RA Publications
Journal of Medical Science & Technology

Case Report Open Access

Atypical Presentation of Hilar Leydig Cell Tumour of ovary with Mullerian anomaly – "A Rare Case Report"

Dr Medabalmi Anitha[1*]

1. Professor & HOD, Narayana Medical College, Nellore, Andhra Pradesh, India.

Abstract

Leydig cell tumors belong to the small group of ovarian steroid tumors, which are hormone producing and derived from specific stromal cells. Morphologically, their endocrine like structure is characteristic, formed of large, polyhedral cells resembling luteal, adrenocortical and leydig cells. They contain crystalloids of Reinke in their cytoplasm which have diagnostic significance. The patients with these tumors present present with androgenic manifestations. We report a case of Leydig cell tumor in 35 years female who presented with infertility and hirsutism.

Key words: Leydig cell tumor, Reinke's crystalloids, Mellerian anomaly, Hirsutism.

*Corresponding Author: Dr M Anitha, Professor and Head, Dept. of Obstetrics and Gynecology, Narayana Medical College and Hospital, Nellore, Andhra Pradesh, India. Email: nitasr1648@gmail.com

Received: March 1, 2013. Accepted: May 2, 2013. Published: May 20, 2013.

Introduction

Hilar leydig cell tumor is a rare androgen secreting ovarian tumor of unknown pathogenesis. The incidence is less than < 0.1% of all ovarian tumors. Hilar leydig cell tumor originates from ovarian stromal sex cords, mostly benign. The coexistence of Hilar leydig cell tumor with mullerian anomalies is very rare.[1] It affects age group of 2-80 yearrs (Hayes & scully) associated with virilizing properties. It can be diagnosed at an early stage. Recurrence is very rare but non -metastatic.

Leydig cells of different origin show the features of lipid cells and they contain crystalloids of Reinke. The presence of crystalloids is specific and of diagnostic value. The case report of Leydig cell tumor in a 35 year old women who presented with history of with infertility and signs of hirsutism and virilization is described here.

Case History

Mrs. X, lady aged 33yrs, married for 16yrs is a house wife attended at OBG department out patient with a c/o secondary amenorrhea, hirsutism, infertility for 2 years. On examination signs of hirsutism and virilization were noticed. Abnormal hair growth is seen inner parts of thighs and buttocks, upper lip & chin. Scalp hair loss and clitoromegaly was noticed. She has male kind of voice change, frontal baldness since 6 months.

Menstrual cycle History: Menarche at the age of 13yrs, Regular cycles for 9 yrs. Irregular menstrual cycle observed since 4yrs. On General Examination we find BMI- 25.57kg/m^2 Breast & axillary hair is normal and pubic hair is male pattern, features like hirsutism & frontal baldness are also seen. Gynaec examination revealed that external Genitalia normal. Clitoromegaly (10x15mm) "clitoral index 150". On bimanual pelvic examination uterus showed less than the normal size, Right ovary is palpable and Left ovary is normal.

Laboratory findings revealed a normal complete blood count and metabolic profile. Endocrinological work up revealed normal hormonal levels except for increased levels of total and free testosterone.

Figure 1: Frontal balding

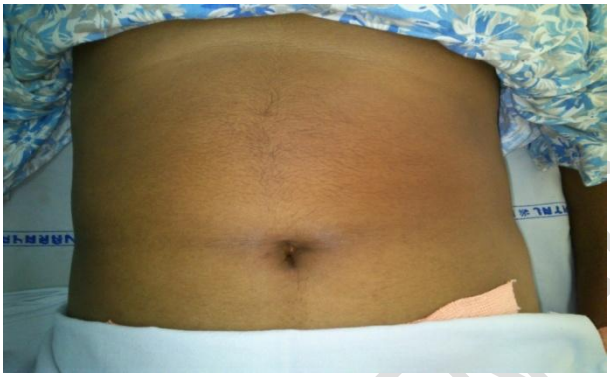

Figure 2: Hair growth on the lower abdomen

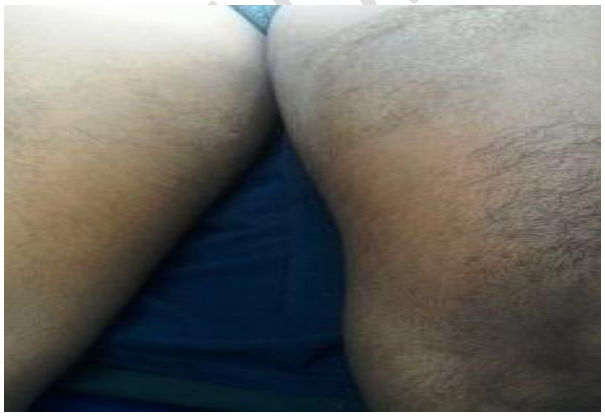

Figure 3: Abnormal hair growth on thighs.

Other Clincal findings: Routine : - WNL ;**TVS :-** Uterus: 5.1x1.3x1.5 cms, ET=3mm. Lt. ovary- 2.2x1.4 cms. Rt. ovary-4.5x2.9cms, enlarged, multiple hyper echoic foci. **X-ray skull :** NAD

MRI & CT: Normal hypothalamus & Pituitary, Rt. Ovary(4.6x3cm)

Estimation of FSH, LH, Prolactin, Estradiol, TSH, Free T3, Free T3, Free T4, 8. am ONDST, Total Testosterone, Free Testosterone, Sr. DHEAS and DHEA. All the hormone levels are normal except for increase in total and free Testosterone levels.

Management

Exploratory Laparotomy Findings:

a) Uterus - Hypoplastic, bicornuate with small right sided non - communicating rudimentary horn.

b) Rt.ovary - Enlarged -4.5x3cms with solid component & increased vascularity.

c) Lt. ovary - Normal size 3x2 cms with abnormal vascularity.

Rt ovary - U/L oophorectomy

Lt ovary - wedge biopsy taken.

Specimen:

Gross:- Right ovary enlarged to 4.5x3x2.5 with well circumscribed nodule confined to the hilum .

Cut section: single well circumscribed greyish yellow nodule of 3x2cms with adjacent normal ovarian parenchyma.

Histopathology report

Right ovary:-

Polygonal tumor cells arranged in lobules with abundant vacuolated cytoplasm containing **Rienkie crystals-** reactive to inhibin- diagnostic of- Leydig cell tumor

Left ovary :- corpus albicantes

Follow up

Post operation period – Uneventful
8th POD - Repeated. Serum Total testosterone - 0.98ng/dl (Norma range- 3 to 46ng/ml)
Since Serum testosterone levels decreased after surgery & maintained later- suggesting ovary as source of androgen excess. Repeat MRI and CT of abdomen & pelvis – normal adrenals & Left ovary. Patient resumed menstrual cycles after 8 wks.

Features of leydig cell tumor of ovary :-

- Androgen secreting , rarely estrogenic.
- Located in the hilum of the ovary.
- Derived from sex cords and mesenchyme.
- Accounts for <0.1- 0.5% of all ovarian neoplasms
- Slow growing, benign, with low malig. potential.
- Mostly unilateral.
- Recurrence - as late as 30yrs after surgery.
- Accounts –15 to 20 % of steroid cell tumors.
- Contains predominantly leydig cells with rienke crystals.
- Size – 1 to 15 cm but mostly <5cms.
- Presents with oligomenorrhea, amenorrhea, infertility, hirsutism, virilization, abd pain, abdomen mass.

Discussion

Most androgen secreting ovarian tumors are sex cord stromal tumors, which constitutes less than 5% of all ovarian neoplasms. Leydig cell tumors are rare ovarian steroid cell neoplasms composed entirely or predominantly of leydig cells that contain crystals of Reinke. These tumors account for 15% to 20% of steroid cell tumors.

The differential diagnosis of the Leydig cell tumors includes ovarian neoplasms containing leydig cells or luteinized stromal cells. Sertoli leydig cell androblastoma occasionally exhibits predominance of leydig cell component. But presence of sertoli cells excludes the diagnosis of pure leydig cell neoplasm.

Summary

Androgen producing tumors should be suspected in women with virilizing clinical symptoms and high testosterone levels. Sertoli leydig cell tumors are larger and usually found easily on imaging, whereas hilar leydig cell tumors are smaller and often difficult to find on imaging. If clinical suspicion is high exploratory laparotomy is indicated. It is noteworthy that in this era where sophisticated and expensive histopathological methods including immunohistochemistry is in use, this rare and benign tumor can be diagnosed with high accuracy on good quality H and E stained slides.

References

1. Paraskevas M, Scully R E. Hilus cell tumor of the ovary. A clinicopathological analysis of twelve Reinke crystal – positive cases and nine crystal negative cases. Int J Gynecol Pathol 1989;8:299-310
2. Zhang J, Young R H, Arseneau J et al. Ovarian stromal tumors containing lutein or leydig cells (Leutinized thecomas and stromal leydig tumors) – a clinicopathological analysis of fifty cases. Int J Gynecol Pathol 1982;1:270-285.
3. Cronje H S, Niemonmd I, Barn R H, Woodruff J D. Review of the granulose – the carcinoma cell tumors from the Emil Novak Ovarian tumor registry. Am J Obstet Gynecol. 1999;180:325 – 327.
4. Tavassoli F A, Devilee p (Eds): World Health organization Classification of tumors. Pathology and Genetics of tumors of the breast and female genital organs. IARC Press: Lyon 2003.
5. Scully R E, Young R H. Atlas of tumor pathology tumors of the ovary. Maldeveloped Gonads, fallopian tubes and broad ligament 3rd ed, AFIP; Washington, DC 1998.
6. Gershenson DM, Young RH. Ovarian sexcord stromal tumors in: Hoskins WJ, editor. Principles and practice of gynecologic oncology. 4th ed. Philedelhic Lipphicott Williams and Wilkins;2005:1024.
7. Ichinohasama R, Teshima S, kishi K, et al. Leydig cell tumor of the ovary associated with endometrial carcinoma and containing 17 beta – hydroxy steroid dehydrogenase. Int J Gynecol Pathol. 1989;8:64 – 71.

