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Ovarian Steroid Cell Tumor: A Case Report

Dr. Manjusha Jindal[(1)], Dr. Viraj Ramesh Naik[(2)], Dr. Mrinalini Sahasrabhojanee[(3)], Dr. Ashwini Naik[(4)], Dr. Saba Anjum Khan[(4)], Dr. Siddhi Naik[(4)]

[(1)] *Associate Professor, Department of Obstetrics and Gynaecology, Goa Medical College, Bambolim Goa.*
[(2)] *Senior Resident, Department of Obstetrics and Gynaecology, Goa Medical College, Bambolim Goa.*
[(3)] *Assistant Professor, Department of Obstetrics and Gynaecology, Goa Medical College, Bambolim Goa.*
[(4)] *Junior Resident, Department of Obstetrics and Gynaecology, Goa Medical College, Bambolim Goa.*

Abstract:
Introduction: *Ovarian steroid cell tumors are a rare subgroup of sex cord stromal tumors of the ovary and comprise less than 0.1% of all ovarian tumors.*
Case report: *A 12 year old patient presented with abdominal pain and found to have right ovarian cyst, right ovarian cystectomy done for the same and histopathology revealed ovarian steroid cell tumor and the same was confirmed by immunohistochemistry with positivity for Vimentin, Calretinin and Inhibin.*
Conclusion: *Ovarian steroid cell tumours, NOS, are rare tumours. Along with clinical correlation, histopathology is the gold standard to confirm the diagnosis in most of the cases. In atypical cases, immunohistochemistry can be helpful for accurate diagnosis. The treatment varies from ovarian cystectomy to total abdominal hysterectomy with bilateral salpingo-oophorectomy depending on histopathological features of malignancy.*
Key words: *Ovarian, Steroid cell tumor, Immunohistochemistry, benign, Malignant*

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I. Introduction:

Ovarian steroid cell tumors belong to a rare subgroup of sex cord stromal tumors of the ovary and form less than 0.1% of all ovarian tumors. They have been divided into three subtypes, according to their cell of origin,: stromal luteoma, leydig cell tumor, and steroid cell tumor not otherwise specified (NOS) [(1,2)] In literature, only a few cases of steroid cell tumors NOS have been described, most with symptoms and bulky tumors at presentation.[(3)] Clinical presentation of steroid cell tumours are due to steroids especially testosterone and generally show androgenic symptoms like amenorrhea, abnormal hair growth in the face and body, hair loss, deepening of voice and increased libido.[(1)] This case report describes a case of steroid cell tumor NOS which was diagnosed retrospectively.

II. Case Report:

12 year old girl presented to gynaecology OPD with complaints of pain in lower abdomen since 5 to 6 months, pain was dull aching type without aggravating or relieving factors. Patient had attained menarche 6 months prior with no history of menstrual irregularities, no history suggestive of virilisation. On clinical examination, no specific findings were elicited. On ultrasound pelvis, cystic mass was noted in right ovary 4.5*4.9*5.9 cm with no septae. A solid component 18*7 mm was noted in inferior aspect of this cystic lesion. Patient was evaluated. CA 125- 8.7, Alpha Feto protein- 1.74, Beta HCG < 1.20, LDH-156, serum testosterone 0.65 (all within normal limits). Patient was taken for laparotomy in view of variegated consistency. OT findings were cystic mass arising from right ovary 5*5 cm with no surface excrescences, or mural nodule however cyst was found to be unhealthy and noted to have hemorrhage within and contralateral ovary was bulky 4*3*2 cm. No free fluid in abdomen was noted. As patient was only 12 years old, cystectomy was done and ovarian reconstruction was done. Biopsy was taken from left ovary. Post operatively course was uneventful and she was discharged. Histopathology report suggested ovarian steroid cell tumor. No mitotic figures were demonstrable. Immunohistochemistry revealed positive result for Inhibin, Calretinin and Vimentin. Patient is adviced to follow up regularly with seum testosterone levels to evaluate for any recurrence.

III. Discussion:

Steroid cell tumours are described under sex cord stromal tumours. These tumors are divided into three subtypes according to their cells of origin: stromal luteoma, leydig cell tumor and steroid cell tumor, not otherwise specified (NOS). Among these subtypes, the steroid cell tumors, NOS constitute for about 56% of steroid cell tumors.[4] The incidence of steroid cell tumors, NOS is highest in women belonging to child bearing age group, especially the third and fourth decades, but in rare circumstances postmenopausal women or children may also get this tumor as seen in our case. Commonly, androgenic manifestations in form of virilisation particularly hirsutism are seen in these tumors as they secrete hormones like androstenedione, α-hydroxyprogesterone, and testosterone.[5] So, in cases wherein there is unexplained hirsutism, ovarian and adrenal tumour association should be ruled out as there may be occult malignancies. [6] However, there may be atypical presentations of these tumours as seen in our case when they do not show any symptoms of virilisation. In these cases, the diagnosis is usually made postoperatively on finding a tumour in ovary as seen in our case.

Macroscopically, steroid cell tumours are mostly yellow orange, indicating their high lipid content, with hemorrhagic and cystic areas rarely seen. The tumour is commonly confined to the ovary, but in few cases extra ovarian steroid cell tumour have been documented.[7]

Definitive diagnosis is arrived by histology. Microscopically, the neoplastic cells have abundant clear vacuolated cytoplasm with rich vascularity, and absence of atypia or necrosis[7]

Steroid cell tumors NOS must be differentiated from other ovarian tumors and other steroid cell tumor types, in which there is proliferation of steroid hormone-producing cells which occur as a secondary event. It includes stromal luteoma, Leydig cell tumor, luteinized thecomas, pregnancy luteomas, and carcinomas. [8] Steroid cell tumors NOS are differentiated from Leydig cell tumors because cytoplasmic Reinke crystals are deficient. Also, usually Leydig cell tumor is located in hilar location and it is frequently associated with Leydig cell hyperplasia. Stromal luteoma is restricted to the ovarian stroma and commonly arise in association with stromal hyperthecosis. The steroid cell tumors NOS may have a fibromatous component like that of thecoma, but this component accounts for less than 10% of the tumor. Pregnancy luteomas are more frequently multifocal, bilateral in one-third of cases, are usually found at the time of Caesarean section, and often regress after pregnancy. [8] In our case, characteristics like stromal hyperthecosis, pseudovascular spaces or Reinke crystals were not present, to suggest the possibility of stromal luteoma and Leydig's cell tumor, and thus, this case was described as steroid cell tumor- NOS.

Figure 1: Haematoxylin and Eosin staining of the surgical specimen showing hyperchromatic nuclei. Cytoplasm is abundant clear and eosinophilic.

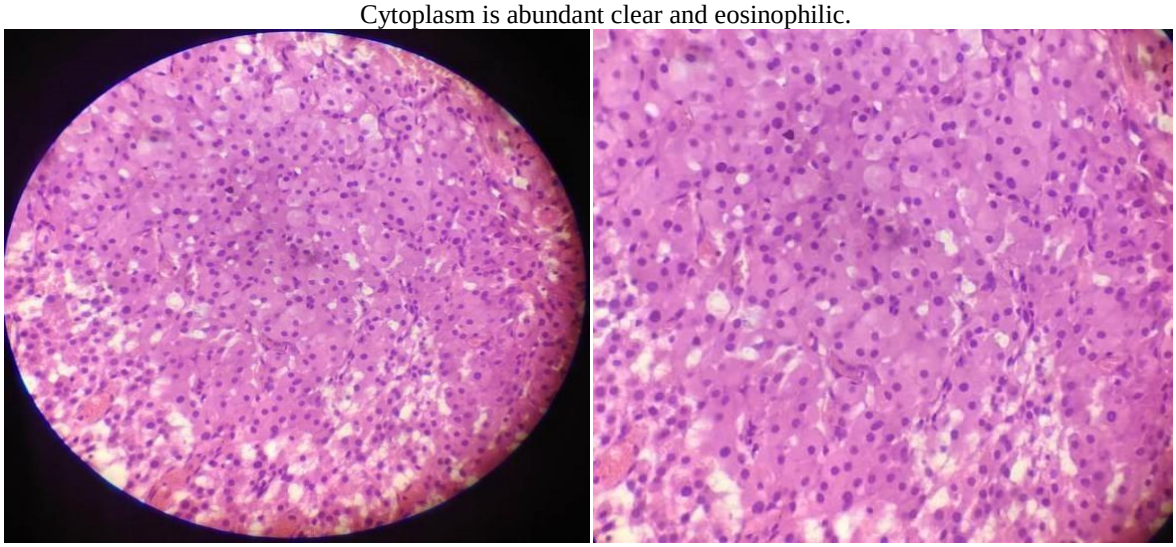
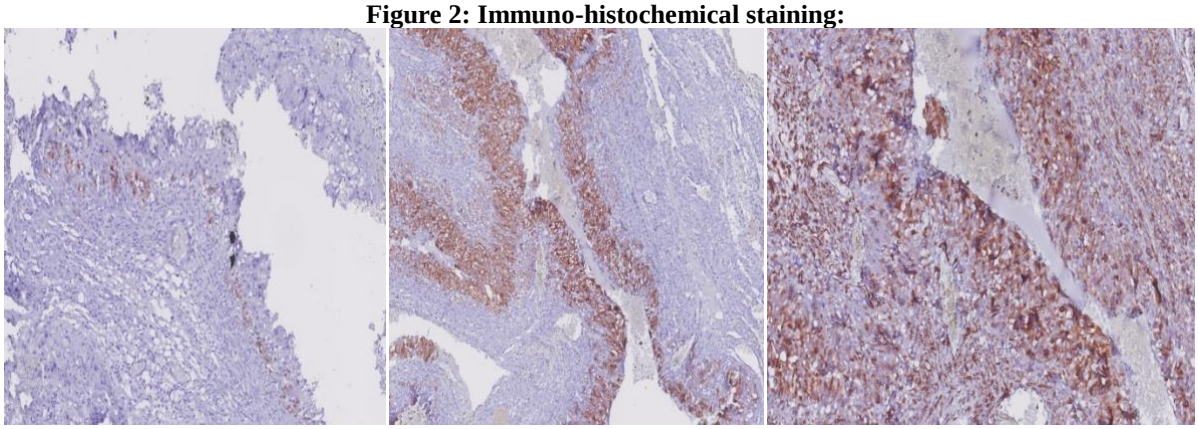


Figure 2: Immuno-histochemical staining:



a) Calretinin b) Inhibin c) Vimentin

In addition to the microscopic features, immunohistochemistry is very useful in coming to diagnosis of these tumors correctly. Calretinin and inhibin stain are identified to be most useful in distinguishing sex cord-stromal from non-sex cord-stromal tumors, because the first is positive for these two markers. [(1,3)] Mostly these tumors are benign, however 25-43% of steroid cell tumors are clinically malignant, with 20% of cases exhibiting metastasis outside of the ovary. [(1,3)] Metastatic lesions commonly occur within peritoneal cavity and occassionally occur at distant sites. [(2)] A study by Hayes and Scully [(9)] documented five pathological features indicative of malignancy as shown in the table below:

Microscopic features	% chance of malignancy
1) Two or more mitosis per 10 high-power fields	92
2) Necrosis	86
3) Size of the tumor (more than 7cm)	78
4) Hemorrhage	77
5) Grade 2 or 3 nuclear atypia	64

In our case, there were no mitotic figures, necrosis was not seen, size of tumor was 5*5 cm, areas of hemorrhages were seen and no atypia was noted. Hence it was benign tumor.

The treatment is guided by age at presentation, clinical presentation, histological features and surgical staging and desire to preserve fertility.[(1,2)] In young age females who desire to preserve fertility, conservative surgery with unilateral oophorectomy can be accepted. [(2, 10)] As in our case patient was very young and hence ovarian cystectomy with biopsy of contralateral ovary was done. A case of laparoscopic cystectomy in steroid cell tumors was already described by Jiang et al. and reported success in 3 years of follow-up without evidence of recurrence of the tumor.[(1)] Similarly Patricia et al [(3)] reported a case wherein nulligravida patient underwent diagnostic laparoscopy as a part of infertility work up, wherein the tumor was an unexpected finding, and laparoscopic cystectomy was done which was found to be ovarian steroid cell tumor on histopathology evaluation and patient monitored with close surveillance of hormonal levels and imaging in the postoperative period. Patricia et al [(3)] believe that performing a cystectomy allows the preservation of the ovarian reserve, is important in a nullipara and probably it is a good approach in circumscribed tumors. For those who have completed childbearing and elderly patients , total hysterectomy, bilateral salpingo-oophorectomy and complete staging are indicated.[(2)]

IV. Conclusion:

Steroid cell tumors, NOS, are usually rare tumors. 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. Disease management should be decided based on tumor pathology, surgical staging and the desire for preserving fertility. The primary treatment is surgical removal of the primary lesion, and radiation or chemotherapy is not much effective.

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