Steroid Cell Tumor of Ovary: An Incidentally Diagnosed Steroid Cell Tumor of Ovary in a Patient of Carcinoma Endometrium: A Very Rare Finding

Authors
Dr Shreena Patidar¹, Dr Niketa Thakur², Dr Sonal Patel Shah³, Dr U. Suryanarayan⁴, Dr Kushboo Jain⁵
Corresponding Author
Dr Shreena Patidar

3rd year Postgraduate Resident, Gujarat Cancer Research Institute (GCRI) Ahmedabad Address- 6, Jamna Niwas, Opposite Gayatri Dairy, Vitthal Nagar Cross road, Shahibaugh, Ahmedabad. 380004, India

Introduction
Steroid cell tumor of the ovary are uncommon sex- cord stromal tumors of ovary with malignant potential. The incidence of steroid cell tumor of the ovary is only 0.1% of all ovarian tumors. There are three subtypes of steroid cell tumor of ovary. These are stroma luteoma, leydig cell tumors and steroid cell tumor NOS. As far as steroid cell tumors, not otherwise specified (NOS) is concerned; it constitutes about 56% of all steroid cell tumors. In literature only a few cases of steroid cell tumors, NOS, have been described. Most of the reported cases described benign cases.

Case Report
We are reporting a case of steroid cell tumor of ovary in a 56 year old postmenopausal woman. Patient presented with history of postmenopausal bleeding ingynaeology opd. There was no clinical abnormality on physical and local examination. CT scan of abdomen and pelvis was suggestive of lesion in endometrium and left adnexal region. There was lesion of size 26x39mm in endometrium involving myometrium to more than 50% thickness. There was also a 32x32mm lesion involving left adnexal region. Biopsy showed endometroid adenocarcinoma of endometrium, moderately differentiated. Patient was referred to gynaeology department where patient underwent staging laparotomy with total abdominal hysterectomy with bilateral salpingo- oophorectomy and bilateral pelvic lymph node dissection. Histopathology report showed endometroid adenocarcinoma of endometrium stageIA1. Tumor size was 5x4x1cm involving less than half thickness of myometrium wall. It was well differentiated that is grade I. Pelvic lymph node dissection showed no nodes were positive. Thirteen lymph nodes were dissected on right side whereas ten lymph nodes were dissected on left side. There was incidental finding of steroid cell tumor in left ovary of size 5x3x2cm. Immunohistochemistry report showed steroid cell tumor of ovary, NOS type. IHC markers vimentin,
calretinin, inhibin, Melan-A synaptophysin and AE1 were positive. Markers ENA, CEA, CK7, CK20, chromogranin and S100 were negative. In this case as ovarian tumor had benign features and endometroid adenocarcinoma is also stage IA1, so no adjuvant treatment is given and patient is kept on observation. Patient’s serum estradiol levels were in normal postmenopausal range. It was less than 12pg/ml.

Computed tomography axial section shows heterogeneously enhancing mass in left adnexa.

H&E stained section shows sheets of tumor cells having abundant eosinophilic granular cytoplasm with few intracytoplasmic vacuoles, hyperchromatic nucleus and a prominent nucleoli. No mitosis or necrosis seen.

Discussion
Steroid cell tumors of ovary are extremely rare tumors. There are three subtypes of steroid cell tumor of ovary. They are stromal luteoma, leydig cell tumors and steroid cell tumor NOS. Stromal luteoma arises from ovarian stroma, leydig cell tumors arise from leydig cells. Rest are steroid cell tumors NOS in which cell of origin is not specific. Most common subtype is steroid cell tumor NOS which accounts for 56% of all these tumors\(^1\). The incidence of steroid cell tumors, NOS is highest in women of child bearing age group, particularly during the third and fourth decades, but in rare cases postmenopausal women or children may also have this tumor. Clinical presentation of these tumors are non specific and include menorrhagia, postmenopausal bleeding, abdominal pain, abnormal growth of hair on body. Androgenic manifestations are common in these tumors as they secrete hormones like androstenedione, α-hydroxyprogesterone, and testosterone\(^2,3\). Estrogen secretion occurs in 6% to 23% of the tumors. Tumors associated with hormonal activity and virilizing properties have more significant presentation. Common signs are hirsutism, acne clitoral enlargement, sterility,
deepening of voice, increased libido and temporal alopecia. So in cases where there is unexplained hirsutism, ovarian and adrenal tumor association should be ruled out as there may be occult malignancies. However, there may be atypical presentations of these tumors also when they do not show any symptoms of virilisation\(^4\). In our patient there were no signs and symptoms of virilization or hormonal changes. In these cases the diagnosis is usually made postoperatively on finding a tumor in ovary\(^5\). The diagnosis of steroid cell tumor of ovary, NOS should be made on the basis of clinical virilizing syndrome, microscopic findings, immune reactivity to various immunohistochemistry markers. Inhibin and Calretinin are sensitive and robust markers in differentiating sex cord stromal tumors from non sex cord stromal tumors\(^6\). Our patient has positive Inhibin and calretinin stain which supported diagnosis of steroid cell tumors, NOS. The most important factor to be determined in steroid cell tumor of ovary is whether tumor has malignant features or not. Most of the steroid cell tumors (NOS) are benign, while malignancy has been reported in as many as 25% to 43% of cases\(^7\). According to Predictive pathological characteristics of malignancy for ovarian steroid cell tumors by Hayes and Scully, features favoring malignancy are Tumor diameter more than 7 cm, two or more mitotic figures per ten high power fields, necrosis, hemorrhage, grade 2 or 3 nuclear atypia. In our patient microscopic features did not reveal any prominent finding in favors of malignancy. There was no mitosis, no hemorrhage/necrosis, tumor size less than 7 cm and only grade 1 nuclear atypia\(^8\). Thus tumor has benign microscopic appearance. The mainstay treatment of these tumors is surgery. For women who do not desire future fertility, Total abdominal hysterectomy with bilateral salpingooophorectomy and complete surgical staging should be done\(^9\). Adjuvant chemotherapy is indicated for malignant steroid cell tumors\(^10\).

**Conclusion**

Steroid cell tumor of ovary are very rare tumors and are difficult to diagnose. Careful clinical history, physical examination laboratory test and Imaging studies are needed for making diagnosis. Regular follow up is necessary for detection of possible recurrences.

**References**

1. Baloglu A, Bezircioglu I, Cetinkaya B, Karci L, Bicer M. Development of secondary ovarian lesions after hysterectomy without oophorectomy versus unilateral oophorectomy for benign conditions: a retrospective analysis of patients during a nine-year period of observation. Clin Exp Obstet Gynecol. 2010;37:299–302.

2. Outwater EK, Wagner BJ, Mannion C, McLarney JK, Kim B. Sex cord-stromal and steroid cell tumours of the ovary. Radiographics. 1998;18(6):1523–46

3. Cserepes E, Szucs N, Patkos P, Csapo Z, Molnar F, Toth M, et al. Ovarian steroid cell tumour and a contralateral ovarian thecoma in a postmenopausal woman with severe hyperandrogenism. Gynecol Endocrinol. 2002;16(3):213–16.

4. Liu AX, Sun J, Shao WQ, Jin HM, Song WQ: Steroid cell tumors, not otherwise specified (NOS), in an accessory ovary: a case report and literature review. Gynecol Oncol 2005; 97: 260–262.

5. Rubido Valle CD, Fuente de la LJC, Martinez CS, Calvo LN, Escarda MR. Ovarian Steroid Cell Tumour Associated to Endometrial Hyperplasia and Presenting as Postmenopausal Vaginal Bleeding. Gynecol Obstet (Sunnyvale) 2015; 5(9):316.

6. Zhao C, Vinh TN, McManus K, Dabbs D, Barner R, Vang R: Identification of the most sensitive and robust immunohistochemical markers in different categories of ovarian sex cord-stromal
tumors. Am J Surg Pathol 2009, 33: 354–366.
7. Duncan TJ, Lee S, Acheson AG, et al. An ovarian stromal tumor with luteinized cells: an unusual recurrence of an unusual tumor. Int J Gynecol Cancer 2008;18:172–5.
8. Hayes MC, Scully RE. Ovarian steroid cell tumors (not otherwise specified). A clinicopathological analysis of 63 cases. Am J Surg Pathol 1987;11:835–45.
9. Swain J, Sharma S, Prakash V, Agrawal NK, Singh SK. Steroid cell tumour: a rare cause of hirsutism in a female. Endocrinology, diabetes and metabolism case reports. 2013;2013:130030.
10. Reedy MB, Richards WE, Ueland F, Uy K, Lee EY, Bryant C, van Nagell JR Jr: Ovarian steroid cell tumors, not otherwise specified: a case report and literature review. Gynecol Oncol 1999, 75:293–297.