

Functional Ovarian Cysts & Benign Ovarian Tumors

JU/ 5th year medical students

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Physiological (functional) Ovarian Cysts

- **1- Corpus Luteal cysts**
- **2- Follicular cysts**
- **3- Theca Lutein cysts**
- **4- Pregnancy Luteoma**

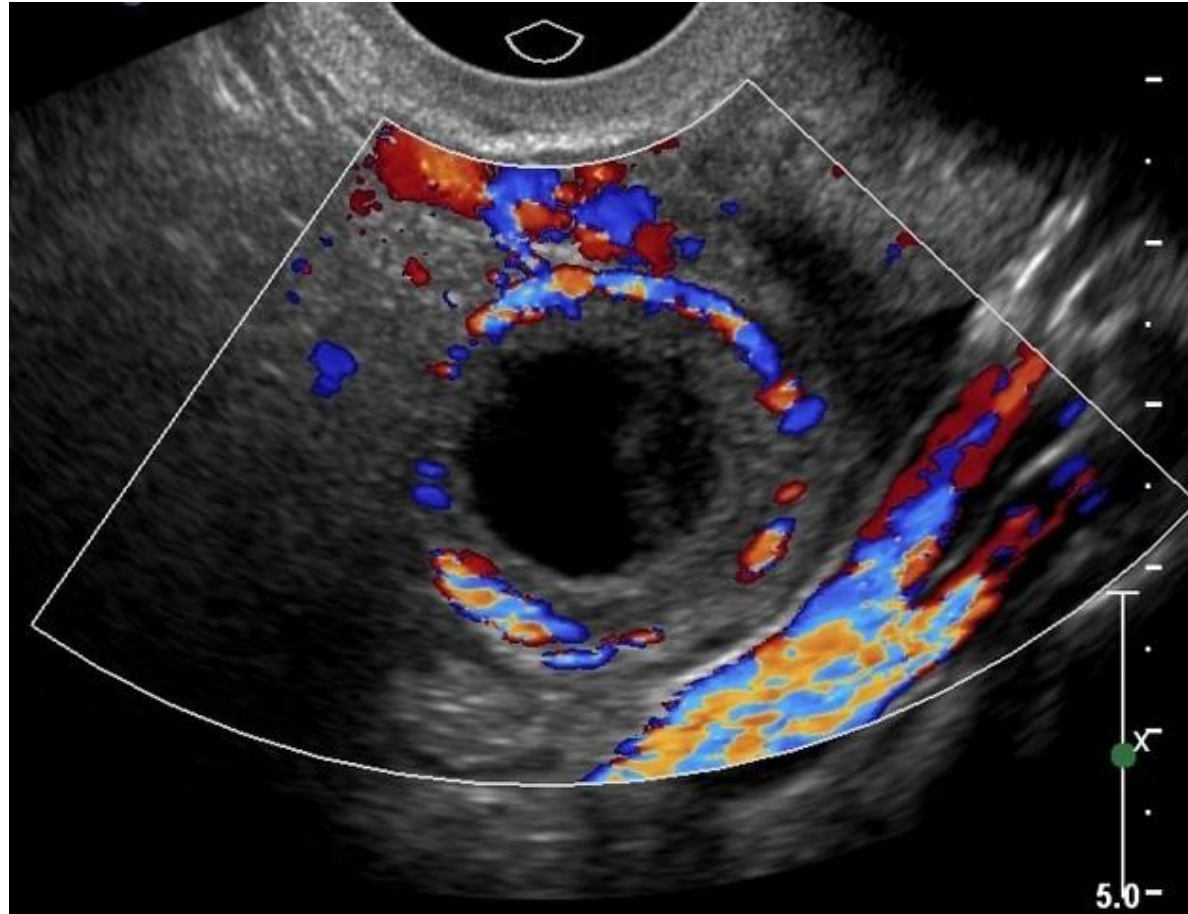
Corpus luteal (CL) cysts

- Corpus luteal (CL) cysts are a type of functional ovarian cyst that results when a corpus luteum fails to regress after ovulation.
- When associated with pregnancy, it is the most common pelvic mass encountered within the 1st trimester.
- The natural history of a normal corpus luteum is to regress by the end of the 1st trimester if pregnancy has occurred. If a corpus luteum fails to regress and instead enlarges with or without hemorrhage, a corpus luteum cyst is formed.
- Generally, they are asymptomatic unless cyst accidents occur (Rupture, Bleeding, Ovarian torsion).
- Usually, small cysts less than 10 cm.
- Most of these cysts resolve **spontaneously** within a few weeks.

Corpus Luteal (CL) cysts

- They have a range of sonographic appearances
- General characteristics include :
- Diffusely thick wall
- peripheral vascularity (Ring of Fire)
- Usually 2-10 cm
- Possible crenulated contour and internal echoes

Color Doppler shows either no vascularity within the cyst or shows low resistance blood flow around the cyst, also known as hypervascular 'ring of fire'



Follicular cysts

- Follicular cysts arise in secondary follicles that fail to ovulate and may be single or multiple.
- They are unilocular, smooth-surfaced, translucent, and thin-walled.
- The characteristic histologic features of follicle cyst are of a cyst that exceeds 3 cm in diameter, lined by attenuated granulosa cell and a theca layer with variable luteinization.
- Most regress spontaneously within a few months.

Follicular cysts are simple unilocular, smooth-surfaced, translucent, and thin-walled.



Theca Lutein cysts (hyperreactio luteinalis)

- Theca lutein cysts (hyperreactio luteinalis) are rare, benign, and typically **bilateral** cystic lesions producing grossly enlarged ovaries during pregnancy.
- They are luteinized ovarian follicle cysts form as a result of overstimulation from high levels of human chorionic gonadotropin (hCG) or hypersensitivity to hCG.
- Occur in a woman with *gestational trophoblastic disease, multiple gestation, ovarian hyperstimulation, or a pregnancy complicated by fetal hydrops.*
- They can also occur in a normal pregnancy due to hypersensitivity to normal levels of hCG.

Theca lutein cysts

- **Most are asymptomatic**, but maternal virilization, hyperemesis gravidarum, preeclampsia, or thyroid dysfunction may occur.
- The cysts **gradually resolve** weeks to months after the source of hCG is eliminated.

Pelvic ultrasound showing *bilateral* multiseptated Theca Lutein cyst



Pregnancy luteoma

- Pregnancy luteomas are rare, benign, solid ovarian tumors composed of hyperplastic masses of large lutein cells, thought to be caused by the hormonal effects of pregnancy.
- They are usually asymptomatic and found incidentally during imaging or surgery.
- It is associated with an increase of sex hormones, primarily progesterone and testosterone.
- The most obvious symptom of a luteoma is masculinization of the mother and the possible masculinization of the fetus.
- The size of the tumor can range from 1 to 25 cm in diameter
- Luteomas regress and disappear after delivery or are adequately treated by a conservative surgical approach

Pregnancy luteomas

- **Risk factors :**

- 1- PCOS

- 2- Past history of luteoma

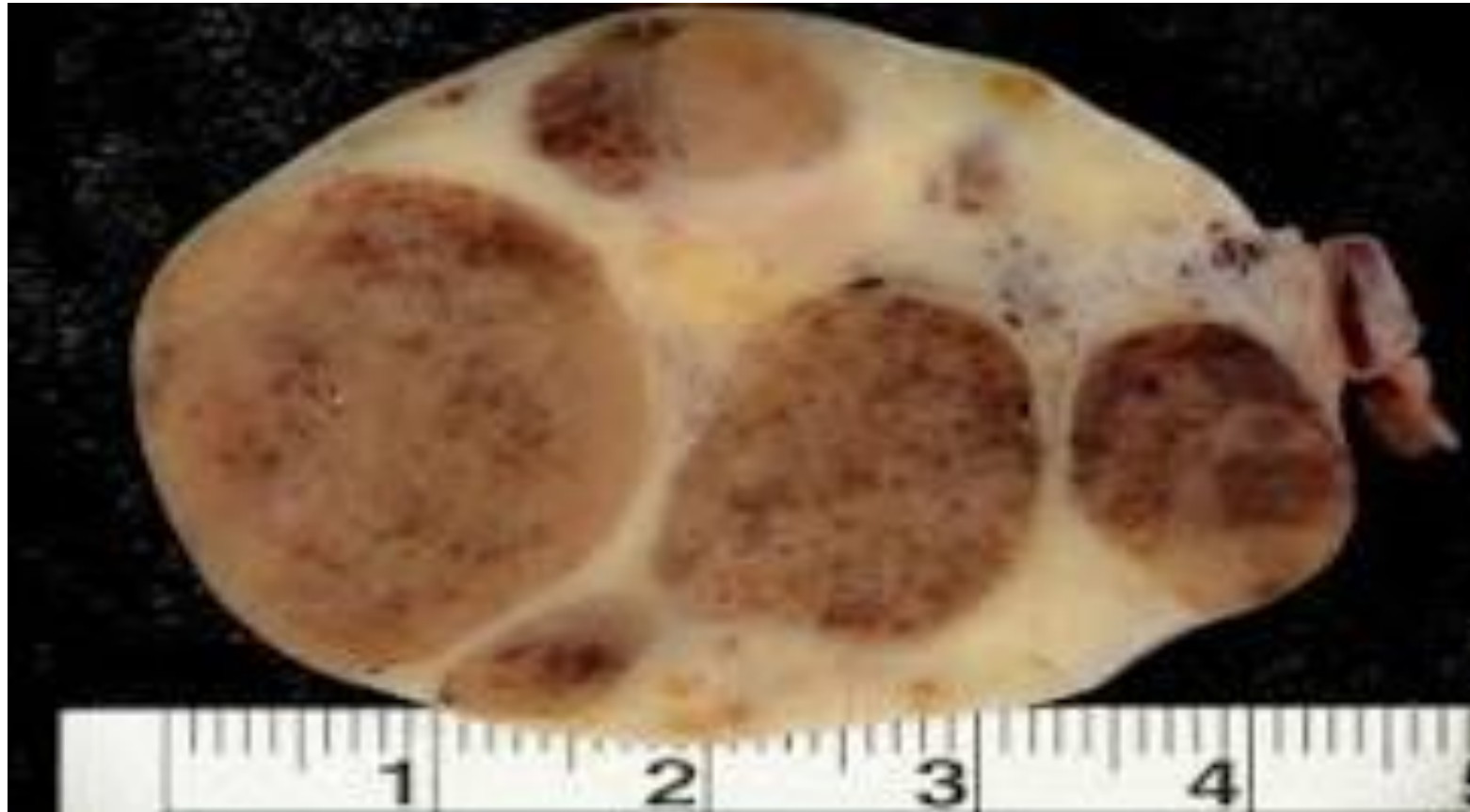
- 3- Multiparity

- 4- Advanced maternal age

- 5- Afro-Caribbean ethnicity

Pregnancy luteomas: benign, solid ovarian tumors

The diagnosis should be suspected in the presence of a solid adnexal mass and maternal hirsutism or virilization during pregnancy.



Pregnancy Luteoma

- **Virilization during pregnancy is rare and it is most commonly caused by:**
- Pregnancy Luteoma
- Theca Lutein cysts (Hyperreactio Luteinalis)
- Both, Luteomas and HL are benign, self-limited conditions

Complications of Ovarian Cysts

- Rupture
- Hemorrhage
- Torsion
- Malignant Transformation

Ovarian Torsion

- Torsion occurs when adnexal structures, including the ovary, the fallopian tube, or paraovarian cysts, twist around the infundibulopelvic and utero-ovarian ligaments
- This twisting can cut off blood flow to the ovary and fallopian tube.
- If left untreated, It can reduce blood supply & lead to ovarian necrosis.
- No risk factors are seen in up to 69% of patients with surgically confirmed torsion.
- **Common risk factors include:** prior pelvic surgery, ovarian cysts, ovulation induction, and pregnancy (particularly in the first trimester).
- The classic presentation of ovarian torsion is the acute onset of pelvic pain (sudden onset sharp & stabbing) often with nausea and vomiting, in a patient with an adnexal mass.

D/D of Acute Pelvic Pain in Female

Table 2: Differential Diagnoses for Acute Pelvic Pain in Females

Gynecologic entities

- Physiologic or hemorrhagic cyst
- Pelvic inflammatory disease
- Ectopic pregnancy
- Torsion
- Endometriosis with acute hemorrhage or rupture
- Ovarian hyperstimulation syndrome
- Fibroid with necrosis or prolapse
- Ovarian vein thrombophlebitis

Nongynecologic entities

- Appendicitis
- Enterocolitis
- Diverticulitis
- Bowel ischemia
- Inflammatory bowel disease
- Irritable bowel syndrome
- Urolithiasis
- Cystitis

Ovarian Torsion

- **Vaginal Ultrasound/CT/MRI:** Imaging features of ovarian edema are the hallmark of adnexal torsion.
- When torsion occurs and ovarian blood vessels become compromised, venous and lymphatic outflows are affected first, as the arteries are initially spared owing to their thicker muscular walls. Venolymphatic compromise results in edema of the ovarian parenchyma, which then further affects arterial inflow, resulting in even greater edema.
- The most common neoplasm to twist is a benign dermoid cyst or mature teratoma.
- Dermoid cyst plus pain equals torsion until proved otherwise

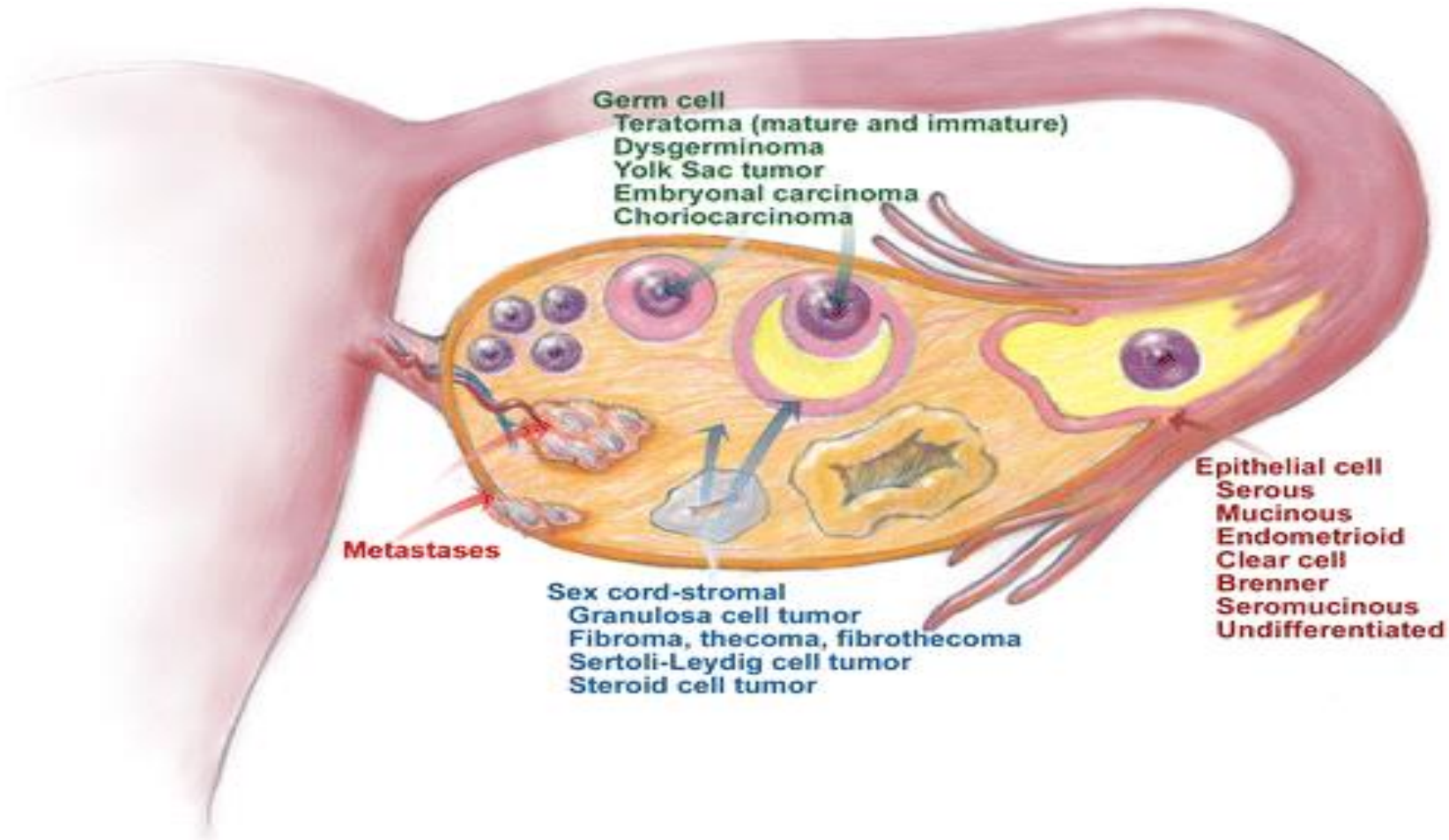
Ovarian Torsion

- **Diagnosis:**
- History, Physical examination
- CBC, CRP, Urine analysis
- Ultrasound: **Ovarian edema, the hallmark feature of torsion**, manifests as asymmetric ovarian enlargement, thicker than expected ovarian parenchyma surrounding a lesion, adjacent free fluid, and stromal changes of heterogeneity at U/S, high attenuation at noncontrast CT, and increased T1-weighted signal intensity at MRI.
- Doppler Ultrasound: Absent color Doppler flow or contrast enhancement
- **Treatment:** The diagnosis should not be delayed because color Doppler flow or contrast enhancement is still present.
- Laparoscopic untwisting + ovarian cystectomy/ Oophorectomy if necrotic

Ovarian Neoplasms

- **Ovarian neoplasms arise from:**
- The Surface Epithelium (The commonest 60-65 %)
- Germ cells (30%)
- Sex-Cord Stromal Tissue (8%)
- Ovarian neoplasms may be benign, borderline or malignant.

Ovarian Neoplasms



Benign ovarian neoplasms

- **A) Epithelial ovarian tumors :**

- 1) Serous cystadenoma
- 2) Mucinous cystadenoma
- 3) Endometrioid cystadenoma
- 4) Brenner tumor

- **B) Germ cell ovarian tumors :**

- 1) Mature cystic teratoma (Dermoid cyst)
- 2) Mature solid teratoma
- 3) Monodermal highly specialized teratomas (e.g., struma ovarii, carcinoid)

- **C) Sex-cord stromal cell ovarian tumors :**

- 1) Fibroma
- 2) Thecoma

Benign ovarian neoplasms

- Predominantly they occur in **premenopausal women**.
- Among premenopausal patients, more than 90% of cases are benign, as opposed to just 60% in the postmenopausal population.

Benign Ovarian Neoplasms

- **Presentation:**

- **Asymptomatic** : incidental finding on examination or imaging.
- **Mild lower abdominal pain.**
- **Ovarian cyst accidents:** torsion, rupture or hemorrhage. Present as acute abdomen.
- **Dyspareunia.**
- **Pressure effects**
- **Ascites** (e.g with Meigs' syndrome).
- **Endocrine:** hormone-secreting tumors may cause virilization, menstrual irregularities or postmenopausal bleeding.

Epithelial ovarian tumors

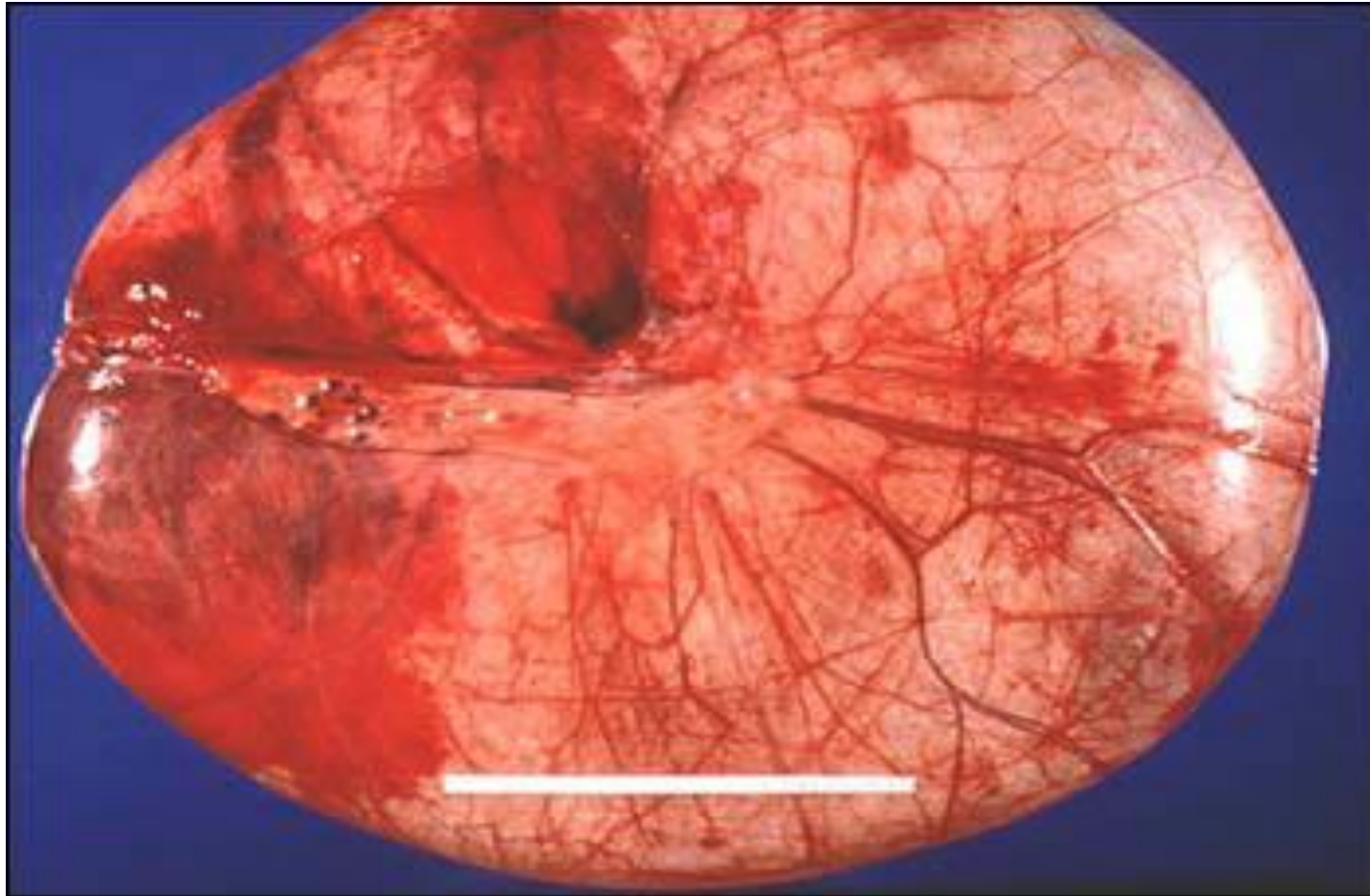
- **1) Serous cystadenoma**
- **2) Mucinous cystadenoma**
- **3) Endometrioid cystadenoma**
- **4) Brenner tumor**

Serous Cystadenoma

- **Ovarian Serous Cystadenoma is the most common ovarian neoplasm.**
- They have a smooth outer surface and contain one or more thin-walled cysts filled with clear, watery fluid.
- They are usually unilocular but may be multilocular.
- Ranges in size from 1 to more than 30 cm in greatest dimension (mean = 10 cm).
- Bilateral in 20% - 25%
- **Psammoma bodies** are present in about 30% of cases
- Most tumors are **asymptomatic** until they reach large size.
- Symptoms include **abdominal discomfort** and **chronic pelvic pain**. Rare tumors may undergo **torsion** and cause **acute abdominal pain**.
- The peak incidence is at the **4th to 5th** decades of life

Serous Cystadenoma

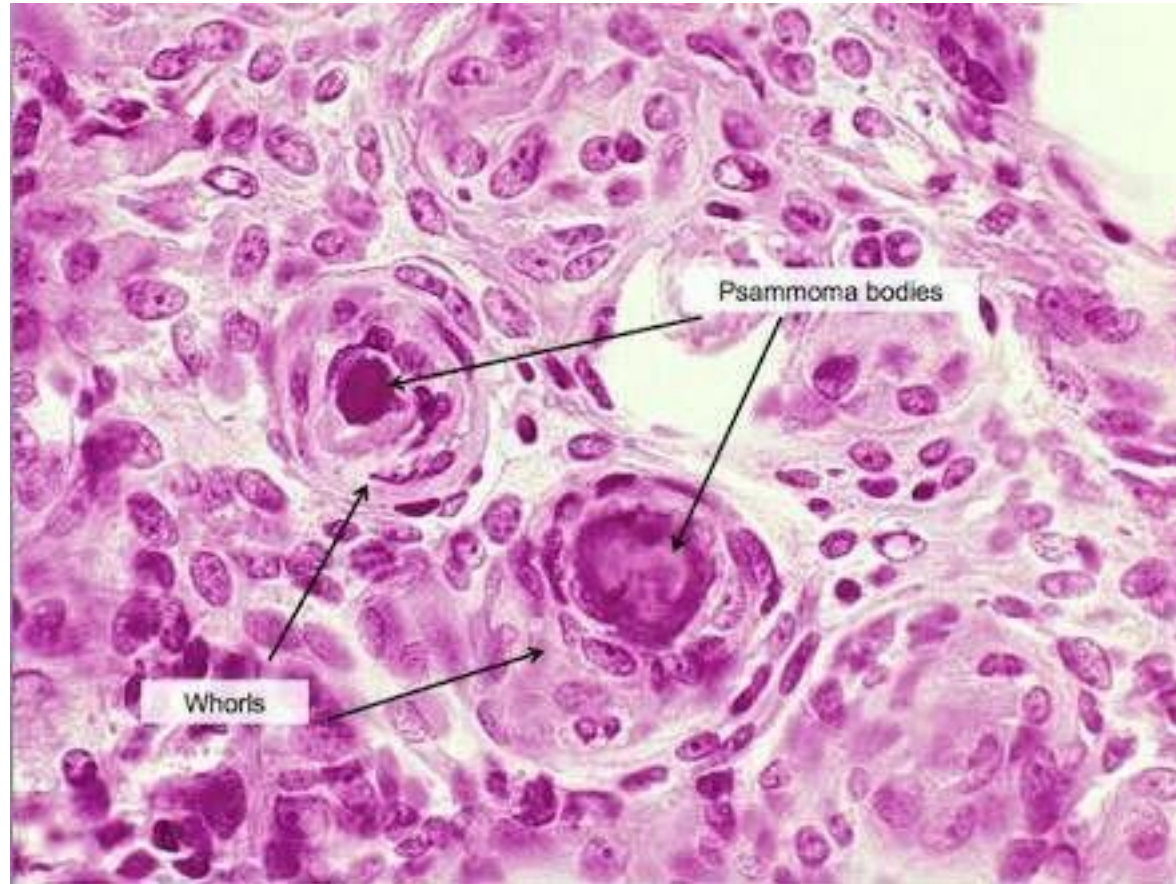
**smooth outer surface, contain one or more thin-walled cysts
filled with clear, watery fluid**



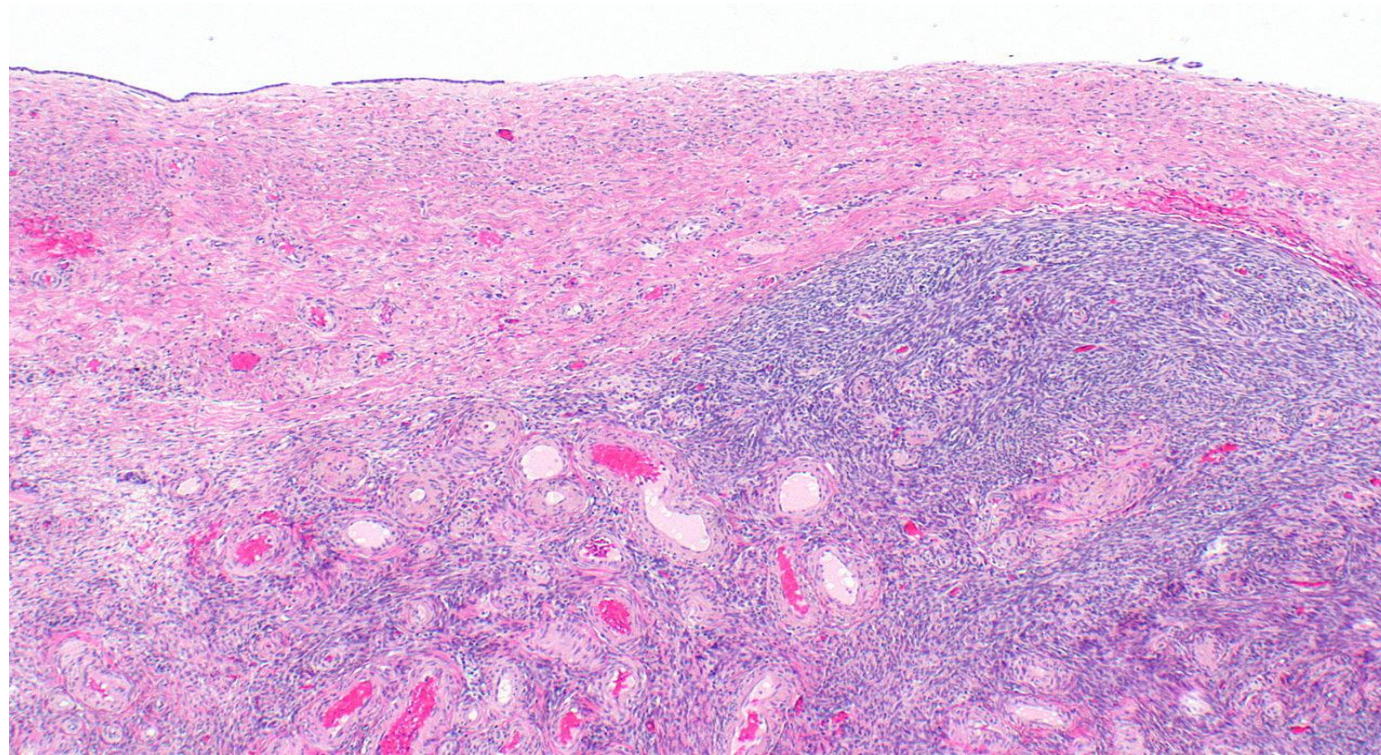
Psammoma bodies

- The term is derived from the Greek word ψάμμος (psámmos), meaning "sand". "sand tumor."
- **Psammoma bodies** are round microscopic calcific collections. It is a form of dystrophic calcification.
- They are generally found in damaged tissues, and are commonly associated with malignant cancers as well as some benign, non-cancerous conditions.
- Although psammoma bodies are classically associated with papillary neoplasms, notably those of the thyroid, ovaries, and lungs, they occur in a wide variety of tumors from many different sites as well as a wide variety of benign conditions, including endosalpingiosis and endometriosis, among others.

Psammoma bodies



Microscopically: the serous cystadenoma lining consists of a simple epithelium, whose cells may be either columnar and tall and contain cilia or cuboidal and have no cilia.



Mucinous Cystadenoma

- **The second most common benign ovarian neoplasms.**
- The peak incidence occurs between 30 and 50 years of age
- They are bilateral in 5–10% of cases.
- **Gross:** These tumors are usually large in size and are often multiloculated. The external surface will be smooth and can be bosselated. They are filled with mucoïd or jelly like fluid which is usually sticky in nature.
- Tend to be **larger** than serous cystadenomas at presentation

Mucinous Cystadenoma

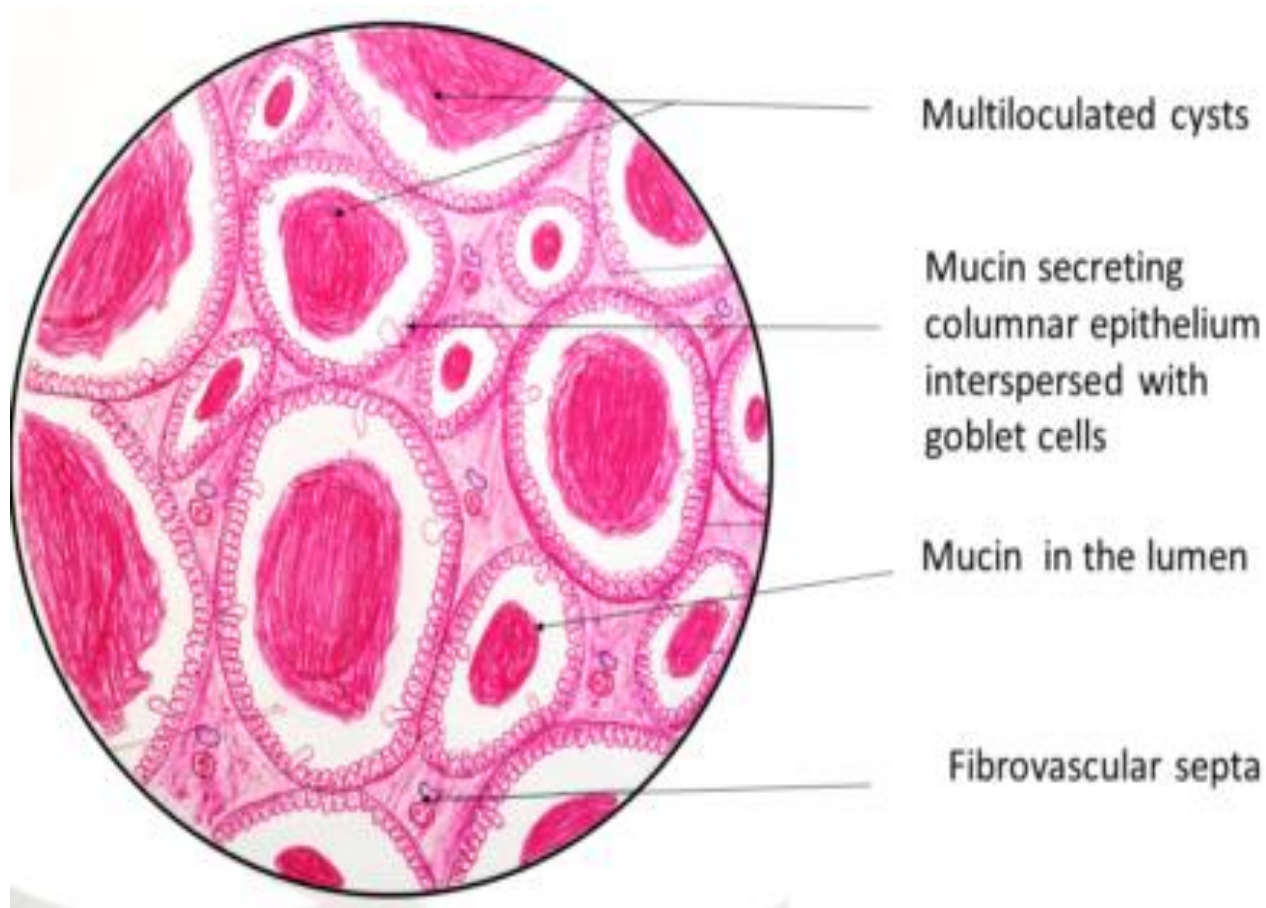
Microscopy: multiloculated cysts, lined by tall columnar epithelium with apical mucin. Cilia are not present.

The epithelium can be of two types

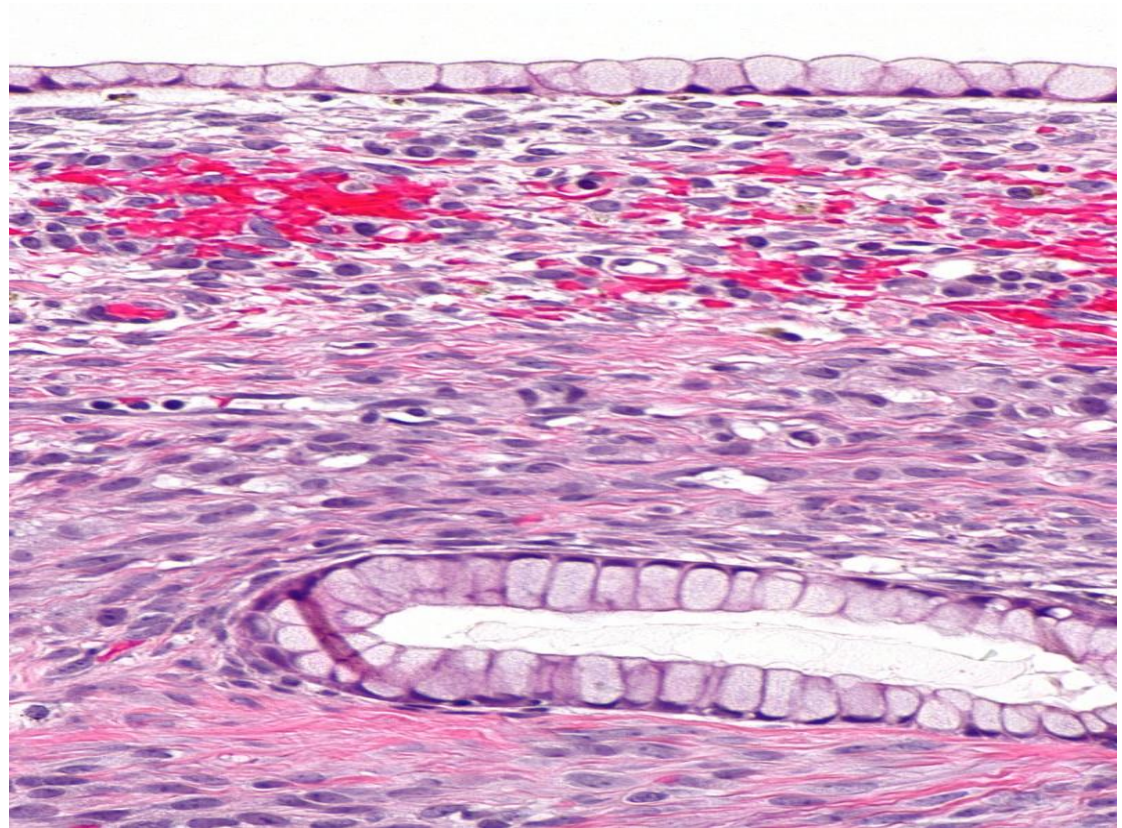
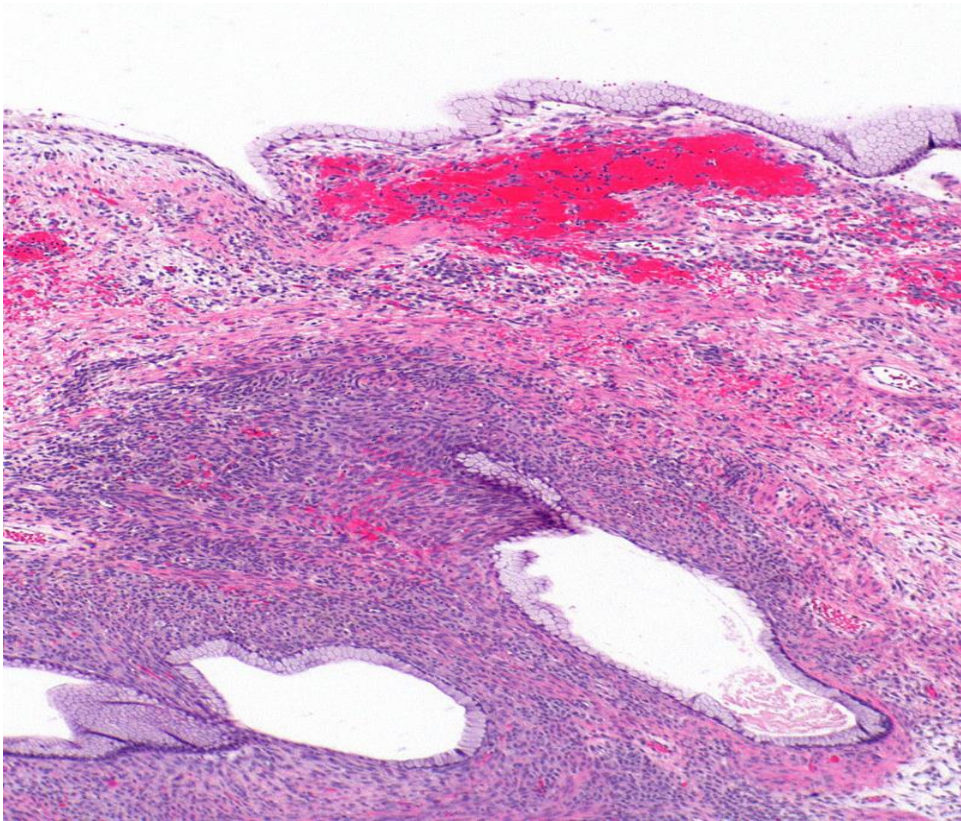
1- *Gastric /intestinal type* – this is most common. They have numerous goblet cells

2- *Endocervical type*: resembling endocervical lining epithelium

Mucinous cystadenoma is composed of multiple cysts and glands lined by simple non-stratified mucinous epithelium resembling gastric foveolar-type or intestinal epithelium containing goblet cells and sometimes neuroendocrine cells or Paneth cells



A single layer of tall columnar mucinous epithelial cells with basally placed nuclei and containing apical mucin, lines the cystic spaces



Endometrioid Cystadenoma

- Benign tumors with endometrioid type glands, sometimes associated with endometriosis.
- Squamous or mucinous metaplasia or dystrophic stromal calcifications can be seen.
- Absence of endometrial type stroma distinguishes endometrioid cystadenoma / adenofibroma from endometriosis.
- Uncommon tumors, most often seen in postmenopausal women.
- Can be an incidental finding.
- Symptoms may include abnormal vaginal bleeding, abdominal pain or pelvic mass.

Brenner tumor

- Brenner tumor is a rare ovarian tumor.
- They are categorized by the World Health Organization as benign, borderline (proliferative), and malignant.
- The vast majority of Brenner tumors are benign.
- Brenner tumors manifest as a multilocular cystic mass with a solid component or as a small, mostly solid mass.
- 90% are unilateral. The tumors can vary in size from less than 1cm to 30 cm.
- Histologically : There are nests of transitional-type epithelial cells with longitudinal nuclear grooves (coffee bean nuclei) lying in abundant fibrous stroma.
- It is usually asymptomatic and most of the times it is an incidental pathological finding.

Germ cell ovarian tumors

- **1) Mature cystic teratoma (Dermoid cyst)**
- **2) Mature solid teratoma**
- **3) Monodermal highly specialized teratomas (e.g., struma ovarii, carcinoid)**

Mature Cystic Teratoma (Dermoid cyst)

- The most common ovarian neoplasm, account for 70% of all benign ovarian masses affecting females in their reproductive years.
- Mature Cystic Teratoma contains representative tissues of the three embryonic germ cell layers: ectoderm, mesoderm and endoderm.
- Typically their diameter is smaller than 10 cm, and rarely more than 15 cm.
- They most commonly affect females ages 20–40 years.
- Ovarian dermoid cysts are almost always benign. However, they can become malignant in rare cases.

Mature Cystic Teratoma (Dermoid cyst)

- They are bilateral in 10-15% of cases
- Uncomplicated ovarian dermoid tend to be asymptomatic and are often discovered incidentally.
- Mature Cystic Teratoma (Dermoid cyst) **has large potential** to ovarian torsion (3-16%), and may then present with acute pelvic pain.
- Most women with dermoid cysts are **asymptomatic**.
- Rupture may occur (1-4%)
- Superimposed infection may occur

Mature Cystic teratoma (Dermoid cyst)

These tumors have a characteristic ultrasound appearance but definitive diagnosis is made at the time of surgical excision.



Mature Cystic teratoma (Dermoid cyst)

- **Grossly:**

- Most cysts are unilocular.
- The inner lining of every mature cystic teratoma contains single or multiple white shiny masses projecting from the wall toward the center of the cysts. When hair, other dermal appendages, bone and teeth are present, they usually arise from this protuberance.
- This protuberance is referred to as the **Rokitansky protuberance**.
- The Rokitansky protuberance is a common site of malignant transformation.

Mature Cystic teratoma (Dermoid cyst)

- **Histopathology:**
- Mature cystic teratomas contain mature tissue of ectodermal (skin, hair follicles & sebaceous glands), mesodermal (muscle & urinary) and endodermal origin (lung & gastrointestinal).

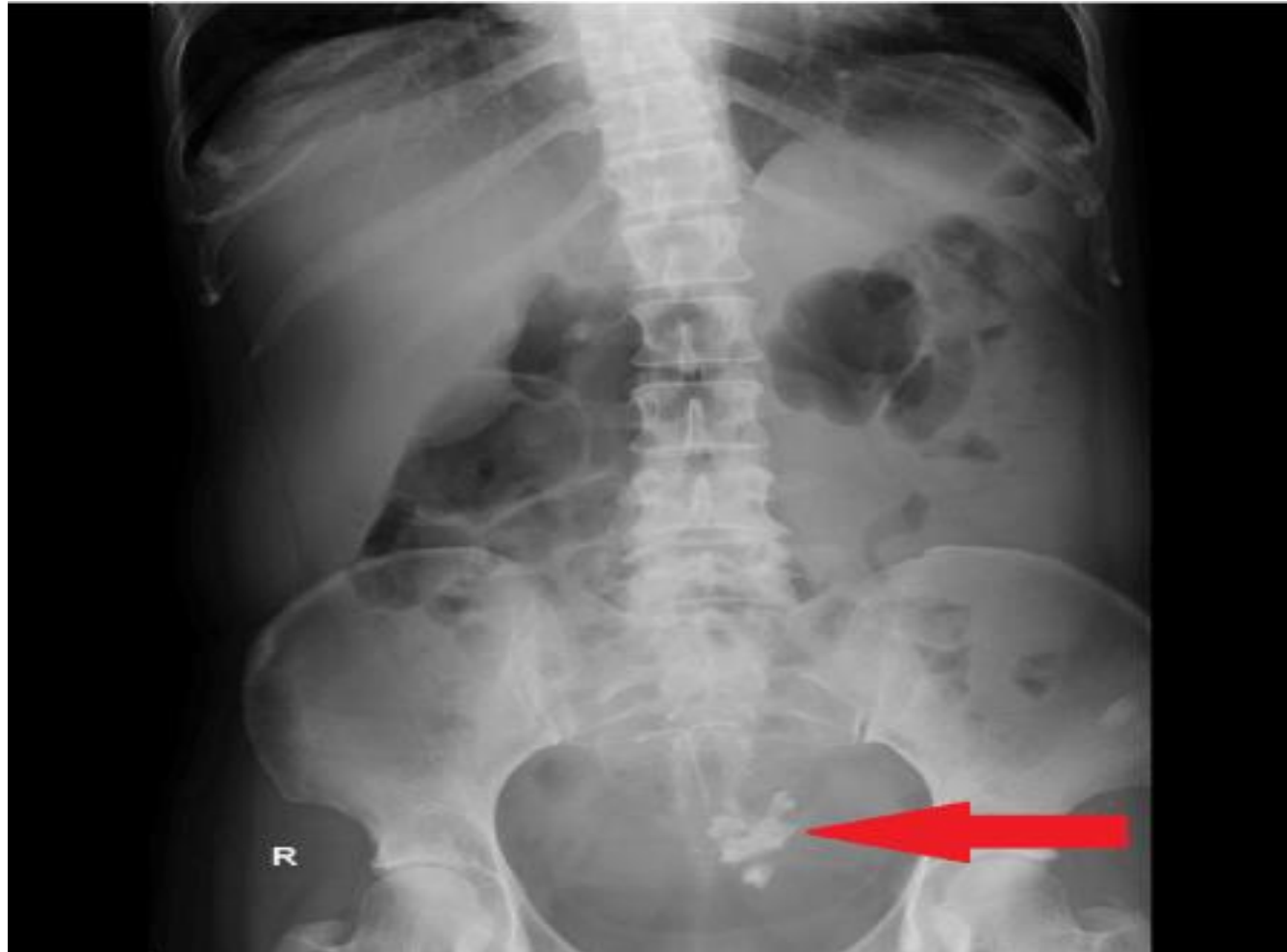
Diagnosis of Mature Cystic Teratoma

- **Plain radiograph**: May show calcification and tooth components of teratoma within the pelvis.
- **Ultrasound**: These tumors have a characteristic ultrasound appearance such as a fat-fluid level, dermoid mesh and tip of the iceberg sign.
- With the presence of two or more of these features a diagnosis of dermoid cyst can confidently be made with reported specificity 98-100 %.

Diagnosis of Mature Cystic Teratoma

- **The spectrum of sonographic features includes:**
- Fat-fluid level; also known as a “hair-fluid level” or “fluid-fluid” level is believed to be the result of layering of serous fluid and sebum.
- Dermoid plug (Rokitansky nodule): this is the most common sonographic feature of a dermoid cyst. It appears as an echogenic mass within the cyst made up of hair, teeth or fat.
- Dermoid mesh: the appearance on ultrasound is of multiple small hyperechoic lines and dots within the cyst forming a “mesh-like” picture (dot-dash pattern)
- Tip of the iceberg sign: the appearance of a hyperechoic area, the base of which cannot be visualised. This is the result of a mass made up of matted hair and sebum casting an echogenic shadow.
- Intracystic floating balls sign is uncommon but is pathognomonic for ovarian mature cystic teratoma
- Color Doppler: no internal vascularity
- Internal vascularity requires further workup to exclude a malignant lesion

Plain radiograph: May show calcification and tooth components of teratoma within the pelvis



Rokitansky Nodule

ROKITANSKY NODULE

- A raised protuberance projecting into the cyst cavity.
- Most of the hair typically arises from this protuberance.
- When bone or teeth are present, they tend to be located within this nodule



The echogenic focus (between markers) and speckled debris within the cyst are characteristic of a benign cystic teratoma



Diagnosis of Mature Cystic Teratoma

• CT SCAN & MRI

- Alternative methods of diagnosis of dermoid cysts include CT and MRI both of which are more sensitive to fat than ultrasound.
- At CT a diagnosis of dermoid cyst can be made when attenuation of fat is imaged.-
- At MRI a dermoid cyst can be reliably differentiated from a haemorrhagic lesion or endometrioma again due to the intensity of the fatty contents on imaging (fat attenuation).

Treatment of Mature Cystic Teratoma

- Mature ovarian teratomas are slow-growing tumor (1-2 mm a year)
- Dermoid cysts no larger than 6 cm can be managed conservatively with annual follow-up, provided the annual growth rate does not exceed 2 cm.
- Larger lesions are often surgically removed.
- Rx: Ovarian cystectomy, via either laparoscopy or laparotomy.
- With either approach, the abdomen should be copiously irrigated to avoid a chemical peritonitis from spillage of the sebaceous cyst fluid.

Malignant transformation of Mature Cystic Teratoma

- **Malignant transformation** occurs in 0.2-2 % of mature cystic teratomas.
- Although any of the components of a mature cystic teratoma may undergo malignant degeneration, squamous cell carcinoma (75%) arising from the ectoderm is the most common secondary neoplasm.
- Other possible malignant neoplasm include: basal cell carcinoma, melanoma, adenocarcinoma, sarcoma and thyroid carcinoma.
- **Risk factors for malignant neoplasm in a mature cystic teratoma include:**
- **Age over 45, tumor diameter > 10 cm, rapid growth, and low-resistance intratumor flow on Doppler.**

Rupture of Dermoid Cysts

- Rupture of dermoid cysts with spillage of sebaceous material into the abdominal cavity can occur, but is uncommon.
- The rate of spontaneous rupture of mature cystic teratomas is low, ranging from 1.2-3.8%. This has been ascribed to the thick capsule surrounding this lesion.
- Shock and hemorrhage are the immediate sequelae of rupture; a marked granulomatous reaction (chemical peritonitis) may subsequently develop and lead to formation of dense adhesions and chronic pain.
- Urgent surgery is needed if rupture occurs.

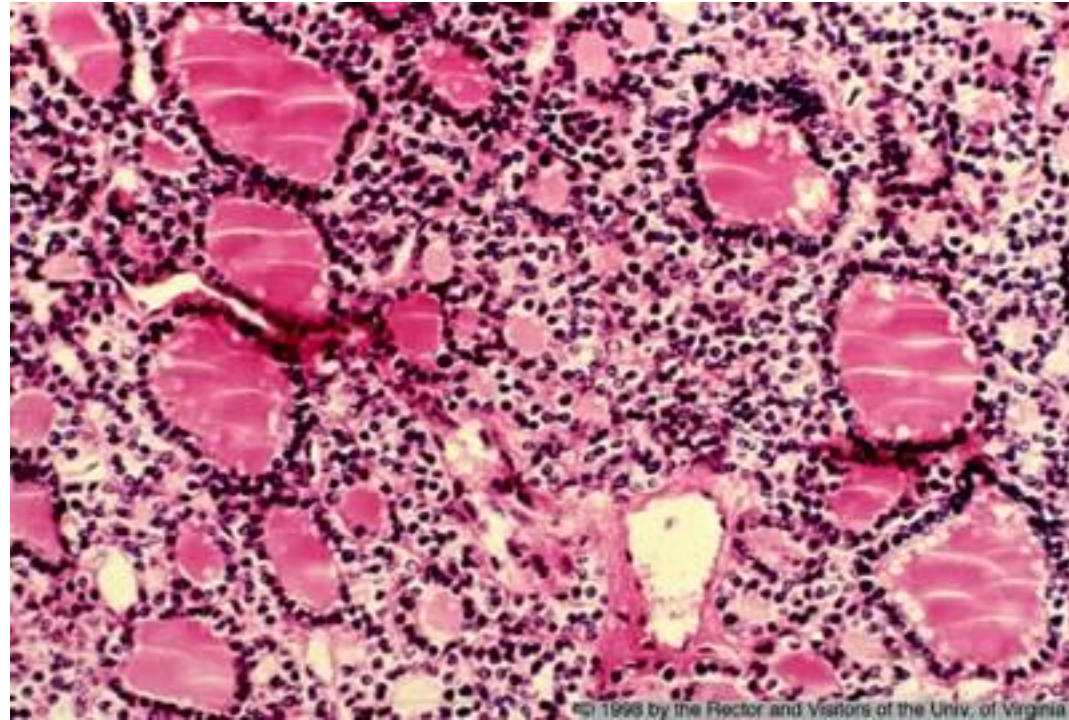
Monodermal highly specialized teratomas

- Specialized teratomas of the ovaries are a rare subtype of ovarian teratomas where there is a monodermal differentiation of tissue element.
- They usually contain only endodermal, ectodermal or mesodermal elements.
- There are three main types of ovarian monodermal highly specialized teratomas:
 - 1- Struma ovarii tumor
 - 2- Ovarian carcinoid tumor
 - 3- Tumors with neural differentiation

struma ovarii tumor

- It is predominantly composed of **mature thyroid tissue**.
- The secretion of thyroid hormones results in clinical hyperthyroidism in **25-35 %** of patients.
- Struma ovarii is uncommon, comprising approximately 2.7 % of ovarian teratomas.
- It is often associated with a mature cystic teratoma and rarely with a cystadenoma.
- Most cases of struma ovarii are benign and can be managed by excision of the ovary or by unilateral salpingo-oophorectomy.
- However, malignant change may occur in struma ovarii, but it is exceedingly rare
- It is difficult to diagnose these cases preoperatively as there are no specific clinical, radiological or serum markers for these rare tumors in the absence of thyroid biology abnormality.
- Most cases are diagnosed on histopathology.

Struma ovarii:
composed entirely of mature thyroid tissue



Mature solid teratoma

- In rare instances, a teratoma is solid but is composed entirely of benign-appearing heterogeneous collections of tissue and organized structures derived from all three cell layers.
- Most mature solid teratomas are **unilateral and benign**, although peritoneal implants have been described.
- Grossly, it may be difficult/impossible to differentiate these neoplasms from malignant solid immature teratomas, which are almost always solid and they therefore require liberal sampling.

Sex-Cord Stromal Cell Ovarian Tumors

- Ovarian fibroma (Benign)
- Theca cell tumor (thecoma) (Benign)
- Sertoli-Leydig cell tumor (Well differentiated Sertoli-Leydig cell tumors are essentially benign)
- Granulosa cell tumor (Malignant)

Ovarian Fibroma

- An **ovarian fibroma** is a rare, benign tumor of the ovary, account for approximately 4% percent of all ovarian tumors.
- The most common sex- cord stromal ovarian tumor.
- Usually unilateral, solid (sometimes mixed), lobulated, firm, white and may have myxoid change.
- They tend to occur mostly during perimenopause and postmenopause, the median age having been reported to be about 52 years.
- Ovarian fibromas are often asymptomatic, but they sometimes can cause pelvic pain or discomfort – especially if they are associated with ovarian torsion.
- Ovarian fibromas can be part of **Meigs Syndrome**, a triad of ovarian fibroma, ascites & pleural effusion.

Meigs Syndrome

- Meigs syndrome is defined as the triad of benign ovarian tumor with ascites and pleural effusion that resolves after resection of the tumor.
- the pleural effusion is classically on the right side
- Ovarian fibromas constitute the majority of the benign tumors seen in Meigs syndrome.
- Meigs Syndrome can be seen with other ovarian tumors: fibrothecoma, Brenner tumor, and occasionally granulosa cell tumor..
- A key feature found in patients with Meigs' syndrome is the resolution of symptoms after tumor resection or after unilateral salpingoopherctomy.

Meigs Syndrome

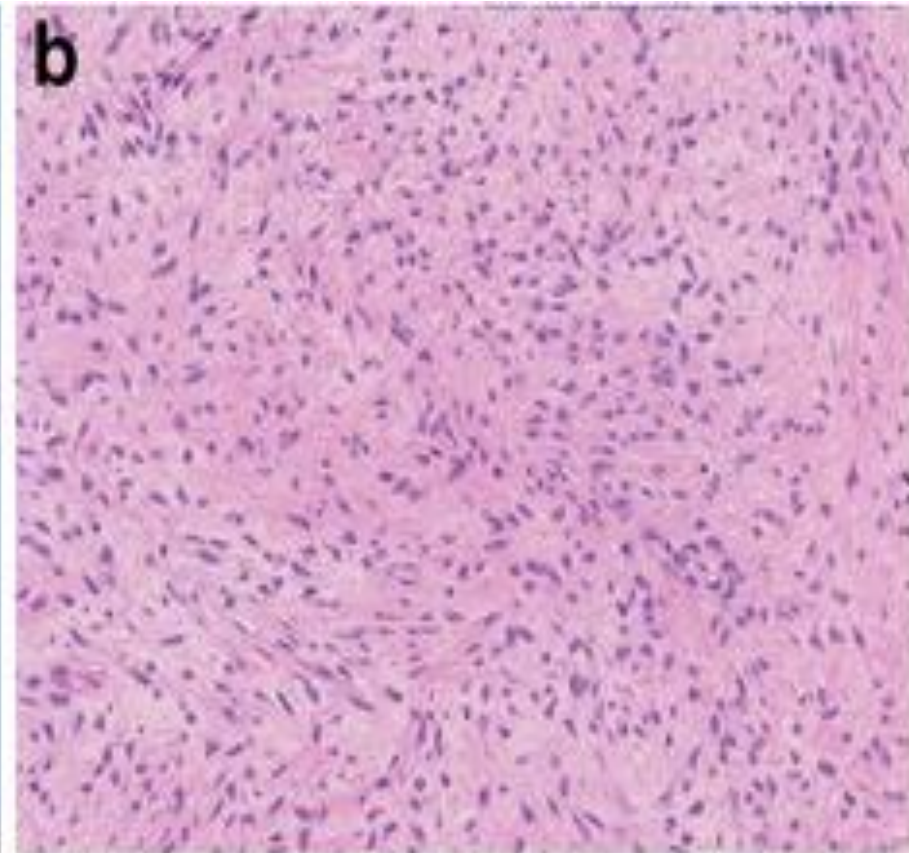
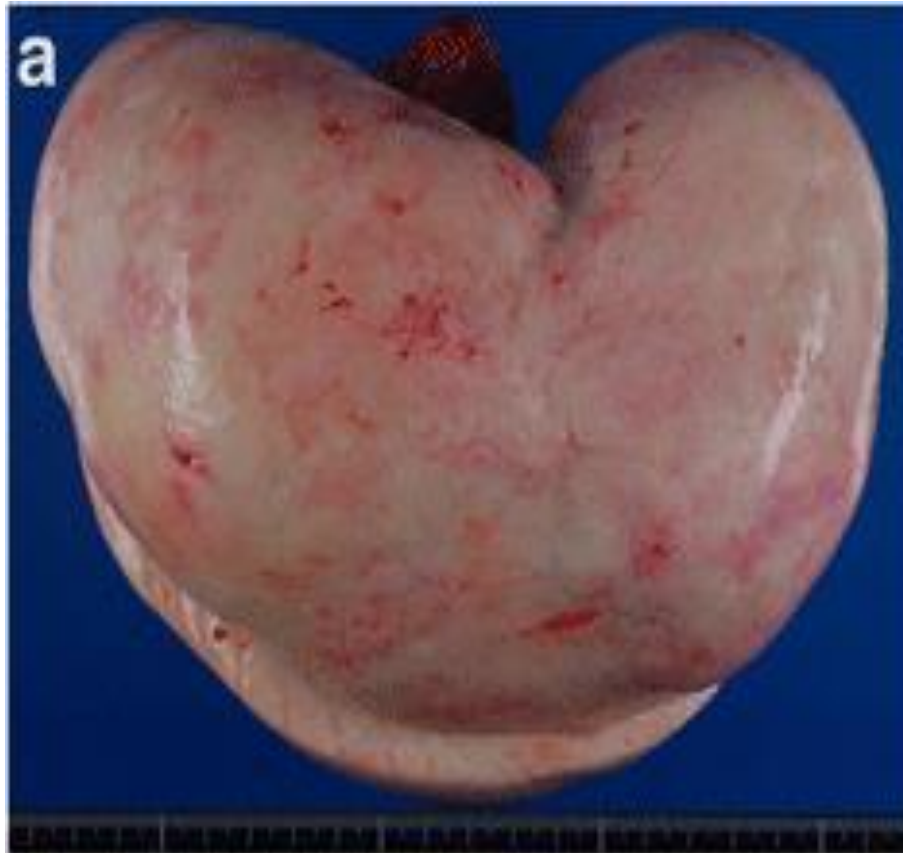
- **D/D:**

- Liver failure
- Congestive Heart Disease
- Renal disease
- Metastatic tumor to the peritoneal surfaces

- **Rx:**

- Thoracocentesis or paracentesis
- Unilateral salpingo-oopherctomy.

Macroscopic appearance showed a solid and smooth mass. Microscopic findings with hematoxylin and eosin staining showed spindle cells that were arranged in intersecting bundle



Thecoma

- Rare benign ovarian stromal tumor, composed of lipid-containing cells resembling theca cells with a variable fibromatous component.
- Cut surfaces are typically solid and firm, with characteristic yellow areas indicative of lipid-containing cells interspersed with gray-white, more fibrotic areas.
- **Unilateral** (95%). They usually range in size between 5 and 10 cm.
- **Age distribution:**
 - 1- Conventional thecoma: mostly postmenopausal women (mean age 59 years)
 - 2- Luteinized thecoma: 30% in patients ≤ 30 years
- These tumors are commonly **estrogen** producing tumors (60%) and most women present with abnormal **vaginal bleeding**.
- Approximately 20% of patients will also develop **concurrent endometrial carcinoma**.
- Luteinized thecomas are **androgenic** in approximately 10% of cases (virilization)
- They could be non-functioning tumors

Thecoma: Prognosis & Treatment

- **Prognosis:**

- Thecomas are [benign tumors](#), and extreme caution should be used if the diagnosis of a malignant thecoma is suspected.
- Rare malignant thecomas have been described; however, these may have actually been [fibrosarcomas](#) or variants of granulosa cell tumor.

- **Treatment:**

- Unilateral salpingo-oophorectomy (unless associated with endometrial adenocarcinoma: [hysterectomy](#) and bilateral salpingo-oophorectomy)

Cut surfaces are typically solid and firm, with characteristic yellow areas indicative of lipid-containing cells interspersed with gray-white, more fibrotic areas

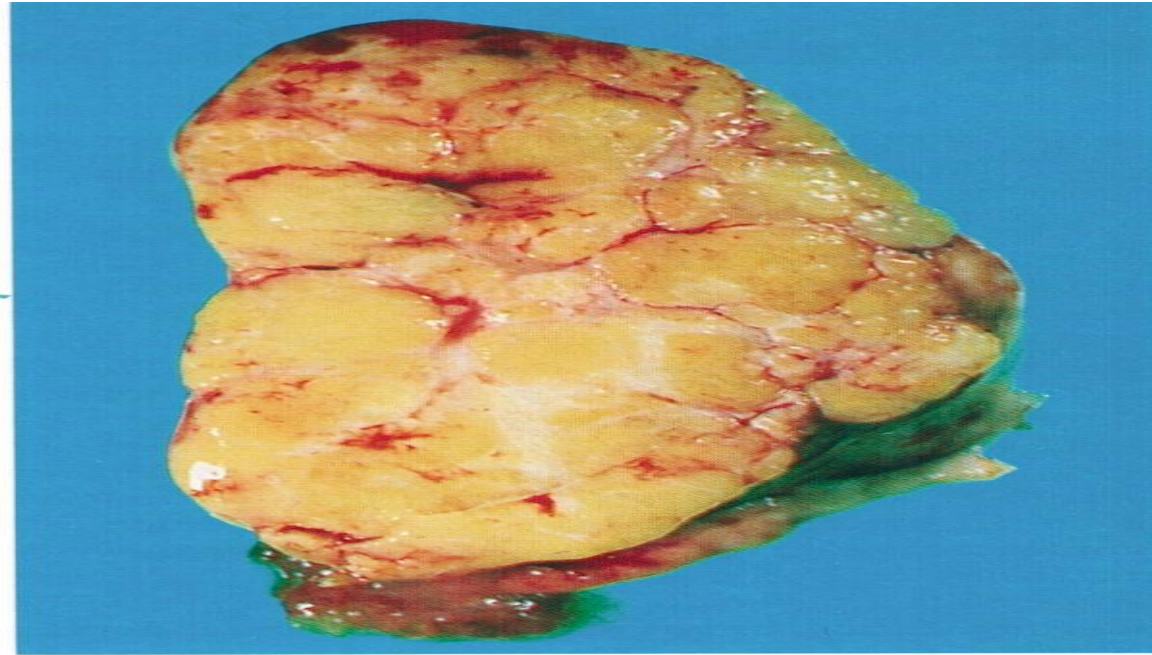


Figure 10-1
THECOMA

The sectioned surface is lobulated and yellow. (Fig. 31 from Young RH, Scully RE. Ovarian sex cord-stromal tumors: problems in differential diagnosis. Pathol Ann 1988;23(Pt 1):237-96.)

The End of the First Part

Thank You

Dr Amal Barakat