University Journal of Surgery and Surgical Specialities

ISSN 2455-2860 2020, Vol. 6(2)

STEROID CELL TUMOR-NOS OF OVARY-A RARE CASE REPORT

DHIVYA C

Department of Obstetrics and Gynaecology, MADRAS MEDICAL COLLEGE AND GOVERNMENT GENERAL HOSPITAL

Abstract : Steroid cell tumors constitute only 0.1 percent of all ovarian neoplasms. They are classified as stromal luteoma, Leydig cell tumor and steroid cell tumors not otherwise specified. The first two categories are usually benign but some in the third group are malignant .We hereby are reporting a case of Steroid cell tumor Not Otherwise Specified (benign) in a postmenopausal women with complaints of lower abdominal pain and excessive growth of hair over the chin and upperlip with mild elevation of serum Testosterone(total) levels.

Keyword : Steroid cell tumor, Testosterone, Hirsutism

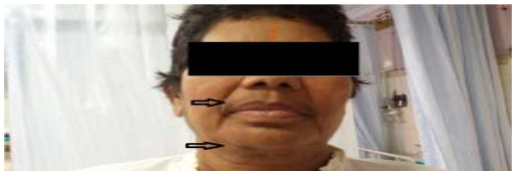
CASE SUMMARY

HISTORY:

A 54 year multiparous women married for 38 years and attained menopause 12 years back, came to our outpatient department with complaints of diffuse lower abdominal pain for the past 3 months was which dull aching type, gradual in onset, not progressive, not radiating, not associated with any other symptoms. History of shaving of hair over the chin and upper lip for the past 6 months was elicited .No associated change in voice, loss of weight, loss of appetite, breast secretion and no history of recent gain in weight. No history of post menopausal bleeding. There was no bladder and bowel disturbance .She has Type 2 Diabetes Mellitus for the past 4 years on Injection Insulin. No history of chronic drug intake for any other diseases. No history of previous surgeries other than puerperal sterilisation. She attained menarche at 13 years ,had regular menstrual cycles .Puerperal sterilisation was done 30 years back.

EXAMINATION:

On examination she was moderately built and moderately nourished with BMI 28 Kg per sq.m. Waist Hip ratio 0.7.Not anemic and no pedal edema. Terminal hair growth seen over chin and upper lip. No evidence of frontal balding or alopecia. Pubic and axillary hair appropriate for her age. Examination of thyroid did not reveal any abnormality. Breasts were atrophic. Temperature ,pulse and BP were normal. Cardiovascular, Respiratory and neurological system were normal. No supraclavicular or inguinal lymphadenopathy. Examination of external genitalia showed atrophic vulva and clitoris. Abdomen showed a 3cm transverse scar of puerperal sterilisation in hypogastrium , was soft on palpation, not tender ,no evidence of organomegaly. No abnormality was detected in Per speculum examination except for the postmenopausal changes. Bimanual pelvic examination showed atrophic uterus, left fornix fullness by a 4*5cm smooth, firm mass, with margins well defined and felt separately from uterus, right fornix was free, no fornicial tenderness. Digital rectal examination didnot reveal any abnormality.



Hirsutism(arrows showing growth of terminal hair over chin and upper lip)

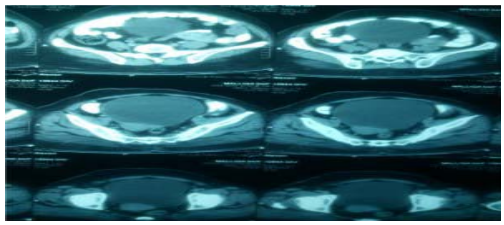
INVESTIGATIONS:

Complete hemogram, Liver function tests , Renal function tests and urine examination were normal .CA 125- 15.54 U/ml .Serum testosterone(Total) -90ng/dl .Serum DHEA and 17-OH Progesterone levels were normal for the age Xray chest normal VIA/VILI- Negative

USG :

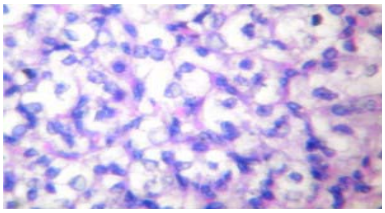
Ill defined hyperechoeic nodule in left adnexa of size 4 * 3 cm Right adnexa normal Uterus atrophic Abdominal organs normal

CECT :



Cyst of size 4.5*3.2 cm from left ovary with solid component which enhances on contrast. Right ovary-normal.Other abdominal organs normal. No Lymph node enlargement
Impression: **Complex Left Adnexal Mass probably malignant**
TREATMENT:
Staging laparotomy was done No ascites. Peritoneal washings were collected Intraoperative findings: Uterus atrophic 4*3 cm left ovarian mass, firm in consistency with intact capsule Right ovary and both fallopian tubes normal All abdominal organs normal Peritoneal biopsy was taken Staging Laparotomy was proceeded to Total Abdominal Hysterectomy with B ilateral Salphingo-oophrectomy with retro peritoneal lymph node dissection with infracolic omentectomy
GROSS APPEARANCE : CUT SECTION

Left ovarian tumor: Shows **solid yellow colored** areas No capsular extension. Right ovary- Normal Uterus, cervix, and both fallopian tubes- normal
HISTOPATHOLOGY:



Neoplasm arranged in sheets with discrete fibrous septae. Cells are oval to polyhedral with centrally placed densely stained nuclei. Abundant clear to eosinophilic cytoplasm. Few congested blood vessels are seen. No nuclear atypia or no reinke crystals seen. Peritoneal fluid shows reactive mesothelial cells in peritoneum. Peritoneum shows mature fibro fatty tissue with congested blood vessels. Lymph nodes shows reactive changes with no evidence of tumor deposits. Omentum showed no evidence of tumor cell deposits. Right ovary, uterus and Cervix-normal. Impression: **Steroid Cell Tumor Not Otherwise Specified** Post operative period was uneventful. Oncologist opinion: No need of Adjuvant Chemotherapy.
FOLLOW UP:
Patient was under regular follow up. Ultrsound of abdomen and pelvis done after 3 months was normal , Serum Testosterone levels decreased to 40ng/dl and no new hair growth over chin and upper lip.
DISCUSSION:
Steroid cell tumours are one of the rare tumors of the ovary accounting for approximately 0.1% of all ovarian tumours. They are ovarian neoplasms composed of steroid hormone-secreting cells. They have been divided into three subtypes according to their cell of origin: stromal luteoma arising from ovarian stroma, Leydig cell tumor arising from Leydig cells in the hilus, and steroid cell tumor not otherwise specified (NOS) when the lineage of the tumor is unknown (1). The last type accounts for approximately 60% of steroid cell tumors. They can occur at any age, but usually develop in adults with an average age of 43 years. Steroid cell tumours often present as unilateral solid tumours and occasionally as cystic tumours. The clinical presentations are not specific, including abdominal pain, distention, and bloating. However, the more significant presentations are those associated with the hormonal activity and virilizing properties of the tumor, which accounts for 56%-77% of patients (7). Signs and symptoms of masculinizing tumors usually take place in two definite phases, an early phase of defeminization and a subsequent phase of masculinization. Typically, a menstruating female will first notice oligomenorrhea or amenorrhea. Common signs of masculinization include hirsutism, acne, clitoral enlargement, increased libido, sterility, enlargement of the larynx, deepening of the voice, and temporal alopecia. Estrogen secretion occurs in 6% to 23% of the tumors, which may be associated with menorrhagia, postmenopausal bleeding, or even endometrial adenocarcinoma. Cushing's syndrome occurs in 6% to 10% of the cases .Approximately 25% of the cases of steroid cell tumours (NOS) are not associated with hormonal disturbances (16). These tumors were called *lipid cell tumors* and *lipoid cell tumors*, composed exclusively of large cells with abundant cytoplasm and central nuclei, resembling lutein, Leydig, and adrenocortical cells. But these terms are not appropriate for all the tumors in this category because approximately 25% contain little or no intracellular lipid and are red to brown on gross examination. The classical gross finding of steroid cell tumor (NOS) is a solid, well-circumscribed ovarian mass.

The cut surface may be yellow to orange if the cells are lipid rich, red to brown if the cells are lipid poor, or dark brown to black if large amounts of intracytoplasmic lipochrome pigment are present. Microscopic findings include diffusely arranged cells, although tumor cells may be present in nests, clusters, cords, or columns. The stroma is most commonly scant, but may be prominent and can occasionally be fibromatous, edematous, or myxoid. The cells are polygonal to round, with distinct cell borders and central nuclei, and they often have prominent nucleoli. The cytoplasm varies from spongy in lipid-rich cells to granular and eosinophilic in lipidpoor or lipid-free cells. Unlike Leydig cell tumors, steroid cell tumors (NOS) lack crystals of Reinke.(1)(2) usually positive for inhibin and calretinin though not specific. Inhibin and calretinin were thought to be sensitive and robust markers in differentiating sex cord-stromal from non-sex cord-stromal tumors (14). (17) In approximately one quarter of the cases, a small number of the neoplastic cells contain crystals of Reinke, identifying the tumor as a Leydig cell tumor. This tumor, which is almost always benign and typically secretes testosterone and virilizes the patient, has two possible origins. It may arise from hilus cells (hilar Leydig cells), which can be identified in the ovarian hilus in more than 80% of the female population.(18) The second origin is from ovarian stromal cells, which rarely exhibit focal differentiation into Leydig cells. When the tumor lies entirely in the hilus, it is usually referred to as a "hilus cell tumor." When it is confined within the ovarian stroma it is called "Leydig cell tumor, nonhilar type."

A second type of steroid cell tumor of identifiable origin is the stromal luteoma, which is composed of a uniform population of steroid-type cells and is situated within the ovarian stroma, often on a background of stromal hyperthecosis (stromal hyperplasia with the formation of lutein cell nests). Stromal luteomas are small benign tumors that are usually associated with estrogenic manifestations but may be nonfunctioning or occasionally androgenic. Steroid cell tumors not otherwise specified are often large neoplasms of which the site of origin is indeterminable because of their size and the absence of

identifiable crystals of Reinke in the cytoplasm of their cells. It is assumed that they arise either from the ovarian stroma or from hilus cells. These tumors are usually androgenic, but occasional examples are estrogenic or are associated with Cushing's syndrome. **Approximately 25% of the cases of steroid cell tumours (NOS) are not associated with hormonal disturbances (16)**. The differential diagnosis includes Leydig cell tumors, luteinizing granulose cell tumors, clear cell carcinoma, metastatic renal cell carcinoma The most important factor to be determined in steroid cell tumors of the ovary is whether the tumor has malignant features or not. Hayes MC1, Scully RE in their study found that the best pathological correlates of malignant behavior were: the presence of two or more mitotic figures per 10 high power fields (92% malignant); necrosis (86% malignant); a diameter of 7 cm or greater (78% malignant); hemorrhage (77% malignant); and grade 2 or 3 nuclear atypia (64% malignant).(1) Metastasis, however, is the only definite sign of malignancy. [2] At least 40% of steroid cell tumors not otherwise specified are clinically malignant.(1) in older women, the frequency of malignancy is greater after than before menopause**. Our patient's tumor did not show any of the above said histologic criteria for malignancy**. The mainstay treatment of ovarian steroid cell tumor is surgery. In general, conservative surgery with unilateral oophorectomy and proper staging should be performed in women with stage I disease who desire future fertility. For women who have completed childbearing, total abdominal hysterectomy with bilateral salpingooophorectomy and complete surgical staging is indicated [15]. Adjuvant chemotherapy should be based on the histologic appearance of the tumor and on its surgical stage. However, there are no well defined chemotherapy guidelines for clinical management of steroid cell tumor. PVB (cisplatin, vincristine, and bleomycin) or BEP (bleomycin, etoposide, and cisplatin) was recommended by some authors [15].

CONCLUSION:

Steroid cell tumors, NOS, are rare ovarian tumors . Careful history and physical examination, in addition to laboratory values and imaging studies, are helpful in making the diagnosis. These tumors should be considered a cause of isosexual precocious puberty in children and virilization in adults though about 25% of them are not associated with hormone secretion. Any patient who presents with virilism should be investigated systematically to determine if the high testosterone levels are of an adrenal or ovarian origin. Therapy should be individualized based on tumor histology, surgical staging, and the desire for fertility preserving. Mainstay of treatment is surgery. Recurrence and metastasis are rare. Malignant steroid cell tumors, NOS, should be managed with surgical removal followed by combination chemotherapy. Follow up evaluation should include sex hormone levels if it was elevated before the removal of tumor.

REFERENCES:

1. Hayes MC, Scully RE: **Ovarian steroid cell tumors (not otherwise specified): a clinicopathological analysis of 63 cases.** Am J Surg Pathol 1987, **11:** 835–845.
2. Zhang X, Lu B: **Ovarian steroid cell tumor, not otherwise specified (NOS): an unusual case with myelolipoma.** Int J Gynecol Pathol 2011, **30:** 460–465.
3. Lee SH, Kang MS, Lee GS, Chung WY: **Refractory hypertension and isosexual pseudoprecocious puberty associated with renin-secreting ovarian steroid cell tumor in a girl.** J Korean Med Sci 2011, **26:** 836–838.
4. Sawathiparnich P, Sitthinamsuwan P, Sanpakit K, Laohapensang M, Chuangsuwanich T: **Cushing's syndrome caused by an ACTH-producing ovarian steroid cell tumor, NOS, in a prepubertal girl.** Endocrine 2009, **35:** 132–135.
5. Flynn A, Ravindranath S: **Steroid cell tumor, NOS, of ovary.** Indian J Pathol Microbiol 2008, **51:** 461.
6. Kim YT, Kim SW, Yoon BS, Kim SH, Kim JH, Kim JW, Cho NH: **An ovarian steroid cell tumor causing virilization and massive ascites.** Yonsei Med J 2007, **48:** 142–146.
7. 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.
8. Deavers MT, Malpica A, Ordonez NG, Silva EG: **Ovarian steroid cell tumors: an immunohistochemical study including a comparison of calretinin with inhibin.** Int J Gynecol Pathol 2003, **22:** 162–167.
9. Bas F, Saka N, Darendeliler F, Tuzlali S, Ilhan R, Bundak R, Gunoz H: **Bilateral ovarian steroid cell tumor in congenital adrenal hyperplasia due to classic 11beta-hydroxylase deficiency.** J Pediatr Endocrinol Metab 2000, **13:** 663–667.
10. Faraj G, Di Gregorio S, Misiunas A, Faure EN, Villabrile P, Stringa I, Petroff N, Bur G: **Virilizing ovarian tumor of cell tumor type not otherwise specified: a case report.** Gynecol Endocrinol 1998, **12:** 347–352.
11. Wang PH, Chao HT, Lee RC, Lai CR, Lee WL, Kwok CF, Yuan CC, Ng HT: **Steroid cell tumors of the ovary: clinical, ultrasonic, and MRI diagnosis–a case report.** Eur J Radiol 1998, **26:** 269–273.
12. Outwater EK, Wagner BJ, Mannion C, McLarney JK, Kim B: **Sex cord-stromal and steroid cell tumors of the ovary.** Radiographics 1998, **18:** 1523–1546.
13. Lin CJ, Jorge AA, Latronico AC, Marui S, Fragoso MC, Martin RM, Carvalho FM, Arnhold IJ, Mendonca BB: **Origin of an ovarian steroid cell tumor causing isosexual pseudoprecocious puberty demonstrated by the expression of adrenal steroidogenic enzymes and adrenocorticotropin receptor.** J Clin Endocrinol Metab 2000, **85:** 1211–1214.
14. 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.
15.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.
16.Malaika W, Natarajan Sathima. Pathologic Quiz Case: A Rare Tumor of the Ovary. Arch Pathol Lab Med. 2003;127:890–892.
17.Young, R, Scully, R, Glob. libr. women's med., (ISSN: 1756-2228) 2008; DOI 10.3843/GLOWM.10249
18.Paraskevas M, Scully RE: Hilus cell tumor of the ovary. A clinicopathological analysis of 12 Reinke crystal-positive and nine crystal-negative cases. Int J Gynecol Pathol 8: 299, 1989

An Initiative of The Tamil Nadu Dr. M.G.R. Medical University
University Journal of Surgery and Surgical Specialities