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Hidden Behind PCOS: An Androgen-Secreting Ovarian Steroid Cell Tumor in a Young Woman

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ABSTRACT

Ovarian steroid cell tumors are rare sex-cord stromal tumors, representing less than 0.1% of all ovarian tumors. They frequently produce steroid hormones and present with endocrine manifestations such as hirsutism, virilization, and menstrual irregularities. We report a case of a 21-year-old unmarried woman presenting with irregular menses and clinical hyper-androgenism. Imaging revealed a right ovarian mass, and serum testosterone was markedly elevated. Laparoscopic cystectomy was performed, and histopathological evaluation confirmed a steroid cell tumor, not otherwise specified (SCT-NOS). The patient recovered uneventfully and was discharged on postoperative day three. This case highlights the importance of considering androgen-secreting ovarian tumors in young women presenting with virilization and elevated testosterone levels predominantly in cases of PCOS.

Keywords: Steroid cell tumor, ovarian tumor, Hyper-androgenism, Virilization, Sex cord stromal tumor

INTRODUCTION

Steroid cell tumors of the ovary are rare neoplasms belonging to the group of sex-cord stromal tumors. Steroid cell tumors are rare ovarian tumors derived from steroid hormone-secreting stromal cells. They account for approximately 0.1% of ovarian tumors.[1,2] In general, testosterone secretion leads to virilization or hirsutism. Clinical manifestations include hirsutism, acne, deepening of the voice, amenorrhea or oligo-menorrhea, while estrogen secretion causes bloating and fibrocystic breast lumps. Only 10%–15% of patients have no clinical signs or symptoms of increased hormone levels.[2] Morphologically, steroid cell tumor-NOS presents as a solid, well-circumscribed yellowish mass in about 89% of cases. Rarely, in about 1.6% of cases, these tumors are completely cystic.[1] They can produce steroid hormones, including androgens, estrogens, and corticosteroids. The subtype steroid cell tumor not otherwise specified (NOS) accounts for the majority of these tumors. Due to their rarity and variable imaging appearance, diagnosis may be challenging before histopathological confirmation.

CASE PRESENTATION

A 21-year-old unmarried woman presented with irregular menstrual cycles for the past one and a half years, followed by two months of amenorrhea, along with progressive hirsutism, weight gain of approximately 15 kg and deepening of the voice over last one year. Patient was known case of PCOS, with menstrual irregularity for which she was on oral contraceptives for past one year, but not relieved. The patient had a low-pitched voice and a body mass index of 33.9 kg/m² (obese class I). Hirsutism was present, with a Ferriman-Gallwey score of 15. There were no features suggestive of Cushing's syndrome, such as moon facies or buffalo hump. There was no history of contraception use or exogenous hormone intake. Her past medical and surgical history was unremarkable. No similar findings were noted in her family. Breast examination revealed symmetrical breasts without masses or nipple discharge. The abdomen was soft and non-tender. Ultrasonography demonstrated a normal-sized ante-verted uterus and a right ovarian echogenic lesion measuring approximately 3.9x3.5x3cm. MRI of the pelvis showed an enlarged right ovary containing a heterogeneous soft tissue lesion measuring approximately 4 × 3.8 cm with cystic septations.

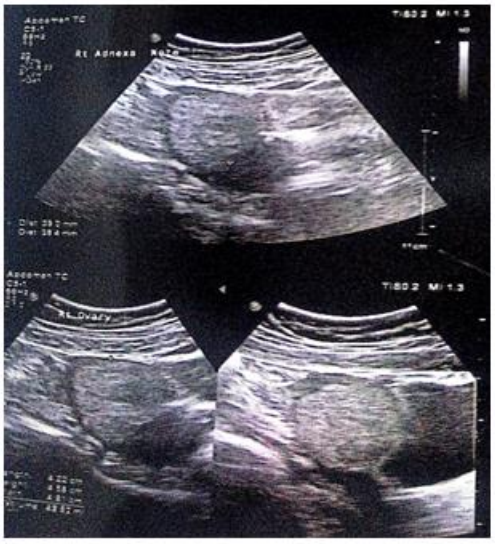

Fig.1: USG image showing homogenous echogenic mass Of 3.9x3.5x3 cm, no ascitis

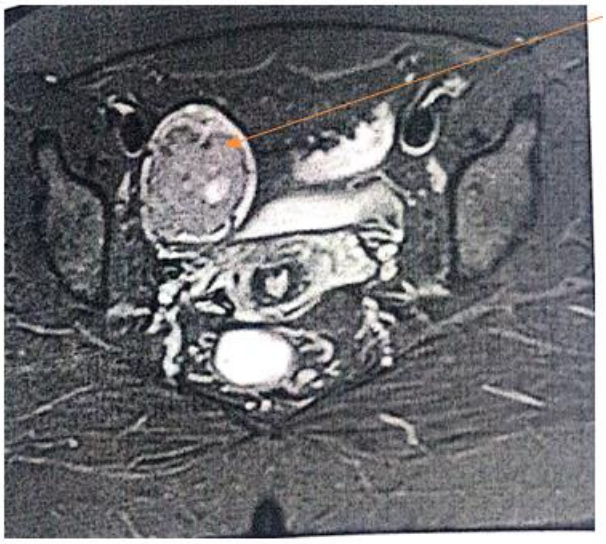

Fig.2: MRI pelvis showing Heterogenous soft tissue lesion~ 4x3.8 cm

Her laboratory findings showed a normal hemogram, electrolytes, liver enzymes, and kidney function test levels. Her beta hCG level was negative, and Ca-125, Ca 19.9, CEA, AFP, cortisol, prolactin, and TSH were within normal limits. Serum testosterone level was markedly elevated at 393 ng/dl, while serum DHEAS was 233 µg/dl. Other tumor markers were within normal limits. The patient underwent laparoscopic right ovarian cystectomy. Intraoperatively, there was no ascites or adhesions. Grossly, the right ovary was cream-tan and glistening, with intact mucosa, measuring 5.2 × 4.5 × 3.5 cm.

The uterus, bilateral fallopian tubes, and left ovary appeared normal. The other organs were also normal. A right-sided cystectomy was performed, the specimen was retrieved in an endobag, and sent for biopsy.

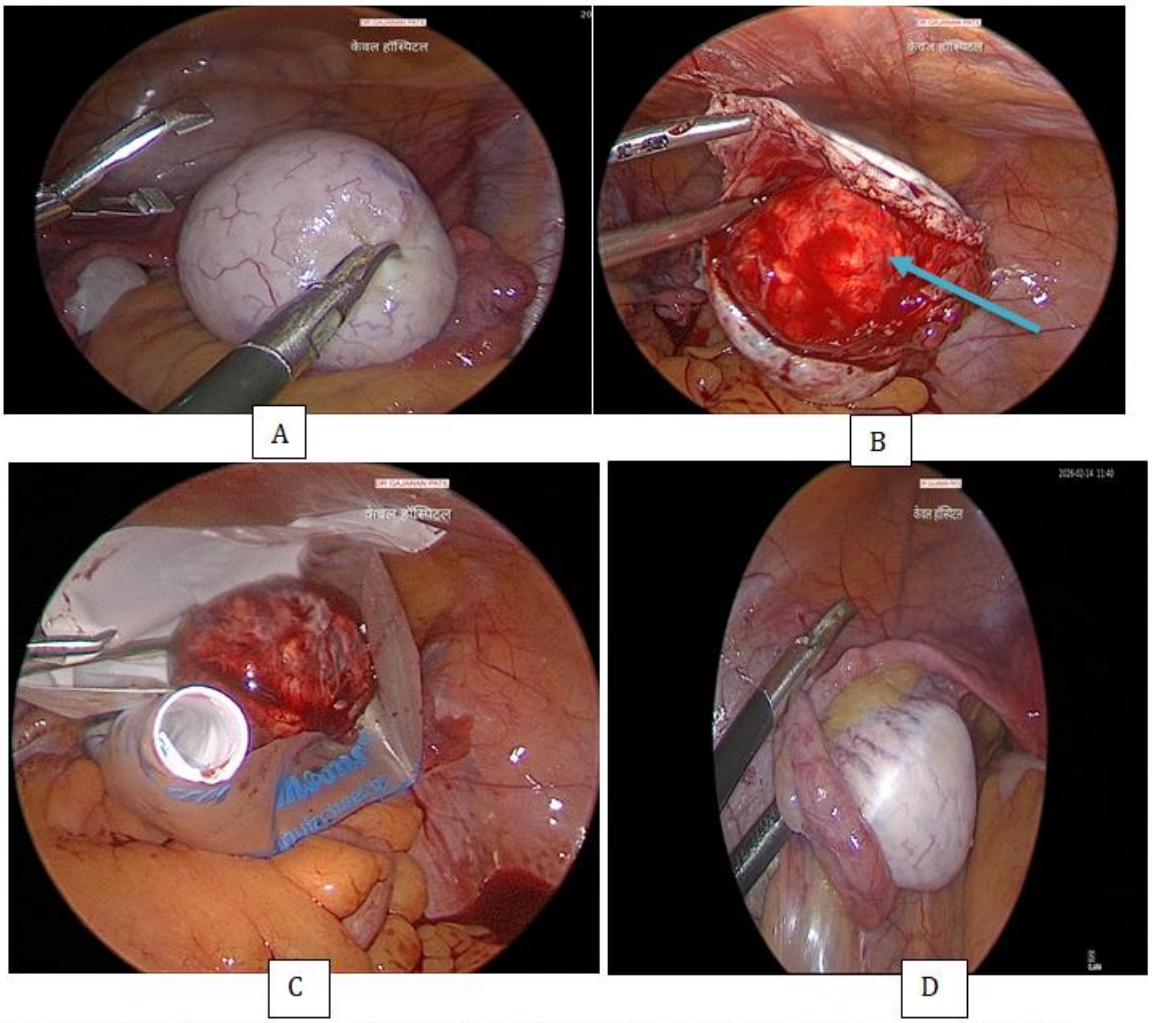

Fig.3: {A} Right ovarian mass, {B}showing right sided Lap. cystectomy, {C}specimen retrieval in endobag {D} shows normal Lt. fallopian tube and bulky Left ovary(PCOS)

Upon sectioning, a yellow, soft to firm, lobulated mass was found occupying the entire ovary and abutting the serosal surface, measuring 4.8 cm × 4.0 cm × 3.5 cm. The specimen was sent for frozen section, which indicated a “steroid cell tumor, NOS.” Microscopy showed solid aggregates of cells with occasional nests. Tumor cells were polygonal, with granular eosinophilic cytoplasm and clear intracytoplasmic vacuoles. No Reinke crystals, nuclear atypia, hemorrhage, or mitosis was detected.

The patient was discharged on postoperative day 3. The postoperative period was uneventful. For follow-up, the patient was called after 2 weeks with repeat values of serum testosterone and DHEAS, which were found to be normal. The patient was relieved of her previous complaints.

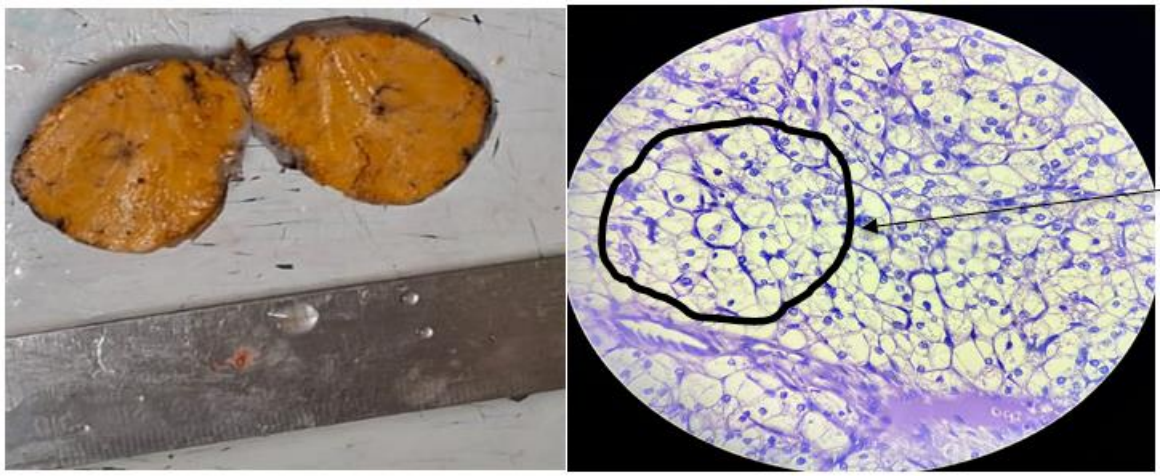

Figure 4: Cut section shows yellow solid mass**.** Histopathology of the right ovary: On microscopy, solid aggregates of cells with occasional nests were seen. Tumour cells were polygonal with cytoplasm that is granular and eosinophilic with clear intracytoplasmic vacuoles (shown by marked area). No Reinke crystals, no nuclear atypia, hemorrhage and mitosis were seen

DISCUSSION

The term "steroid cell tumors" was first introduced by Hayes and Scully in 1987 to replace the terms "lipid cell tumor" and "lipoid cell tumor," which previously referred to "morphologically similar ovarian neoplasms of diverse cellular origin," composed exclusively of cells resembling typical steroid hormone-secreting cells.[3,7] These tumors are associated with characteristic virilizing clinical syndromes. All contain steroid hormone-secreting cells such as lutein cells, Leydig cells, and adrenal cortical cells. Therefore, Hayes and Scully proposed using the term "steroid cell tumors" for this unique group of ovarian sex cord stromal neoplasms. Their proposal further sub-classified steroid cell tumors as (1) stromal luteoma, arising from the ovarian stroma; (2) Leydig cell tumor –hilus and non-hilus type, arising from Leydig cells; and (3) not otherwise specified, if the lineage of the tumor is unknown.[1,4]

The majority produce androgens, leading to clinical manifestations of hyper-androgenism and, menstrual irregularities, insulin resistance. They often mimic with similar features as PCOS. However, Elevated serum testosterone levels greater than 200 ng/dl strongly suggest the presence of an androgen-secreting tumor. Imaging typically reveals a unilateral solid ovarian mass, although findings may mimic a dermoid cyst or other ovarian neoplasms, as in our case. Histopathological examination remains the gold standard for diagnosis. Most steroid cell tumors not otherwise specified are unilateral, benign, and well-circumscribed. The size varies from 1.2 to 45 cm.[3] Grossly, these tumors are commonly solid; however, a combination of solid and cystic forms or a predominantly cystic form may also be seen. The color of the cut surface may range from yellow to orange, red, or brown depending on the lipid content. Areas of hemorrhage and necrosis may also be seen.[3,5] The tumor in our case was completely solid with no cystic areas. The cut surface was typically yellow and lobulated. Surgical excision is the treatment of choice. In young patients, fertility-preserving surgery is recommended when

malignancy is not suspected. In many cases, the diagnosis is made postoperatively upon finding a tumor in the ovary incidentally, where these tumors do not show any symptoms of virilization.[6] Small tumors may be missed in most of PCOS cases by imaging. Therefore, patient with presumed diagnosis of PCOS not responding to standard treatment, or if androgen levels continue to rise sharply testing for an androgen-secreting tumor is recommended.

CONCLUSION

Steroid cell tumors represent less than 0.1% of ovarian tumors and frequently cause virilization in patients of all ages, with an average age of 43 years. It should also consider in young women with PCOS, not respondent to treatment, or rapid virilizing symptoms. Markedly elevated testosterone suggests an androgen-secreting tumor. Histopathology is essential for definitive diagnosis. Fertility-preserving surgery is appropriate in young patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Authorship contributions

Gajanan G. Patil - Involved in conceptualization, resources, reviewing, editing, visualization, funding, supervision.

Shalini Mishra - Involved in conceptualization, reviewing, writing draft.

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Conflicts of interest

There are no conflicts of interest.

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