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Case Report: Steroid Cell Ovarian Neoplasm in a Patient with Polycystic Ovarian Syndrome

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Abstract

Introduction:

Polycystic ovarian syndrome (PCOS) is the predominant cause of hyperandrogenism in women of reproductive age. Ovarian steroid cell tumors are rare, accounting for less than 0.1% of all ovarian tumors, with approximately half of these classified as not otherwise specified (NOS). These tumors frequently present with hyperandrogenic symptoms and elevated serum androgen levels, and generally follow a benign course post-surgery.

Case Description:

We report the case of a 31-year-old female managed as PCOS, who presented with amenorrhea, hirsutism, and virilization. Initially managed as having a broad ligament fibroid, her condition was re-evaluated due to worsening symptoms. Laboratory tests showed elevated testosterone (8.6ng/mL) and high 17-OH progesterone (12.12 ng/mL), and imaging revealed a right-sided ovarian mass. Surgical intervention revealed a highly vascular right ovarian tumor, later confirmed as a steroid cell tumor (NOS) through histopathology. Postoperative recovery was notable for the resolution of hyperandrogenic symptoms and the return of regular menstrual cycles. The patient conceived within six months post-surgery and is currently 20 weeks pregnant without complications.

Conclusion:

This case underscores the importance of considering tumorous causes of hyperandrogenism in women with PCOS, especially when there is a rapid onset of symptoms and elevated testosterone levels. Prompt clinical suspicion and early identification are crucial to avoid delays in appropriate treatment.

Keywords: steroid cell tumour, polycystic ovary, hyperandrogenism

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Introduction

Polycystic ovarian syndrome (PCOS) is the commonest cause for hyperandrogenism in women at reproductive age. Other causes include congenital adrenal hyperplasia, Cushing's syndrome, androgen-secreting ovarian and adrenal tumors, and iatrogenic causes. Ovarian steroid cell tumors are very rare. They make up less than 0.1% of all ovarian tumors [1]. Approximately half of all steroid cell tumors are not otherwise specified (NOS). In most cases, steroid cell tumors (NOS) present with hyperandrogenic symptoms and increased serum androgen levels. After surgery, hyperandrogenic symptoms normalize as these tumors follow a benign course most of the time [2]. Herein, we present a case of a 31-year-old female who was initially managed as PCOS found to have NOS subtype of steroid cell tumor, who presented with amenorrhea, hirsutism and virilization.

Case Presentation

This is a case of a 31-year-old mother of one child who was previously investigated for hirsutism, oligomenorrhea, and subfertility almost three years ago. She underwent imaging study with ultrasonography (USS) and computed tomography (CT) of the abdomen and pelvis. The results revealed polycystic ovaries and a solid mass lesion measuring 5.3 x 5.8 x 4.2 cm within the right-side pelvic cavity. Unfortunately, this was diagnosed as broad ligament fibroid. The diagnosis of PCOS was made based upon Rotterdam criteria (clinical or biochemical hyperandrogenism, evidence of oligo-anovulation, polycystic appearing-ovarian morphology on ultrasound), and managed as PCOS due to its common occurrence.

Recently, she sought medical advice when her symptoms intensified with escalating hirsutism, muscle mass, and deepening of the voice.

Physical examination was significant for overweight with a BMI of 24 kg/m^2, acanthosis nigricans, and a male pattern of coarser hair distribution in the beard region, anterior chest wall, abdomen and upper back and arms with modified Ferriman-Gallwey score of 32 indicating severe hirsutism (Figure 1 & Figure 2). She was normotensive and did not have typical clinical features of Cushing syndrome.

Laboratory investigations showed significantly elevated total testosterone of 8.6 ng/ml (<1 for female) and high 17-OH progesterone (17-OHP) with normal dehydroepiandrosterone sulfate (Table 1). Low dose dexamethasone suppression test revealed non-suppressed testosterone level of less than 40% of basal value suggesting a tumoral origin from ovary.

Subsequent imaging conducted during recent assessments, prompted by intensified symptoms, revealed significant changes. Ultrasonography of the abdomen and pelvis identified a solid mass in the right adnexa measuring 4.7 x 5.9 cm, displaying notable vascularity within the lesion. Additionally, CT abdomen and pelvis depicted a right-sided ovarian tumor measuring 7 x 6.9 cm, with significant vascularity, suggestive of a secreting tumor.

Treatment and further management

The patient underwent a planned laparoscopic cystectomy, which was converted to laparotomy due to surgical complexities. Intra-operatively, a right-sided ovarian lesion measuring 5 x 3 cm was observed, while the left ovary and other pelvic structures appeared normal (Figure 3). The cyst was extremely vascular and posed a huge technical challenge intraoperatively. The tumor was excised and submitted for histopathological examination. The cut

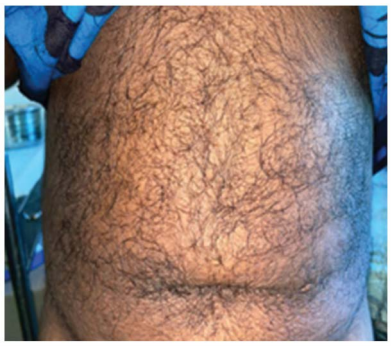

Figure 1: Hirsutism in the abdomen

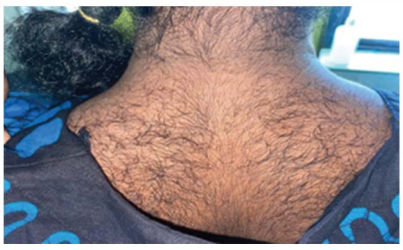

Figure 2: Hirsutism in the upper back

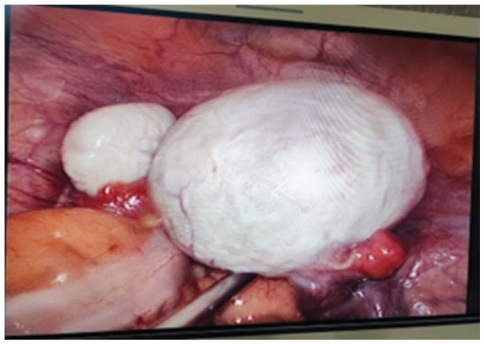

Figure 3: Laparoscopic image of an enlarged right ovary

Figure 4: Cut section of the tumor

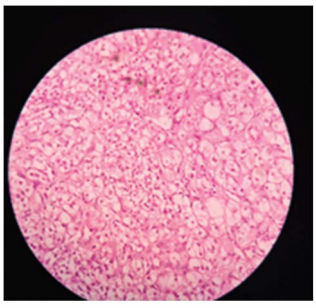

Figure 5: Histopathological appearance of the tumor

Table 1: Laboratory Investigations

Tests	At presentation	Postoperatively	Reference ranges
Total testosterone	8.6 ng/mL	<1	<1 for female
17-OHP	12.12 ng/mL	1.7	0.6-2.3
DHEA-S	327.3 µg/dL		18-391

section of the tumor showed a dark yellowish mass measuring 5.5x4.3x4.0 cm in size (Figure 4). Histopathology of the neoplastic tumor was composed of polygonal cells with abundant vacuolated cytoplasm, minimally pleomorphic nuclei, and cells arranged in diffuse pattern separated by fibrous septae containing many vascular channels. There were no mitosis, necrosis or foci of haemorrhage (Figure 5). These appearances support the diagnosis of sex cord stromal tumour steroid cell tumour (NOS).

Her postoperative recovery was unremarkable. Her excessive hair on the back, chest and abdomen completely disappeared in three months and facial hair was lost significantly, but not completely. Her periods returned and more importantly, she conceived within six months after surgery and currently, she is 20 weeks pregnant without any complications so far.

Discussion

Androgen-secreting tumors are rare causes of hyperandrogenism in women (0.2%)[3]. Androgen-secreting tumors, on the other hand, present with progressive and more severe hyperandrogenism leading to signs of virilization, such as clitoromegaly, deepening of voice, and male-pattern hair loss, as opposed to PCOS, where hyperandrogenism is mostly evident as hirsutism. In the case of rapidly progressing virilization and amenorrhea, neoplastic origin of hyperandrogenism should be excluded by doing total testosterone and DHEA-S levels [4]. Once the elevated testosterone in the presence of normal DHEA-S is established that is more suggestive of an ovarian tumor rather than adrenal origin. In our patient, elevated levels of 17-OHP were observed; however, further investigation was deferred due to the confirmation of an ovarian neoplasm through both the low-dose dexamethasone suppression test and CT imaging. Nevertheless, post-surgical normalization of 17-OHP levels strongly suggests the ovarian origin of the hormone[3].

Steroid cell tumors are classified into three subtypes: steroid cell tumor (NOS), stromal luteoma, and Leydig cell tumor[5]. NOS steroid cell tumors can generate hormones other than testosterone, with estradiol secretion being recorded in 6-23% of patients. In 6-10% of cases, these tumors have been linked to Cushing's syndrome[6]. Other hormones that may be secreted include androstenedione, dehydroepiandrosterone sulfate (DHEA-S), estrogens, 17-hydroxyprogesterone, ACTH, cortisol, renin, and others.

Surgery has successfully preserved ovarian function and fertility in young patients with ovarian steroid cell tumors (NOS). Given the low malignant potential of these tumors, fertility-preserving surgeries should be considered for patients under 40 years old, provided there are no malignant features present[5].

Conclusion

This case illustrates that tumorous cause for hyperandrogenism should be considered even in a woman with a known PCOS in the setting of rapid onset of hirsutism and elevated testosterone. Clinical suspicion and early identification will avoid unnecessary delay in the treatment.

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