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Comprehensive review of imaging features of sex cord-stromal tumors of the ovary

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Abstract

Sex cord-stromal tumors of the ovary (SCST) are uncommon ovarian tumors arising from sex cord and/or stromal cells of the ovaries. They may be nonfunctional and asymptomatic or functional presenting with hyperestrogenic, hyperandrogenic or cushingoid symptoms. They present in a wide age group of women, mostly in early stages and follow a nonaggressive clinical course after surgical resection. They differ from more prevalent epithelial ovarian tumors which tend to present in older women in advanced stages with poor prognosis. Some of SCSTs are associated with clinical syndromes. We will review imaging features on ultrasound, computed tomography and magnetic resonance imaging, epidemiology and clinical presentations of these tumors.

Keyword Sex cord-stromal tumors of the ovary · Fibroma · Granulosa cell tumor · Sertoli cell tumor · Thecoma

Introduction

Sex cord-stromal tumors of the ovary (SCST) are uncommon ovarian tumors, representing only 7–8% of ovarian neoplasms [1]. The incidence of SCSTs was reported as 0.2 per 100,000 women in the United States with higher incidence rate in African American women compared to white women (0.44 versus 0.18 per 100,000) [2].

SCSTs consist of a heterogeneous group of tumors. The World Health Organization (WHO) classified sex cord-stromal tumors of the ovary into 3 main categories, based on the cell type that they arise from including pure sex cord tumors, pure stromal tumors and mixed sex cord-stromal tumors (Table 1) [3]. These tumors are further subdivided and some are associated with specific genetic syndromes or result in systemic disorders (Table 2).

Sex cords include granulosa and Sertoli cells, while stromal cells include theca cells, Leydig cells and fibroblasts. SCSTs can present in any age group and have a variable clinical presentation. They may be diagnosed incidentally as nonfunctioning asymptomatic ovarian masses or present as hormone-producing tumors associated with estrogenic, androgenic or cushingoid symptoms [4]. Tumors arising from ovarian granulosa and theca cells often produce estrogen and may result in abnormal uterine bleeding, endometrial hyperplasia or carcinoma in adults or precocious puberty in children. In contrast, tumors arising from Leydig and Sertoli cells produce testosterone and may present with hirsutism, acne, deepening of the voice, clitorial hypertrophy, amenorrhea or irregular menstrual cycles [4, 5].

SCSTs differ epidemiologically, radiographically, histopathologically and clinically from the more prevalent epithelial tumors which represent approximately 90% of ovarian tumors [6]. The mean age at diagnosis for SCSTs is 50 years compared to 61 years for epithelial ovarian tumors [2]. Unlike epithelial tumors, SCSTs are not associated with BRCA gene mutations and do not demonstrate predisposition to breast cancer except for Granulosa cell tumors which are more frequently seen in patients with family history of breast and ovarian cancers [7]. Furthermore, epithelial tumors commonly present at advanced stages (III or IV) at the time diagnosis and are treated with chemotherapy and surgical debulking. SCSTs most commonly present in younger patients at early stages (I) and are mainly treated surgically with overall better prognosis [8]. Some

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Table 1 WHO classification of ovarian sex cord-stromal tumors (5)

Pure sex cord tumors	Pure stromal tumors	Mixed sex cord-stromal tumors
Adult Granulosa cell tumor	Fibroma	Sertoli-Leydig cell tumors
Juvenile Granulosa cell tumor	Cellular fibroma	
Sertoli cell tumor	Thecoma	
Sex cord tumor with annular tubules	Luteinized thecoma associated with sclerosing peritonitis	
	Fibrosarcoma	
	Sclerosing stromal tumor	
	Signet-ring stromal tumor	
	Microcystic stromal tumor	
	Leydig cell tumor	
	Steroid cell tumor	
	Steroid cell tumor, malignant	

WHO World Health Organization, *NOS* Not otherwise specified

Table 2 Sex cord-stromal tumors of the ovary with associated genetic syndromes and systemic disorders

Sex cord-stromal tumors	Associated syndromes and systemic disorders
Juvenile granulosa cell tumor	Ollier's disease, Maffucci syndrome
Fibroma	Meigs syndrome and Gorlin syndrome
Sex cord with annular tubules	Peutz–Jeghers syndrome
Sertoli cell tumor	Peutz–Jeghers syndrome
Steroid cell tumor	Cushing's syndrome, VHL disease
Sclerosing stromal tumor	Gorlin syndrome

SCSTs demonstrate characteristic imaging features, which in conjunction with clinical symptoms can point to accurate diagnosis.

More common sex cord-stromal tumors of the ovary

Granulosa cell tumors

Granulosa cell tumors (GCTs) are the most common potentially malignant sex cord-stromal tumors of the ovary, accounting for 90% of malignant SCSTs and 5% of all malignant ovarian tumors [9, 10]. These tumors are further divided into adult and juvenile types. The adult type which constitutes of 95% of Granulosa cell tumors affects postmenopausal women (peak age of 50–55 years), whereas juvenile type affects children and younger women (< 30 years old), although only 5% of the GCTs occur in prepubertal girls [5, 11]. These tumors typically result in hyperestrogenicity symptoms, although up to 20% are reported to be asymptomatic [5]. Juvenile GCTs have an association with Ollier's disease and Maffucci's syndrome [12]. Most common presenting symptoms are vaginal bleeding and palpable pelvic masses [5]. GCTs are commonly unilateral (right-sided predominance). If associated with hyperestrogenic symptoms, they may be discovered when smaller in size. Otherwise, they typically tend to be large in size at diagnosis [13]. Mean tumor size of 9.2–10.4 cm (range, 0.2–40 cm) among adult GCTs and 12.4 cm (range, 2–26 cm) among juvenile GCTs has been reported [5]. Large SCTs may compress adjacent organs and result in abdominal symptoms [5]. They commonly present at early stages (74–95% at stage I versus only 0.5–9% at stage IV) with favorable prognosis [5]. In a study of 37 women with stage I disease, survival rates at 5, 10 and 20 years were reported as 94, 82 and 62%, respectively [14]. Recurrence, most commonly presenting as peritoneal disease, can occur several years after diagnosis [5, 11].

The adult and juvenile GCTs demonstrate similar imaging features despite differences in age of onset, clinical

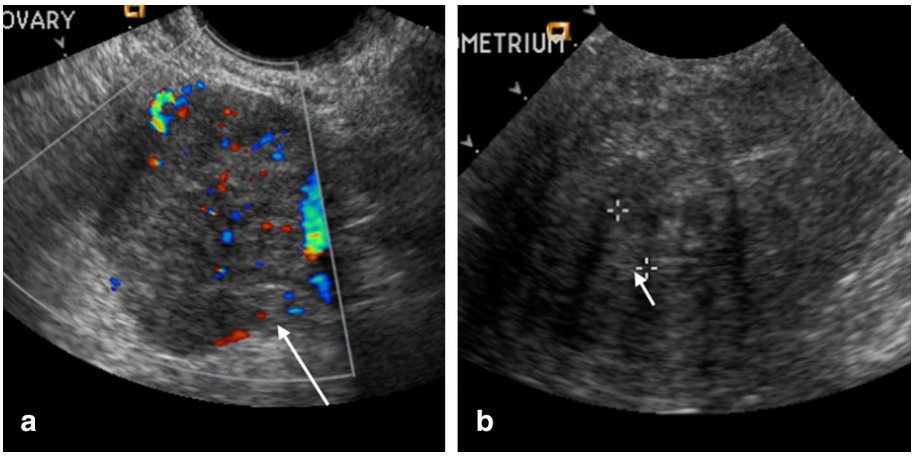

Fig. 1 A 54-year-old woman with ovarian granulosa cell tumor. Ultrasound image **a** demonstrates a hypoechoic solid ovarian mass (long arrow) consistent with GCT and **b** demonstrates a thickened endometrial stripe (short arrow) in the same patient which was biopsy proven endometrial cancer

presentation and histologic appearance. On ultrasound (US) and computed tomography (CT), they most commonly appear as multicystic masses with solid components or solid heterogeneous masses (Fig. 1) [15, 16]. Thick- or thin-walled unilocular cystic or homogeneous solid GCTs are uncommon [13]. On magnetic resonance imaging (MRI), they have a "sponge-like" appearance due to multicystic nature of the mass [17]. On T2-weighted images, the cystic components are most likely hyperintense with hypointense septations and solid components may be isointense (Fig. 2). Fluid–fluid levels may be seen due to internal hemorrhagic content which demonstrates T1 hyperintense signal [17]. Solid component usually demonstrates heterogeneous contrast enhancement on CT and MRI due to intra-tumoral hemorrhage, infarct or degeneration [8]. GCTs may be associated with uterine changes like uterine enlargement, hemorrhage or endometrial thickening due to hyperplasia, polyps or less commonly carcinoma secondary to hormonal stimulation [18]. Multicystic GCTs may mimic epithelial tumors of the ovary but unlike epithelial tumors, they tend to be unilateral and peritoneal carcinomatosis is rare at the time of diagnosis [18].

The treatment of choice for GCTs is early and complete surgical resection with surgical staging; however, the extent of surgery remains a topic of discussion, particularly when fertility preservation is desired [5]. A large study of 2680 women with GCTs did not demonstrate increased survival with adjuvant chemotherapy [7].

Fibroma, cellular fibroma, fibrosarcoma

Fibromas are the most common sex cord-stromal tumors of the ovary, accounting for 4% of all ovarian tumors. They most commonly occur in women in their 4th decade of life [11]. They arise from nonfunctioning fibroblast cells of the ovarian stroma and do not demonstrate hormone-mediated symptoms. They are considered benign tumors, usually asymptomatic and discovered incidentally. However, larger tumors may cause pelvic or acute abdominal pain due to ovarian torsion [19]. Different degrees of ascites have been reported in 63% of patients with fibromas [20]. Fibromas are associated with Meigs' syndrome (ascites, hydrothorax and benign ovarian tumor) which may demonstrate elevated CA-125 level [21], mimicking malignancy. Ascites and

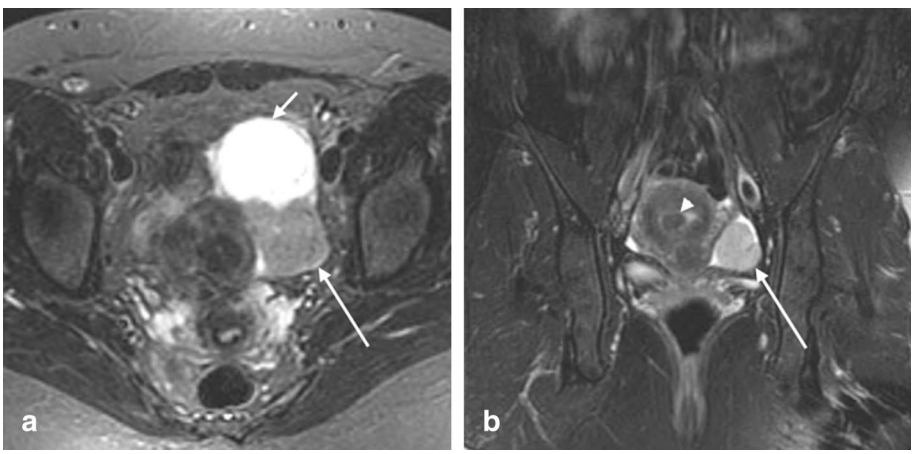

Fig. 2 A 58-year-old woman with ovarian granulosa cell tumor. Axial T2-W MR Image of the pelvis **a** demonstrates a solid left ovarian mass with T2 intermediate signal (long white arrow) with a T2 hyperintense cystic component (short white arrow) consistent with GCT. Coronal T2-W MR Image **b** demonstrates a T2 hypointense filling defect in the endometrial cavity (arrow head) consistent with biopsy proven polyp as well as the left ovarian GCT (white arrow)

pleural effusion may often resolve after tumor resection [22]. Fibromas are also associated with Gorlin syndrome (nevoid basal cell carcinoma), a rare autosomal dominant disease. In these cases, fibromas can be bilateral [23].

Since fibromas have fibrous and collagenous contents, they usually appear as a homogeneous solid mass on imaging. Chung et al. reported that 84% of the fibromas are predominantly solid masses, 11% mixed solid and cystic, and only 5% occur as predominantly cystic lesions [20]. On US, they are usually hypoechoic, although hyperechoic appearance has also been reported. They demonstrate a homogeneous delayed enhancement on CT. Rarely, calcifications may be seen. On MRI, they demonstrate a hypointense signal on T1- and T2-weighted images with delayed enhancement on post-contrast images (Fig. 3) [10]. A study demonstrated that fibromas had the lowest ADC value (0.470×10^{-3} mm^2/s) compared to malignant ($0.825 \pm 0.129 \times 10^{-3}$ mm^2/s) or benign ($1.343 \pm 0.528 \times 10^{-3}$ mm^2/s) SCSTs of the ovary [24]. The time-signal intensity curve on perfusion-weighted MR images were classified as type I or II consistent with benign etiology of these tumors [20]. Large fibromas (> 6 cm) and fibrosarcomas can demonstrate areas of increased signal intensity on T2-weighted images due to necrosis, hemorrhage or cystic degeneration [10, 20]. Twisted ovarian fibroma may also demonstrate increased signal intensity on T1- and T2-weighted images due to passive congestion of the mass [19]. Given that fibromas are solid masses, detection of hemorrhagic infarction after torsion can be difficult, in these cases, area of high signal intensity in the periphery of the tumor on T1-weighted images can be helpful in identifying a hemorrhagic infarction of the fibroma [25]. In cases of exophytic fibromas that grow from periphery of the ovary, the shape of the ovary can be preserved, particularly in premenopausal women [26]. Given that fibrous tumors including non-degenerated pedunculated uterine and broad-ligament leiomyomas similarly demonstrate low signal intensity on T1- and T2-weighted images, they can be mistaken for ovarian fibromas. Identifying a pedicular attachment to the uterus and vascular signal voids between the uterus and leiomyoma and detecting feeding ovarian vessels can help distinguishing these entities [10].

Cellular fibromas account for 10% of ovarian fibromas and demonstrate low malignant potential. They are characterized by mild increased cellular density, mild nuclear atypia and 3 or fewer mitotic figures per 10 high power fields (HPF). If the mitotic figures are more than 4 per 10

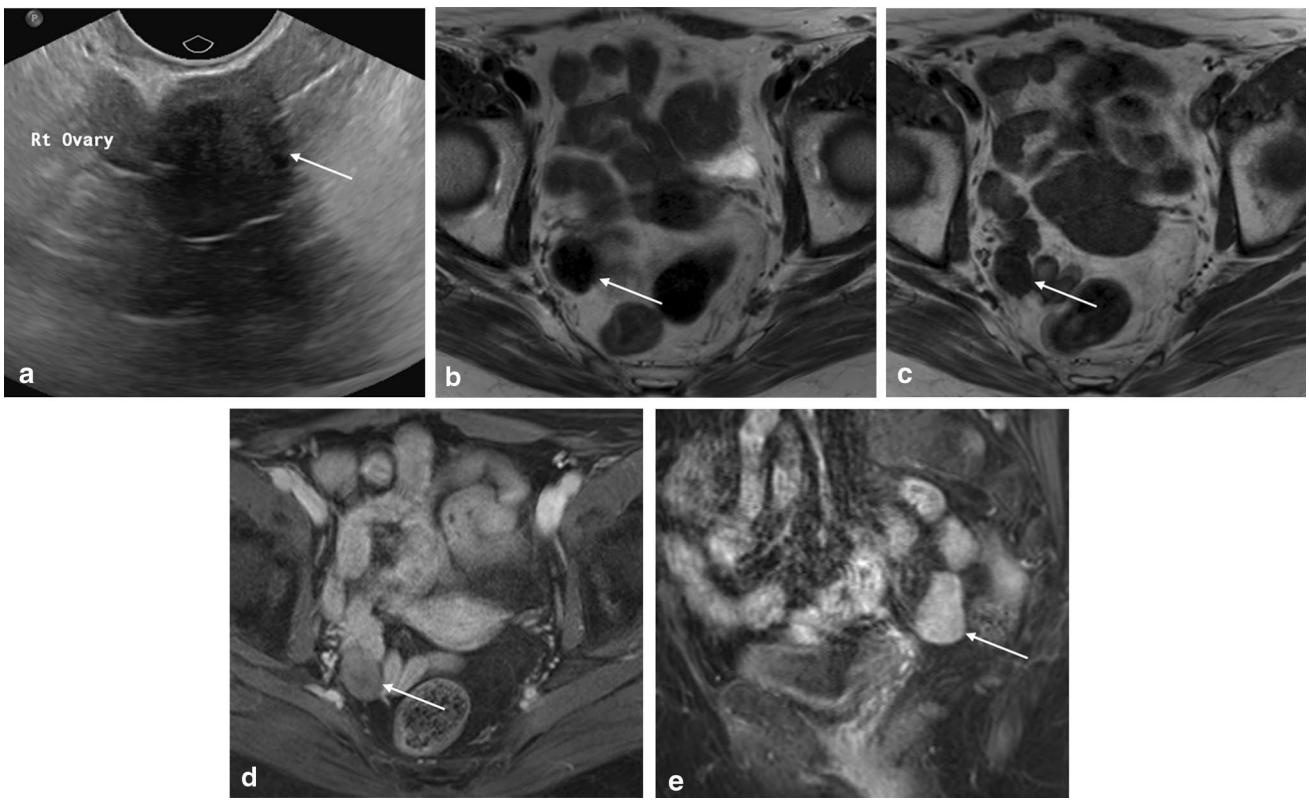

Fig. 3 A 65-year-old woman with ovarian fibroma. Ultrasound image of the pelvis (**a**) demonstrates a solid hypoechoic right ovarian mass (arrow). Axial T2-W (**b**) and T1-W (**c**) MR images of the pelvis demonstrate a right ovarian hypointense homogenous mass (arrows) with enhancement on axial T1-W post-contrast image (**d**) and more intense enhancement on delayed phase image on sagittal T1-W post-contrast image (**e**)

HPF, they are considered mitotically active cellular fibromas and need to be differentiated from fibrosarcomas which are identified by 4 or more mitotic figures per 10 HPF with significant cellular density and nuclear atypia [27]. They have similar clinical presentations as fibromas but tend to be larger resulting in increased necrosis, hemorrhage or torsion (Fig. 4) [19]. Fibrosarcomas are very rare malignant tumors [28]. The number of mitoses, mitotic activity and degree of anaplasia correlates with aggressiveness [28]. Fibrosarcomas are managed surgically and adjuvant chemotherapy with improved outcome [28].

Except rare cellular fibromas and fibrosarcomas, fibromas are benign tumors and can be cured with surgical resection.

Thecoma and luteinized thecoma associated with sclerosing peritonitis

Thecomas are composed of theca cells and are rare, benign tumors, most commonly seen in postmenopausal women [3]. They account only for 1% of all primary ovarian tumors [29]. They can demonstrate signs and symptoms of hyperestrogenism including endometrial hyperplasia and carcinoma in 15% and 20–25%, respectively [4]. Although, rarely, they may demonstrate hyperandrogenic symptoms [29]. The term "fibrothecoma" refers to rare tumors with intermediate features between a fibroma and a thecoma; however, fibrothecoma is not part of the WHO classifications of ovarian tumors [30].

Thecomas usually tend to be unilateral, solid ovarian masses and have similar imaging appearance to other solid ovarian tumors. A study by Zhang et al. demonstrated that thecomas/fibrothecomas were mostly homogenous solid masses with similar signal intensity to myometrium on DWI and similar ADC values to leiomyomas [31]; however, they demonstrated higher signal intensity on T2-weighted images and more avid enhancement on post-contrast images compared to predominantly fibrous tumors [31]. Another study reported large tumors (> 5 cm) demonstrate hyperintense or mixed hyperintense signals on DWI as well as cystic changes and pelvic fluid, whereas small tumors (< 5 cm) demonstrate a hyperintense signal on DWI but cystic changes were uncommon [32]. Malignant tumors are usually detected on positron emission tomography-computed tomography (PET/CT) by demonstrating increased fluorodeoxyglucose F^{18} (^{18}F-FDG) activity; however, thecomas occasionally can produce false-positive results confounding the diagnosis [33].

Luteinized thecomas associated with sclerosing peritonitis are extremely rare and have been reported in younger premenopausal patients [34]. They typically present with symptoms like abdominal pain and distention, ascites and pleural effusion and sometimes with small bowel obstruction [35]. Acute abdomen due to hemorrhagic necrosis and rupture of the tumor as well as death due sclerosing peritonitis have also been reported [34, 36]. Bilateral luteinized thecoma of the ovaries have been linked to anticonvulsant therapy [37]. Sclerosing peritonitis which is described as chronic fibro-inflammatory disease of the peritoneum, results in thick fibrous membrane formation causing recurrent small bowel obstruction [36]. On X-ray, air-fluid level in bowels can be seen. On US, fibrous membranes are demonstrated as hyperechoic linear structures encasing bowel loops. On CT and MRI, clustered small bowel loops due to a thin membrane surrounding and tethering the bowel loops are seen. Fibrous membranes are hypointense on T2-weighted MR images [38]. It is crucial to biopsy the omentum in these cases to establish the correct diagnosis. Surgical excision is considered the treatment of choice for these tumors.

Less common sex cord-stromal tumors of the ovary

Sertoli cell tumor

Sertoli cell tumors are very rare tumors. They are associated with Peutz-Jeghers syndrome (PJS), a rare autosomal

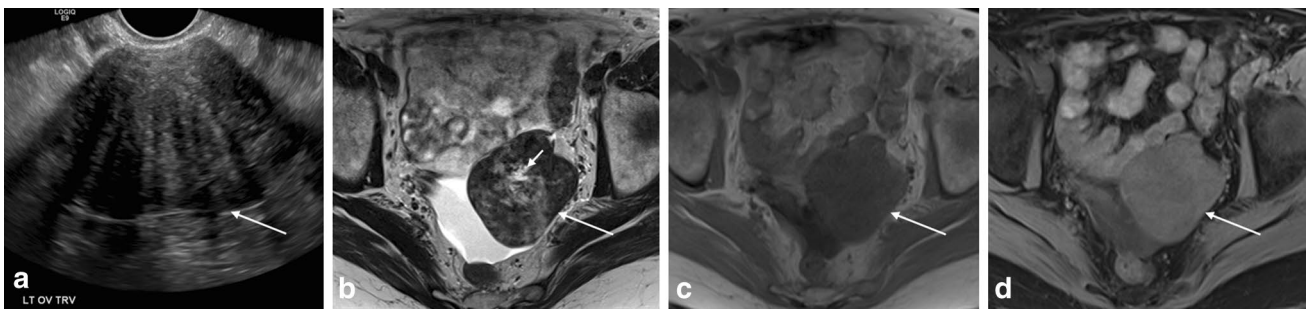

Fig. 4 A 56-year-old woman with cellular fibroma. Ultrasound image of the pelvis (**a**) demonstrates a large heterogeneous hypoechoic left ovarian mass (arrow). Axial T2-W (**b**) MR image of the pelvis demonstrates a T2 hypointense heterogenous mass (long arrow) with cystic components (short arrow). The mass (arrow) is hypointense on axial T1 (**c**) with enhancement on axial T1 post-contrast (**d**) MR images

dominant condition defined by gastrointestinal polyps and mucocutaneous dark spots [39]. They produce estrogen and result in menstrual irregularities and postmenopausal bleeding in women and precocious puberty in girls. They are usually unilateral solid masses ranging 4–12 cm in size [40]. On US, they are echogenic solid masses and on CT and MRI, they appear as encapsulated solid enhancing masses [39]. They have an excellent prognosis, and are associated with low recurrence rate after resection [41].

Sex cord tumor with annular tubules

Sex cord tumor with annular tubules is a rare tumor occurring in women of reproductive age. It accounts only for 6% of sex cord-stromal tumors of the ovary. However, they have a strong association with Peutz–Jeghers syndrome. Approximately 30% of these tumors occur in women with PJS [42] and they tend to be bilateral in two thirds of the cases, whereas in non-PJS cases, the tumors tend to be unilateral, large and malignant in 20% of the cases [11, 42].

On imaging, they are solid tumors with cystic spaces [43]. On US, they have been reported to be multiple confluent, lobulated echogenic masses or tumorlets (Fig. 5) [44].

Sclerosing stromal tumor

Sclerosing stromal tumors (SST) are rare, benign tumors seen most commonly in young women. They are usually unilateral masses, although bilateral tumors have been described in a premenarchal girl and a pregnant woman with Gorlin syndrome [45–47].

On US, SSTs are usually unilateral solid masses, isoechoic to myometrium with cleft-like hypo- or anechoic cystic areas [48, 49]. On color Doppler images, they demonstrate peripheral and internal intercystic hypervascularity with low-impedance flow [48]. On CT, they show a nonhomogeneous solid appearance in the periphery and star-shaped low density internally (Fig. 6) [49]. On MRI, they present as a heterogeneous solid mass with hyperintense cystic components on T2-weighted MR images. Due to slow growing nature of these tumors, a thick peripheral hypointense rim can be seen on T2-weighted images due to compressed ovarian tissue. On post-contrast images on CT and MRI, marked early peripheral enhancement with centripetal progression is seen, differentiating them from fibromas which show delayed enhancement [17]. Given benign nature of SCTs, these tumors can be cured with surgical resection [45].

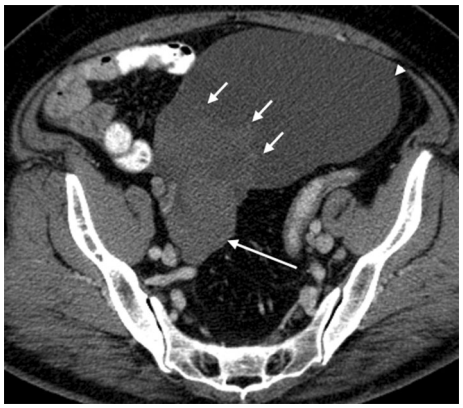

Fig. 6 A 32-year-old woman with ovarian sclerosing stromal tumor. Axial CT image of the pelvis demonstrates a large right ovarian nonhomogeneous solid mass (long arrow) with star-shaped low density internally (short arrows) and a cystic component (arrow head)

Signet-ring stromal tumor and microcystic stromal tumor

Signet-ring stromal tumors of the ovary are extremely rare. They are benign ovarian tumors and can be mistaken for metastatic signet-ring adenocarcinomas (krukenberg tumor), particularly if bilateral and multinodular in appearance [50, 51]. They appear as solid masses with nonspecific or unestablished imaging features [52].

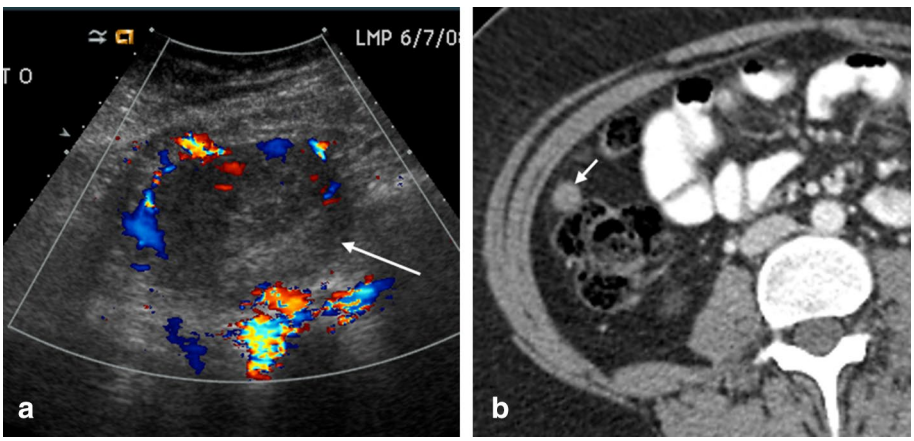

Fig. 5 A 15-year-old girl with sex cord tumor with annular tubules. Ultrasound image of the pelvis (**a**) demonstrates a heterogenous solid ovarian mass with cystic changes (arrow). Axial CT image of the abdomen (**b**), 6 months after surgical resection demonstrates a right paracolic gutter implant (short arrow)

Microcystic stromal tumors of the ovary are also very rare tumors with favorable clinical course. A few case reports describe them as unilateral predominantly cystic masses with solid components that demonstrate mild FDG activity on PET/CT [53, 54]. The cystic component appears as fluid density on CT, anechoic on US and hyperintense on T2-weighted MR images [54]. Although, the imaging features are nonspecific and can be seen in malignant epithelial tumors, malignant potential has not yet been associated with microcystic stromal tumors [54].

Steroid cell tumor and leydig cell tumor

Steroid cell tumors of the ovary are rare tumors accounting for less than 0.1% of all ovarian tumors [55]. They are usually seen in women in 5th and 6th decades of life and present with hyperandrogenic symptoms including virilization and rarely hyperestrogenic symptoms and Cushing's syndrome [11, 56]. The symptoms related to hyperandrogenism can be masked in pregnancy [57]. They tend to present as unilateral solid ovarian tumors but multilocular cystic mass with mural nodule appearance have also been reported [58]. On MRI, they demonstrate increased signal on T1-weighted images and drop of signal in out-of-phase images compared to in-phase images on dual echo gradient-echo (GRE) MRI (chemical shift artifact) due to significant intracellular fat with strong enhancement on post-contrast images due to hypervascularity (Fig. 7) [59, 60]. They have been reported to be associated with von Hippel-Lindau (VHL) syndrome [59]. One third of the steroid cell tumors are thought to be malignant [55]. The larger size tumors and higher stage are associated with poor prognosis in one study [55].

Leydig cell tumors of the ovary occur in older women who may present with similar hyperandrogenic symptoms. However, unlike steroid cell tumors, leydig cell tumors are benign [61]. They also tend to present as unilateral solid masses but are typically smaller in size (average size of 2.4 cm) compared to Steroid cell tumors (average size of 8.4 cm) [8]. On US, both Steroid cell tumors and Leydig cell tumors are isoechoic to myometrium and on CT they are hypodense [58]. On MRI, the signal intensity on T2-weighted images depends on the amount of fibrous

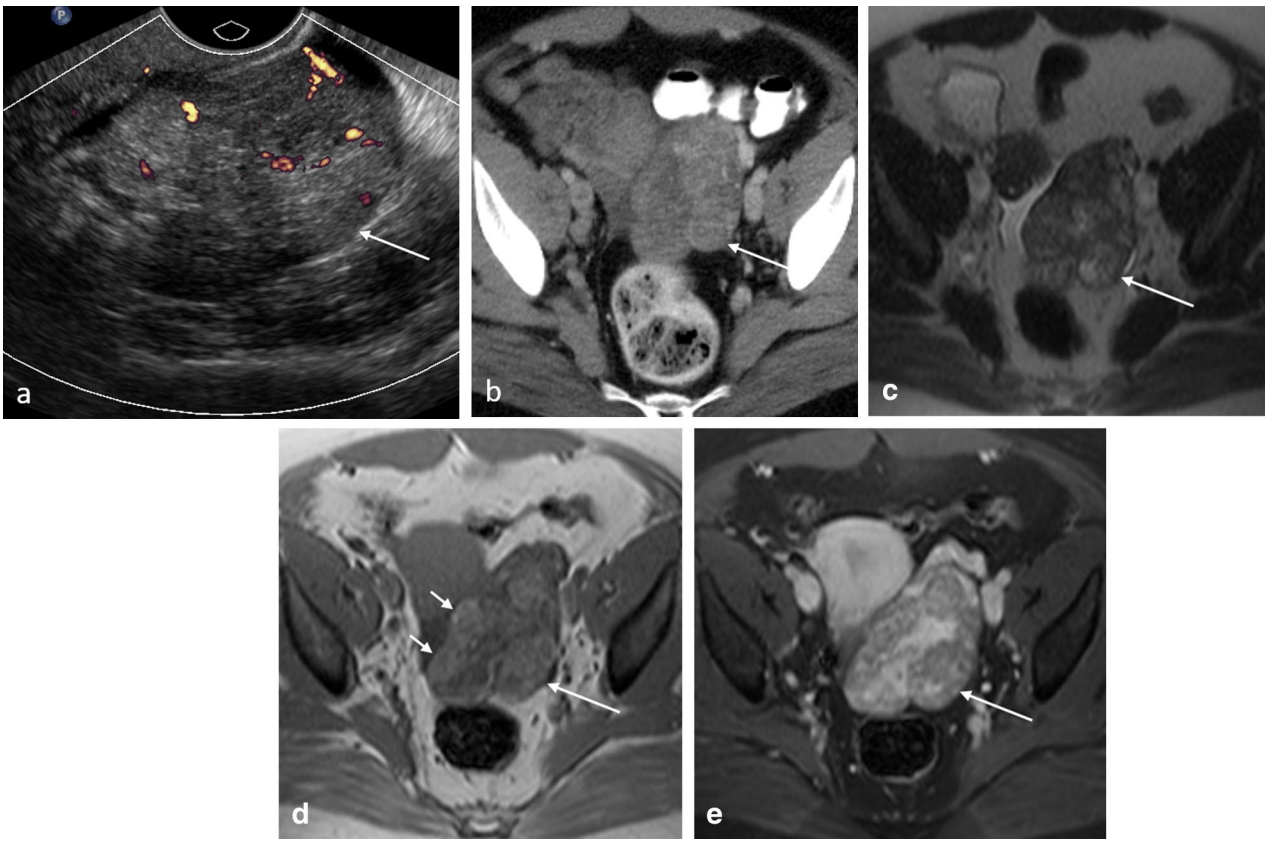

Fig. 7 A 42-year-old woman with ovarian steroid cell tumor. Ultrasound image (**a**) demonstrates a hypoechoic solid vascular ovarian mass (arrow). Axial CT image of the pelvis (**b**) shows a lobular heterogenous solid enhancing left ovarian mass (arrow). Corresponding axial T2-W (**c**) MR image of the pelvis demonstrates a T2 hyperintense heterogenous solid left ovarian mass (arrow). On T1-W image (**d**), areas of hyperintense signal (short arrows) is suggestive of fat. The mass is heterogeneously enhancing (arrow) on T1 post-contrast (**e**) MR image

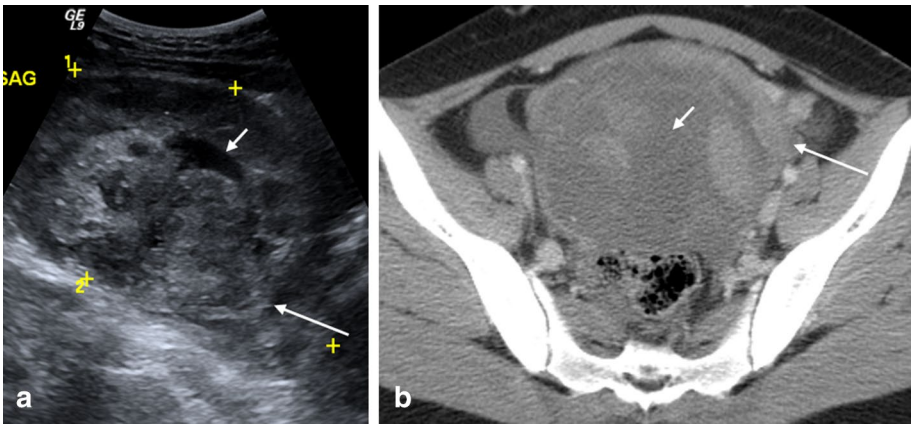

Fig. 8 A 50-year-old woman with Sertoli-Leydig cell tumor of the ovary. Ultrasound image of the pelvis (**a**) demonstrates a hypoechoic heterogenous solid ovarian mass (long arrow) with cystic areas (short arrow). Axial CT image of the pelvis (**b**) shows soft tissue density mass with marked enhancement (long arrow) and cystic areas (short arrow)

content [58, 60]. Leydig cell tumors, demonstrate gradual increasing and delayed enhancement. Given small size and solid appearance, they can be difficult to differentiate from ovarian tissue. In these case, DWI sequence can be helpful, since the tumor demonstrates slightly higher signal intensity compared to ovarian tissue [62].

The treatment for both tumors is surgical resection. In cases of malignant steroid cell tumors, chemotherapy is also warranted [55].

Sertoli-Leydig cell tumors

Sertoli-Leydig cell tumors (SLCT) are rare ovarian tumors, most commonly seen in young women (< 30 years of age) [11]. SLCTs are the most common hyperandrogenic tumors of the ovary and most patients develop virilization symptoms [11]. They are usually unilateral solid masses (98%) with nonspecific imaging appearance [8]. On US, they appear as a hypoechoic mass or a heterogeneous solid mass with cystic areas and on CT, they may have soft tissue density with marked enhancement on post-contrast images (Fig. 8). On MRI, they demonstrate low signal on T1- and intermediate signal on T2-weighted images and depending on the amount of fibrous content of the tumor, they may demonstrate hypointense signal on T2-weighted images [63]. Hyperintense areas on T2-weighted images represent cystic changes. Multilocular cystic tumors with irregular septa and mural nodules have also been reported [63]. Similar to CT, they demonstrate marked enhancement on post-contrast images [63].

Most SLCTs are detected in the early stages and have favorable outcome with conservative surgeries, given the young age of the patients. Adjuvant chemotherapy is recommended in moderate and poorly differentiated types [64].

Conclusion

Sex cord-stromal tumors of the ovary are rare tumors that arise from sex cords or stromal cells and constitute the majority of the hormone-producing ovarian tumors. The demographics, histopathology, clinical features and prognosis of these tumors differ significantly from the more common epithelial ovarian tumors.

The radiologic appearances are variable but some of these tumors may present with specific clinical symptoms and imaging findings. Knowledge of pertinent clinical symptoms in conjunction with the hormones they produce and the imaging findings may help in making a correct diagnosis, directing to appropriate management including conservative and fertility-sparing surgeries in younger patients.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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