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A Rare Case Series Of Sex Cord Stromal Cell Tumour

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ABSTRACT

Sex cord-stromal tumors (SCSTs) are a heterogeneous group of rare neoplasms arising from the sex cords and stromal components of the gonads, encompassing both the ovaries and testes. They account for approximately 5-8% of malignant potential is 3-4% of ovarian neoplasms and a smaller fraction of testicular tumors. These tumors are unique in that they often secrete sex steroids such as estrogen, progesterone, or androgens, contributing to diverse endocrine manifestations, including menstrual irregularities, virilization, or feminization depending on the hormone produced. Common subtypes include granulosa cell tumors, Sertoli-Leydig cell tumors, fibromas, and thecomas, each with varying clinical behavior and prognosis. While many SCSTs tend to follow a benign course, certain types exhibit malignant potential, necessitating long-term surveillance and intervention. Diagnosis is largely reliant on histopathological evaluation supported by immunohistochemistry, which helps differentiate them from other gonadal tumors. Imaging modalities such as ultrasound, CT, or MRI assist in initial localization and characterization but are not definitive. Treatment primarily involves surgical excision, often followed by adjuvant therapies like chemotherapy or radiotherapy, depending on factors such as histological subtype, tumor grade, and stage at presentation. Fertility-sparing surgery may be considered in younger patients with early-stage disease. Prognosis is generally favorable for benign tumors, but malignant variants may recur and require close follow-up. Advances in molecular genetics and biomarker discovery are contributing to improved diagnostic accuracy and individualized management strategies. Understanding the clinical, hormonal, and pathological spectrum of sex cord-stromal tumors is essential for timely diagnosis and optimal patient outcomes.

Keywords: *Sex cord-stromal tumors, steroid cell tumours, Case series*

1. INTRODUCTION

Sex cord-stromal tumours are a rare and diverse group of neoplasms originating from the sex cords (precursors of Sertoli and granulosa cells) and stromal tissue of the gonads (ovaries or testes). They account for approximately 5-8% of malignant potential is 3-4% of ovarian neoplasms and a smaller fraction of testicular tumors. They can be hormonally active, producing oestrogen, progesterone, or androgens, leading to endocrine manifestations. The most common subtypes include granulosa cell tumors, Sertoli-Leydig cell tumors, and thecomas.

Ovarian steroid cell tumours are a rare subtype of sex cord-stromal tumours, representing approximately 0.1% of all ovarian neoplasms. These tumours are further classified into three categories: stromal luteomas, Leydig cell tumours, and steroid cell tumours not otherwise specified (NOS), with the latter being the most common. Despite their rarity, these tumours hold clinical significance due to their ability to secrete steroids, particularly androgens, which can result in symptoms of hyperandrogenism such as hirsutism, virilization, acne, oligomenorrhea or amenorrhea, deepening of voice, and clitoromegaly. Occasionally, they may also present with estrogenic or corticosteroid excess, leading to menstrual irregularities or Cushingoid features, respectively[1].

The global burden of ovarian sex cord-stromal tumours is relatively low compared to epithelial ovarian cancers. However, their potential for hormonal activity, local invasion, and occasional malignancy makes early recognition and management

CASE 1

The case report details a clinical scenario involving Mrs. X, a 46-year-old multiparous woman (P3L3) with a history of tubal ligation performed 26 years ago at Cheyyar Government Hospital. The patient presented with intermittent, localized, pricking-type abdominal pain for one week, along with a single episode of painless hematuria. She denied any history of burning micturition, fever, or chills. Menstrual history revealed that she had attained menopause 4 years ago. Her past medical history was unremarkable, with no history of diabetes, hypertension, epilepsy, thyroid disorders, or other chronic illnesses.

On general examination, Mrs. X was afebrile, with no pallor, icterus, cyanosis, clubbing, pedal edema, or lymphadenopathy. Systemic examination was within normal limits: S1 and S2 heart sounds were normal, respiratory system showed normal vesicular breath sounds, and there were no focal neurological deficits on CNS examination. On abdominal palpation, a firm, tender, mobile mass approximately 5x6 cm was identified in the hypogastric region extending into the right iliac fossa. Per speculum examination revealed a hypertrophied cervix with visible erosion, and on per-vaginal examination, the uterus was anteverted and enlarged to 14-16 weeks size. The previously noted abdominal mass was palpable per vaginum, mobile and non-tender, and it did not move with the cervix.

Intra-operatively, dense adhesions were found between the anterior peritoneum, bowel, and bladder. During dissection, a bladder rent was identified with the Foley's catheter bulb visible, prompting the involvement of a urologist and general surgeon, who proceeded with a total abdominal hysterectomy. A subserosal fibroid (6x5 cm) was found on the right side of the uterus, densely adherent anteriorly to the abdominal wall and posteriorly to the bowel. Following excision, the specimen was sent for histopathological examination (HPE). The bladder injury was repaired, and both a suprapubic catheter (SPC) and an intraperitoneal drain were placed.

Gross pathological examination revealed a specimen from total abdominal hysterectomy with bilateral salpingo-oophorectomy, including the right adnexal mass. The uterus, already sectioned, measured 11x7x5 cm, and there was no fundal pedunculated fibroid noted. The right ovarian mass measured 6x5x3.5 cm, with a grey-black external surface. The cut surface displayed a grey-yellow to orange solid mass with grey-black speckling, and the residual right ovary measured 3x1.7x1 cm.

Microscopically, the ovarian mass revealed a benign neoplasm arranged in a diffuse pattern composed of a dual cell population:

1. Polygonal cells with vacuolated cytoplasm and centrally located nuclei.
2. Round cells with moderately eosinophilic cytoplasm and prominent nucleoli.

The background showed prominent vascular congestion and areas of hemorrhage. Importantly, there was no evidence of nuclear atypia, mitotic activity, or necrosis. The residual ovary showed features of stromal hyperthecosis.

The final histopathological diagnosis was Steroid Cell Tumor, Not Otherwise Specified (NOS) of the Right Ovary, a rare subtype of sex cord-stromal tumors of the ovary. These tumors are known for their endocrine activity, although this case presented without overt hormonal symptoms.

Fig 1:Grey yellow to orange solid mass.

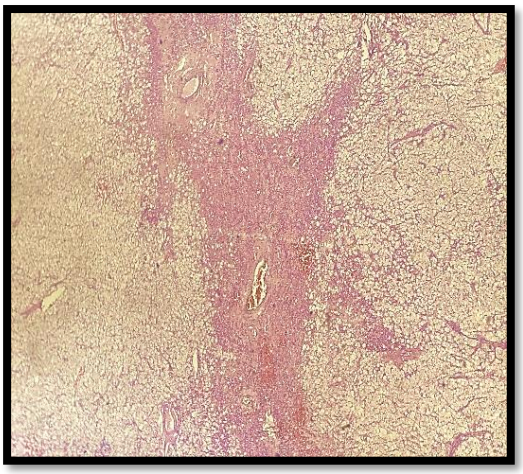


Fig 2: Dual population of cells – polygonal and round cells.

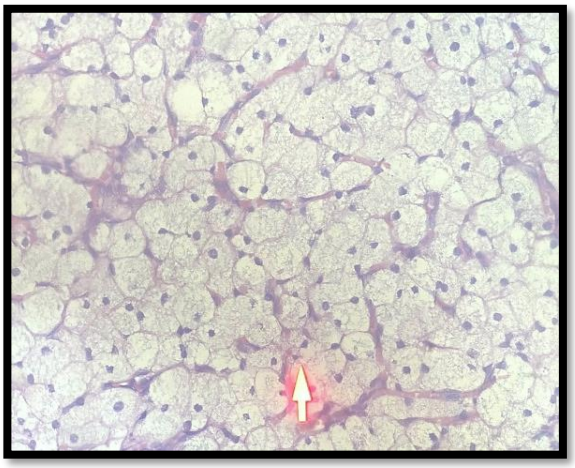


Fig 3: Polygonal cells with vacuolated cytoplasm and centrally placed nuclei.

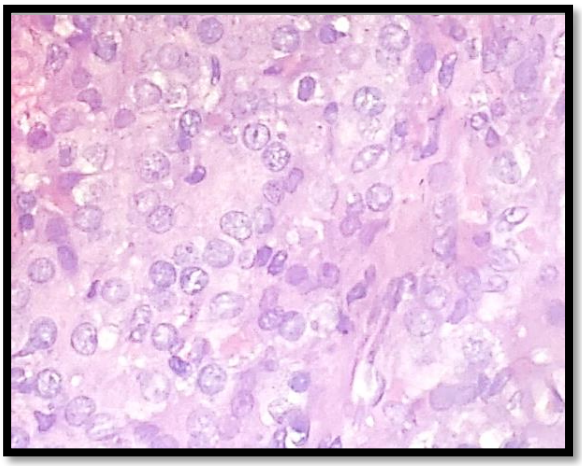


Fig 4: Round cells with moderately eosinophilic cytoplasm and prominent nucleoli.

CASE 2

The patient, a 20-year-old unmarried woman, of socio-economic class 3, presented with frequent menstrual cycles occurring every 15 days, with normal flow, lasting 4–5 days, for the past five months. She reported a bloating sensation after meals for one week but denied any pain, clots, appetite loss, weight loss, or other systemic symptoms. Her menstrual history revealed menarche at 13 years, with previously regular cycles. Current cycles were irregular with two episodes per month, but without clots or dysmenorrhea.

She had no significant past medical, surgical, or family history of gynecological malignancy, with normal bowel and bladder habits, and a normal sleep-wake cycle. On general examination, she was moderately built, with mild pallor, no pedal edema, and stable vital signs (PR 84 bpm, BP 110/70 mmHg, Temp 98.4°F, RR 14/min). Abdominal examination revealed a non-tender, cystic-to-firm pelvic mass, about 18–20 weeks in size, occupying the hypogastric and right iliac region, with mobility and smooth surface, suggesting a pelvic origin. On percussion, there was a dull note over the mass area and bowel sounds were audible. Per rectal examination was unremarkable.

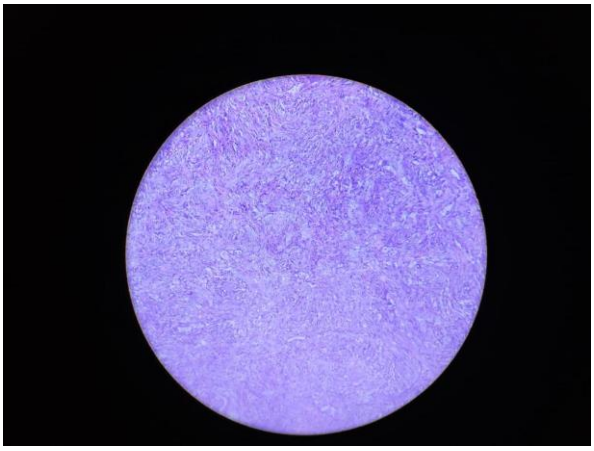
Ultrasound (22/11/23) revealed a large, multiloculated cystic lesion in the right abdominopelvic area, measuring 12.7 x 12.5 x 7.5 cm, with echogenic contents and peripheral vascularized stroma, likely representing mucinous cystadenoma. The right ovary was not visualized separately, and minimal free fluid was noted in the peritoneal cavity and pouch of Douglas. Based on these findings, a diagnosis of right complex ovarian cyst was made, and the patient was planned for staging laparotomy with right salpingo-oophorectomy under general anesthesia, which was conducted on 28/11/2023.

Intraoperative findings included about 60 ml of peritoneal fluid (sent for cytology), normal left ovary and fallopian tube, and a 12x10 cm cyst arising from the right ovary, which had undergone torsion twice. The cyst was adherent to the broad ligament, and 250 ml of straw-colored fluid was aspirated from it for cytology. A right salpingo-oophorectomy was performed, and an omentectomy was done for histopathological examination.

Histopathological examination of the excised ovarian mass (12x9x3.8 cm) revealed solid and cystic areas without papillary excrescences, and microscopy showed a benign neoplasm arranged in pseudolobules, with spindle-shaped and round cells, vacuolated eosinophilic cytoplasm, round to oval nuclei, some with prominent nucleoli, and dilated thin-walled blood vessels. The stroma was collagenous and hyalinized, with areas of myxoid degeneration, and abundant thin-walled capillaries. The right fallopian tube showed normal histology, and the omentum displayed fibrofatty tissue with congested vessels. Cytology of peritoneal and cystic fluids showed proteinaceous content with RBCs and few inflammatory cells, ruling out malignancy. Final diagnosis was Sclerosing Stromal Tumor (SST) of the right ovary, a rare benign ovarian sex cord-stromal tumor.

The discussion emphasizes that SST is extremely rare, with fewer than 200 cases documented globally. It typically affects women in their 20s to 30s, presenting with menstrual irregularities and pelvic pain, and is usually unilateral and hormonally inactive, though rarely it may cause estrogenic or androgenic symptoms. SSTs are often misdiagnosed preoperatively as malignancies due to solid-cystic appearance and vascularity. Diagnosis is confirmed via histopathology and immunohistochemistry, with positivity for inhibin, calretinin, smooth muscle actin (SMA), and vimentin. The etiology remains unclear but may involve pluripotent stromal cells of the ovarian cortex and possibly evolve from preexisting fibromas. Some cases show elevated CA-125 and ascites, with rare association with Meigs syndrome or ovarian torsion.

Fig 5: Intra operative image



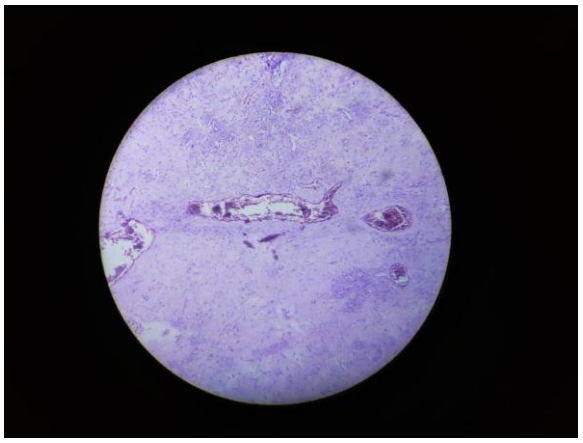


Fig 6 & 7: 1.Pseudolobules 2.Spindle Shaped Round Cells 3.Vaculated Eosinphilic Cytoplasm 4.Dilated Thin Walled Blood Vessels 5.Collagenous Stroma 6.Hypocellular & Hypercellular Areas

CASE 3

The patient is a 22-year-old unmarried woman, currently a first-year BA student from Nemmili, categorized under socio-economic class 4 according to the Modified Kuppuswamy Scale. She presented with a gradually enlarging abdominal mass over 2 months without associated abdominal pain, weight loss, gastrointestinal or urinary symptoms, hormonal disturbances, or systemic features suggestive of infection or malignancy.

Her menstrual history revealed regular cycles since menarche at age 14, with no dysmenorrhea or abnormal bleeding. She had no significant medical history (no diabetes, hypertension, thyroid disorders, epilepsy, or malignancy) and no family history of ovarian or colorectal cancers. She had a past surgical history of a right-sided modified radical mastoidectomy for CSOM 4 years ago.

On general examination, she was thin built with a BMI of 18.1, but was otherwise stable and without lymphadenopathy or systemic abnormalities. Per abdominal examination revealed a firm to hard, well-defined suprapubic mass, approximately the size of a 26-week gravid uterus, occupying both iliac fossae and infraumbilical region. The mass was smooth, mobile, non-tender, with no signs of ascites or organomegaly. Per vaginal exam was not performed due to intact hymen, and per rectal examination was normal.

Imaging studies revealed significant findings. CT scan showed bilateral adnexal cystic lesions suggestive of mucinous cystadenoma, with elevated tumor markers (CA-125: 147.6 U/ml and CA 19.9: 52.62 U/ml). MRI abdomen and pelvis demonstrated a large, solid-cystic lesion in the right adnexa, crossing the midline, compressing and displacing adjacent structures, and associated with moderate ascites and bilateral mild hydronephrosis.

The patient underwent a fertility-sparing staging laparotomy with right salpingo-oophorectomy on 18/11/2024 under general anesthesia. Intraoperative findings included a 15x15 cm right ovarian cyst, normal left ovary and tube, and normal uterus, pouch of Douglas, omentum, and other organs. A prophylactic appendectomy was also performed, and 50 ml of peritoneal fluid was sent for cytology.

Gross pathology revealed a solid-cystic ovarian mass measuring 26x16x12 cm, with the solid portion measuring 17x16x12 cm. The mass showed papillary excrescences and contained serous fluid. Another smaller cyst, 8.5x7x2.5 cm, was also attached.

Microscopically, the tumor showed both hypocellular and hypercellular areas. Hypocellular zones displayed spindle cells with vacuolated cytoplasm, while hypercellular regions had clear to eosinophilic cytoplasm and spindle nuclei. No nuclear atypia, necrosis, or increased mitotic activity was observed, but multiple papillary projections were present within the cyst lumen.

The histopathological diagnosis confirmed a benign collision tumor consisting of Sclerosing Stromal Tumor along with a Papillary Serous Cystadenoma in the right ovary. The right fallopian tube and pelvic lymph nodes were tumor-free. The peritoneum exhibited fibrocollagenous tissue with hemorrhage, the omentum showed congested vessels, and the appendix was normal with a faecolith inside the lumen.

The discussion highlighted that Sclerosing Stromal Tumor is a rare benign ovarian sex cord-stromal tumor, typically seen in young women aged 20-30 years, with fewer than 200 cases reported. It presents with menstrual irregularities or pelvic pain but is often hormonally inactive. Due to its solid-cystic nature and vascularity, it can mimic malignancies on imaging, making histopathology and immunohistochemistry (positive for inhibin, calretinin, SMA, and vimentin) crucial for diagnosis. SSTs may arise from pluripotent stromal cells and can sometimes coexist with ascites, elevated CA-125, and rare cases of Meigs syndrome or ovarian torsion.

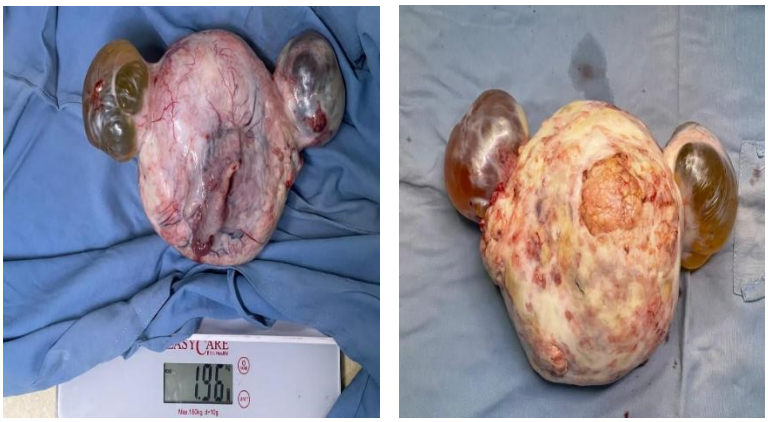


Fig 8 & 9: Intra operative image

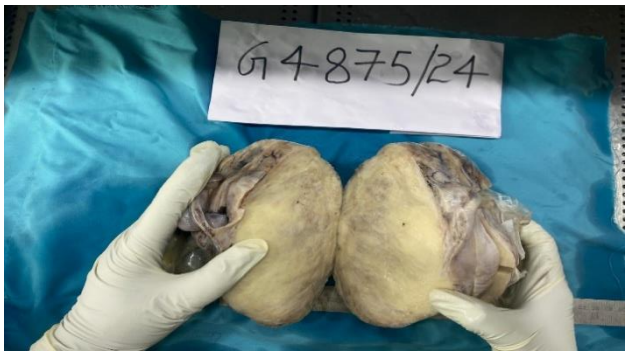


Fig 10: Macroscopic Image

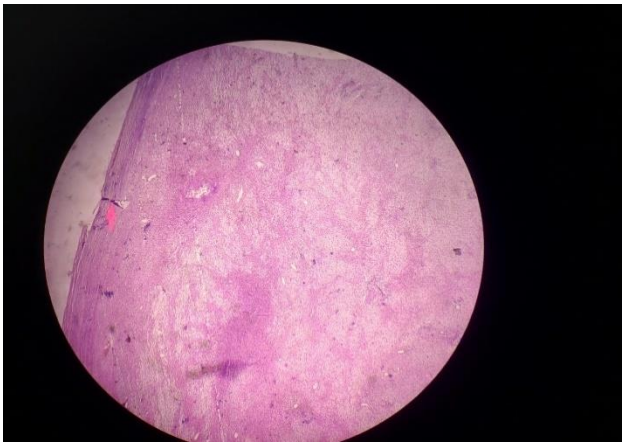


Fig 11: Microscopic features
Hypocellular Areas In Low Power

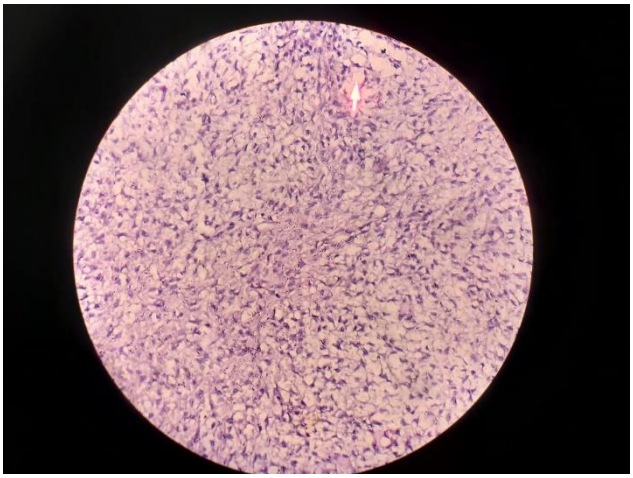


Fig 12: Microscopic features
1. Hypocellular Areas In High Power Showing Spindle Cells With Vacuolated Cytoplasm And Spindled Nuclei

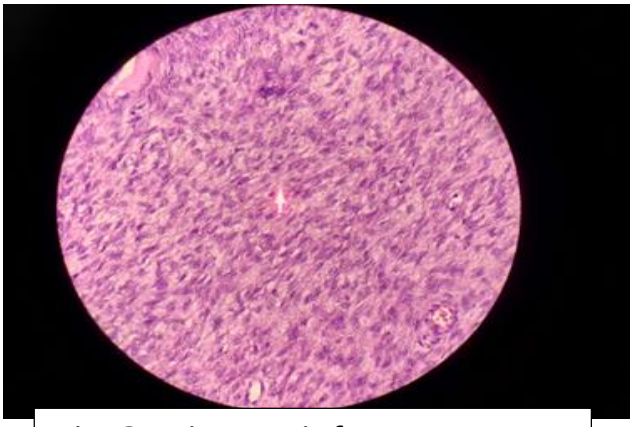


Fig 13: Microscopic features Hypercellular area with clear to eosinophilic cytoplasm and spindled nuclei.no evidence of nuclear atypia/increased mitotic activity.mutiple papillary projections into the lumen.

2. DISCUSSION

Steroid cell tumours of the ovary are uncommon neoplasms classified under sex cord-stromal tumours, comprising less than 0.1% of all ovarian tumours (1). The most frequently encountered subtype is the Steroid Cell Tumour, Not Otherwise Specified (NOS), representing about 56% of all steroid cell tumours (2).

These tumours are clinically significant due to their hormone-secreting potential and the possibility of malignant transformation in a subset of cases. They can occur in a wide age range but are typically diagnosed in women between 30 and 60 years of age (3). The present case is consistent with this demographic, as the patient is a 46-year-old postmenopausal woman.

Clinical Presentation and Hormonal Activity

The majority of steroid cell tumours NOS are functional and produce androgens, leading to hyperandrogenic symptoms. Hayes and Scully, in their clinicopathologic study of 63 cases, reported that 56-77% of patients presented with virilization symptoms such as hirsutism, acne, oligomenorrhea, amenorrhea, deepening of voice, and clitoromegaly (1). However, our patient did not report significant androgenic symptoms, which is consistent with reports by Roth and Tsubura (2), who identified that around 25-30% of these tumours are non-functional.

In some cases, excess secretion of estradiol (in 6-23% of patients) or cortisol may lead to abnormal uterine bleeding or Cushingoid features (4). Irshad et al. (5) also presented a case where the tumour secreted both testosterone and estradiol, leading to mixed clinical features. The absence of overt virilization or estrogenic symptoms in this patient demonstrates the tumour's hormonal behavior heterogeneity.

Radiological and Intraoperative Findings

Radiologically, steroid cell tumours are generally solid, well-circumscribed masses. Outwater et al. (6) described these tumours as hypoechoic on ultrasound and hyperintense on T1-weighted MRI due to their lipid-rich cytoplasm. Intraoperatively, our patient had a right adnexal mass (6x5 cm) with dense adhesions involving the bowel and bladder, features commonly seen in larger or older tumours.

Additionally, Malpica et al. (7) suggested that masses larger than 7 cm are more likely to be malignant. Our patient's tumour measured 6x5 cm, and histology revealed no features of malignancy, which is in line with benign cases reported by Roth and Tsubura (2).

Histopathological Characteristics

Histologically, steroid cell tumours NOS show a dual population of polygonal and round cells arranged in sheets, with abundant eosinophilic or vacuolated cytoplasm (8). Our case showed these classical features, along with the absence of

nuclear atypia, mitoses, or necrosis, all of which are markers suggestive of benign behavior.

According to Young and Scully (9), criteria for malignancy include necrosis, high mitotic index (>2 mitoses/10 HPF), marked nuclear atypia, and tumour size >7 cm. The absence of these criteria in our case further confirms its benign nature.

Differential Diagnosis

The histologic differential diagnosis includes Leydig cell tumour, which typically contains Reinke crystals, absent in steroid cell tumour NOS. Stromal luteoma is another differential, often smaller and located within the ovarian stroma without significant androgen production (9,10). Immunohistochemically, most steroid cell tumours are positive for inhibin, calretinin, and Melan-A, which are useful markers for diagnosis (11).

Comparison with Other Studies

In a review by Gheorghisan-Galateanu et al. (12), the authors emphasized that most benign steroid cell tumours NOS are managed with surgical excision alone, especially when fertility preservation is not a concern. Similar findings were reported by Obermair et al. (13) and Huang et al. (14), where total abdominal hysterectomy with bilateral salpingo-oophorectomy was curative in postmenopausal women.

However, Brown et al. (15) discussed a rare case where recurrence occurred in a malignant variant after conservative surgery, suggesting the importance of long-term surveillance.

Management and Prognosis

Given the absence of malignancy indicators, surgery remains the gold standard in such cases. Fertility-sparing surgery may be considered in younger women with unilateral disease and benign histology. Long-term prognosis is excellent for benign steroid cell tumours, as highlighted by Roth and Tsubura (2) and Young and Scully (9).

These tumours may also produce other hormones such as androstenedione, dehydroepiandrosterone sulfate (DHEAS), and cortisol, which could present as virilization or Cushingoid features depending on the hormone secreted.

3. CONCLUSION

The management of ovarian steroid cell tumours, NOS, should be individualized based on several factors, including the histopathological characteristics of the tumour, comprehensive surgical staging findings, and the patient's reproductive desires. In younger women who wish to preserve fertility and where the tumour is confined to one ovary with benign features, unilateral salpingo-oophorectomy is considered the optimal approach.

In contrast, for postmenopausal women or when malignancy cannot be excluded, a more definitive procedure such as total abdominal hysterectomy with bilateral salpingo-oophorectomy is preferred. In the present case, given the patient's postmenopausal status and the intraoperative findings of a unilateral mass without evidence of metastasis or malignancy, a total abdominal hysterectomy with bilateral salpingo-oophorectomy was performed. This approach not only ensures complete tumour removal but also eliminates the risk of recurrence and addresses the patient’s long-term oncologic safety.

Hence, a multidisciplinary decision-making strategy, taking into account the tumour's biological behavior, patient’s age, hormonal activity, and fertility considerations, is crucial for optimal management and prognosis.

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