

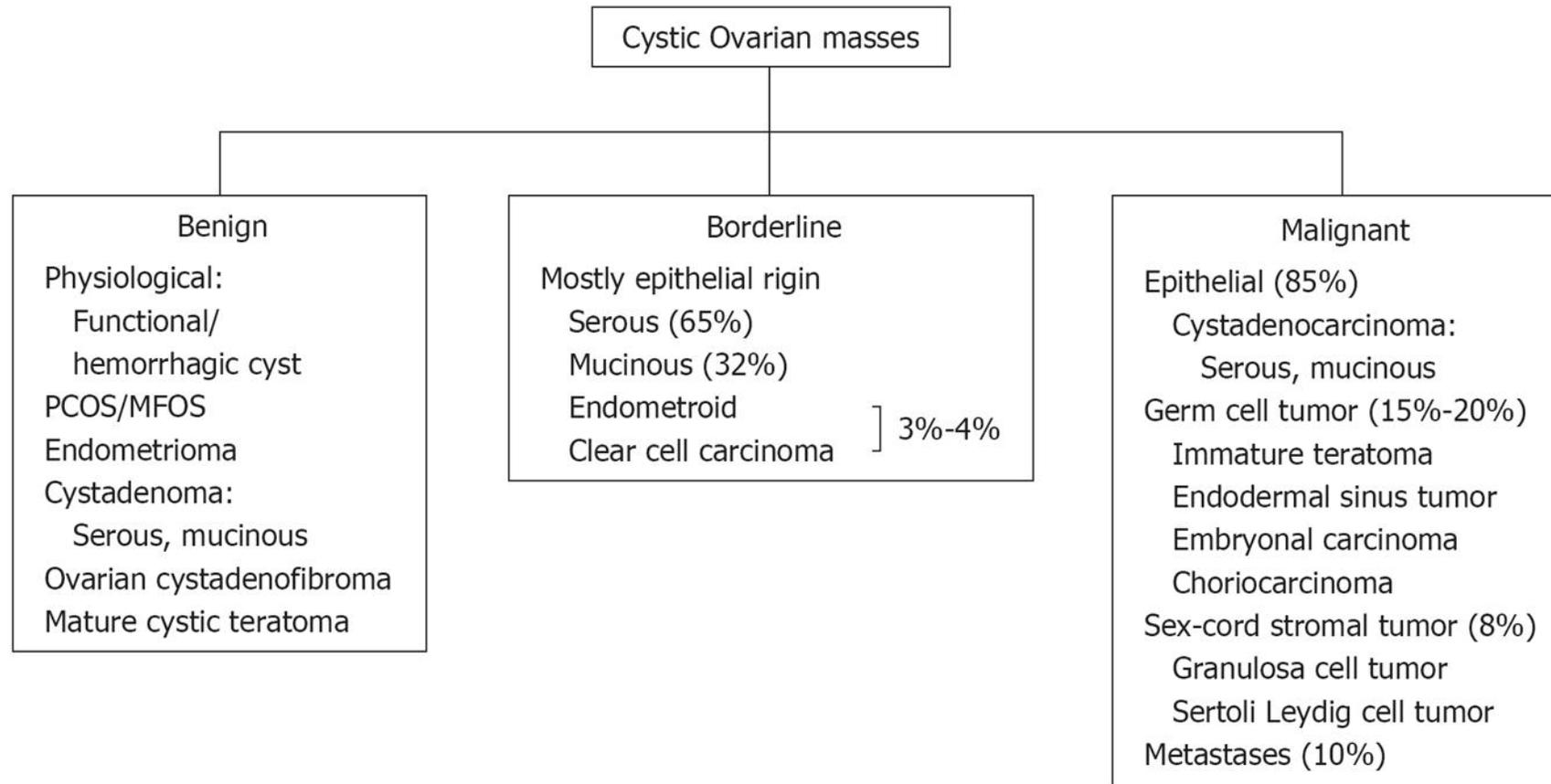
RADY 403: Ovarian Teratoma

Megan Foster, MS4
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Focused Patient History and Workup

- JC is a 7-year-old female with a history of asthma who presented to UNC via a referral from her endocrinologist with a history of vaginal bleeding which started 2 months prior to presentation and breast development beginning a year ago concerning for precocious puberty.
- At OSH a year ago, an abdominal X-ray was performed and was notable for an area of calcification in the LLQ concerning for a **swallowed tooth vs. teratoma**. The patient was lost to follow-up.
- A transabdominal US at an OSH was performed a couple of months prior showing a **9.6 cm cystic mass in the left adnexa with internal calcifications likely consistent with a germ cell tumor**. She was also found to have **elevated LH and FSH** with no detectable estrogen or testosterone.
- She was referred to UNC pediatric surgery for r/o of **malignant vs. benign ovarian mass**.
- Upon admission:
 - Vitals: unremarkable
 - ROS: Endorsed vaginal bleeding and vaginal discharge. No N/V, wt. loss, constipation
 - PE: Normal weight. Umbilical hernia.
 - Labs: AFP, CA 125, bHCG, amylase, lipase, CBC, Chem 10 are within normal limits.

Differential Diagnosis of Cystic Ovarian Mass

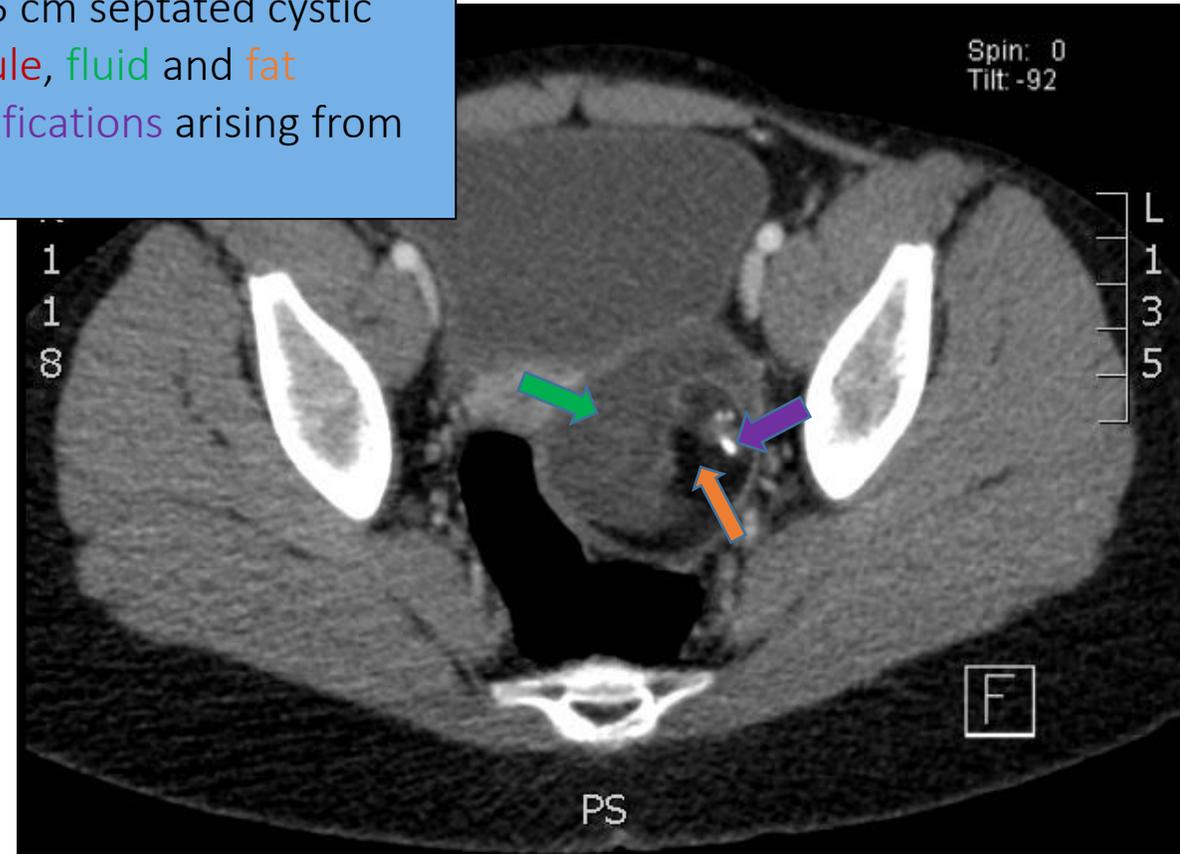
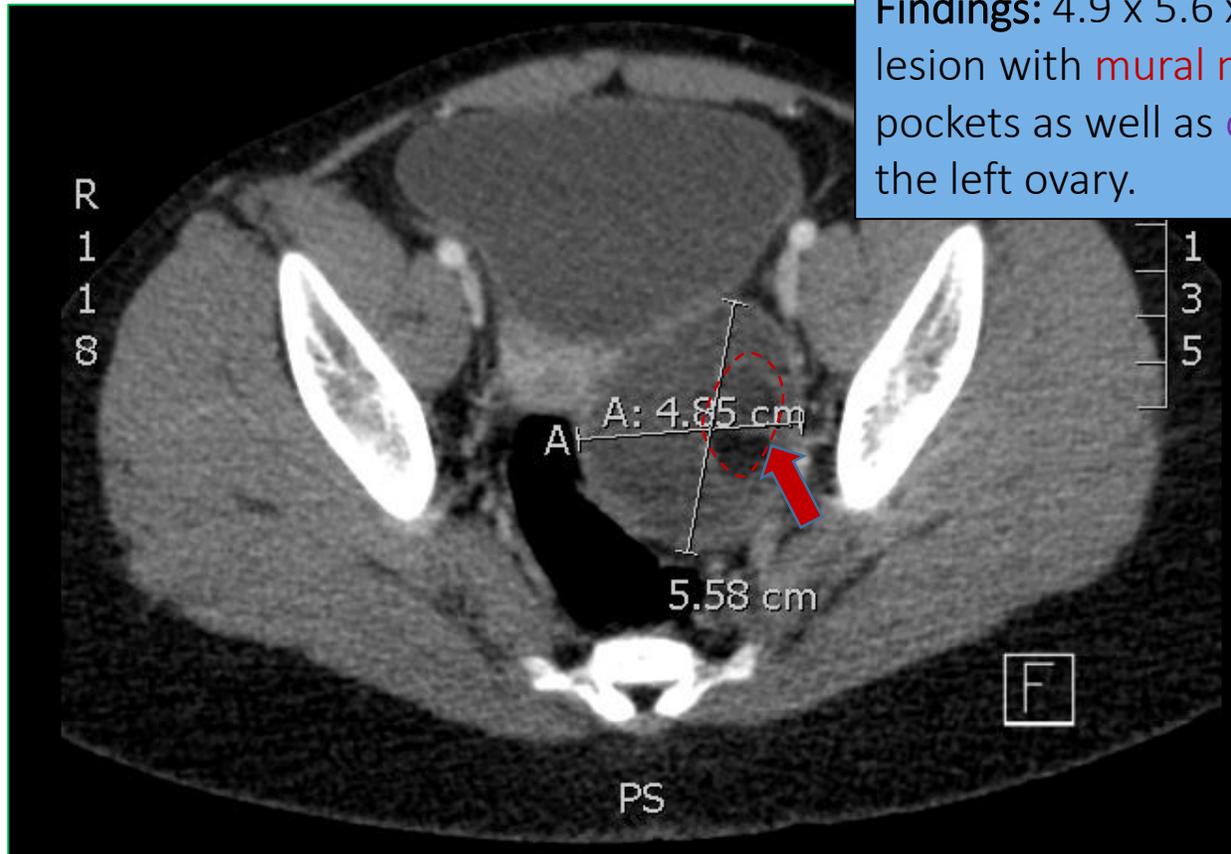


List of imaging studies

- KUB- done at OSH a year prior
- Transabdominal US- done at OSH 2 months prior
- CT abdomen and pelvis with contrast
- CT chest with contrast- **normal without any suspicious findings suggestive of metastatic disease**

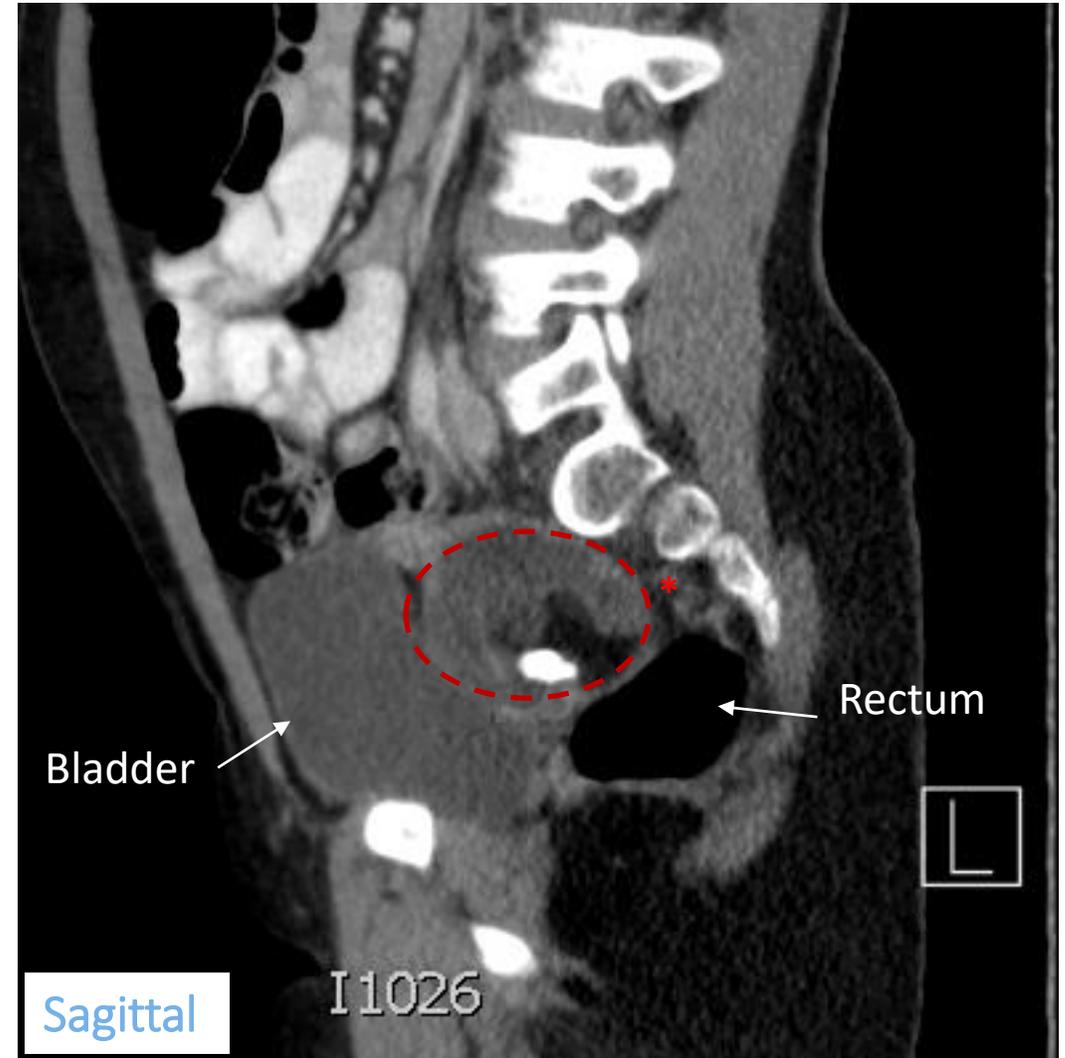
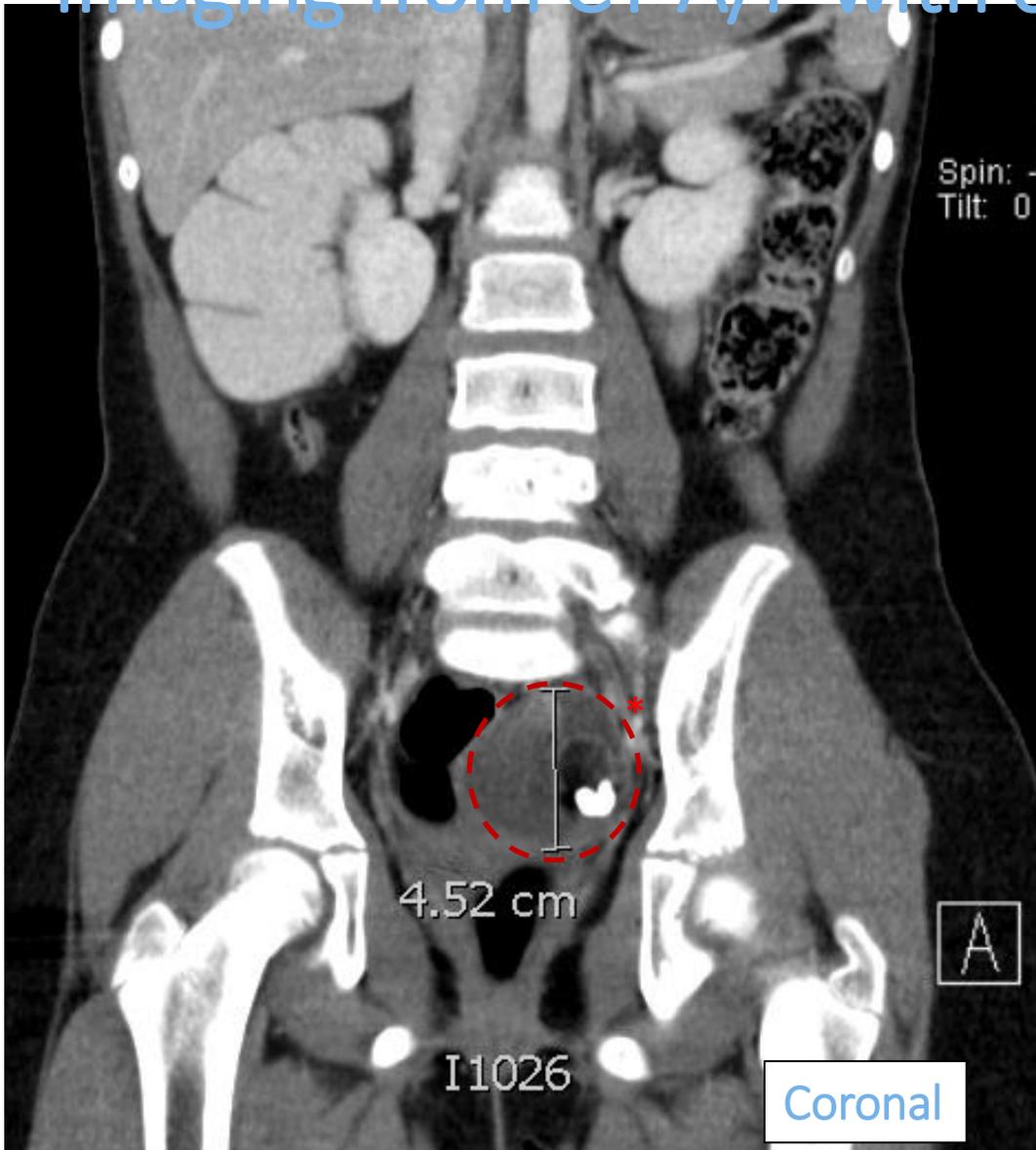
Imaging from CT A/P with contrast - axial

Findings: 4.9 x 5.6 x 4.5 cm septated cystic lesion with mural nodule, fluid and fat pockets as well as calcifications arising from the left ovary.



Impression: Left ovarian mass consistent with imaging characteristics of mature cystic teratoma of the ovary.

Imaging from CT A/P with contrast - axial



* Red-dotted line: consistent with mature cystic teratoma

Treatment and Outcome

- Pediatric heme/onc was consulted and discussed concern for adrenal cortical carcinoma due to her presenting with precocious puberty.
- Baseline labs and imaging most supportive of teratoma and exploratory laparotomy and left oophorectomy were performed 1 day after admission.
- She was ready for discharge on POD2.
- Pediatric (ultrasensitive) LH, pediatric FSH, pediatric testosterone and ultra sensitive estradiol, inhibin B were all ordered on day of discharge and came back as normal.
- **Surgical pathology report:** Mature cystic teratoma (dermoid cyst).
- **Cytology report:** Degenerating mesothelial cells. No malignant cells identified.
- Upon chart review, there has been no further work-up for JC's precocious puberty.

Discussion Precocious puberty

- Defined as pubertal changes before the age of 8 in females and 9 in males.²
- Precocious puberty may be central (gonadotropin-dependent) or peripheral (gonadotropin-independent).²
- Central precocious puberty (CPP) is due to early maturation of hypothalamo-pituitary gonadal (HPG) axis and may be due to CNS tumors/ lesions, environmental estrogens, or idiopathic (most common).²
- Peripheral PP is caused by excess estrogen and may be caused by McCune-Albright syndrome, primary hypothyroidism, ovarian tumors, or idiopathic.²

Discussion D Dx Ovarian mass and precocious puberty

- *Remember, JC presented after a year long history of premature vaginal bleeding and breast budding with high LH and FSH.*
- Children with precocious puberty and ovarian mass have a high likelihood of malignancy.⁴
- A tumor marker panel: AFP, hCG, CA-125 is performed and if positive is concerning for malignancy, but if normal does NOT exclude malignancy.⁴
- *Remember, those tumor markers were all normal for JC.*
- Premenarchal girls may present with signs of isosexual precocious puberty (as seen in JC with breast development and vaginal bleeding) which is concerning for sex cord-stromal tumors, gonadoblastomas, and rarely malignant germ cell tumors.⁴

Discussion Mature cystic teratoma (MCT) of ovary

- Ovarian tumors are rare in the pediatric population accounting for 1-2% of all tumors in this population.³
- Mature in this context refers to being benign, as opposed to the immature, malignant teratoma composed of neuronal tissue.³
- Histopathology: Contain mature tissue of ectoderm (eg, hair follicles, skin, sebaceous glands), mesoderm (eg, muscle and urinary system), and endoderm (eg, gastrointestinal, lung).⁴
- Ovarian germ cell tumors (OGCTs) (e.g. **mature teratoma [benign]**, immature teratoma [malignant], gonadoblastoma [benign], dysgerminoma [malignant]) are most common of the ovarian tumors.⁴
- OGCTs arise primarily in women 10-30 years of age.⁴

Discussion OGCTs³

- OGCTs often may produce hormones such as beta subunit of human chorionic gonadotropin (hCG) or alpha-fetoprotein (AFP).
- Clinical manifestations may include:
 - Enlarged abdomen- from large mass, ascites, or both
 - Abdominal pain- from rupture or ovarian torsion
 - Precocious puberty, abnormal vaginal bleeding- assumed to be due to beta- hCG production
 - Symptoms of pregnancy- from hCG production
 - Nausea, vomiting, loss of appetite, weight loss, constipation, urinary frequency

Tumor Marker	Associated Ovarian Tumors
AFP	Yolk sac tumor
	Immature teratoma
	Embryonal carcinoma
	Sertoli-Leydig cell tumor (rare)
β-hCG	Choriocarcinoma
	Embryonal carcinoma
	Dysgerminoma (rare)
LDH	Dysgerminoma
CA-125	Epithelial tumors
Inhibin	Granulosa cell tumor

GCTs are the most common ovarian tumor, and embryonal carcinoma should be on the differential of child presenting with precocious puberty and an adnexal mass. In the case of JC, beta-hCG was negative, but remember that tumor markers are not always present.

ACR Appropriateness Criteria¹

Variant 7:

Adnexal mass, highly suspicious for malignancy, no acute symptoms. Premenopausal and postmenopausal. Initial follow-up.

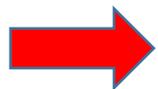
Although JC had imaging findings on US suggestive of a mature teratoma, with the history of precocious puberty, the adnexal mass was highly suspicious for malignancy.

Use of CT in this case was appropriate and beneficial in surgical planning.

Procedure	Appropriateness Category	Relative Radiation Level
CT abdomen and pelvis with IV contrast	Usually Appropriate	☼☼☼
MRI pelvis without and with IV contrast	Usually Appropriate	○
US pelvis transabdominal	May Be Appropriate	○
US pelvis transvaginal	May Be Appropriate	○
CT pelvis with IV contrast	May Be Appropriate (Disagreement)	☼☼☼
CT pelvis without and with IV contrast	May Be Appropriate (Disagreement)	☼☼☼☼
FDG-PET/CT skull base to mid-thigh	May Be Appropriate	☼☼☼☼
MRI pelvis without IV contrast	May Be Appropriate	○
US duplex Doppler pelvis	May Be Appropriate (Disagreement)	○
CT abdomen and pelvis without IV contrast	May Be Appropriate	☼☼☼
CT abdomen and pelvis without and with IV contrast	Usually Not Appropriate	☼☼☼☼
CT pelvis without IV contrast	Usually Not Appropriate	☼☼☼

Discussion Transabdominal US

- Initial choice in children and adolescents because of lack of ionizing radiation, ease of use, wide availability, and does not require sedation.^{1,5}
- Useful in determining tumor size and whether the tumor is solid, cystic, or complex.⁵
- Endovaginal US may be used in sexually active adolescents.¹

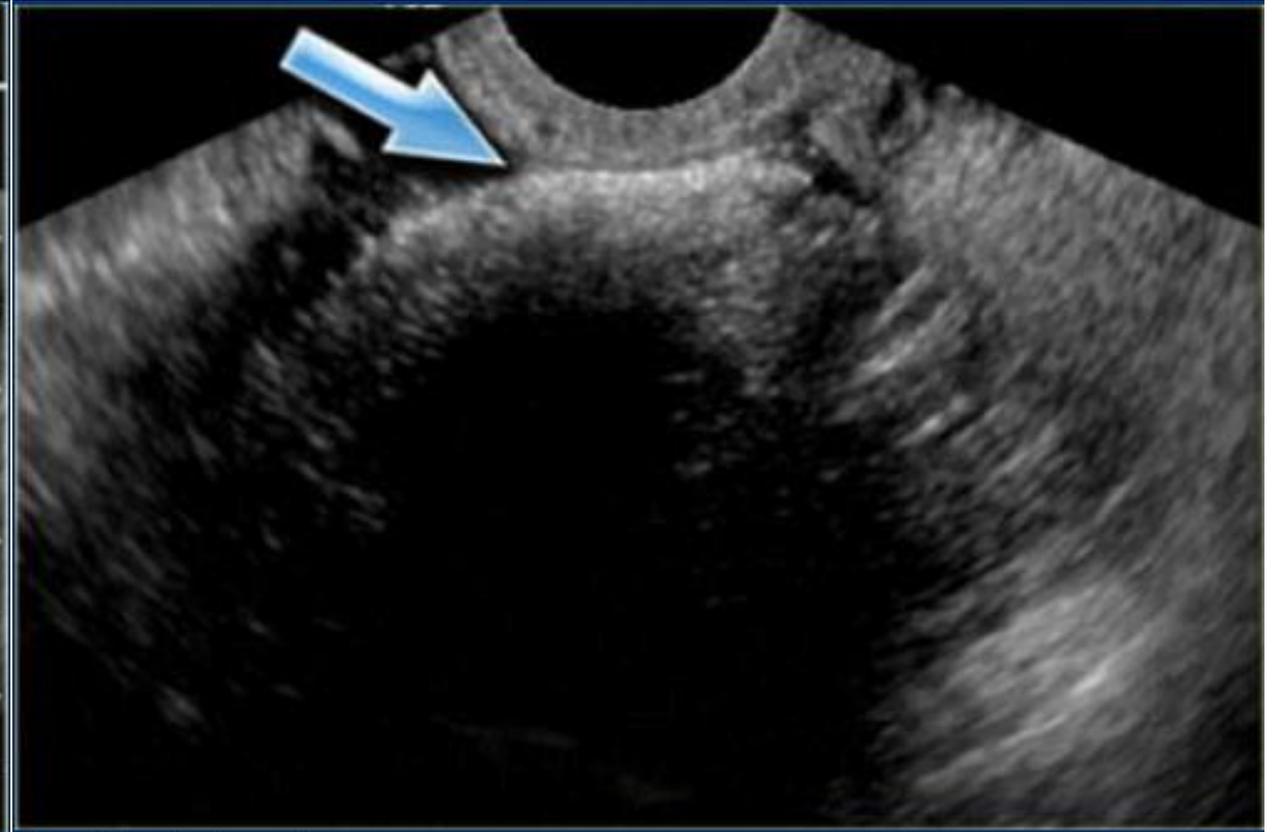
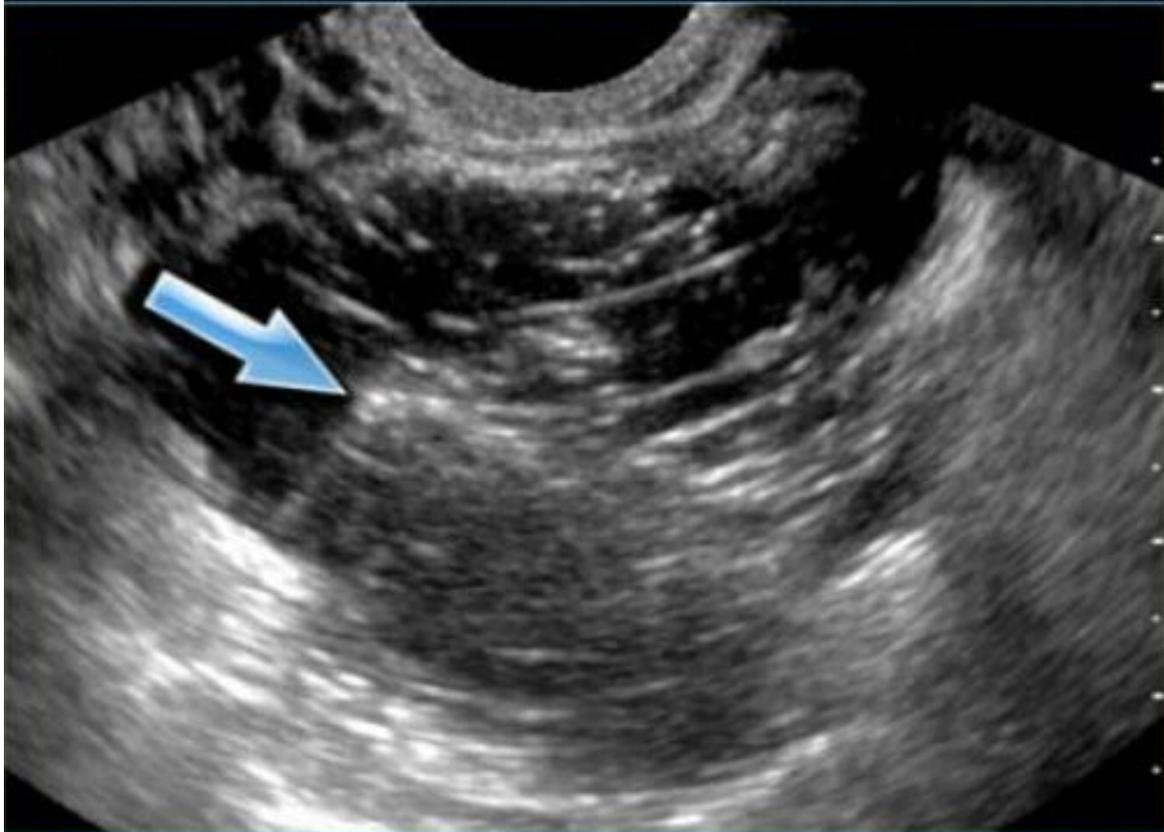


Relative Radiation Level*	Adult Effective Dose Estimate Range	Pediatric Effective Dose Estimate Range	Example Examinations
○	0 mSv	0 mSv	Ultrasound; MRI
⊕	<0.1 mSv	<0.03 mSv	Chest radiographs; Hand radiographs
⊕⊕	0.1-1 mSv	0.03-0.3 mSv	Pelvis radiographs; Mammography
⊕⊕⊕	1-10 mSv	0.3-3 mSv	Abdomen CT with IV contrast, Nuclear medicine bone scan
⊕⊕⊕⊕	10-30 mSv	3-10 mSv	Abdomen CT without and with contrast; Whole body PET/CT
⊕⊕⊕⊕⊕	30-100 mSv	10-30 mSv	CTA chest abdomen and pelvis with contrast; Transjugular intrahepatic portosystemic shunt placement

Table 1. Incidence and appearance of morphological features and signs of ovarian teratomas in different imaging modalities⁵

Imaging feature/ sign	Incidence	US	CT	MRI
Rokitansky nodule (dermoid plug)	81–86% [5, 6]	Shadowing echodensity: a densely echogenic tubercle projecting into the cystic lumen	A rounded structure protruding into to the cystic lumen, mural thickening, a bridge across the cyst, a cystic structure or sometimes only tooth	A rounded structure protruding into to the cystic lumen, mural thickening, a bridge across the cyst or a cystic structure
Tip of the iceberg sign	4% [7]	Mixture of fatty fluid, hair and cellular debris creating an echogenic focus with acoustic shadowing behind it		
Dot-dash sign	61% [5]	Hyperechoic lines and dots arising from hairs in different orientations within the imaging plane		
Fat-fluid / fluid-fluid level	8–12% [5, 6]	Anechoic sebum layered above hyperechoic aqueous/debris containing layer or less frequently, supernatant hyperechoic sebum layer above hypoechoic aqueous fluid	Supernatant fatty layer with lower attenuation and dependent fluid layer with higher attenuation	High SI of supernatant fatty layer on T1-W images and low SI on fat-suppressed T1-W images
Floating balls sign	Uncommon [4]	Intracystic floating hyperechoic globules moving with changing position of the patient	Floating globules in gravity- independent position within the cyst fluid or in the interface of fat- fluid level	Floating globules in gravity-independent position within the cyst fluid or in the interface of fat-fluid level
Comet tail appearance	12% [7]	Hypoechoic hair balls with posterior acoustic shadowing		
Intratumoral fat	93% [6]	Diffuse or regional high amplitude echoes	A component with density between –144 and –20 HU in Rokitansky nodule or cyst wall, a layering component or a floating mass mixed with hair	A component with high SI on T1-W images and signal drop on fat-saturated T1-W images
Tooth/ calcification	56% [6]	Regional high amplitude echoes with shadowing	Curvilinear or globular calcification in the Rokitansky nodule, in the tumour wall or in/near the septa	
Chemical shift artefact	86% [8]			Foci or areas of very high SI on T2-W images or a boundary artefact with high and low SI bands on opposite sides of the tumour
Tuft of hair	65% [6]	Diffuse or regional high amplitude echoes		A component with chemical shift artefact in the gravity-dependent part of the cyst
Palm tree-like protrusion	21% [8]			Polipoid mass protruding into cyst cavity with internal pattern resembling a palm tree
Intratumoral keratinoid material	75% [9]			A component with low SI on T1-W and high SI on T2-W images and diffusion restriction

Mature Cystic Teratoma on US example⁶ - key findings



Mature cystic teratoma with a Rokitansky nodule or dermoid plug

Tip-of-the-iceberg sign

Tip-of-the-iceberg sign: acoustic shadowing from the hyperechoic part of the dermoid cyst. This may be misinterpreted as bowel gas and the lesion may be overlooked.⁶

Discussion CT A/P with contrast

- CT and MR may be used to predict if a tumor is malignant based on the fact that malignant tumors appear solid or heterogeneous, and tend to be larger.⁶
- CT is important for the staging of ovarian tumors and surgical planning and in certain cases should be performed in children despite the radiation exposure.⁶
- In the case of mature cystic teratomas, intratumoral fat is diagnostic.⁶



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Case Wrap-Up

- The cause of the precocious puberty in this case was indeterminate as the patient was lost to follow-up.
- Transabdominal US is the preferred method of characterizing an adnexal mass as benign vs. malignant and appearance.
- In the case of suspicious appearing adnexal masses or symptoms concerning for malignancy, CT A/P with contrast or MR with and without contrast are reasonable choices for further work-up.
- CT A/P with contrast is preferred for ovarian mass staging. In the case of a mature cystic teratomas, intratumoral fat is diagnostic alongside presence of a mural nodule and calcifications.

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