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Laparoscopic management of an androgen secreting ovarian tumor: A case report of a rare ovarian tumor

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Abstract

Ovarian steroid cell tumors are fewer than 5% of sex-cord stromal tumors and 0.1% of all ovarian tumors. The average age of diagnosis is the mid-20s, but patients can present at virtually any age. We present a case of 28-year-old P1L1 with history of amenorrhea, secondary infertility and clinical signs and symptoms of virilization developed over past 2 years. With elevated serum testosterone level, clinical and radiological findings of a right solid ovarian mass; the patient was suspected to have a virilizing tumor of right ovary. Laparoscopic right ovariotomy was performed. Histopathology revealed pure leydig cell tumor-NOS variety. After surgery, patients' virilization reduced and she resumed her periods after 2 weeks and is on regular follow up and is now in the second trimester of pregnancy.

Keywords

ovarian steroid cell tumor; pure leydig cell tumor

Abbreviations

NOS: Not otherwise specified

Introduction

The spectrum of ovarian neoplasms covers an extremely wide range of tumors. The best recognized of these are the surface epithelial cell tumors. Amongst the less common variants, lipid or steroid cell tumors are very rare sex-cord stromal tumors comprising less than <0.1% of all ovarian tumors [1,2].

Steroid cell tumors are composed entirely or predominantly (>90%) of cells that resemble steroid hormone-secreting cells and have been sub classified as stromal luteomas, leydig cell tumors, and steroid cell tumors NOS [3].

Pure leydig cell tumor is a rare member of this group. They are usually benign, unilateral and characterized by the presence of Reinke crystals in the steroid cells; sub classified as hilar, non-hilar and not otherwise significant variety [4].

Leydig cell tumors are functional tumors and one of the ovarian virilizing tumors. They produce testosterone, leading to hyperandrogenism and virilization at any age group including reproductive age

group. In this case report we present a case of right ovarian tumor who presented with functional amenorrhea, secondary infertility and virilization diagnosed to be steroid hormone secreting tumor-leydig cell tumor NOS variety based on presentation, histology and reporting the management and prognosis of Leydig cell tumor-NOS variety.

Case Report

A 28-year-old P1L1 came to our hospital who presented with complaints of amenorrhea for 4 years and hirsutism since 2 years. She also had history of frontal balding and change in voice since 2 years.

On examination she had male pattern alopecia and increased facial hair. Her breasts were normal and she had severe hirsutism affecting the chest, anterior abdominal wall, and thighs. On per-abdomen examination there were no palpable masses. On pelvic examination revealed– mobile 4*4 cm firm mass in right adnexal region.

The blood investigations (DHEA- 219 µg/dL, CA125-12 U/mL, TSH- 4 mIU/L and 17 hydroxy progesterone-50 ng/dL) were found to be normal. The Serum testosterone level was increased (406.31 ng/dl). Ultrasound features showed minimal ascites, 5.2*4.8 cm solid right ovarian mass with increased vascularity. Uterus and the other ovary were found to be normal. Endometrial thickness was fpund to be 10 mm. Virilizing ovarian tumor was suspected and a diagnosis of androgen secreting ovarian tumor was made. Laparoscopic right ovariotomy was performed and cytology, omental biopsy and multiple peritoneal biopsies were taken and ovary extracted from pouch of douglas using endobag. Endometrial biopsy was also taken during the procedure.

Grossly, cut surface show solid yellowish tumor measuring 4.5*3.5*3 cm with no areas of hemorrhage or necrosis. Immunohistochemistry showing tumor cells positive for inhibin and negative for CD99andfinalHistopathology showed pure Leydig cell tumor-NOS variety. Endometrial biopsy showed proliferative endometrial cells.

After surgery her virilization and hirsutism were improved and she resumed her periods after 2 months. Her serum testosterone was found to be within normal limits (70 ng/dL). Patient is on regular follow-up for one year, ultrasound and clinical examination was being done every 3rd month and the patient now is pregnant and into her second trimester.

Discussion

Most androgen-secreting ovarian tumors are sex cord-stromal tumors, which constitute less than 5% of all ovarian neoplasms [5]. According to the World health Organization histologic classification of ovarian tumors, Leydig cell tumors are rare ovarian steroid cell neoplasms composed entirely or predominantly of Leydig cells that contain crystals of Reinke which may be identified in approximately 50% of these neoplasms. Pure leydig cell tumor is a rare member of this group. They are usually benign, unilateral and characterized by the presence of Reinke crystals in the steroid cells sub classified as hilar, non-hilar and not otherwise significant variety [4].

Leydig cell tumors typically seen in postmenopausal women (average age 58 years) but may occur in young women, pregnant women or children. They are usually associated with androgenic

manifestations, but occasionally produce estrogenic effects and are associated with endometrial carcinoma. These are mostly benign tumors in nature unless cells produce estrogen. Few reports also stating ovarian leydig cell tumors have been associated with MEN syndrome and congenital adrenal hyperplasia.

Leydig cell tumors of all types are intensively positive for alpha-inhibin and vimentin. The clinical behavior of all neoplasms in the pure Leydig cell category is benign, and neither clinical recurrence nor metastasis has been documented.

In case of larger tumors when it may not possible to determine whether the tumor arose from ovarian parenchyma or in the hilus, and these are referred to as Leydig cell tumors-NOS type like in our case. These are large tumors, well circumscribed, yellow to brown-black on cross section and associated with hemorrhage and necrosis if tumor size is very large.

Hirsutism and virilization are the most common symptoms occurring in 56-77% of patients and hormones other than testosterone can be elevated like estrogen and progesterone. Estradiol secretion by these tumors is not uncommon, occurring in 6-23% of patients. This excess estrogen production can result in menorrhagia and postmenopausal bleeding; in addition, endometrial adenocarcinoma has been described [6].

The proportion of tumors that are clinically malignant is more in leydig cell tumor-NOS variety than other pure Leydig cell tumors like non-hilar Leydig cell tumor or hilar Leydig cell tumor. The pathologic features which indicate malignancy or which indicate that the tumor can turn malignant are presence of two or more mitotic figures per 10 high-power fields associated with 92% of malignancy, necrosis with 86%, diameter of >7 cm with 78% of malignancy, hemorrhage with 77% and grade 2 or 3 atypia with 64% of malignancy [7]. None of the above mentioned features found in our patient.

Although the clinical course is not exactly known, it is recommended that leydig cell tumor-NOS, are managed surgically like other ovarian stromal tumors. Conservative surgery with unilateral oophorectomy and proper staging is performed in women with stage 1 disease who desire future fertility. Complete surgical staging including total abdominal hysterectomy and bilateral salpingo-oophorectomy, an accepted treatment for older women who do-not want to preserve their fertility.

The presented case emphasizes the importance awareness of a patient who presents with secondary amenorrhea and hirsutism and should be investigated systematically and to figure out if the high testosterone levels are of adrenal or ovarian origin and expedite appropriate management to achieve oncologic control of a rare tumor with malignant potential.

Conclusion

Pure Leydig cell tumor is a rare benign androgen secreting tumor. Laparoscopic management of this condition is feasible and reduces virilizing symptoms and restores fertility.

Table

Table 1: Pre operative and post operative hormonal profile

	Pre Surgery	Post Surgery
Serum Testosterone	406.31 ng/dL	70 ng/dL
DHEA	219 µg/dL	198 µg/dL
CA-125	12 U/mL	20 U/mL
Serum TSH	4 mIU/L	Not done
17-hydroxyprogesterone	50 ng/dL	Not done
Serum FSH	3 mIU/mL	Not done
Serum LH	4.11 mIU/mL	Not done
Serum Prolactin	11.94 ng/mL	Not done

Figures

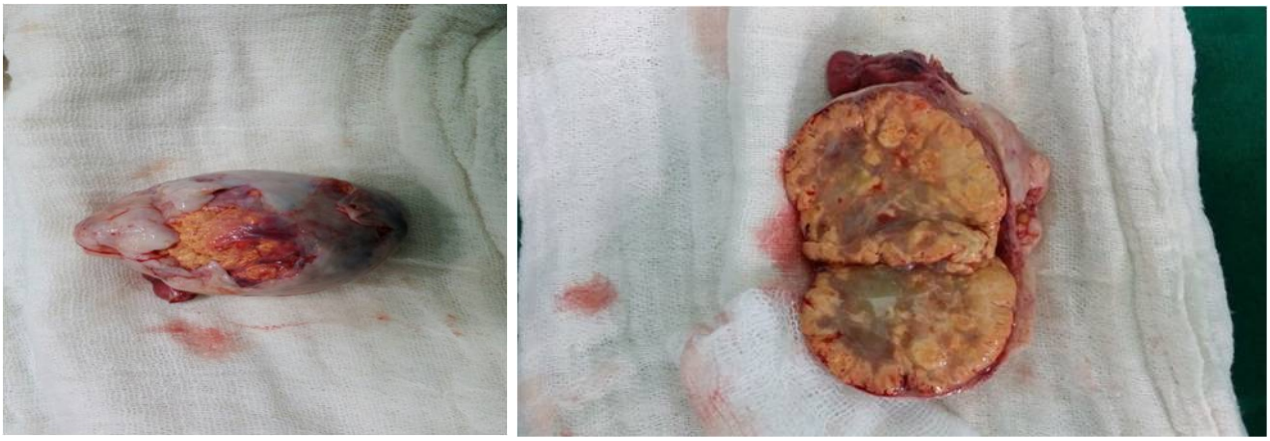

Figure 1 & 2: A photograph of gross and cut section of the patient's right ovary demonstrating a Leydig cell tumor measuring 4.5*3.5*3 cm with no hemorrhage and necrosis.

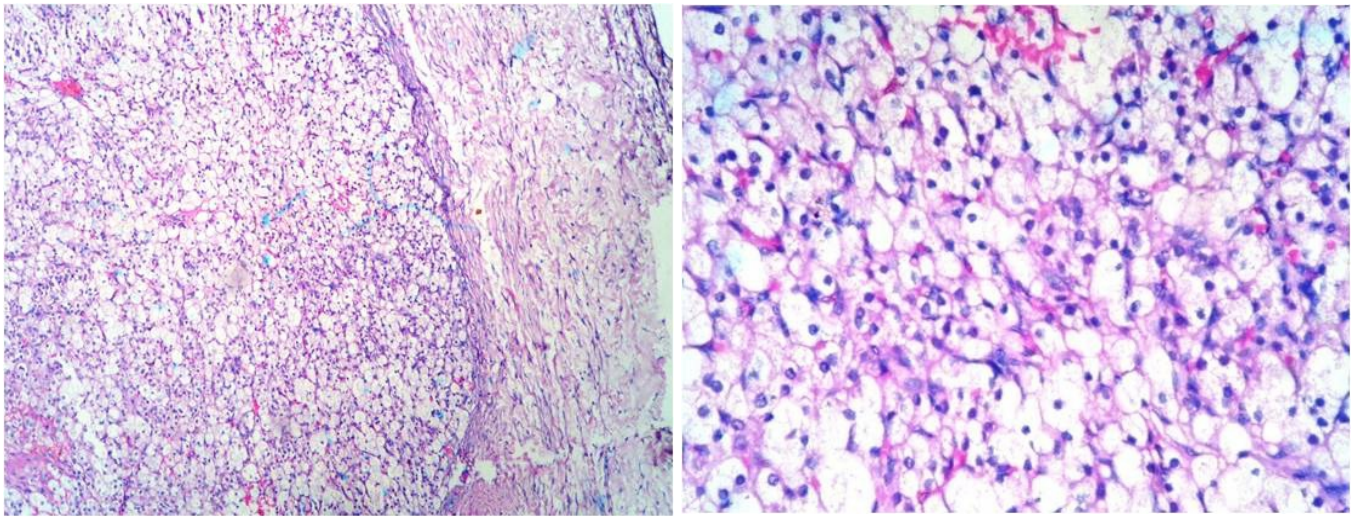

Figure 3 & 4: Low power (H&E X100) and high power (H&E X400) photomicrograph showing a pure Leydig cell tumor- NOS type

Sections show a well circumscribed tumor confined by the ovarian capsule, comprising of sheets of monomorphic polyhedral cells possessing abundant clear to pale eosinophilic cytoplasm and exhibiting minimal nuclear atypia. No foci of necrosis/atypia/increased mitotic activity.