

AMSER Rad Path Case of the Month:

64 yo female presents with facial flushing

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Patient Presentation

HPI:

- 64-year-old female with no significant PMH presents with intermittent facial flushing, heart palpitations, and fatigue for 5 years duration. ROS negative for changes in bowel habits.

PSH:

- Total abdominal hysterectomy (2007) for uterine leiomyoma. Benign pathology confirmed.

Physical Exam:

- Left adnexal mass and fullness present. No tenderness observed.

Pertinent Labs

- 5 HIAA (urine) = **25.2** mg/24hr (*Ref* <0.6)

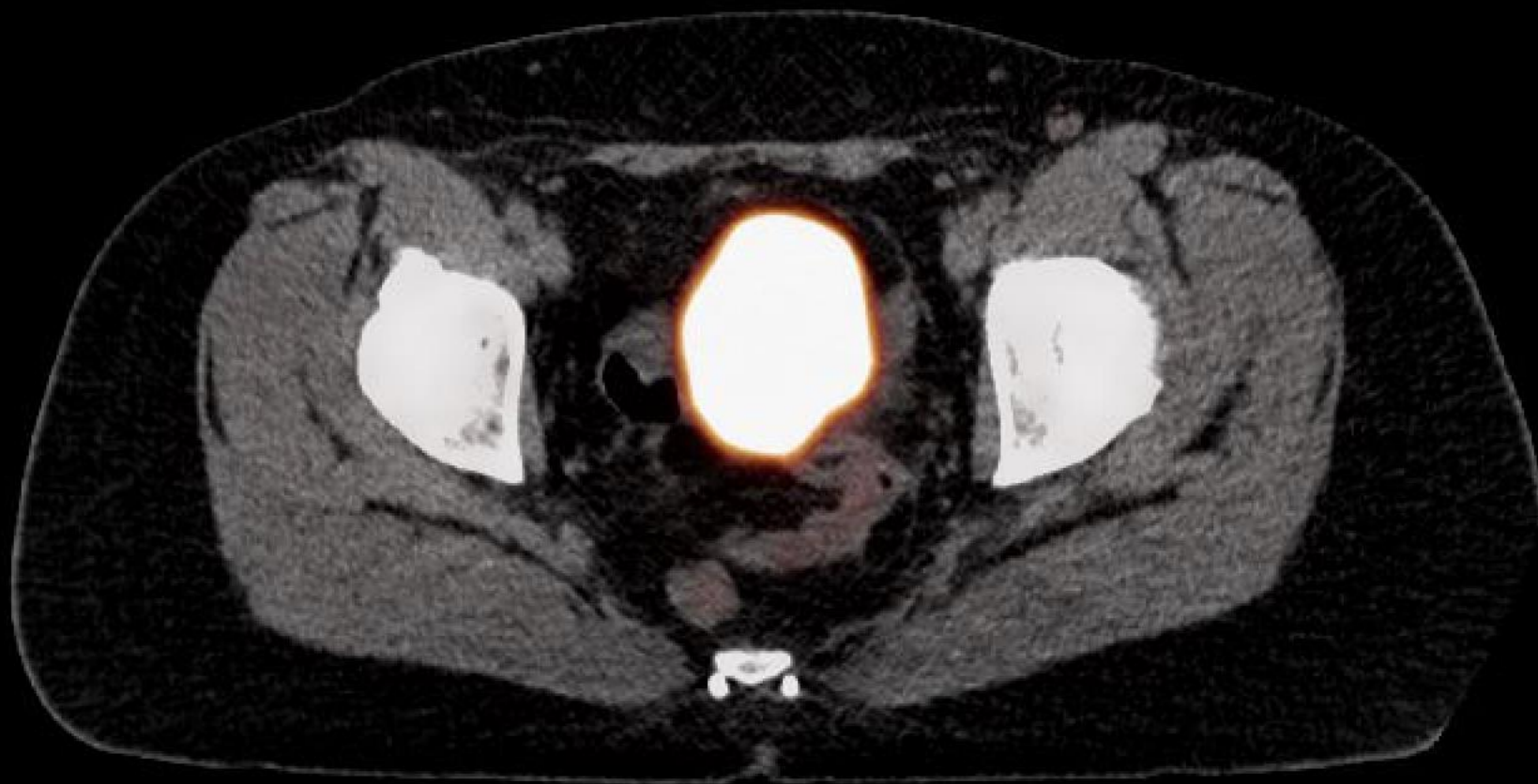
What Imaging Should We Order?

Due to high suspicion for a neuroendocrine tumor given elevated 5 HIAA, the following modalities were ordered by the oncologist:

1st: 68-Gallium Dotatate PET CT

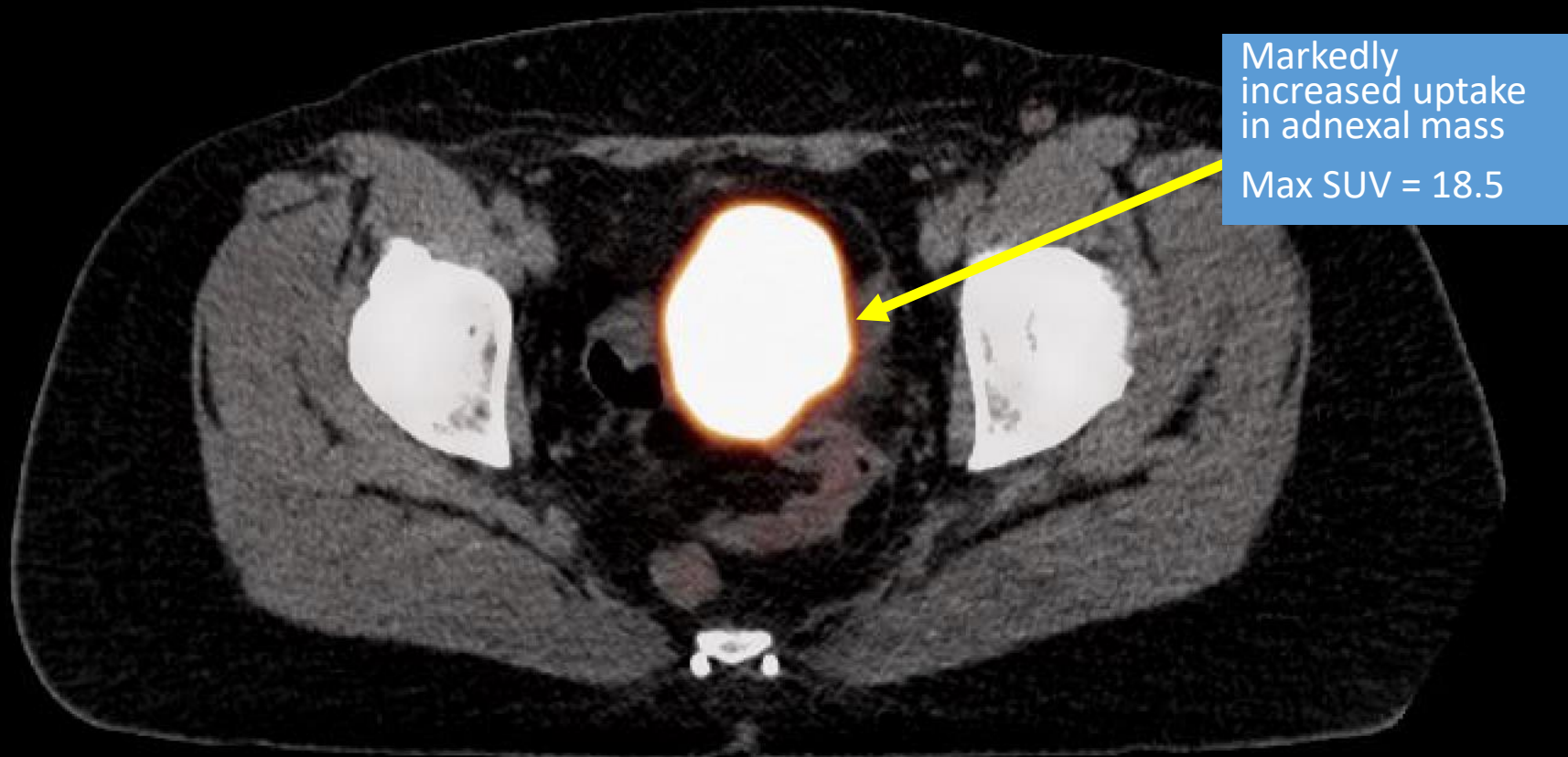
2nd: MRI with and without IV contrast

Findings (unlabeled)



Axial 68-Gallium DOTATATE PET CT

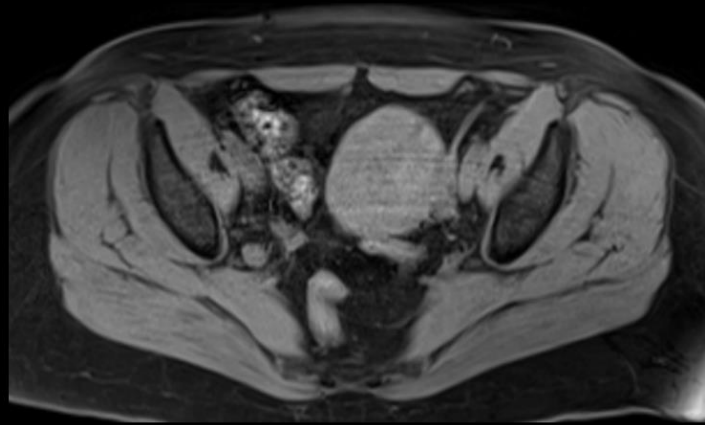
Findings (labeled)



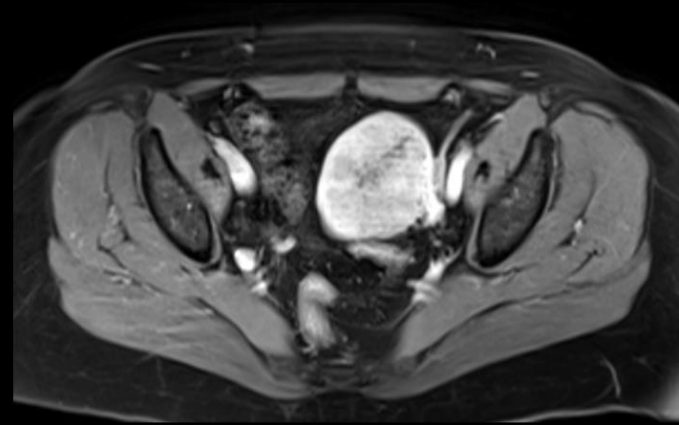
Markedly increased uptake in adnexal mass
Max SUV = 18.5

Axial 68-Gallium DOTATATE PET CT

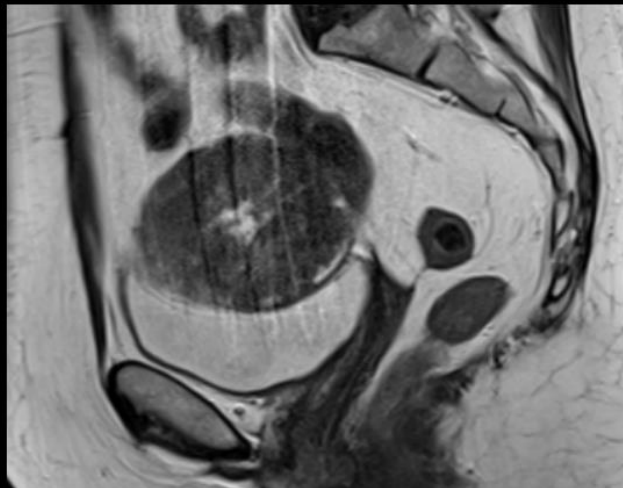
Findings (unlabeled) – MRI



Axial T1



Axial T1 with contrast

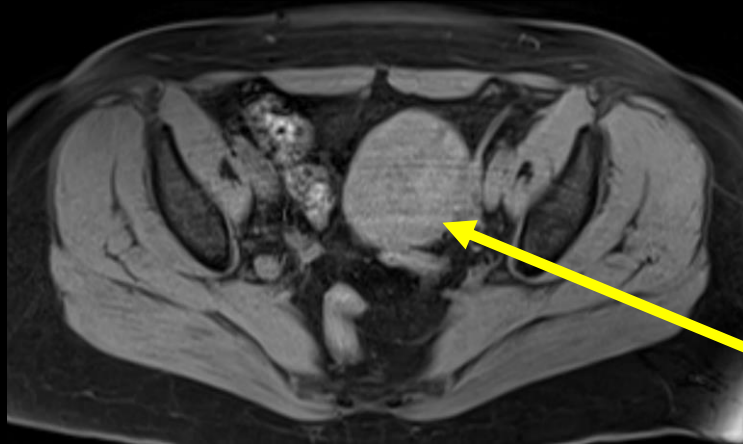


Sagittal T2



Axial T2 Sigmoid Colon

Findings: (labeled) - MRI



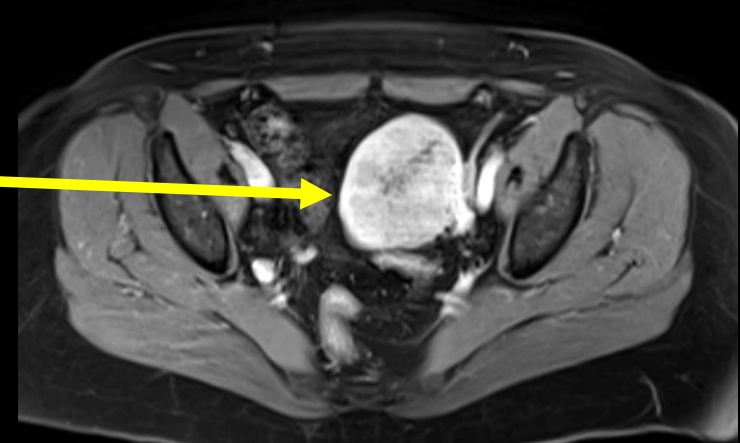
Axial T1

Heterogeneous well-circumscribed solid enhancing left adnexal mass

Peripheral T1 hyperintense component

Predominantly T2 hypointense with central cystic hyperintensity

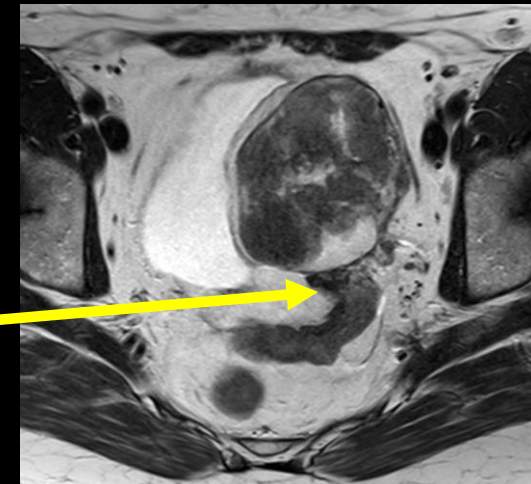
Mass directly abuts sigmoid colon and left vaginal cuff apex



Axial T1 with Contrast



Sagittal T2



Axial T2

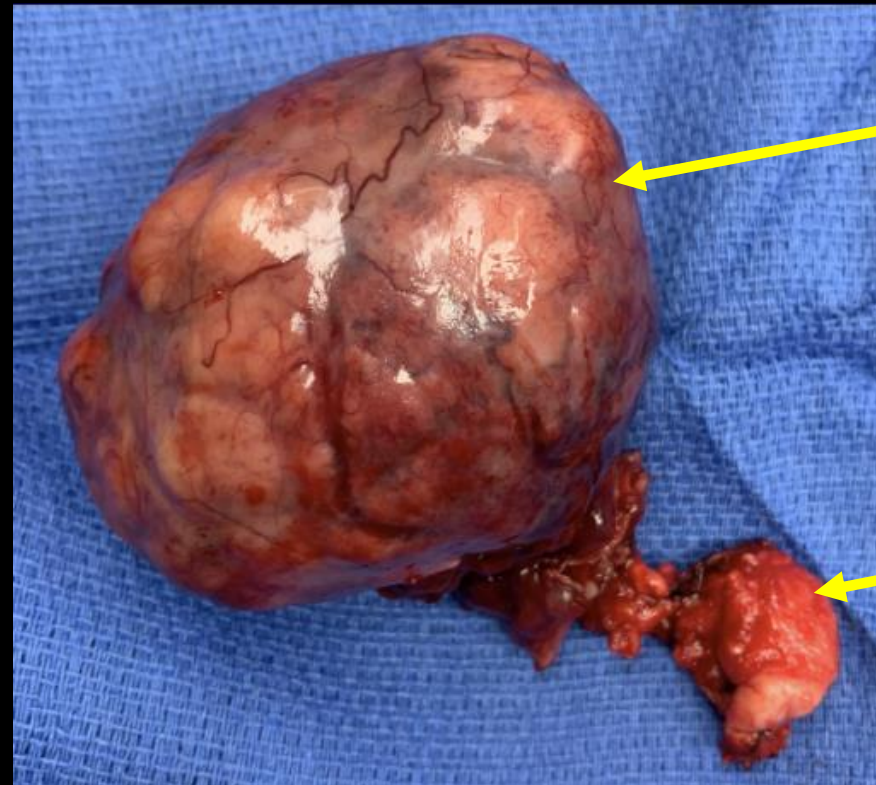
Gross Pathology

Surgery Performed:

Exploratory laparotomy - resection of pelvic mass
Bilateral salpingo-oophorectomy
Trachelectomy
Bilateral pelvic and para-aortic lymphadenectomy
Omentectomy
Repair of rectal serosal laceration
Cystoscopy

Intraoperative Findings:

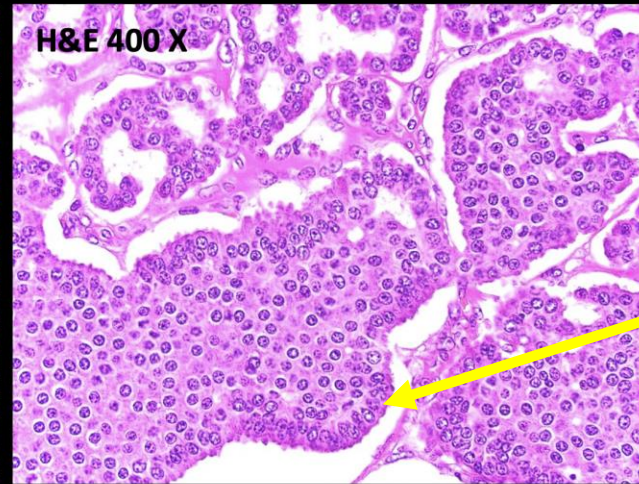
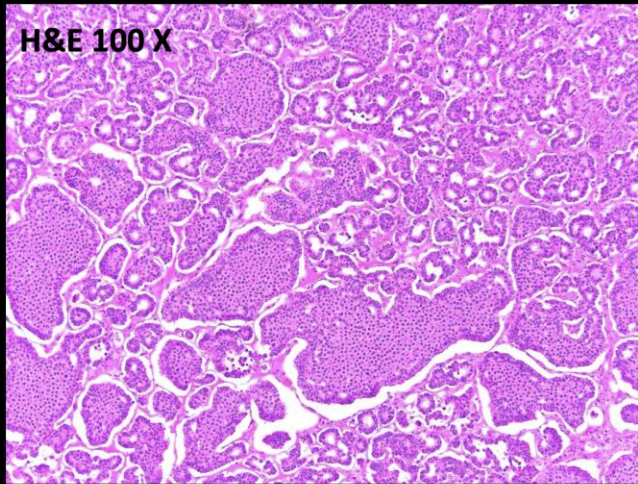
Survey of the abdomen revealed pelvic mass 10+cm in diameter, encapsulated, and originating from the left tubo-ovarian structures tightly adhered to rectosigmoid colon



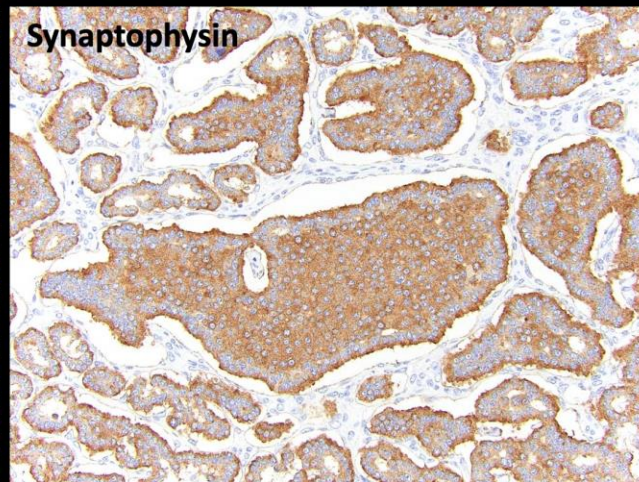
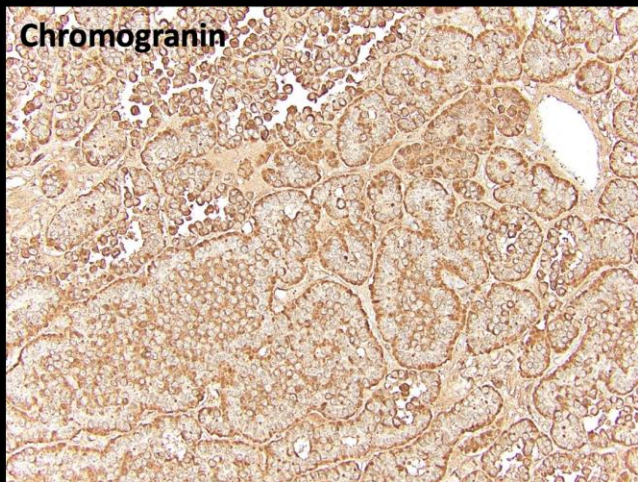
Adnexal mass

Residual cervix

Surgical Pathology



Salt-and-pepper chromatin
Argentaffin present (reddish brown cytoplasmic granules)



Positive immunoreactivity
Negative stain for PAX-8, TTF-1, and inhibin

Final Dx:

Stage IA Insular Ovarian Carcinoid

Case Discussion – Ovarian Carcinoid

- Primary ovarian carcinoid tumors are rare (<0.1% of all ovarian carcinomas)
- Insular (most common ~50%), trabecular, strumal, and mucinous subtypes
- Often arise within a cystic teratoma
- Associated with a high expression of somatostatin receptors
- Pertinent labs = elevated 5HIAA
- 1/3 associated with carcinoid syndrome: flushing, diarrhea, wheezing, right sided valvular heart disease (due to excess production of serotonin)
 - Ovarian carcinoids drain directly into the systemic circulation, bypassing the liver, which inactivates intestinal carcinoids
- Treated with total abdominal hysterectomy, bilateral oophorectomy, and omentectomy

Case Discussion – Ovarian Carcinoid: Rad/Path

Nuclear Medicine Findings:

- Octreotide scan (old) = somatostatin receptor scintigraphy with DTPA-octreotide
 - Increased uptake in ovarian carcinoid tumors
- ⁶⁸Ga-DOTATATE PET/CT (new) = Gallium labeled somatostatin ligands
 - Increased uptake in ovarian carcinoid tumors

MR Findings:

- T1 = low signal intensity solid mass
- T1 C+ = variable enhancement of solid mass
- T2 = intermediate signal intensity solid mass

Gross Pathology:

- Well circumscribed, firm yellow-tan colored mass

Microscopic Features:

- Positive immunohistochemistry for neuroendocrine markers: chromogranin A, synaptophysin, CD56
- Insular Subtype (most common ~50%)
 - Uniform polygonal cells
 - Argentaffin granules
 - Hyalinized surrounding connective tissue due to fibrogenic effect of serotonin

Differentiation from other solid malignant ovarian tumors may be difficult thus nuclear medicine scans and pathology are essential to diagnosis

References:

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Díaz-Montes TP, Rosenthal LE, Bristow RE, Grumbine FC. Primary insular carcinoid of the ovary. *Gynecol Oncol*. 2006;101(1):175-178. doi:10.1016/j.ygyno.2005.10.015

Bozkurt, M.F., Virgolini, I., Balogova, S. *et al*. Guideline for PET/CT imaging of neuroendocrine neoplasms with ⁶⁸Ga-DOTA-conjugated somatostatin receptor targeting peptides and ¹⁸F-DOPA. *Eur J Nucl Med Mol Imaging* **44**, 1588–1601 (2017). <https://doi.org/10.1007/s00259-017-3728-y>