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

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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 ingynaecology 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 gynaecology department where patient underwent staging laparotomy with total abdominal hysterectomy with bilateral salpingooophorectomy 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 5x3x2cm. tumor in left ovary of size Immunohistochemistry report showed steroid cell tumor of ovary, NOS type. IHC markers vimentin,

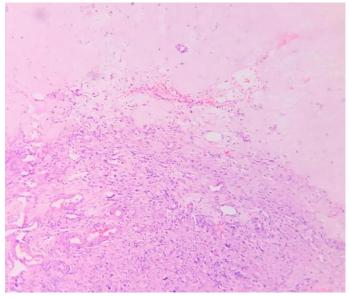
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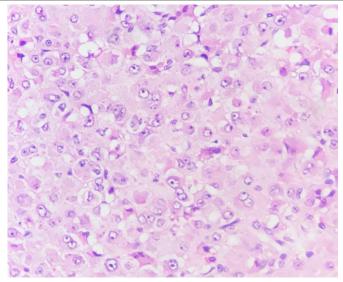
calretinin, inhibin, Melan-A synaptophysin and AE1 were positive. Markers ENA, CEA, CK7, CK20, chromogranin and S100 were negative. In this case as ovarian tumor had benign features and endometroid adenocarcinoma is also stage IA1, so no adjuvant treatment is given and patient is kept on observation. 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.



10x view of H&E stained section shows nests of tumor cells admist normal ovarian stroma.



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,

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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 7cm, two or more mitotic figures per ten high power fields, necrosis, hemorrhage, grade 2or 3 nuclear atypia. In our patient microscopic features did not reveal any prominent finding in favors of was malignancy. There no mitosis. no hemorrhage/necrosis, tumor size less than 7cm 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.

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