AN OVARIAN STEROID CELL TUMOUR CAUSING VIRILISATION- A RARE CASE REPORT

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INTRODUCTION: Steroid cell tumor of the ovary a very rare androgen secreting tumor accounts for less than 0.1% of ovarian tumors. These tumors may present at any age ranging from 2 to 80 yrs & associated with hormonal activity & virilisation. These tumors are most commonly unilateral, may be both benign or malignant, most of these tumors are usually diagnosed in early stages because of virilisation caused by these tumors.

In an extensive literature search (medline & pubmed) from 1979, only around 80 cases of ovarian steroid cell tumors have been reported. As ovarian stromal tumors are infrequent, little attention was given to their response to radiotherapy and chemotherapy. A recent report stated gonadotropin releasing hormone agonist was effective in treating steroid cell tumors.

Here we present the case of a rare ovarian steroid cell tumor in a 32 yrs old woman who presented with virilisation.

CASE REPORT: A 32 yrs old woman with P2L1D1A1 presented with history of amenorrhea of 7 months duration, hoarseness of voice since 6 yrs, decrease in breast size since 6 yrs, lower abdominal pain which is dull aching type, coarse hair growth over the body since 2 yrs. She attained menarche at the age of 12 yrs. Her previous menstrual cycles were normal.

Physical examination revealed male pattern of coarse hair distribution in the beard region, face, chest wall, abdomen, arms and thighs. Both breasts were small in size, no mass palpable per abdominally.

Local examination shows clitoromegaly. On per vaginal examination, there was right fornicis fullness, and a mass felt through the right fornix.

INVESTIGATIONS: Ultrasonography of abdominopelvic region revealed a solid right adnexal mass of 4.3 x 4.3 cm, Doppler study shows minimal vascularity in the tumor. CT scan revealed a right adnexal solid mass of 6.1x4.3 cm. No ascites.

PRE-OPERATIVE HORMONAL ASSAY:

	VALUES	NORMAL
S. Total Testosterone level	197.14 ng/dl	15-70 ng/dl
S. Free Testosterone level	79 ng/dl	0.06-3.18 ng/dl
17-OH Progesterone	5.67 ng/dl	0.11-5 ng/dl
DHEAS04	160 microgm/dl	35- 430 microgm/dl
S. Prolactin level	5.25 ng/dl	
FSH	2.42mIU/ml	1.9-12.5
LH	4.07mIU/ml	2.5-10.2

Intra-operatively, the right ovarian mass is about 6 x 4 cms, tan brown, well encapsulated solid mass, firm in consistency. Left ovary is normal in size and appearance. Hysterectomy with bilateral salpingo-oophorectomy was done.

HISTOPATHOLOGICAL REPORT: Grossly the ovary was enlarged and measured 6.5 x 4 x 3.5 cm, external surface was smooth. Cut surface showed a circumscribed, lobulated tumor which was yellow in colour.

Microscopy revealed a tumor composed of lobules of tumor cells arranged in sheets and nests. These cells were polyhedral and showed abundant clear to eosinophilic granular cytoplasm (Fig 2). No mitosis or necrosis was seen. The histopathological diagnosis was steroid cell tumor.

POST SURGICAL FOLLOW-UP OF THE PATIENT: Patient was followed regularly after surgery for 3 yrs.

	Free testosterone levels	hair distribution	Breast changes	clitoromegaly	Recurrence of the disease(BY ULTRASOUND)
1st visit after 2 months	0.45 ng/dl	Coarse hair became thin	Mild enlargement in the breast	No changes	No
2nd visit after 6 months	0.14 ng/dl	Almost feminine	Mild to moderate enlargement seen	No changes	No
3rd visit after 1 yr	0.12 ng/dl	Feminine	Breasts enlarged	No changes	No
4th visit after 1 ½ yr	0.16 ng/dl	Feminine	Enlarged almost to normal size	No changes	No
5th visit after 2 yrs	0.09 ng/dl	Feminine	Enlarged almost to normal size	No changes	No
6th visit after 3 yrs	0.01 ng/dl	Feminine	Enlarged to normal size	No changes	CT scan study shows no recurrence

DISCUSSION: Ovarian steroid cell tumor, a rare tumor accounts < 0.1 % of all ovarian tumors. They are usually benign, unilateral and characterised by steroid cell proliferation.

This term was first described by Scully. Prior to this, it was called as lipoid cell tumor.

Majority of these tumors are benign, about 20% of patients developed metastatic lesions within the peritoneal cavity. Heyes and Scully identified pathologic features that are highly associated with malignancy – more than 2 mitoses per high power field, necrosis, size of 7 cms or larger and grade 2 or 3 nuclear atypia.

The primary treatment is surgery. Surgical treatment using total abdominal hysterectomy with bilateral salpingo-oophorectomy and complete surgical staging is appropriate management for

older women, who are not willing to preserve their fertility. However such practices require mandatory follow-up and evaluation.

In virilised patient, serum testosterone levels more than 2 ng/dl, normal DHEA-S and no evidence of 21 alfa hydroxylase deficiency are strong indicators of steroid cell tumor of ovary, as in our case.

CONCLUSION: Steroid cell tumors of the ovary are rare tumors, more commonly benign with virilising property with distressing symptoms to the patient because of abnormal pattern of hair distribution, voice changes. One has to be aware of this condition in women who present with above symptoms, so that one can prevent the spread of tumors, unwanted signs and symptoms like hair growth , voice changes, & clitoromegaly.

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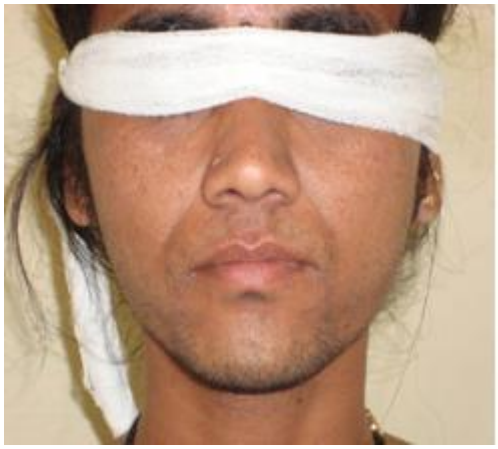
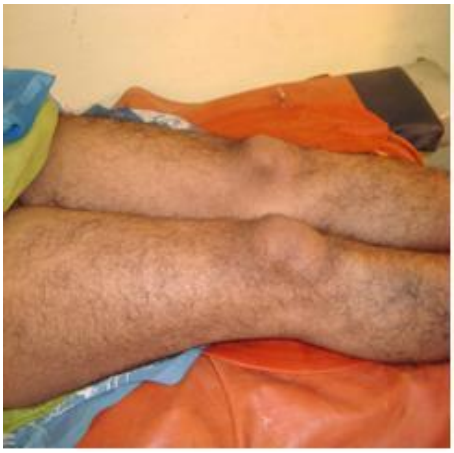
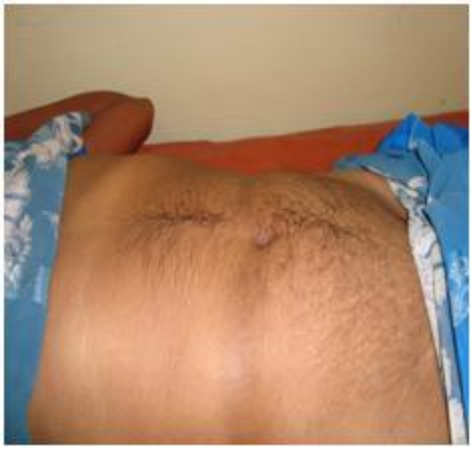
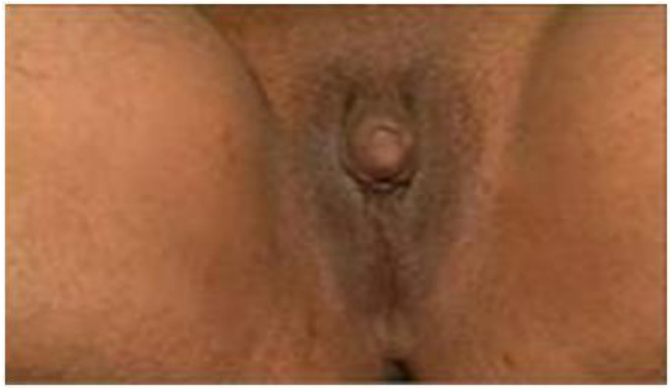
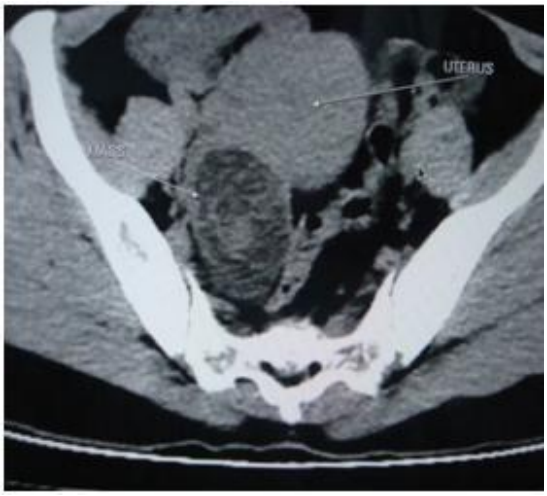


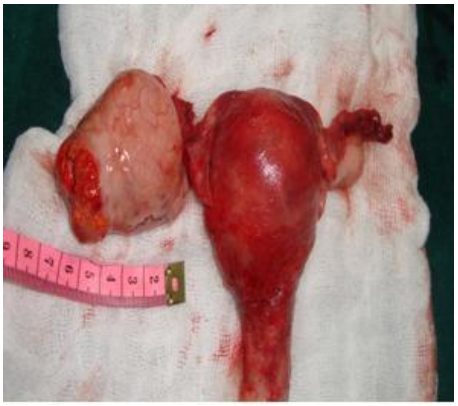


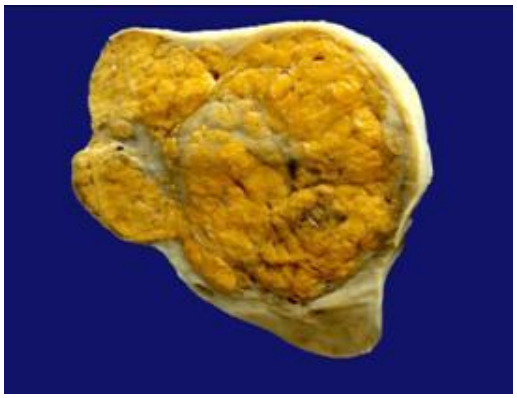






CT SCAN IMAGE





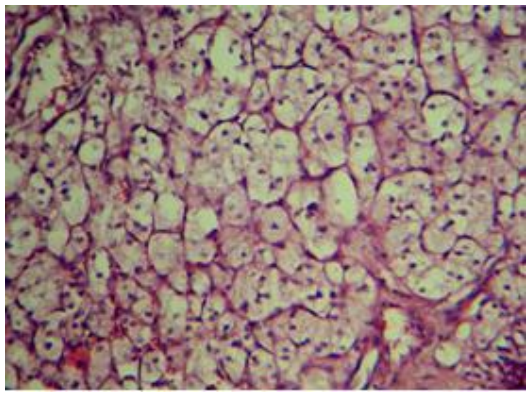


Fig 1: Gross appearance of Steroid cell tumor. Cut section of the ovary showing a yellow lobulated, circumscribed tumor.

Fig 2: Microphotograph showing polyhedral cells with clear to eosinophilic granular cytoplasm (H&E x 400).

CASE REPORT

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