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ANDROGEN SECRETING STEROID CELL TUMOR OF OVARY: A RARE CASE REPORT

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ABSTRACT

Steroid cell tumors of the ovary are rare an uncommon sex-hormone secreting tumors. These tumors are characterized by a steroid cell proliferation. The incidence of steroid cell tumor of the ovary is only 0.1% of all ovarian tumors. Here we present one case of 40-year-old female came with complaints of Hirsutism, Voice changes, amenorrhea since 6 months. Testosterone levels was increased {4.15 ng/ml}. Histopathology confirmed the diagnosis of steroid cell tumor with no cytological atypia. On follow up after removal of tumor, patient became free of virilisation and had normal level of testosterone.

INTRODUCTION

Steroid cell tumors of ovary are very rarely found and they constitute only 0.1% of all ovarian tumors. These tumors are divided into 3 subtypes according to their cell of origin as follows:

- Stromal Luteoma
- Leydig cell tumor
- Steroid cell tumor – Not otherwise specified

Most Steroid cell tumors of ovary are most often associated with secretion of steroid hormones, which causes symptoms that lead to clinical diagnosis. In general testosterone secretion lead to virilisation, hirsutism and amenorrhea; Estrogen secretion lead to bloating, fibrocystic lumps in breast and only 10 -15% of patients have no clinical signs or symptoms of increased hormone levels.

MATERIAL AND METHODS

A 40-year-old female presented with hirsutism, amenorrhea and voice change since 6 months. Testosterone levels were increased {4.15 ng/ml}. Past medical history was otherwise unremarkable. Family history was noncontributory.

On per vaginum examination a firm mobile left adnexal mass palpated. Ct scan showed bulky left ovary with thick walled centrally placed cysts.

On gross, A well circumscribed, unilateral, yellow lobulated Ovarian tumour located in left ovary with characteristic golden yellow colour with no areas of hemorrhage and necrosis. [Figure 1].

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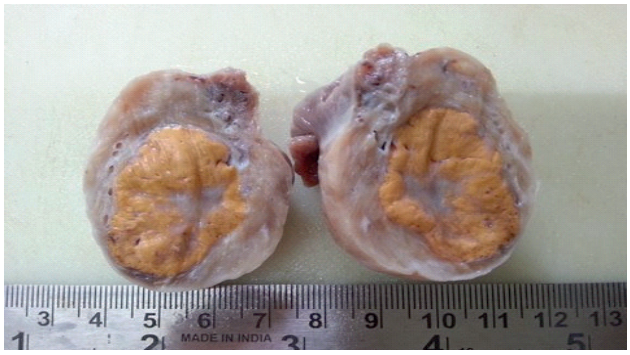
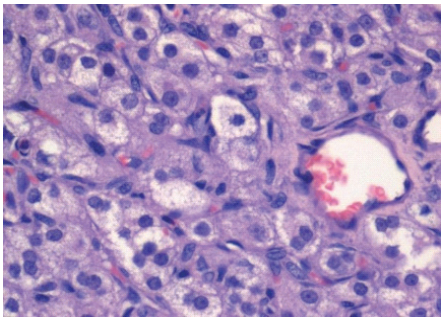


Figure 1

Figure (1.): Steroid cell tumor: Gross specimen of steroid cell tumor showed well circumscribed solid lesion with a characteristic golden yellow colour with no areas of hemorrhage and necrosis.



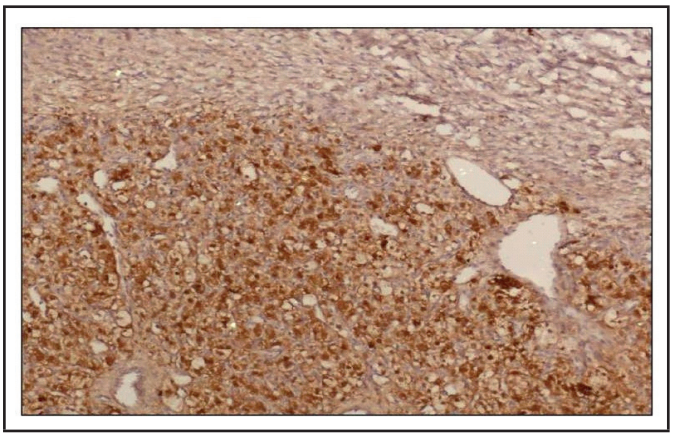
Sections from ovarian tumour showed well circumscribed tumour comprising of large polygonal to round cells with centrally placed nuclei, prominent nucleoli and abundant amount of eosinophilic cytoplasm {Figure 2a & 2b}. There was no necrosis, atypia or mitotic figures noted in the tumour. Histological features were consistent

with steroid cell tumour NOS type.

Figure 2a- Large polyhedral cells with vacuolated cytoplasm and smaller cells with eosinophilic granular cytoplasm in vascular stroma (H&E,10X).

Figure 2b –High power showing polygonal tumor cells with central nuclei, prominent nucleoli and abundant cytoplasm (H&E,40X).

A calretinin stain was performed in this case but was found to be positive {Figure 3}.



On follow up after removal of tumor, patient became free of virilization and had normal level of testosterone {0.54 ng/ml}.

DISCUSSION

Scully was the first person who have described ovarian steroid cell tumor, and he reported 63 cases ranging from age 2 to 80 years[1].Formerly, these tumors were referred as lipid or lipoid cell tumors of the ovary.

Steroid cell tumors of the ovary are rarely identified and constitute only about 0.1% of ovarian tumor

These tumors are divided into three subtypes according to their cells of origin as follows: Stromal luteoma, Leydig cell tumor, and Steroid cell tumor-not otherwise specified (NOS)[2,3] Of these subtypes, the steroid cell tumors-NOS account for about 56% of steroid cell tumors. Androgenic manifestations are common in these tumors as they secrete hormones like testosterone, α-hydroxyprogesterone and and rostenedione.

These tumours are known to produce symptoms of virilisation particularly hirsutism and amenorrhoea. So in cases when there is unexplained hirsutism, ovarian and adrenal tumor association should be ruled out as there may be occult malignancies.[4]

However, there might be atypical presentations of these tumours also when they do not show any symptoms of virilisation. In these cases, the diagnosis is usually made postoperatively on finding a tumour in ovary.

A majority of steroid cell tumor-NOS are generally unilateral, benign and well-circumscribed. The size varies from 1.2 to 45 cm[1] . Grossly, these tumors are commonly solid; however, a combination of solid and cystic form or predominantly cystic form may also be seen. The color of the cut surface may range from yellow - orange to red or brown depending on the lipid content. Areas of hemorrhage and necrosis may also be seen.

The tumor in our case was completely solid with no cystic area. The cut surface was typically yellow.

There are certain clinicopathologic parameters which correlate with worse behavior of the tumor such as older age at the time of presentation, tumor size more than 7.0 cm, mitosis more than 2/10 HPFs, nuclear atypia grade 2–3, hemorrhage and necrosis[5].

In the present case, even though the patient is of middle age group, she had a good prognosis as the size of the tumor was < 7 cm and on microscopy, there were no necrosis, mitosis and nuclear atypia.

A recent study analysed ovarian sex-cord stromal tumours for calretinin positivity [6]. All the six steroid cell tumours in this study were positive for the marker. Calretinin can be considered a sensitive marker for ovarian sex-cord stromal tumours in general, but is less specific [7].

The patient was not offered any further medications or chemotherapy and was advised only follow-up after her tumor removal.

CONCLUSION

Steroid cell tumors-NOS are very rare ovarian sex cord-stromal tumors which are usually associated with various symptoms such as pain in the abdomen, hirsutism and irregular menstrual cycles. Along with clinical correlation, histopathology is the gold standard which can confirm the diagnosis in most of the cases. In atypical cases, immunohistochemistry can be helpful for accurate diagnosis[8,9].

Declaration of patient consent

The patient understand that name and initials will not be published and due efforts will be made to conceal identity.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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