Neuroendocrine Tumors

NAACCR 2018-2019 WEBINAR SERIES

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Q&A

Please submit all questions concerning the webinar content through the Q&A panel.
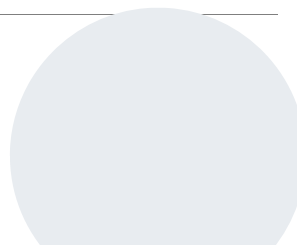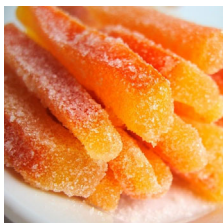
If you have participants watching this webinar at your site, please collect their names and emails

We will be distributing a Q&A document in about one week. This document will fully answer questions asked during the webinar and will contain any corrections that we may discover after the webinar.
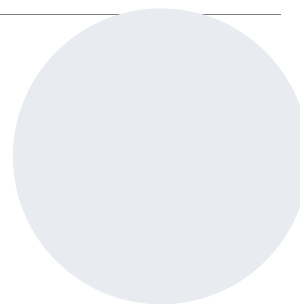
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Fabulous Prizes

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Guest Speaker

Tonya Brandenburg, MHA, CTR
◦ QA Manager Casefinding
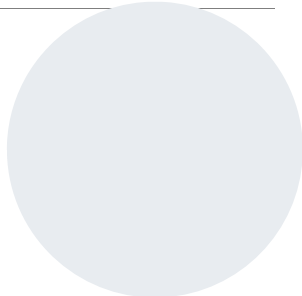◦ Kentucky Cancer Registry

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Agenda

Overview

Solid Tumor Rules

Staging
- ◦ Grade
- ◦ SSDI
- ◦ AJCC
- ◦ EOD
- ◦ Summary Stage

Treatment

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Overview

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Neuroendocrine Cells

Neuroendocrine cells receive messages from Neurons.
◦ The electrical signals from the neuron is converted into hormonal signals and hormones or peptides are produced.

Neuroendocrine cell are found in almost every organ in the body.
◦ Mainly found in the GI tract, the gallbladder, pancreas, and thyroid.

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Types of Neuroendocrine Tumors (NETS)

GI NETS
◦ Well differentiated NET
◦ Neuroendocrine *carcinoma*

Lung NETS
◦ Typical
◦ Atypical
◦ Large Cell
◦ Small Cell

Pancreatic NETS (pNETS)
◦ Well differentiated pNETS
◦ Poorly differentiated pancreatic neuroendocrine *carcinoma*
◦ Functional pNETS
  ◦ Gastrinoma
  ◦ Insulinoma
  ◦ Glucagonoma
  ◦ Somatostatinoma
  ◦ VIPoma
  ◦ ACTHoma

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Assigning Histology

SOLID TUMOR RULES

2018 ICD-O UPDATE

2014 ICD-O UPDATE

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ICD-O Updates

2018

8041 High-grade neuroendocrine carcinoma (C54._, C55.9)

8041 Neuroendocrine carcinoma, poorly differentiated (C50._)

8246 Neuroendocrine tumor, well differentiated (C50._)

Implementation guidelines
https://www.naaccr.org/implementation-guidelines/

2014

8240 Neuroendocrine tumor grade 1
 ◦ Neuroendocrine carcinoma, low grade
 ◦ Neuroendocrine carcinoma, well differentiated

8249 Neuroendocrine tumor grade 2
 ◦ Neuroendocrine carcinoma, moderately differentiated

8244 Mixed adenoneuroendocrine carcinoma (MANEC)

Previous guidelines
https://www.naaccr.org/implementation-guidelines/#PrevICDO3

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Well Differentiated, Neuroendocrine Tumor

8240/3 Well differentiated neuroendocrine tumor G1

8249/3 Well differentiated neuroendocrine tumor G2

8240/3 Well differentiated neuroendocrine tumor G3

8246/3 Neuroendocrine carcinoma

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Solid Tumor Rules Colon/Rectum

8240/3-Neuroendocrine **Tumor** Grade 1
- Well Differentiated Neuroendocrine **Tumor**, G1
- Low Grade Neuroendocrine **Tumor**
- NE**T** G1
- Carcinoid, NOS

Subtypes
- 8249/3-Neuroendocrine **Tumor** Grade 2
- 8156/3-Somatostatin-producing NE**T**
- 8241/3-EC cell serotonin-producing NE**T**/enterochromaffin cell carcinoid

8246/3 Neuroendocrine *Carcinoma (NEC)*
- 8013/3 Large Cell NEC
- 8041/3 Small Cell NEC

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Solid Tumor Rules

Breast
- 8041/3 Neuroendocrine Carcinoma, poorly differentiated
- 8246/3 Neuroendocrine Tumor, well-differentiated

Urinary
- 8041/3 Small cell neuroendocrine carcinoma

Lung
- 8041/-Neuroendocrine Tumor
- 8013/3 Large cell neuroendocrine carcinoma/Combined Large Cell Neuroendocrine Carcinoma
- 8249/3 Atypical Carcinoid
- 8240/3Typical Carcinoid

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Pop Quiz 1

A patient was found to have a single tumor in their **colon**. The tumor was resected.

Pathology showed:
- **Well differentiated neuroendocrine tumor, G1**

What histology would be assigned?
- Rule H7 tells us to code the histology when only one is present and refers us to Table 1
- Table 1 tells us to code the histology to **8240**
- **Grade, AJCC Stage, EOD, Summary Stage will all be based on the NET Colon schema not the Colon Schema.**

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Pop Quiz 2

A patient was found to have a single tumor in their **breast**. The tumor was resected.

Pathology showed:
◦ Well differentiated neuroendocrine tumor

What histology would be assigned?
◦ Rule H12 tells us to code the histology when only one is present and refers us to Table 3
◦ Table 1 tells us to code the histology to **8246/3**

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Pop Quiz 3

A patient was found to have a single tumor in their **Lung**. The tumor was resected.

Pathology showed:
◦ Neuroendocrine tumor

What histology would be assigned?
◦ Rule H4 tells us to code the histology when only one is present and refers us to Table 3
◦ Table 1 tells us to code the histology to **8041/3**

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Pop Quiz 4

A patient was found to have a single tumor in their **colon**. The tumor was resected.

Pathology showed:
◦ **Neuroendocrine carcinoma**

What histology would be assigned?
◦ Rule H7 tells us to code the histology when only one is present and refers us to Table 1
◦ Table 1 tells us to code the histology to **8246/3**
  ◦ Grade, AJCC Stage, EOD, Summary Stage will all be based on the Colon schema not the NET Colon Schema.

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Other NET Tumors

Use the Other chapter in the 2007 MP/H rules for Sites other than:
◦ Head and Neck
◦ Colon
◦ Lung
◦ Breast
◦ Kidney
◦ Urinary
◦ CNS Malignant
◦ CNS Non-malignant

Review the 2018 ICD-O Updates Table prior to assigning histology.

May also need to review the 2014 ICD-O Updates

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Pop Quiz 5

A patient was found to have a single tumor in their **pancreas**. The tumor was resected.

Pathology showed:
- **Neuroendocrine tumor, G1**

What histology would be assigned?
- Rule H11 tells us to code the histology when only one is present
- The 2018 ICD-O Updates Table does not include this term
- **The 2014 ICD-O Updates Table tells us to code this 8240**
- Grade, AJCC Stage, EOD, Summary Stage will all be based on the Net Pancreas schema not the Exocrine pancreas schema.
  - Neuroendocrine carcinoma (8246/3) would go to to the exocrine pancreas schema

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Case 1

STOMACH, BODY, BIOPSY: ANTRAL TYPE GASTRIC MUCOSA WITH:
- Well-differentiated neuroendocrine tumor, grade 1, measuring 4 mm maximally on the slide.

How many primaries?
- Other Chapter: 1 primary per rule M2

What histology should be assigned?
- Other Chapter Rule H11 tells us to code the histology when only 1 histology is present
- Next step is to check the 2018 ICD-O updates table.
  - Histologic term is not part of in table
- Check 2014 ICD-O updates
  - 8240/3 Neuroendocrine tumor, grade 1

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Case 2

Tumor Size:
- 2 x 1.5 x 1.3 cm

Tumor Focality:
- Multifocal (specify number of tumors: 2)

Histologic Type and Grade:
- G2 Well-differentiated neuroendocrine tumor

How many primaries?
- Other Chapter: 1 primary per rule M18

What histology should be assigned?
- Other Chapter: Rule H23 tells us to code the histology when only 1 histology is present
- Next step is to check the 2018 ICD-O updates table.
  - Histologic term is not part of in table
- Check 2014 ICD-O updates
  - 8249/3 Neuroendocrine tumor, grade 2

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Case 3

Tumor Site: Pancreatic tail

Histologic Type and Grade:
- G2: Well-differentiated neuroendocrine tumor

Mitotic Rate: 2-20 mitoses / 2 mm2

Ki-67 Labeling Index: 3% to 20%

Tumor Size: 2 Centimeters (cm)

Tumor Focality: Unifocal

Tumor Extension: Tumor is limited to the pancreas

How many primaries?
- Other Chapter: 1 primary per rule M2

What histology should be assigned?
- Other Chapter Rule H11 tells us to code the histology when only 1 histology is present
- Next step is to check the 2018 ICD-O updates table.
  - Histologic term is not part of in table
- Check 2014 ICD-O updates
  - 8249/3 Neuroendocrine tumor, grade 2

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Staging

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NET AJCC Chapters

NET of the Stomach (Ch 29 Pg 351 - 359)

NET of the Duodenum and Ampulla of Vater (Ch 30 Pg 360 - 374)

NET of the Jejunum and Ilelum (Ch 31 Pg 375 - 388)

NET of the Appendix (Ch 32 Pg 389 - 394)

NET of the Colon and Rectum (Ch 33 Pg 395 - 406

NET of the Pancreas (Ch 34 Pg 407 - 420)

Adrenal Gland NET (Ch 77 Pg 927 - 935)

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AJCC Staging Reminder

Topography ➕ Histology

For Every Chapter in the AJCC Manual

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AJCC Staging

The clinical and pathological classification of NETs is based on size, functionality, site, grade, and invasion.

NET may be Grade 1, Grade 2, or Grade 3

- NET Grade 1 represent 80-90% of gastric NET tumors, rarely metastasize, and have a 100% 5 year survival rate.
- NET Grade 2 represent 5-7% of gastric NET tumors, 10-30% metastasize, have a 60-90% 5 year survival rate.
- NET Grade 3 represent 10-15% of gastric NET tumors, 50% metastasize, and have less than 50% 5 year survival rate.

- Amin, Mahul B.; Gress, Donna M.; Meyer Vega, Laura R.; Edge, Stephen B.. AJCC Cancer Staging Manual, Eighth Edition (Page 371). American College of Surgeons. Kindle Edition.

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Grade and SSDIs

Data Item	Coded Value
Clinical Grade	
Pathological Grade	
Post Therapy Grade	

No SSDIs for NET Tumors

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NET Grade Tables

Code	Description
1	G1: Mitotic count (per 10 HPF) less than 2 AND Ki-67 index (%) less than 3
2	G2: Mitotic count (per 10 HPF) equal 2-20 OR Ki-67 index (%) equal 3-20
3	G3: Mitotic count (per 10 HPF) greater than 20 OR Ki-67 index (%) greater than 20
A	Well differentiated
B	Moderately differentiated
C	Poorly differentiated
D	Undifferentiated, anaplastic
9	Grade cannot be assessed (GX); Unknown

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Pop Quiz 6

Colonoscopy, small bowel biopsy:
well differentiated neuroendocrine G2 tumor.

Distal, ileum, and cecum resection:
0.8 cm well differentiated neuroendocrine tumor G1.

What is the Clinical Grade?

- 1
- 2
- A
- B

What is the Pathological Grade?

- 1
- 2
- B
- A

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AJCC Staging

Rule of Thumb
TWO (2) *STAGES are* **usually** defined per case…..however….
sometimes just one (1) will be defined…..but….
NEVER ALL 3
GOT IT!

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Prognostic Factors

Ki-67 proliferative index
- Measured using MIB1 antibody 500 to 2,000%

Mitotic count
- Measured using mitoses per 10 high-power fields (HPF)
- Has to be at least 50 HPFs

Gastrin level
- Elevated in type I and II, normal in type III
- Normal levels mean worse prognosis in gastric NETs

Chromogranin A (CgA) level
- Higher levels mean worse prognosis

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EOD and Summary Stage 2018

Data Item	Coded Value
Summary Stage 2018	
EOD Primary Tumor	
EOD Regional Nodes	
EOD Mets	
Regional Nodes Pos	
Regional Nodes Ex	
Lymphovascular Inv	

AJCC T and EOD/Summary Stage 2018 for NET of the Stomach
See your AJCC 8th edition Manual for code definitions

T Category	Things to look for…
TX	Primary tumor cannot be assessed
T0	No evidence of primary tumor
T1	Invades the lamina propria, not bigger than 1 cm
T2	Invades the muscularis propria, bigger than 1 cm
T3	Invades into subserosa doesn't go to serosa
T4	Invades serosa or other close organs

EOD T	Explanation	SS 2018
000	In situ, intraepithelial, noninvasive	IS
100	Intramucosal, lamina propria, mucosa, confined to stomach, NOS, localized, NOS	L
200	Invasion of musclaris propria	L
400	Extension through wall, NOS, Invasion of muscularis propria, perimuscular tissue ivaded, sub(serosal) tissue/fat invaded	L
500	Extension to adjacent connective tissue w/o perforation of visceral peritoneum covering gastric artery, ligaments, omentum, perigastric fat	RE
600	Invasion of/through serosa, visceral peritoneum, pancreas, small intestine	RE
700	Further contiguous extension	D

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AJCC N and M for NET of the Stomach
See your AJCC 8th edition Manual for code definitions

N Category	Things to look for…
NX	Regional nodes not assessed
N0	No regional node mets
N1	Regional node mets

EOD N	Description	SS2018
000	No Regional LN involvement	None
300	Most Nodes	RN
700	For lesser curvature only – hepatoduodenal	D
800	Regional Nodes, NOS	RN

M Category	Things to look for…
M0	No distant mets
M1	Distant Mets
M1a	Mets confined to liver
M1b	Mets in at least one extrahepatic site (lung, bone, lymph nodes)
M1c	Both liver and extrahepatic mets

EOD M	Description	SS2018
00	No Distant Mets	None
10	Liver	D
20	Bone, Lung, Ovaries, Peritoneum	D
30	Distant Lymph Nodes W/ or W/ code 20	D
50	Code 20 + 30, carcinomatosis	D
70	Distant mets, NOS	D

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Pop Quiz 7

Stomach biopsy:
- well differentiated neuroendocrine G2 tumor.

Segmental resection:
- 0.8 cm well differentiated neuroendocrine tumor G2 invading muscularis propria into subserosal tissue, not into serosa.

What is the EOD T
- 100
- 200
- **400**
- 500

What is the AJCC T?
- cT2
- **pT3**
- pT4
- cT4

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Case Scenario 1

Clinical Stage	Code	Pathological Stage	Code	PostTx Stage	Code
cT	cTX	pT		PostT xT	
cT Suffix		pT Suffix		PostTxT Suffix	
cN	cN0	pN		PostTxN	
cN Suffix		pN Suffix		PostTxN Suffix	
cM	cM0	pM		PostTxM	
cStage	99	pStage	99	PostTxStage	

Data Item	Code
Summary Stage 2018	1 - Local
EOD Primary Tumor	100
EOD Regional Nodes	000
EOD Mets	00
Regional Nodes Pos	98
Regional Nodes Ex	00
Lymphovascular Inv	0

Data Item	Code
Clinical Grade	1
Pathological Grade	9
Post Therapy Grade	

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AJCC T and EOD/Summary Stage 2018 for NET of the Jejunum
See your AJCC 8th edition Manual for code definitions

T Category	Things to look for…
TX	Primary tumor not assessed
T0	No evidence of primary tumor
T1	Invades the lamina propria, not bigger than 1 cm
T2	Invades the muscularis propria, bigger than 1 cm
T3	Invades into subserosa doesn't go to serosa
T4	Invades serosa or other close organs

EOD T	Explanation	SS 2018
000	In situ, intraepithelial, noninvasive	IS
100	localized, NOS or less than 1 cm	L
200	Invasion of musclaris propria or greater than 1 cm	L
400	Extension through wall, NOS, Invasion of muscularis propria, perimuscular tissue ivaded, sub(serosal) tissue/fat invaded	L
500	Extension to adjacent connective tissue w/o perforation of visceral peritoneum covering gastric artery, ligaments, omentum, perigastric fat	RE
600	Invasion of/through serosa, visceral peritoneum, pancreas, small intestine	RE
700	Further contiguous extension	D

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AJCC N and M for NET of the Jejunum
See your AJCC 8th edition Manual for code definitions

N Category	Things to look for…
NX	Regional nodes not assessed
N0	No regional node mets
N1	Regional node mets less than 12 nodes
N2	Greater than 12 nodes

EOD N	Description	SS2018
000	No Regional LN involvement	None
300	Cecal, ileocecal, mesenteric, superior mesenteric	RN
400	Large mesenteric greater than 2 cm	RN
800	Regional Nodes, NOS	RN

M Category	Things to look for…
M0	No distant mets
M1	Distant Mets
M1a	Mets confined to liver
M1b	Mets in at least one extrahepatic site (lung, bone, lymph nodes)
M1c	Both liver and extrahepatic mets

EOD M	Description	SS2018
00	No Distant Mets	None
10	Liver	D
20	Distant lymph nodes	D
30	At least one extrahepatic site, distant mets w/ or w/o distant lymph nodes, carcinomatosis	D
50	Any combination of codes 10, 20, or 30	D
70	Distant mets, NOS	D

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Case Scenario 2

Clinical Stage	Code	Pathological Stage	Code	PostTx Stage	Code
cT	cTX	pT	pT3	PostT xT	
cT Suffix		pT Suffix		PostTxT Suffix	
cN	cN0	pN	pN1	PostTxN	
cN Suffix		pN Suffix		PostTxN Suffix	
cM	cM1c	pM	pM1c	PostTxM	
cStage	IV	pStage	IV	PostTxStage	

Data Item	Code
Summary Stage 2018	7 – Distant
EOD Primary Tumor	400
EOD Regional Nodes	300
EOD Mets	50
Regional Nodes Pos	2
Regional Nodes Ex	6
Lymphovascular Inv	1

Data Item	Code
Clinical Grade	9
Pathological Grade	2
Post Therapy Grade	

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AJCC T and EOD/Summary Stage 2018 for NET of the Pancreas
See your AJCC 8th edition Manual for code definitions

T Category	Things to look for…
TX	Primary tumor cannot be assessed
T1	Limited to pancreas smaller than 2 cm
T2	Limited to pancreas 2-4 cm in size
T3	Limited to pancreas larger than 4 cm, or a tumor that invade duodenum or cbd
T4	Invades adjacent organs or wall of large vessels

EOD T	Explanation	SS 2018
000	In situ, intraepithelial, noninvasive	IS
100	localized, NOS	L
200	Peripancreatic fat	RE
300	CBD, Bile Duct, NOS, Duodenum	RE
500	Fixation to adjacent structures	RE
600	Further contiguous extension to other major arteries	RE
700	Further contiguous extension to other organs	D

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AJCC N and M for NET of the Pancreas
See your AJCC 8th edition Manual for code definitions

N Category	Things to look for…
NX	Regional nodes not assessed
N0	No regional node mets
N1	Regional node mets

EOD N	Description	SS2018
000	No Regional LN involvement	None
300	Most Nodes	RN
700	Celiac nodes	D
800	Regional Nodes, NOS	RN

M Category	Things to look for…
M0	No distant mets
M1	Distant Mets
M1a	Mets confined to liver
M1b	Mets in at least one extrahepatic site (lung, bone, lymph nodes)
M1c	Both liver and extrahepatic mets

EOD M	Description	SS2018
00	No Distant Mets	None
10	Liver	D
20	Distant lymph nodes – celiac, cbd, porta hepatic, pyloric	D
40	Distant lymph nodes – pancreaticosplenic, splenic, distant nodes, nos	D
50	Extra hepatic sites, carcinomatosis, distant mets w/ or w/o distant lymph nodes	D
60	Hepatic and extrahepatic mets, code 10 with any combo of 20-50	D
70	Distant mets, NOS	

Pop Quiz 8

Pancreatic FNA:
- C/W Pancreatic NET Ki-67 less than 1%

LAP Subtotal Pancreatectomy and LN dissection:
- neuroendocrine tumor, mitoses 15 per 2mm^2 (per 10hpf), Ki-67 index: 17%

What is the Clinical Grade?
- 1
- 2
- 3
- **9**

What is the Pathological Grade?
- 1
- **2**
- 3
- 9

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Case Scenario 3

Clinical Stage	Code	Pathological Stage	Code	PostTx Stage	Code
cT	cT2	pT	pT2	PostT xT	
cT Suffix		pT Suffix		PostTxT Suffix	
cN	cN0	pN	pN0	PostTxN	
cN Suffix		pN Suffix		PostTxN Suffix	
cM	cM0	pM	cM0	PostTxM	
cStage	II	pStage	II	PostTxStage	

Data Item	Code
Summary Stage 2018	1 - Local
EOD Primary Tumor	100
EOD Regional Nodes	000
EOD Mets	00
Regional Nodes Pos	0
Regional Nodes Ex	9
Lymphovascular Inv	1

Data Item	Code
Clinical Grade	9
Pathological Grade	2
Post Therapy Grade	

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Questions?

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Treatment

CASE 1
CASE 2
CASE 3

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Case 1

3/28/18-Endoscopy
◦ EGD
◦ Colonoscopy

No further treatment.
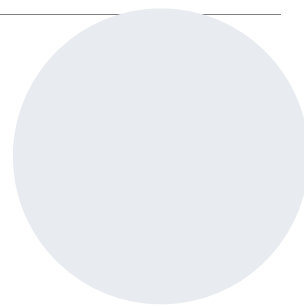
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Treatment Case 1

Diagnostic Staging Procedure	02
Surgery Codes	
Surgical Procedure of Primary Site	00
Scope of Regional Lymph Node Surgery	0
Surgical Procedure/ Other Site	0
Systemic Therapy Codes	
Chemotherapy	00
Hormone Therapy	00
Immunotherapy	00
Hematologic Transplant/Endocrine Procedure	00
Systemic/Surgery Sequence	0

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Case 2

02/06/2018
- ◦ Segmental resection, small intestine
- ◦ Omentum, excision

3/1/2018
- ◦ Patient started on Gleevec post-operatively.

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Treatment Case 2

Diagnostic Staging Procedure	00
Surgery Codes	
Surgical Procedure of Primary Site	30
Scope of Regional Lymph Node Surgery	5
Surgical Procedure/ Other Site	4
Systemic Therapy Codes	
Chemotherapy	02
Hormone Therapy	00
Immunotherapy	00
Hematologic Transplant/Endocrine Procedure	00
Systemic/Surgery Sequence	3

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Case 3

7/21/18

◦ Pancreas, Body and Tail, Mass, Endoscopic Ultrasound Guided Fine Needle Aspiration

8/3/18

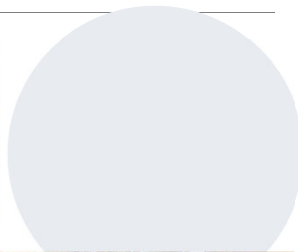◦ Laprascopic Subtotal Pancreatectomy and Laparascopic Retroperitoneal Lymph Node Dissection

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Treatment Case 3

Diagnostic Staging Procedure	02
Surgery Codes	
Surgical Procedure of Primary Site	30
Scope of Regional Lymph Node Surgery	5
Surgical Procedure/ Other Site	4
Systemic Therapy Codes	
Chemotherapy	00
Hormone Therapy	00
Immunotherapy	00
Hematologic Transplant/Endocrine Procedure	00
Systemic/Surgery Sequence	0

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Questions?

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Fabulous Prize Winners

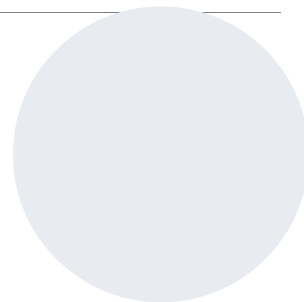# Coming UP…

Collecting Cancer Data: Ovary
- 06/06/2019

Collecting Cancer Data: Hospital Topic
- 07/11/2019

CE Certificate Quiz/Survey

Phrase

Link

https://www.surveygizmo.com/s3/4981301/Neuroendocrine-2019

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