Ovarian mucinous lesions: Common diagnostic dilemmas

Karuna Garg, MD
University of California San Francisco

Intestinal or usual type
Seromucinous (Endocervical mucinous or Mullerian mucinous) type

Ovarian mucinous lesions: problematic issues

- Mucinous cystadenoma versus borderline tumor
- Mucinous borderline tumor versus carcinoma
- Primary mucinous tumors versus metastasis
- Pseudomyxoma peritonei – site of origin, classification and clinical outcomes
- Effective handling of mucinous ovarian lesions at frozen section
- Mural nodules

Mucinous cystadenoma versus borderline tumor
Mucinous cystadenoma versus borderline tumor

**Significance**
Borderline tumors are usually staged.

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**Assess for epithelial proliferation**
- Sample well
- If less than 10% - cystadenoma with focal epithelial atypia/proliferation
- Clinical outcome similar to cystadenoma
- If >10% - borderline tumor
Mucinous borderline tumor versus carcinoma
Mucinous borderline tumor versus carcinoma

Significance
- Carcinomas always staged
- Small risk of extra-ovarian disease

- Mucinous borderline tumors are benign and should be stage 1A (if considering an advanced stage borderline tumor or borderline tumor with implants-exclude metastasis)

Mucinous borderline tumor

Microinvasion
• No single invasive focus should measure >5 mm or 10 mm²
• Multiple foci can occur
• Prognosis similar to borderline tumors (one recent study shows higher risk of recurrence)
Mucinous borderline tumor

Intraepithelial carcinoma

- High grade nuclear atypia
- Prognosis similar to borderline tumors (one recent study suggests higher risk of recurrence)
Ovarian mucinous carcinoma

- Unequivocal invasion >5 mm (10 mm)
- Two patterns of invasion:
  - Expansile (confluent cribriform/glandular pattern) more common
  - Infiltrative (worse prognosis) (should exclude metastasis)
**Immunophenotype of primary ovarian mucinous tumors**

- CK7++
- CK20+ (83%)
- CDX2+ (40%-less frequently positive in primary ovarian mucinous tumors)
- PAX-8+ (50%)
- ER/PR-
- WT1-

**Primary versus metastatic mucinous neoplasms of the ovary**

<table>
<thead>
<tr>
<th>Tumor type</th>
<th>Primary ovarian mucinous tumors (n=42)</th>
</tr>
</thead>
<tbody>
<tr>
<td>CK7+/CDX2-</td>
<td>25 (60%)</td>
</tr>
<tr>
<td>CK7+/CDX2+</td>
<td>15 (36%)</td>
</tr>
<tr>
<td>CK7-/CDX2-</td>
<td>2 (5%)</td>
</tr>
<tr>
<td>CK7-/CDX2+</td>
<td>1 (5%)</td>
</tr>
<tr>
<td>CK7+/CK20-</td>
<td>7 (17%)</td>
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<tr>
<td>CK7+/CK20+</td>
<td>33 (79%)</td>
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<tr>
<td>CK7-/CK20+</td>
<td>2 (5%)</td>
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<tr>
<td>CK7-/CK20-</td>
<td>0 (-)</td>
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</table>

Vang, et al. Mod Pathol 2006
<table>
<thead>
<tr>
<th>Primary versus metastasis</th>
<th>Ovarian mucinous carcinoma</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Can be challenging</td>
<td>• Uncommon</td>
</tr>
<tr>
<td>• Metastatic mucinous carcinomas to the ovary can be highly deceptive</td>
<td>• &lt;3% of all ovarian carcinomas</td>
</tr>
<tr>
<td>• Sometimes metastatic involvement of the ovary is the primary presentation of a mucinous tumor without clinical evidence of an extra-ovarian lesion</td>
<td>• Predominantly stage 1</td>
</tr>
<tr>
<td>• Immunohistochemistry of limited help</td>
<td>• Extra-ovarian disease rare</td>
</tr>
</tbody>
</table>

**Ovarian mucinous carcinoma**

- Metastatic mucinous carcinomas are more common
- Metastatic mucinous carcinomas can simulate ovarian mucinous carcinoma, borderline tumor or cystadenoma by radiology and pathology

**Ovarian mucinous carcinoma**

- 44 cases of advanced stage mucinous carcinomas
- 61% reclassified as likely metastasis
- True advanced stage primary ovarian mucinous carcinomas rare (0.5-1.5%)
- Advanced stage ovarian mucinous carcinomas highly lethal and survival shorter than serous

HER2 in ovarian mucinous carcinomas

- Low response rates to carbotaxol
- Her2 amplification in 18% mucinous carcinomas and borderline tumors
- No association with prognosis
- Patients may respond to trastuzumab therapy

McAlpine et al, BMC Cancer 2009

Primary versus metastasis

Significance
- Prognosis
- Therapy

Primary versus metastasis: prognosis

5 year survival:

Primary versus metastasis: therapy

- Surgery
  - Primary ovarian cancer: comprehensive surgical staging and debulking
  - Metastasis: No staging

- Chemotherapy: Different agents (move toward treating mucinous tumors by histology rather than site of origin)
Primary versus metastasis

Gross features

<table>
<thead>
<tr>
<th>Laterality</th>
<th>Primary</th>
<th>Metastasis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bilateral</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Unilateral</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Size</th>
<th>Primary</th>
<th>Metastasis</th>
</tr>
</thead>
<tbody>
<tr>
<td>&gt;10 cm</td>
<td></td>
<td></td>
</tr>
<tr>
<td>&gt;12 cm</td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt;10 cm</td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt;12 cm</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Surface involvement</th>
<th>Primary</th>
<th>Metastasis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Absent</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Present</td>
<td></td>
<td></td>
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</table>

<table>
<thead>
<tr>
<th>Stage</th>
<th>Primary</th>
<th>Metastasis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Usually stage I</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Advanced stage</td>
<td></td>
<td></td>
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</table>

Lee et al, Am J Surg Pathol 2003

Primary versus metastasis: pitfalls

Gross:
Metastatic mucinous tumors can be
- Unilateral
- Large
- Grossly multicystic
- Smooth surface

Primary versus metastasis

Gross algorithm:
- Bilateral tumors of any size, unilateral <13 cm: Metastatic
- Unilateral > 13 cm: Primary

Application of this algorithm correctly identified 98% of primary tumors and 82% metastases

Common exceptions: Colorectal and endocervical carcinomas

Primary versus metastasis

Microscopic features

<table>
<thead>
<tr>
<th>Microscopic feature</th>
<th>Primary</th>
<th>Metastasis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pattern of growth</td>
<td>Expansile</td>
<td>Nodular</td>
</tr>
<tr>
<td>Destructive stromal invasion</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Ovarian hilar involvement</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Lymphovascular invasion</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Microscopic surface mucin</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Signet ring cells</td>
<td>No</td>
<td>Yes/No</td>
</tr>
<tr>
<td>Pseudomyxoma peritonei and ovarii</td>
<td>No</td>
<td>Yes</td>
</tr>
</tbody>
</table>
Metastatic colon carcinoma: Nodular, desmoplasia, infiltrative growth pattern

Metastatic colon carcinoma: Surface involvement

Metastatic gastric carcinoma: Signet ring cells

Metastatic colon carcinoma: Lymphovascular invasion
Primary versus metastasis: pitfalls

Microscopic:
“Maturation phenomenon”
Metastatic mucinous carcinomas can simulate
- Mucinous cystadenoma
- Borderline mucinous tumor
- Borderline mucinous tumor with intraepithelial carcinoma
- Borderline mucinous tumor with microinvasion

Primary versus metastasis

- Heterogeneous
- Highly differentiated areas adjacent to malignant areas
- Look for foci that may be suggestive of metastasis
- Consider submitting 2 sections per cm in difficult cases

Sampling is key!
Pseudomyxoma peritonei (PMP)

Two broad categories:
1. Low grade: Disseminated peritoneal adenomucinosis (DPAM)
2. High grade: Peritoneal mucinous carcinomatosis (PMCA)

Pseudomyxoma peritonei (PMP)

- Clinical entity
- Abundant extracellular peritoneal mucin
- May or may not contain epithelial cells
- Associated with a heterogeneous group of pathologic lesions

<table>
<thead>
<tr>
<th></th>
<th>DPAM (Disseminated peritoneal adenomucinosis)</th>
<th>PMCA (Peritoneal mucinous carcinomatosis)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cellularity</td>
<td>Scant</td>
<td>Moderate to abundant</td>
</tr>
<tr>
<td>Cytologic atypia</td>
<td>Minimal</td>
<td>Moderate to marked</td>
</tr>
<tr>
<td>Mitotic activity</td>
<td>Rare</td>
<td>Occasional to abundant</td>
</tr>
<tr>
<td>Lymph node involvement</td>
<td>Rare</td>
<td>Frequent</td>
</tr>
<tr>
<td>Parenchyma organ involvement</td>
<td>Rare</td>
<td>Frequent</td>
</tr>
<tr>
<td>Site of origin</td>
<td>Appendix - Cystadenoma - Hyperplastic polyp - Villous adenoma</td>
<td>Appendix/colon - Carcinoma</td>
</tr>
<tr>
<td>5 and 10 year survival</td>
<td>75% and 68%</td>
<td>14% and 3%</td>
</tr>
</tbody>
</table>

Ronnett et al, Cancer 2001
Pseudomyxoma peritonei (PMP)

Problems:
1. Pathologic terminology
   - Lack of consensus
   - Disseminated peritoneal adenomucinosis (DPAM) = Well differentiated adenocarcinoma = Low grade appendiceal mucinous neoplasm (LAMN) = Pseudomyxoma peritonei
2. PMP frequently involves ovaries, but virtually never originates there
Pseudomyxoma peritonei (PMP)

Take home message:
- Most cases of PMP are of appendiceal or intestinal origin
- Ovarian involvement is secondary

Exception: Mucinous tumors arising in ovarian teratomas

Ovarian mucinous tumors associated with mature cystic teratomas

- 2-11% of mature cystic teratomas
- Teratoma component may be focal
  1. Mucinous cystadenoma
  2. Mucinous borderline tumor
  3. Appendiceal type low grade mucinous neoplasm
  4. Goblet cell carcinoid
  5. Mucinous adenocarcinoma

- Variable immunophenotypes (may stain like a lower GI primary)
- May closely resemble appendiceal mucinous tumors
- Can lead to pseudomyxoma peritonei (PMP) and pseudomyxoma ovarii


Patient with pseudomyxoma peritonei, unilateral low grade mucinous tumor and no identifiable GI primary?

Additional sections from the ovary to identify a teratomatous component may be helpful!
Teratoma (caseous material and hair)

Microscopic: Ovarian mucinous tumors associated with mature cystic teratomas
Primary sites for mucinous neoplasms involving ovary

1. Appendix
2. Large intestine
3. Pancreas
4. Gallbladder
5. Stomach
6. Cervix
7. Urachus
8. Lung
9. Others

Appendiceal mucinous neoplasms

1. Mucinous adenoma:
   - Low grade, confined to appendix
2. Low grade mucinous neoplasm with low risk of recurrence:
   - Low grade, extra-appendiceal acellular mucin, no invasion
3. Low grade mucinous neoplasm with high risk of recurrence:
   - Low grade, extra-appendiceal neoplastic epithelium, no invasion
4. Mucinous adenocarcinoma:
   - High grade, complex, invasive


Low grade appendiceal mucinous neoplasms

Appendix:
- Low grade cytology
- Appearance similar to mucinous adenoma but with extra-appendiceal mucin (with or without neoplastic epithelium)
- Submit entire appendix

Low grade appendiceal mucinous neoplasms involving ovary

Gross:
- Bilateral (but may be unilateral)
- Large
- Multilocular
- Surface involvement
- Gross mucin

Microscopic:
- Tall columnar cells with mucin
- Minimal cytologic atypia and mitotic activity
- Mucin dissection into ovarian stroma (pseudomyxoma ovarii)

Can mimic ovarian mucinous cystadenoma or borderline tumor
Low grade appendiceal mucinous neoplasms involving ovary: distinction from primary

- Bilateral
- Surface involvement
- Pseudomyxoma peritonei
- Pseudomyxoma ovarii
- CK20>>CK7
- More CDX2
Low grade appendiceal mucinous neoplasm involving ovary: pseudomyxoma ovarii

Low grade appendiceal mucinous neoplasm

Low grade appendiceal mucinous neoplasm

CK20

CK7
**Intestinal adenocarcinoma metastatic to ovary**

- Predominantly large intestine (colorectal)
- Most common metastatic tumor involving ovary
- Can mimic endometrioid and mucinous carcinomas of the ovary
- May present with elevated CA-125
- May first present with an ovarian mass

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**Intestinal adenocarcinoma metastatic to ovary: distinction from primary**

- Small, bilateral with surface involvement
- But frequently large, unilateral with smooth surface
- Solid and cystic

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**Intestinal adenocarcinoma metastatic to ovary: distinction from primary**

- Nodular
- Confluent/cribriform glandular
- “Garlanding”
- Dirty necrosis
- Infiltrative growth and desmoplasia (often focal)
- Lymphovascular invasion
- High grade cytology- but may have foci of extremely well differentiated mucinous epithelium

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**Immunohistochemistry**

- CK20 >> CK7
- CDX2:
  - Not specific but typically more compared to ovarian primary

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Metastatic colon carcinoma: Nodular, desmoplasia, infiltrative growth pattern
Pancreatobiliary system

- Ovarian tumor can present first or synchronously with the pancreatic tumor
- Can simulate an ovarian primary grossly and microscopically
- Frequently unilateral, cystic and large, with smooth surface
- Microscopically can simulate mucinous cystadenomas or borderline tumors (can show mixture of benign to malignant epithelium)


Pancreatobiliary carcinoma: distinction from ovarian primary

- Bilaterality (89%)
- Small size (mean size <12 cm)
- Multinodular growth (63%)
- Surface implants
- Focal infiltrative invasion
- Presence of extra-ovarian disease

**Immunohistochemistry**

- CK7>>CK20
- Loss of SMAD4 (DPC4)
- Positive for CA-125

**DPC4/SMAD4**

- SMAD4 (DPC4) somatic alterations in 55% of pancreatic cancers and 10-35% of colon carcinomas
- Immunohistochemical loss of expression
  - Pancreatic carcinoma (46%-61%)
  - Colon carcinoma (11%)
  - Ovarian mucinous carcinoma (0%)

-Loss of DPC4 expression is very helpful to make a diagnosis of pancreatobiliary carcinoma
-But retained DPC4 expression does not exclude pancreatobiliary origin

*Ji et al, Int J Gynecol Pathol 2002
Meriden et al, Am J Surg Pathol 2011*
Metastatic pancreatic carcinoma

Metastatic pancreatic carcinoma: Omentum

Metastatic pancreatic carcinoma: Loss of SMAD4/DPC4
Krukenberg tumor of the ovary

- Signet ring cells 10%
- Tubular type
- Average age 45 years

- Most common primary sites include (may be occult):
  - Stomach
  - Appendix
  - Colon/rectum
  - Gall bladder/biliary system
  - Breast

Ovarian primary (extremely rare and diagnosis of exclusion!)
Metastatic gastric carcinoma

- HPV associated and non HPV (minimal deviation adenocarcinoma) associated
- Metastases to ovary uncommon
- Usually associated with previously or concurrently diagnosed cervical carcinomas but sometimes cervical tumor diagnosed subsequently
- Typically deeply invasive cervical primary
- Also reported in cases with microinvasion and cases of AIS alone without unequivocal stromal invasion

Chang et al, Int J Gynecol Pathol 2009

Uterine cervix

- Frequently mimic primary ovarian mucinous neoplasm on gross and microscopic evaluation
  - Gross: Large, unilateral, multicystic, smooth capsule (frequently violate algorithm)
  - Microscopic: Borderline tumor like growth pattern, expansile/cribriform growth pattern

Only 3/29 cases bilateral with infiltrative growth pattern

Uterine cervix: distinction from ovarian primary

- Gross and/or microscopic features of metastatic tumor

HPV associated cervical tumors:
  - Appropriate clinical history
  - Cytologic features: nuclear characteristics, apical mitoses and apoptotic bodies
  - Ancillary studies: p16, HPV DNA in situ hybridization

Non-HPV associated tumors:
  - Clinical history
  - Morphologic similarity with cervical primary

Immunohistochemistry

- CK7>>CK20
- HPV-ISH (limited sensitivity)

- p16
  - HPV associated endocervical adenocarcinomas: Strong diffuse staining (90-100%)
  - Non HPV associated endocervical carcinomas: Negative or focal staining
  - Primary ovarian tumors and metastases from noncervical primary sites: Predominantly p16 negative or focal

Metastatic endocervical adenocarcinoma

Determining site of origin for mucinous tumors: Immunophenotype

<table>
<thead>
<tr>
<th></th>
<th>Ovary</th>
<th>Appendix/colorectal</th>
<th>Pancreatobiliary tract</th>
<th>Uterine cervix</th>
</tr>
</thead>
<tbody>
<tr>
<td>CK7</td>
<td>++</td>
<td>-/+</td>
<td>++</td>
<td>++</td>
</tr>
<tr>
<td>CK20</td>
<td>+/-</td>
<td>++</td>
<td>-/+</td>
<td>-/+</td>
</tr>
<tr>
<td>CDX2</td>
<td>+/-</td>
<td>++</td>
<td>+/-</td>
<td>+/-</td>
</tr>
<tr>
<td>SMAD4</td>
<td>+</td>
<td>+</td>
<td>Loss in ~50%</td>
<td>+</td>
</tr>
<tr>
<td>p16</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>++</td>
</tr>
</tbody>
</table>
Tumor type

- Primary ovarian mucinous tumors (n=42)
- Metastatic lower gastrointestinal tract adenocarcinomas (n=29)
- Metastatic upper gastrointestinal tract adenocarcinomas (n=19)

<table>
<thead>
<tr>
<th>Tumor type</th>
<th>Primary ovarian mucinous tumors</th>
<th>Metastatic lower gastrointestinal tract adenocarcinomas</th>
<th>Metastatic upper gastrointestinal tract adenocarcinomas</th>
</tr>
</thead>
<tbody>
<tr>
<td>CK7+/CDX2-</td>
<td>25 (60%) 2 (7%) 12 (26%)</td>
<td>1 (26%)</td>
<td>2 (11%)</td>
</tr>
<tr>
<td>CK7+/CDX2+</td>
<td>5 (17%) 6 (21%)</td>
<td>14 (83%)</td>
<td>12 (63%)</td>
</tr>
<tr>
<td>CK7-/CDX2+</td>
<td>1 (5%)</td>
<td>10 (57%)</td>
<td>5 (26%)</td>
</tr>
<tr>
<td>CK7+/CK20-</td>
<td>1 (5%)</td>
<td>0 (-)</td>
<td>11 (57%)</td>
</tr>
<tr>
<td>CK7-/CK20-</td>
<td>2 (5%)</td>
<td>25 (86%)</td>
<td>11 (57%)</td>
</tr>
</tbody>
</table>

Determining site of origin for mucinous tumors: Immunophenotype

Equivocal cases (overlapping features)

- Diagnose as “Mucinous carcinoma involving ovary” and discuss the differential of primary versus metastasis
- “May be accepted as an ovarian primary if the possibility of extra-ovarian origin is clinically and radiologically excluded”

Intraoperative assessment of ovarian mucinous lesions

- Radiologic
- Clinical
- Gross
- Microscopic

Communication between surgeon and pathologist is key
**Intraoperative assessment of ovarian mucinous lesions**

**Clinical history**
- Prior history of mucinous neoplasm: Consider metastasis

**Radiology**
- Bilateral ovarian involvement
- Evidence of extra-ovarian disease
- Lesion in another organ

**Operative findings**
- Status of contralateral ovary
- Ovarian surface status
- Mucin or tumor in peritoneal cavity
- Appearance of appendix

**Gross/microscopic features**

- Consider metastasis

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**Gross features:**
- Bilateral tumors of any size, unilateral <13 cm: Metastatic
- Unilateral > 13 cm: Primary

**Microscopic features - consider metastasis:**
- Nodular growth
- Desmoplasia
- Infiltrative growth
- Signet ring cells
- Mucin dissection
- Surface involvement

If features suggestive of metastasis: Ask surgeon to examine for another primary site particularly in the gastrointestinal tract (appendix)

**Implications of FS diagnosis:**
- Mucinous cystadenoma: No staging
- Mucinous cystadenoma with focal borderline features: May or may not stage
- Mucinous borderline tumor: Staging (extent may depend on patient age)
- Mucinous carcinoma: Staging
- Metastatic tumors: No staging
Intraoperative assessment of ovarian mucinous lesions

- Ovarian mucinous lesion
  - Large, unilateral
  - Small, bilateral
  - Low grade
  - High grade
  - Favor primary
  - Favor metastasis
  - Borderline/carcinoma
  - Cystadenoma

Staging
- No staging

Mural nodules in ovarian mucinous neoplasms

- Rare
- Can occur in cystic mucinous tumors (cystadenomas, borderline tumors and carcinomas)
- Single or multiple, variable size
- Heterogeneous entities

Benign: Sarcoma like mural nodules (SLMNs)
Malignant:
1. Sarcomas
2. Anaplastic carcinomas
3. Carcinosarcomas

Mural nodules in mucinous cystic neoplasms

<table>
<thead>
<tr>
<th></th>
<th>Sarcoma like mural nodules</th>
<th>Anaplastic carcinoma</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of nodules</td>
<td>1 to many</td>
<td>Usually 1</td>
</tr>
<tr>
<td>Size</td>
<td>Small</td>
<td>Large</td>
</tr>
<tr>
<td>Circumscription</td>
<td>Good</td>
<td>Poor</td>
</tr>
<tr>
<td>Necrosis</td>
<td>Uncommon</td>
<td>Common, extensive</td>
</tr>
<tr>
<td>Cell composition</td>
<td>Heterogeneous</td>
<td>Homogeneous</td>
</tr>
<tr>
<td>Inflammation</td>
<td>Marked</td>
<td>Rare</td>
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<tr>
<td>Epulis type giant cells</td>
<td>Common, abundant</td>
<td>Uncommon, focal</td>
</tr>
<tr>
<td>Spindle cells</td>
<td>Common</td>
<td>Occasional (spindle cell carcinoma)</td>
</tr>
<tr>
<td>Cytokeratin staining</td>
<td>Negative/weak</td>
<td>Positive</td>
</tr>
</tbody>
</table>


Anaplastic carcinoma in ovarian mucinous neoplasms

- Prognosis dependent on stage
- No correlation with histology, size, type of mucinous tumor, presence of lymphovascular invasion
- May not be associated with adverse prognosis when stage Ia
- May recur as high grade carcinoma/sarcoma

Mucinous cystadenoma with sarcoma-like mural nodule

Mucinous cystadenoma with sarcoma-like mural nodule

Mucinous cystadenoma with sarcoma-like mural nodule

Mucinous cystadenoma with sarcoma-like mural nodule – CD68
Summary

- Focal epithelial proliferation can be seen in mucinous cystadenoma
- Mucinous borderline tumors are benign and stage 1A (exclude metastasis if considering a diagnosis of advanced stage mucinous borderline tumor!)
Summary

- Primary ovarian mucinous carcinomas are uncommon
- Metastatic mucinous carcinomas to the ovary are more common
- Metastatic mucinous tumors to the ovary can simulate ovarian primaries
- Distinction has therapeutic and prognostic significance

Summary

- Gross and microscopic evaluation very helpful to differentiate between primary and metastasis (but remember the pitfalls)
- Adequate sampling important
- Immunophenotype helpful in some settings

Summary

- Pseudomyxoma peritonei is frequently of appendiceal origin and virtually never of ovarian origin (exception mucinous tumor arising in teratoma)
- Can be high grade or low grade (no consensus on terminology)
- Examine entire appendix carefully in cases of pseudomyxoma

Summary

- Mural nodules can be associated with mucinous cystic tumors
- These can be sarcoma-like or malignant (sarcomatoid carcinoma, anaplastic carcinoma or carcinosarcoma)
Thank you!