive mucoid material. The appendiceal serosa has mucoid adhesions. The remaining bowel mucosa has slightly irregular folds. The wall is up to 0.2 cm thick. The distal small bowel and large bowel serosa has scattered mucinous and hemorrhagic adhesions.

Representative sections are submitted:

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E1-mucosal margins (perpendicular)

E2-E3-appendiceal orifice with appendix (perpendicular)

E4-E7-remaining appendix entirely submitted (tip in E7)

E8-radial margin closest to firm area of appendix

E9-small bowel with serosal adhesions, and ileocecal valve

E10-large bowel with serosal adhesions

E11-mucin loosely surrounding appendix E12-E 13-possible ileocolic lymph nodes.

F. Diaphragm, biopsy or tissue. Received in formalin labeled, "6. RIGHT DIAPHRAGM AND MORRISON'S POUCH PERITONEUM" is a 26.0 x 17.0 x 1.8 cm greatest dimension portion of membranous tissue. 1 surface is carpeted by a mucinous friable neoplasm. Representative section is submitted in cassette F1.

G. Capsule. Received in formalin labeled, "7. GLISSON'S CAPSULE" is a 5.0 x 2.5 x 1.0 cm aggregate of heavily congested fibromembranous tissue. 1 surface is fairly smooth, tan-pink. The opposite surface is red-tan and granular with scattered nodularity. Representative sections are submitted in G1-G2.

H. Peritoneum. Received in formalin labeled, "8. PORTO-HEPATIS TUMOR" is a 2.0 x 0.8 x 0.5 cm aggregate of red-tan soft tissue fragments. Entirely submitted in H1.

I. Fallopian Tube, Left. Received in formalin labeled, "left fallopian tube and ovary" is a 3.0 x 3.0 x 2.0 cm ovary. There are thin congested fibrous adhesions overlying approximately 10%. The cut surface has a 2.0 cm multiloculated cyst filled with pink-tinged watery fluid and a 2.5 cm focally disrupted cyst filled with red-tinged watery fluid. No excressences are identified. The attached questionable ovary is 1.0 cm long by 0.4 cm diameter and ending in possible fimbria. Representative sections are submitted:

I1-I3-ovary

I4-ovary entire possible fallopian tube and fimbria.

J. Fallopian Tube, Right. Received in formalin labeled, "right fallopian tube and right ovary" is a 3.2 x 2.0 x 1.5 cm ovary partially surrounded by hemorrhagic adhesions. The ovary has 3 cysts filled with hemorrhagic material that are up to 1.0 cm greatest dimension the ovary has a 6.0 x 4.0 x 1.0 cm of attached heavily congested fibromembranous tissue, and yellow lobulated adipose. Embedded in the adipose are multiple vessels and a questionable fallopian tube that is 1.5 cm long by 0.5 cm diameter. Fimbria is not identified. The fibromembranous tissue has adherent congested mucoid material representative sections are submitted:

J1-J2-ovary

J3-entire possible fallopian tube

J4-vessels, fibromembranous tissue with mucoid material.

K. Peritoneum. Received in formalin labeled, "pelvic peritoneum " is an 8.0 x 2.0 x 0.5 cm aggregate of congested fibromembranous tissue. 1 surface is partially lined with yellow lobulated adipose. The opposite surface has hemorrhagic adhesions. Cut surface is focally tan-white nodular over an area of approximately 1.0 cm. Representative sections are submitted in K1.

L. Duodenum. Received in formalin labeled, "12. PERIDUODENAL TUMOR" is a 3.5 x 3.0 x 0.3 cm flattened portion of yellow lobulated adipose partially surrounded by congested fibrous membrane with scattered hemorrhagic adhesions. The tissue is sectioned and entirely submitted in L1-L2.

M. Peritoneum. Received in formalin labeled, "13. LEFT DIAPHRAGM PERITONEUM" is a 16.0 x 6.0 cm ragged portion of fibromembranous tissue partially surfaced by soft dark red slightly nodular adhesions. Representative sections are submitted in M1.

N. Peritoneum. Received in formalin labeled, "14. EPIPLOIC APPENDAGES" is a 3.5 x 2.5 x 1.0 cm aggregate of yellow lobulated soft tissue with scattered hemorrhagic adhesions. Sectioned and entirely submitted in N1-N2.

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