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Case Report

Rare ovarian tumor in post hysterectomized status: case report

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ABSTRACT

Ovarian tumors are known for their silent nature, non-availability of definitive screening methods and varied clinico pathological variations**.** Conservation of normal looking ovaries macroscopically during hysterectomy is recommended to benefit the future health of the woman. Still the development of new issues related to ovaries need to be counseled to the patient and stringent vigilance by the clinician is mandatory. Steroid cell tumor is rare ovarian tumor and histopathology is the gold standard for diagnosis. Any patient with high testosterone levels should be investigated meticulously to define the origin adrenal/ovarian with an awareness about this rare entity with malignant potential. Lifetime follow up of the ovaries in women are crucial at all ages. Here we report a case of rare ovarian tumor in post hysterectomized patient.

Key words: Ovarian tumor, Malignancy, Steroid cell tumor, Hirsuitism, Ovary

INTRODUCTION

Ovarian tumors are known for their silent nature, non-availability of definitive screening methods and varied clinic pathological variations. Conservation of normal ovaries macroscopically during hysterectomy is recommended to benefit the future health of the woman. Still the development of new issues related to ovaries need to be counseled to the patient and stringent vigilance by the clinician is mandatory. Development of ovarian neoplasm in these situations warrants thorough clinic pathological correlation not to miss the aggressiveness of the tumor. Steroid cell tumors are the terminology given to ones which are not characterized as Luteomas or Leydig cell tumor. They are coined as Steroid cell tumors not otherwise specified (NOS).[1] Here we report a rare ovarian tumor in post hysterectomized patient.

CASE REPORT

Mrs. A, 62 years old, multiparous woman presented with the complaint of vague lower abdomen on and off for one month duration., She has undergone Hysterctomy for (AUB-E) excessive bleeding 30 years ago. Bowel and bladder habits normal and no other associated complaints. She is a known case of Diabetes mellitus on treatment for 8 years. On examination obese, afebrile, comfortable, normotensive cardiovascular and respiratory system clinically normal. Abdominal examination revealed no area of tenderness or mass and vaginal vault healthy.

On evaluation normal blood counts, HbA1C 8.3 with normal thyroid, liver and renal function tests. MRI Pelvis with contrast and CT Abdomen screening revealed intensely enhancing lobulated solid lesion in the left adnexa with extensions with possibility of malignant neoplasm. CA 125 level normal ad Carcino embryonic

antigen level 5.44 ng/ml (CEA). Planned for surgery after counseling and arranging for frozen section. Abdomen opened hrough subumbilical midline incision and peritoneal lavage aone and sent for cytology.

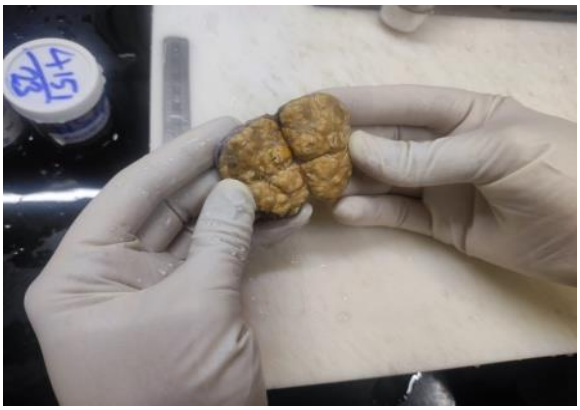

Figure 1: Steroid cell tumor-macroscopic appearance.

Bowel and omental adhesions present between abdominal wall and left adnexal region. Left ovary. replaced by a solid mass of 4x4 cm mass external surface is nodular and raw focally. Right ovary and other viscera normal. Proceeded to Left Ovariotomy and sent for frozen section. Right oopherectomy and Infracolic omentectomy done. Pelvic peritoneum removed and sent for HPE. Frozen section reports not suggestive of malignancy. Postoperative period uneventful. Patient discharged on 4th Postoperative day. Final HPE- revealed Steroid cell tumor NOS, with absent Reinke crystals, tumor cells with clear cytoplasm, powdery chromatin, trabecular pattern with thin blood vessels in between. No necrosis or nuclear atypia evidenced, sparse mitosis, no significant pathology in right ovary, unremarkable omentum and peritoneum and peritoneal washing negative for malignant cells. Follow up of the patient will be done periodically both clinically and radiologically.

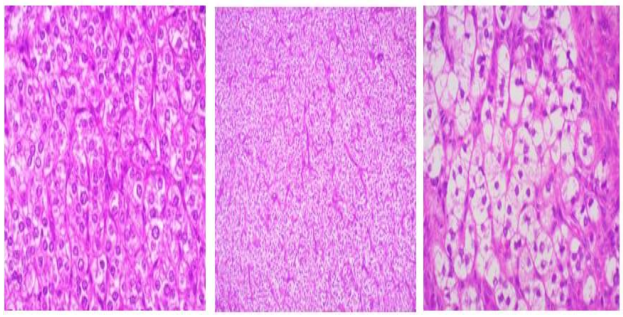

Figure 2: Steroid cell tumor-microscopic appearance.

DISCUSSION

Steroid ell tumor-NOS is a rare type of ovarian sex cord stromal tumor coined by Scully in 1979, constituting <0.1 % of ovarian tumors, previously coined as lipoid cell tumors and reclassified later.[2] In Scully series of 63 cases age distribution ranged from 2 to 80 years but the mean age of occurrence is 43 years and rare before puberty.[2,3] It is a paraneoplastic endocrinal tumor and more than 50% will present with hyperandrogenic features like Hirsuitism, hoarseness of voice, etc. 6% will present with estrogenic features and 25% are non-functioning tumors.[4,5] Our patient did not have any symptoms related to hormones and not evaluated preoperatively.

Diagnosis is by clinicopathological features with Tumor markers, testosterone levels, histology and immunohistochemistry. Imaging by MRI is useful in identifying the solid component distinctly. Most of them are benign but malignant behavior noticed in 25-40%.and only 6% of them are bilateral. Our patient had unilateral disease. Histologically solid, well circumscribed lobulated enlarged ovary, yellow-orange-red-brown tinge. Absence of Hyperthecosis, endovascular spaces, Reinke crystals will distinguish these tumors from Leydig cell tumors and Luteoma. In 2014 Ovarian sex-cord stromal tumors revised by WHO and regrouped into three clinicopathological entities-Pure stromal tumors, Pure sex cord tumors and Mixed Sex cord-stromal tumor. Steroid cell tumor belongs to pure stromal tumor group.

Pleomorphism, High mitotic activity and necrosis are suggestive of malignant nature of the neoplasm.[2] Five pathological criteria to predict maliganacy will be more mitotic figures/10HPF; 92%, necrosis (86%), Diameter >7 cm (78%), haemorrhage (77%) and grade2 &3 nuclear atypia (64%). In our patient no necrosis or nuclear atypia and sparse mitotic activity. In general, they are positive for Inhibin, Calretinin and sterogenic factor, Melan A and negative for FOX1.2.[7,8]

Radical Surgery is the treatment of choice and stringent postoperative vigilance. Recurrence of tumors in different organs as metastasis have been reported following surgery to consider the malignant potential of the tumor. Adjuvant chemotherapy using etoposide, bleomycin and cisplatin are tried.[9] GnRH agonists are used in case of recurrent tumors with careful follow up of sex hormone levels along with clinical surveillance.[9] If the patiens had features of hyperandrogenism or oestrogenic features initially, postoperative follow up with sex hormone levels are required.[10]

CONCLUSION

Steroid cell tumor is rare ovarian tumor and histopathology is the gold standard for diagnosis. Any patient with high testosterone levels should be investigated meticulously to define the origin-adrenal/ovarian with an awareness about this rare entity with malignant potential. Lifetime follow up of the ovaries in women are crucial at all ages.

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