Exploration of the Impact of Ovarian Sex Cord-Stromal Tumor (SCST) Classification on Diagnostic Coding

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Abstract: Ovarian sex cord-stromal tumors (SCSTs) are relatively rare with numerous subtypes, making preoperative diagnosis difficult. Treatment plans should be individualized based on pathological types. In practice, diagnostic coding errors frequently occur with ovarian SCSTs. According to pathology report results and combined with the WHO Classification of Female Genital Tumors, 5th edition, coding analysis involves disease diagnostic codes C56.x, D27.x, D39.1, and pathological diagnostic codes M8632/1, M8634/1, M8634/3. As key molecular alterations affecting tumor classification continue to be discovered, the integration of morphological and molecular classification has gradually replaced the previously pure morphological classification of female genital tumors, impacting diagnosis and treatment. Coders must master the latest WHO version of ovarian SCST morphological and histological classifications, promptly review differences between new and old editions, strengthen communication with pathologists and clinicians, and derive accurate coding through comprehensive analysis.

***Keywords*:** sex cord-stromal tumor; morphological and histological classification; diagnostic coding

1. Introduction

Ovarian sex cord-stromal tumors (SCSTs) are a relatively rare group of ovarian tumors, accounting for 5% to 8% of ovarian tumors, with an incidence rate of (0.09~0.10) per 100,000 annually. They predominantly affect adolescent and reproductive-age women, while adult granulosa cell tumors (GCTs) are mostly seen in perimenopausal women aged 50–55 years. SCSTs comprise numerous subtypes, making preoperative diagnosis difficult; histopathological differential diagnosis is also challenging. Some malignant subtypes have a wide age range compared to other ovarian malignancies. Based on the 2014 classification, new tumor subtypes have been added, and key molecular genetic alterations of ovarian SCSTs updated [1]. Morphological coding for this disease is complex, as morphological features may overlap. Immunohistochemical staining assists in confirming sex cord-stromal differentiation but is difficult to clearly distinguish specific subtypes. Besides mastering tumor disease coding lookup rules, comprehensive consideration of morphological-molecular classification results combined with clinical context is necessary, alongside continuous learning of updates in the WHO tumor histological classification.

2. Changes in the Names of Ovarian Sex Cord-Stromal Tumors (SCST) Between the New and Old Versions

(1) Sex cord-stromal tumors are still divided into three major types: pure stromal tumors, pure sex cord tumors, and mixed sex cord-stromal tumors. Among pure stromal tumors, the luteinized thecoma with sclerosing peritonitis was renamed to luteinized theca cell tumor, removing the association with sclerosing peritonitis; the thecoma was renamed to theca cell tumor, adding the word "cell."

(2) In pure sex cord tumors, adult granulosa cell tumor and juvenile granulosa cell tumor were renamed to adult granulosa cell tumor and juvenile granulosa cell tumor, respectively; the name of the tubular sex cord tumor was changed to sex cord tumor with tubular structures.

3. Updates in the Classification of Ovarian Sex Cord-Stromal Tumors (SCST)

The 5th edition of the WHO Classification of Female Genital Tumors, based on the 2014 edition, added several new tumor subtypes, emphasized the differing pathogenesis between HPV-related and non-HPV-dependent tumors in lower genital tract epithelial tumors and their relationship with clinical treatment and prognosis, updated key molecular genetic alterations of tumors including ovarian SCSTs, and provided a more detailed description of the histological features and patient prognosis evaluation for each tumor [2].

(1) The 5th edition WHO ovarian SCST classification reintroduced gonadoblastoma, which contains female

components (including AGCT or JGCT) and male components (including Sertoli cell tumor or Sertoli-Leydig cell tumor). The most common combination is a predominance of Sertoli-Leydig cell tumor components with fewer JGCT components. Immunohistochemical staining shows that both tumor components usually express sex cord-derived tissue markers (such as inhibin and FOXL2) positively. Most gonadoblastomas are benign with rare recurrence [2]. However, the added histological classification of gonadoblastoma remains dynamic and uncertain; the pathological diagnostic code is M8632/1.

(2) In the new version, the subtype of Sertoli-Leydig cell tumor with heterologous elements as a separate classification was removed, and the pathological diagnostic codes M8634/1, M8634/1, and M8634/3 were cancelled.

4. Case Analysis

(1) A search was conducted in a tertiary general hospital for cases from January 1, 2019 to June 1, 2025, with a primary diagnosis of ovarian malignant tumor, totaling 159 cases. Among them, 58 cases were judged to have incorrect primary diagnosis selection based on clinical course records and pathology results; 13 cases were ovarian sex cord-stromal tumors, all adult granulosa cell tumors.

Patient: Female, 45 years old. In 2011, she presented with lower abdominal pain. Ultrasound and CT revealed a mixed tumor above the uterus. Under general anesthesia, a "left fallopian tube and tumor resection" was performed. Pathology reported thecoma with granulosa cell components. After consultation at the First Affiliated Hospital of China Medical University, a sex cord-stromal tumor was considered, and the tumor was regarded as benign. In August 2015, the patient developed chest pain, followed by abdominal pain and distension, mainly in the right upper abdomen. She visited the First Affiliated Hospital of China Medical University and underwent ultrasound-guided abdominal tumor biopsy. Postoperative pathology confirmed a granulosa cell–theca cell tumor in the left lower abdomen. After chemotherapy, the tumor progressed. In November 2016, she was readmitted and definitively diagnosed with a sex cord-stromal tumor. Four cycles of chemotherapy were administered, with the last cycle in March 2017. Since January 2019, abdominal distension and pain worsened. After further chemotherapy, severe chemotherapy-induced bone marrow suppression occurred, and she was readmitted to our hospital in June 2019 for treatment.

Analysis: In the 3rd edition (2003) WHO Classification of Female Genital Tumors, the histological classification of theca cell tumor was 8600/0; granulosa cell tumor was classified as a tumor of uncertain malignant potential, with borderline tumor components requiring more than 10% of the tumor to be classified as borderline, otherwise considered benign [3]. Therefore, the postoperative pathology report in 2011 considered the tumor benign, histological classification 8600/0, primary diagnosis code D27.x, pathological diagnosis named “adult granulosa cell tumor,” pathological diagnosis code M8600/0.

Ovarian sex cord-stromal tumors originate from sex cords and stromal tissue in primordial gonads, differentiating into male and female-specific cell types. Female sex cords differentiate into granulosa cell tumors (epithelial differentiation) and stromal differentiation forms theca cell tumors and fibromas; mixed tumors such as granulosa-theca cell tumor or fibrothecoma may also occur. Granulosa cell tumors account for 5% of ovarian malignancies, commonly associated with estrogen secretion, rarely with distant metastasis but possible local spread, thus mostly exhibiting low-grade malignancy [4]. Between 2015 and 2016, the patient’s disease progressed, and pathology confirmed sex cord-stromal tumor. According to the 4th and 5th edition (2014) WHO Classification of Female Genital Tumors, the codes should be 8620/3 and 8600/0, so the primary diagnosis code should be C56.x. The 4th edition renamed granulosa cell tumors as "adult granulosa cell tumor and juvenile granulosa cell tumor," and the 5th edition changed the pathological histological names back to "adult granulosa cell tumor and juvenile granulosa cell tumor."

The 2019 admission was for treatment of leukopenia and thrombocytopenia caused by chemotherapy-induced bone marrow suppression. According to the “Quality Specifications for Filling Inpatient Medical Record Front Pages (Interim)” (National Health Office Medical [2016] No. 24), Chapter 2 Filling Specifications Article 13, when hospitalization is for treatment of tumor complications or diseases other than tumors, the complication or disease should be selected as the primary diagnosis. Therefore, the primary diagnosis code should be D61.1.

(2) A search of discharged cases from January 1, 2019 to June 1, 2025 identified 537 cases with a primary diagnosis of benign ovarian tumor, including 28 cases of ovarian sex cord-stromal tumors: 10 theca fibromas, 5 ovarian fibromas, 1 sclerosing stromal tumor, 1 cellular fibroma, 1 steroid cell tumor, and 2 granulosa cell tumors; among these, 12 cases had incorrect pathological histological classification of granulosa cell tumor, fibroma, steroid cell tumor, or theca cell tumor; 1 case of cellular fibroma had an incorrect primary diagnosis code selection; 2 cases of granulosa cell tumor had incorrect primary diagnosis code selection.

① Patient: Female, 60 years old, presented with persistent lower abdominal pain for 3 days. Ultrasound at an external

hospital showed a pelvic cystic-solid mass. Further abdominal CT indicated a pelvic lesion closely related to the bilateral adnexal area, suspected cystadenocarcinoma. Under general anesthesia, bilateral salpingo-oophorectomy was performed. Intraoperative frozen section suggested a left adnexal sex cord-stromal tumor, most likely theca fibroma. Postoperative paraffin pathology and immunohistochemistry confirmed left ovarian fibroma with focal cellular fibroma. Immunohistochemistry results: WT1 negative, CD56 positive, inhibin positive, calretinin (CR) focal positive, SMA focal positive, ER negative, PR positive, Ki-67 1%. The physician diagnosed left ovarian fibroma; coder assigned diagnostic code D27.x, pathological diagnostic code M8440/0.

Analysis: According to the WHO 4th and 5th edition histological classifications of ovarian tumors, cellular fibroma is classified as a tumor of uncertain malignant potential. Therefore, the correct pathological diagnostic code should be M8810/1. Referring to ICD-10 Diseases and Related Health Problems Volume 1, the correct diagnostic code is D39.1.

② Patient: Female, 14 years old, presented with amenorrhea for over 3 months. Ultrasound revealed a 4.6×4.4 cm uniformly hyperechoic, regular-shaped, well-defined mass in the left ovary with detectable blood flow signals. Under general anesthesia, laparoscopic ovarian tumor resection was performed. Intraoperative frozen pathology suggested juvenile granulosa cell tumor, with luteinized theca cell tumor not excluded. Postoperative paraffin pathology and immunohistochemistry confirmed benign left ovarian steroid cell tumor. Physician diagnosed left ovarian steroid cell tumor; coder assigned diagnostic code D27.x, pathological diagnostic code D27.x.

Analysis: Ovarian steroid cell tumor (SCT) is a subtype of ovarian sex cord-stromal tumor, relatively rare, accounting for less than 0.1% of all ovarian tumors. Most are benign but have malignant potential, approximately 25%–43%. Prognosis is generally good, but malignant cases tend to recur and metastasize. Metastatic lesions usually occur within the abdominal cavity and rarely at distant sites. In 2013, WHO classified ovarian steroid cell tumors under the pure stromal tumor category of sex cord-stromal tumors [4]. The pathological diagnosis in this case is clearly benign ovarian steroid cell tumor, with histological classification code 8670/0 in both the 4th and 5th edition ovarian tumor classifications. Therefore, the pathological diagnostic code should be M8670/0.

(3) A search of discharged cases from January 1, 2019 to June 1, 2025 identified 56 cases with a primary diagnosis of ovarian borderline tumor, including 8 cases of ovarian sex cord-stromal tumors. Among the ovarian sex cord-stromal tumors, 3 cases of theca cell tumor had incorrect diagnosis and pathological diagnostic codes. The pathological diagnostic code was recorded as M86210/1, and the primary diagnosis code for theca cell tumor was D39.1.

Patient: Female, 57 years old, postmenopausal with irregular vaginal bleeding. Ultrasound showed a cystic mass below the left side of the abdomen near the umbilicus measuring approximately 7.8×8.3×7.3 cm, with clear borders. Inside the cyst, a hyperechoic mass about 2.4×1.5 cm was seen, with no obvious blood flow signal. Endometrial thickness was about 0.6 cm. Pelvic CT showed an irregular cystic-solid mass in the pelvic cavity measuring about 10.5×9.6 cm in largest cross-sectional area, with uneven density, septations, punctate calcifications, and cystic low-density areas; the boundary with pelvic tissues was unclear. After completing preoperative preparation, laparoscopic right adnexectomy was performed under combined intravenous and inhalation anesthesia. Intraoperative frozen section suggested a benign tumor. Postoperative paraffin pathology diagnosed right ovarian theca cell tumor. The coder assigned pathological diagnostic code M86210/1 and primary diagnosis code D39.1.

Analysis: Ovarian theca cell tumor is a rare benign functional ovarian tumor derived from sex cords and stromal tissue in the primordial ovary, composed of follicular cells of the ovarian sex cords and stromal derivatives. According to the WHO 4th and 5th edition histological classifications of ovarian tumors, the histological code for theca cell tumor is 8600/0, a benign tumor. Therefore, the correct pathological diagnostic code for this case should be M8600/0, and the primary diagnosis code should be D27.x.

5. Discussion

(1) The main differences in ovarian sex cord-stromal tumor classification between the two versions are as follows: First, granulosa cell tumors have different biological behaviors between adult type (AGCT) and juvenile type (JGCT). Second, Sertoli-Leydig cell tumors are divided into three different subtypes: DICER1 mutation type, FOXL2 mutation type, and DICER1/FOXL2 wild type. Additionally, microcystic stromal tumors contain CTNNB1 or the less common APC mutations and may represent an extra-colonic manifestation of familial adenomatous polyposis. Molecular testing is helpful for precise classification when diagnosis is difficult. Third, gonadoblastoma was reintroduced; it is a sex cord-stromal tumor with both male and female differentiation and is relatively rare. Its classification was removed in the 4th edition but remained controversial, and was reinstated under mixed sex cord-stromal tumors in the 5th edition [2].

(2) The 5th edition classification emphasizes the integration of pathological diagnosis with clinical treatment, providing

effective prognostic information for precise patient management. Coders mastering the latest WHO ovarian sex cord-stromal tumor histological classification, reviewing pathology reports, and communicating with clinicians can improve the accuracy of data reported by medical institutions to the national tumor registry. This contributes to advancing research, diagnostic and treatment quality of ovarian sex cord-stromal tumors nationwide, reducing incidence, improving cure rates, alleviating patient financial burden, and saving government expenditure, thus having significant guiding value.

Acknowledgments

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