Learning objectives

1. Describe the various clinical and radiologic manifestations of sex cord-stromal ovarian tumor.
2. Identify benign sex cord-stromal ovarian tumor.
3. Identify malignant sex cord-stromal ovarian tumor.
Background

Ovarian tumors are classified as epithelial tumors (serous and mucinous tumors, endometrioid and clear cell carcinomas, Brenner tumor), germ cell tumors (mature and immature teratomas, dysgerminoma, endodermal sinus tumor, embryonal carcinoma), sex cord-stromal tumors (fibrothecoma; granulosa cell, sclerosing stromal, and Sertoli-Leydig cell tumors), and metastatic tumors on the basis of tumor origin. Along them, sex cord-stromal tumors of the ovary are rare ovarian neoplasm. These tumors arise from two groups of cells in the ovary: stromal cells and primitive sex cords. This group of tumors represents approximately 5-10% of ovarian neoplasms and affects all age groups. Sex cord-stromal tumors are of interest partly because of their hormonal effects (estrogen effect or virilization), which are rare in other ovarian neoplasms. The majority of them are either benign or largely confined to the ovary at diagnosis. Consequently, the surgical excision is treatment of choice primarily and the prognosis is generally good. However, treatment of choice may often be problematic, especially in young reproductive women. Therefore, precise knowledge of clinical and imaging features, especially MR imaging is crucial in establishing an accurate diagnosis and determining treatment.

The purpose is to present the clinical and imaging features for the spectrum of sex cord-stromal ovarian tumor, from benign to malignant and from cystic to solid and correlate the imaging features with the clinical and pathologic features.

1. Granulosa cell tumor (GCT).

GCT is most commonly malignant sex cord-stromal tumor as well as the most common estrogen secreting tumor. It is usually unilateral. Adult granulosa cell tumor (GCT) is much more common than juvenile type and accounts for 95% of all GCT and 5-10% of solid ovarian tumors. GCTs are usually occur in peri- and postmenopausal women (50-55 years). Irregular bleeding, endometrial hyperplasia, polyps, or carcinoma may occur due to hyperestrigenemia. Most patients have an excellent prognosis (>90% having 10-year survival rate) but there is a tendency of late recurrence even 20 years after diagnosis. Juvenile GCTs are more common in younger than 30 years old (mean age, 13 years old) and presents precocious puberty. They account about 10% of precocious puberty in girls.

The imaging features of GCT (Fig. 1 and 2): varies widely and has spectrum from solid mass to completely cystic tumor. GCTs are most commonly large, encapsulated, unilateral, multicystic mass with thick, irregular septa and solid component. A characteristic feature of GCTs is multiple blood filled cysts within the tumor. GCTs have no intracystic papillary projection, have less propensity for peritoneal seeding and are confined to the ovary at the time of diagnosis in contrast to the more common epithelial tumors.

2. Fibrothecoma
Fibroma and thecoma are forms of a spectrum of benign tumors and have spectrum of fibroma, fibrothecoma, and thecoma. Lipid-rich thecoma can show estrogenic activity. Fibroma is the most common sex cord tumor and associated with Meigs' syndrome—ascites, an ovarian tumor and right-sided pleural effusion.

The Imaging Features: are typical(Fig. 3 to 7). CT scan shows homogeneous solid tumor with dense calcification and delayed slight enhancement. MRI shows low SI on T1 and T2WI. Observation of the interface vessels between the uterus and adnexal masses seems to be useful in differentiating leiomyoma from ovarian fibroma(Fig.5).

3. Sclerosing stromal tumor

Sclerosing stromal tumor is rare benign tumor predominantly in young women, much younger than fibrothecoma(> 80% , less than 30 years old). The clinical symptom is menstrual irregularity.

Imaging Features(Fig.8): The mass is predominantly fibrotic and shows hemangioma-like enhancing pattern(early peripheral enhancement with centripetal progression). Peripheral striking early enhancement is due to cellular areas with prominent vascular network and central prolonged enhancement is due to collagenous hypocellular area.

4. Sertoli-Leydig cell tumor

Sertoli-Leydig cell tumor is the most common virilizing ovarian tumor having androgenic activity but very rare and usually occurs in young women. Sertoli-Leydig cell tumor is more common than Sertoli cell tumor.

Imaging Features(Fig.9 to 10): The tumors mostly behave in a benign fashion. They are almost always unilateral and solid or solid with cystic mass is more common than larger multicystic appearance. They tend to recur relatively soon after initial diagnosis. Calcification is unusual. They do not commonly contain the hemorrhagic cysts of granulosa cell tumor.

5. Steroid cell tumor

Steroid cell tumor is very rare and most commonly occurs in the 5th or 6th decade of life. Most are virilizing and approximately one third of them behave in a clinically malignant fashion.

Imaging Features: They are usually unilateral small solid tumor (< 3cm). Small area of cystic change or necrosis may be seen. The high signal intensity on T1WI was attributed to its lipid content. Intense Gd-enhancement reflects rich vascularity.

6. Specific consideration of functional ovarian tumors
Functional ovarian tumors have unique clinical manifestations related to hormone overproduction and may give rise to a broad spectrum of clinical syndromes. Sex cord-stromal tumors, the most common functional ovarian neoplasms, are associated with either hyperestrogenism (as in granulosa cell tumor and thecoma) or hyperandrogenism (as in Sertoli-Leydig cell tumor and Leydig cell tumor). Other, less common ovarian neoplasms that may have endocrine or nonendocrine syndromic manifestations include germ cell tumors associated with the excessive production of human chorionic gonadotropin (eg, choriocarcinoma, dysgerminoma), monodermal teratomas (eg, carcinoid tumor, struma ovarii) associated with carcinoid syndrome and hyperthyroidism, and primary epithelial ovarian cancers associated with paraneoplastic syndromes.
**Fig. 1:** F/42 Left ovarian granulosa cell tumor with endometrial hyperplasia (Hyperestrogenism). Large multicystic mass (M) is seen in lower abdomen and pelvis, which contains hemorrhage. Associated endometrial hyperplasia (arrow) is also noted.

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**Fig. 2:** F/46 Left ovarian granulosa cell tumor. Large solid and multicystic mass (M) with hemorrhage (arrow) is seen in transvaginal US and MRI.

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**Fig. 3:** F/33 Right ovarian fibroma. Typical homogeneous mild enhancing solid mass (M) is seen in enhanced CT scan and MRI. The mass shows low SI on T2WI.

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**Fig. 4:** F/27 Right ovarian fibroma. Typical homogeneous mild enhancing solid mass (M) with T2 low SI. Crescent-shaped ipsilateral right ovary (arrows) are evident. Time-to-SI curve from DCE MR shows mild enhancement (blue).

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Fig. 5: F/33 Subserosal leiomyoma. Typical homogeneous well enhancing mass (M) with T2 low SI. Associated bridging or interfaced vessel (arrow) between mass and uterus. Time-to-SI curve from DCE MR shows strong enhancement comparable myometrium (yellow).

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Fig. 6: F/22 Left ovarian fibroma (Typical feature). T1 and T2 typical low SI mass with mild enhancement (M). Abutting and crescent-shaped ipsilateral left ovary (arrow) is also noted.

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**Fig. 7:** F/57 Left ovarian thecoma. Large heterogeneous hemorrhagic mass mimic malignant tumor (Atypical feature). Large heterogeneous enhancing solid mass (arrows) with hemorrhage. Associated subserosal leiomyoma (M) with bridging vessels (short arrow) are also noted. Microphotographs show solid sheets of round to spindle cells with pale cytoplasm intermixed with collagenous stroma. Hemorrhage and necrosis are also evident.

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**Fig. 8:** F/12 Right ovarian sclerosing stromal tumor. Large solid and cystic mass (M) with characteristic peripheral strong enhancement (arrows).

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**Fig. 9:** F/27 Right ovarian Sertoli-Leydig cell tumor. She suffered from amenorrhea and virilizing symptoms with elevation of male sex hormone (testosterone). Solid appearing right ovarian mass (M). Microphotographs show intermediate differentiated sertoli and Leydig cells.

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**Fig. 10:** F/23 Right ovarian sertoli cell tumor. She suffered from virilizing symptoms. Large multilobulated solid and cystic mass (M). Enhancing solid portion (arrow) is evident.

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Fig. 11: F/74 Right ovarian Brenner tumor and mucinous borderline tumor (Hyperestrogenism). Large heterogeneous mass including low SI solid portion (M) and complex cystic lesion (arrows). Increased uterine size due to hyperestrogenism probable due to ovarian tumor. Photograph of gross specimen with cut section and microphotograph reveal solid and cystic area. Cystic lesion is filled with mucinous material (M, mucinous tumor). The solid lesion appears bright yellow to white-tan, mutinodular (B, Brenner tumor).

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Findings and procedure details

In this presentation, we describe imaging features of the benign (fibroma, fibrothecoma, thecoma, sclerosing stromal tumor, sertoli-leydig cell tumor, and steroid cell tumor) and malignant (granulosa cell tumor) sex cord-stromal ovarian tumor. We also discuss possible differential point from complex multicystic lesion (central enhancing granulosa cell tumor, peripheral enhancing sclerosing stromal tumor) and functioning or hormonal active tumor. We also correlate imaging features with clinical and pathologic features.
Conclusion

The major teaching points of this exhibit are:

1. Sex cord-Stromal ovarian tumors are broad spectrum, from benign to malignant and cystic to solid. It is important to know these conditions for narrowing the differential diagnosis.

2. Familiarity with the clinical setting and imaging feature of sex cord-stromal ovarian tumor as depicted with US, CT and MRI will facilitate prompt and accurate diagnosis and treatment.
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References