10.5005/jp-journals-10006-1150

CASE REPORT

A Rare Case of Lipoid Cell Tumor of Ovary

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ABSTRACT

Lipoid cell tumors are rare hormone producing ovarian neoplasm. Majority of patients with these tumors are diagnosed at an early stage and unlike common epithelial tumors, these tumors have more specific symptoms due to the production of hormones and are associated with signs of virilism (hirsutism, amenorrhea, changed voice, breast atrophy and clitoromegaly).

Reported is a case of patient with sudden virilization caused by the benign androgen-producing lipoid-cell tumor of the ovary, very infrequently described in the literature.

Keywords: Lipoid cell, AVBRH, Ovary.

INTRODUCTION

Lipoid cell tumors are rare ovarian sex cord-stromal tumors with malignant potential. The majority of these tumors produce several steroids, particularly testosterone. Various virilizing symptoms, such as hirsutism, temporal balding, amenorrhea, clitoral enlargement and change of voice are common in these patients.

We are hereby presenting a case where the patient presented with rapidly progressive virilization and normal pelvic findings on clinical and sonographical examination producing a dilemma in diagnosis and management.

CASE REPORT

A 50-year-old lady, P3L3 presented at the Dermatology Outpatient Department on Dec 2008, for excessive hair growth over the face, coarse skin and change of voice of sudden onset, for 6 to 7 months. On investigations, her serum testosterone levels were high hence opinion of surgeons and gynecologist was saught (Fig. 1).

She was menopausal since last 10 years and on general examination, she was having generalised hirsutism, frontal baldness, coarse skin and husky voice. On per abdominal examination, no abnormality detected. On local examination, clitoromegaly was present. On per speculum examination, cervix was hypertrophied with multiple nabothian follicles, and on bimanual examination, uterus was bulky and adnexa clear.

On ultrasonography, uterus and ovaries were normal with slight increase in size of left ovary and increase in solid component alongwith doubtful adrenal gland adenoma. On CT scan solid ovarian enlargement of left side of size 4 × 3.5 cm was seen with no evidence of adrenal enlargement.

Fig. 1: Clinical photograph of patient showing hirsutism

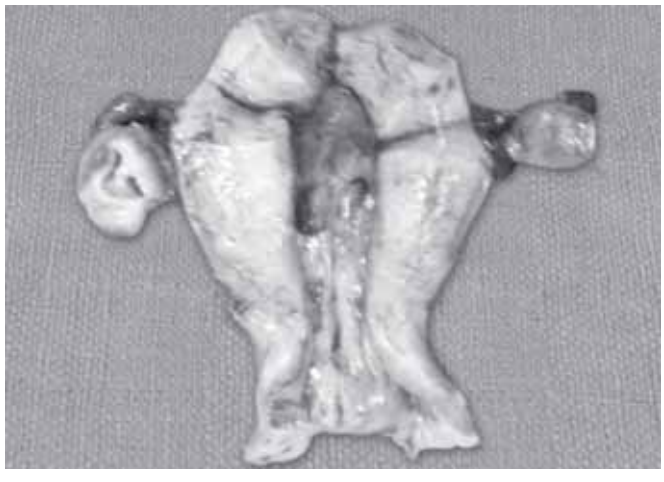


Fig. 2: Gross picture of specimen of total abdominal hysterectomy with bilateral salpingectomy

Inv: T3—120.48 normal: (60-200 µg/dl)
T4—8.5 normal: (4.5-12 µg/dl)
TSH—1.39 normal: (0.30-5.5 µg/dl)
FSH—1.39 MIU/ml
LH—9.24 MIU/ml
Serum testosterone—206.9 normal: (6-82 ng/ml)

Her laparotomy was planned and in view of chronic cervicitis and solid enlargement of right ovary, her total abdominal hysterectomy and bilateral salpingo-oophorectomy was done, though on gross appearance this enlargement was not much significant and uterus was size of a 6 weeks gravid uterus with myohyperplasia and endometrial atrophy. Right ovary was slightly enlarged, approximately 3 × 3.5 cm in size. Adenexae was absolutely normal looking without any adhesions. Cut section of right ovary showed mild solid enlargement with excessive yellow cortex (Fig. 2).

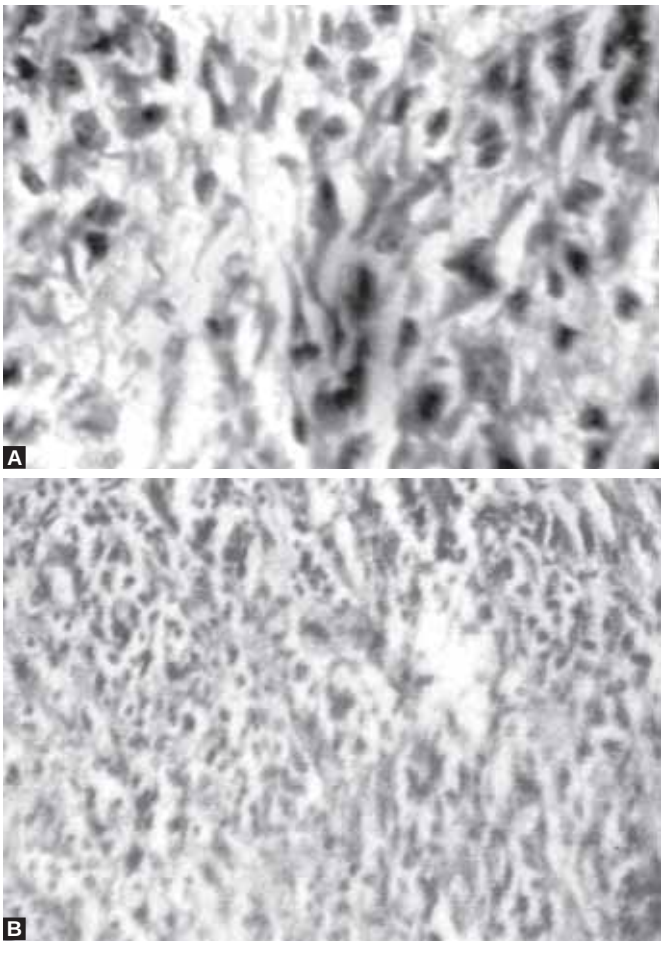
According to general surgeon, no surgical intervention was required for suspected adrenal mass.

On histopathological examination, right ovary had (Figs 3A and B) lipoid cell tumor (benign) and cervix had mild dysplasia.

Her hormone levels were normal after 48 hours, and post-operative period was uneventful and she was discharged after 8 days.

At the time of her first postoperative visit to the outpatient clinic 3 weeks after surgery, her virilization and hirsutism were improved and her hormone levels were within the normal range: testosterone, 21 ng/ml and androstenedione, 2.7 ng/ml (normal range, 1.1-3.9 ng/ml).

She currently remains entirely asymptomatic, 5 months after surgery.



Figs 3A and B: Histopathological slides showing lipoid cell tumor of ovary

DISCUSSION

Hormone producing ovarian neoplasms account for 5 to 8% of all ovarian neoplasms. They are composed of cells that differentiate in the direction of sex cords and/or the specialized ovarian neoplasm. They account for most of the hormonally active ovarian neoplasms.

Granulosa cell tumors and Sertoli-Leydig cell tumors are the most common. Others include thecomas, fibroma, gynandroblastoma, sex-cord tumor with annular tubular unclassified tumors.

Lipoid cell tumors are rare hormone producing ovarian neoplasm.

Unlike patients with common epithelial tumors, in which 75% are considered to be at advanced stages at diagnosis, majority of patients with these tumors are diagnosed at an early stage. And unlike common epithelial tumors, these tumors have more specific symptoms due to the production of hormones and are associated with signs of virilism.[1]

The histopathological term "lipoid cell tumors of the ovary" was used to describe "steroid cell carcinoma" prior to Scully's description.[2] Such terminology was obscure because some tumors have little or no lipid present.[3]

There is debate over the genesis of lipoid cell tumors of the ovary, i.e as to whether they are primarily ovarian or adrenal in origin. However, as adrenal and ovarian steroid producing cells are both derived from a common primitive mesenchymal cell, it can also be argued that when these cells become neoplastic, previously represented genes become functional, resulting in the subsequent activation of enzyme systems. Thus under these circumstances ovarian tumors have the potential to function like adrenocortical tissue.

According to WHO, lipoid cell tumors are defined as tumors composed of cells that resemble leydig, lutein or adrenal cortical cells, but cannot be identified specifically as any one of the three types as illustrated by the existence of not less than 28 a different names under which these tumors appear in the medical literature.[4]

NOS tumors can occur at any age, but usually develop in adults with an average age of 43 years. It has been reported in 93-year-old lady with virilization[5] as well as in 8-year-old prepubertal girl with androgenic pseudoprecocity.[6]

The major symptoms detected in 56 to 77% of patients are hirsutism and virilization. Reedy et al reported a case of an undifferentiated NOS steroid cell tumor with hirsutism, amenorrhea, clitoromegaly and temporal baldness.[7] Massive ascites and elevated CA125 levels are infrequent present .[3]

A study by Hayes and Scully reported the most predictive malignant features of ovarian steroid cell tumors. According to

their criteria, the most accurate predictor of malignant behavior in these tumors is the presence of two or more mitotic figures per 10 high-power fields. In addition, the majority of malignant tumors also demonstrate grade 2-3 nuclear atypia, necrosis, hemorrhage and a tumor diameter greater than 7 cm. Most tumors are unilateral (94%) and the majority are capable of sex steroid hormone production. In adults, approximately a quarter of steroid cell tumors are malignant.[3]

Grossly, lipoid cell tumors are solid ovarian neoplasms whose cut surfaces are bright orange or yellow in color and range in diameter from 0.5 to 24 cm. They are frequently unilateral and usually compress rather than invade the adjacent ovarian stroma.

Microscopically, these tumors are composed of polygonal cells with abundant vacuolated or eosinophilic cytoplasm with abundant lipid and in addition to virilization, produce polycythemia, disturbances in carbohydrate metabolism, hypertension, features of cushing's syndrome, hyperostosis frontalis, abnormal bleeding due to progestational endometrial changes, amenorrhea due to androgens and precocious puberty. Their diagnosis is traditionally based on the finding of elevated plasma testosterone and elevated urinary 17-keto-steroid excretion. They are usually benign and slow growing, and the symptoms are frequently present for many years before the diagnosis is made. The propensity for malignant degeneration of an ovarian lipoid cell tumor is low (20%).

CT and MRI characteristics of steroid cell tumors are variable owing to the amount of lipid components and fibrous stroma. However, the tumors show intense enhancement on MRI, reflecting the hypervascularity. Tumors causing virilization are often small. Some recommend performing chemical shift MRI in differentiating testosterone producing ovarian tumors.

The mainstay of ovarian steroid cell tumor treatment is surgery.

Surgical treatments using total abdominal hysterectomy, bilateral salpingo-oophorectomy and complete surgical staging are an appropriate management option for old women who do not want to preserve their fertility, as was our case. However, in young patients, unilateral salpingo-oophorectomy is adequate most of the time due to the low bilateral frequency of 6%. However, such practices require a mandatory follow-up evaluation and should include a measurement of sex hormone levels, particularly for those patients who demonstrated elevated levels before removal of the primary tumor. Additionally, a gonadotropin-releasing hormone agonist could be used as postoperative adjuvant therapy. In adults, approximately 25 to 43% of steroid cell tumors are malignant.

Reported is a case of lady with sudden virilization caused by the benign unilateral, apparently normal appearing, androgen-producing lipoid cell tumor of the ovary, rarely described in the literature.

ACKNOWLEDGMENTS

We are extremely thankful to Department of Dermatology and Department of General Surgery of our institute for their support and cooperation in presenting this case report.

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