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Uncommon cause of secondary amenorrhea and hirsutism: Steroid cell tumor of ovary (case report)

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Abstract: *Ovarian steroid cell tumors are rare functioning sex cord stromal tumors. They accounts <0.1% of all ovarian tumors. A subtype of this tumor also known as not otherwise specified (NOS) that accounts for approximately one-half of all the steroid cell tumors. Approximately one-third of steroid cell tumors in adults have been reported to be malignant. Previously, these tumors were referred to as lipid or lipoid cell tumors of ovary. The mean age of presentation of these tumors at around 40 years. They are usually small (<3-cm) nodules and virtually always unilateral.*
Herein, we are reporting a case of an 18 year old female with NOS subtype of steroid cell tumor, who presented with hirsutism and virilisation.

I. Case presentation

An 18 years old unmarried girl presented with complaints of pelvic pain, acne, progressive deepening of voice, hirsuitism, an increase in facial [Figure-1] and body hair, and oligomenorrhea for 3 years and amenorrhea from last 6 months. There was no history regarding anorexia and weight loss. Her medical and family history was unremarkable. Patient attained menarche at the age of 14 years, and had regular menses with interval of 28-31 days lasting 3 to 4 days with regular flow.

On physical examination, the patient was moderately built with body weight of 56 kg. Patient had a low piched female voice, male pattern of facial and pubic hairs, clitoromegaly and a firm and mobile solid mass in the left adnexa.

She has been amenorrheic for the past six months. There was no history suggestive of hyperprolactinemia or hypothyroidism. Her medical and family history was not significant.

Laboratory tests showed elevated serum testosterone of 9.26 ng/mL (normal range 0.2-1.2 ng/mL) and normal CA-125 of 17.16 U/mL (normal range <35 U/mL). All other laboratory tests including complete blood count (CBC), renal, bone, hepatic and coagulation profiles, alkaline phosphatase, follicle stimulating hormone (FSH) and luteinizing hormone (LH) levels were within the normal range.

Further, the patient was referred for imaging workup to our department. Ultrasound showed a large, well-circumscribed, well-marginated heterogeneous hypoechoic predominantly solid lesion with few cystic areas in left adnexa with internal vascularity on color Doppler [Figure-2]. The right ovary and the uterus were normal.

Nonenhanced magnetic resonance imaging (MRI) of the pelvis - T2-weighted images showed heterogeneous signal intensity lesion with few central cystic areas in the left adnexa. The lesion showed multiple signal voids. The uterus and right ovary appeared unremarkable [Figure-3a]. The T1-weighted image demonstrated isointense signal and multiple signal voids [Figure-3b]. On contrast enhanced MRI left ovarian mass shows intense contrast enhancement [Figure-3c].

On the basis of the above clinical, biochemical and imaging findings, a diagnosis of androgenic neoplastic primary ovarian tumor was made. Sertoli stromal tumors were kept First differential diagnosis and second were steroid cell tumors, because sertoli-stromal tumors are more common virilising tumor incidence wise and most common age of presentation is less than 30 year.

The patient underwent for exploratory laparotomy. The left ovary was replaced by a large capsulated solid grey-white mass [Figure-4A]. The right ovary and uterus were found unremarkable. Left salpingo-oophorectomy was performed. The specimen was sent for histopathological examination.

A cut-section of the specimen revealed solid mass with mutiple nodular yellowish areas [Figure-4B]. The histopathological examination disclosed granular to eosinophilic tumor cells and a clear appearance, with a moderate amount of cytoplasm. Few tumor cells had a vacuolated clear appearance suggestive of NOS subtype of steroid cell tumor [Figure-4C].

Final diagnosis suggested benign steroid cell tumor (Not otherwise specified) on the basis of histopathology. Tumor Capsule was intact and there was no evidence of vascular invasion. Peritoneal fluid

cytology did not show any malignant cells. Intraoperatively, lymph nodes were not significant. Thus, tumor was staged T1N0M0.

Serum testosterone level of the patient was 0.185 ng/ml during the first month of follow up. Her menstrual cycles were resumed within 2 months of the surgery. Four month postoperatively, ultrasonography of abdomen and pelvis areas was performed and no abnormal finding was observed.

II. Discussion

Ovarian steroid cell tumors are quite rare functional sex cord stromal tumors. They account for <0.1% of all ovarian tumors [1]. Sex cord stromal tumors are derived from the sex cord and stromal components of the developing gonads. Steroid cell tumors are often virilizing, frequently secreting testosterone; However, they may be endocrinologically inactive or estrogenic. As many as 25% of steroid tumors exhibit malignant behaviour [2].

Steroid cell tumors are further sub-grouped into stromal luteoma, Leydig cell tumors and steroid cell tumors that are not otherwise specific [3].

Steroid cell tumors; NOS, usually occur in adults with an average age at the time of diagnosis is of 47 years [4]. The clinical presentations are not very specific. Most of the patients (56%-77%) present with the symptoms which are associated with the hormonal activity and virilizing properties of the tumor [5].

The masculinizing tumors often have two definite phases of signs and symptoms, first early phase of defeminization and followed by phase of masculinization. As obvious a menstruating female will first notice oligomenorrhea or amenorrhea. There is devolution of the breasts and external genitalia, atrophy of the uterus and adnexa, and loss of the female body contour. This is followed by acne, hirsutism, clitoral enlargement, increased libido, sterility, enlargement of the larynx, deepening of the voice, and temporal alopecia [6, 7].

With regard to blood investigations, detecting the source of the androgenic tumor is a process of exclusion. In our patient, the high serum testosterone confirmed the presence of a virilizing neoplasm.

On radiological imaging, appearances of virilizing tumors of the ovary depend, to some extent, on the tumor type. Virilizing steroid cell tumors of the ovary are mostly one sided and usually very small, appears only mild bulky than the normal ovary [6, 7]. They are mostly confined to the ovary when become symptomatic, predominantly solid, noncalcified, and not associated with ascites. Small steroid cell tumors have been described as slightly hypoechoic or hyperechoic relative to the ovary with high diastolic flow on Doppler imaging. They may be difficult to identify on radiological imaging because they are small in size and isoechoic to the uterus on ultrasound and isoattenuating on CT [8, 9]. However MRI with phased array coils and color Doppler imaging can detect smaller tumors than more conventional imaging methods [8, 10].

The histological and /or imaging criteria for malignancy are grade 2 or 3 nuclear atypia, tumour diameter (> 7 cm), mitotic rate (> 2 high-power fields), necrosis and haemorrhage. Capsular penetration and vascular invasion also point to malignancy, although metastasis is the only definite sign of malignancy [1, 11].

The first line treatment is surgery. In older women, total abdominal hysterectomy and bilateral salpingo-oophrectomy are the adequate management options, while in young women; unilateral salpingo-oophrectomy is appropriate in most cases if histology shows no malignant features. Regular follow up evaluation with measurement of serum testosterone level is mandatory [12]. Malignant NOS steroid cell tumors should be followed by a combination of chemotherapy and radiotherapy after surgical removal [12]. Most of these tumors are diagnosed in early stage due to signs and symptoms and do not metastasize. So, the therapeutic value of chemotherapy and radiotherapy is controversial [12].

III. Conclusion

Steroid cell tumors, NOS, are rare ovarian sex-cord tumor of the ovary with good prognosis postoperatively. Therefore, imaging findings may help in the early detection and preoperative differentiation and staging of steroid cell tumor in young females. A patient who presents with secondary amenorrhea and hirsutism should be investigated systematically to figure out if the high testosterone levels are of an adrenal or ovarian origin its management depends on the degree of differentiation and staging of tumor, which mostly depend on histopathology.

Teaching point

Ovarian steroid cell tumors are rare functioning sex cord stromal tumors usually presents as secondary amenorrhea and hirsutism should be investigated systematically. Nevertheless, they should be included in the differential diagnosis when an adult female presents with secondary amenorrhea and hirsutism. Imaging and histopathology findings help in staging and the degree of differentiation of tumor, which play an important role in management.

IV. Consent

Written informed consent was obtained from the patient for publication of this Case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

V. List of abbreviations

SCT: Steroid cell tumor; NOS: Not otherwise specified CT: Computer tomography; FSH: Follicle stimulating hormone; LH: luteinizing hormone; MRI: Magnetic resonance imaging

Conflicts of interest

The authors declare that they have no competing interests.

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Figure & figure ligends:-

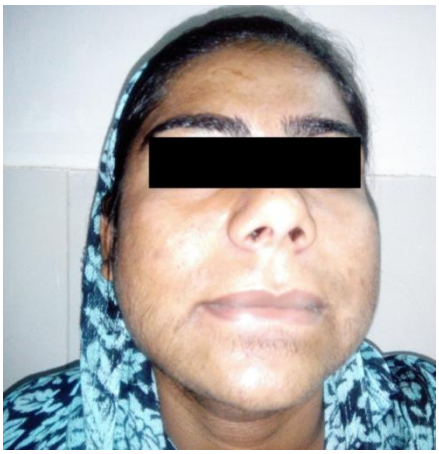

Figure 1- growth of coarse hairs over the face in 18 year old girl

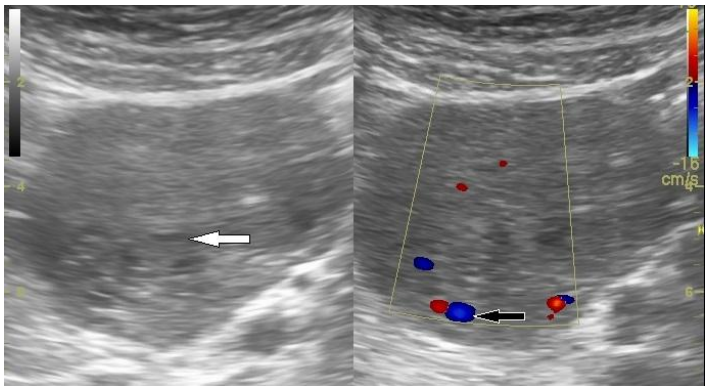

Figure 2- Ultrasound shows heterogenous hypoechoic solid mass with few cystic areas (white arrow) in the left adnexa whereas the left ovary is not visualized separately; on color Doppler ultrasound, the left adnexal lesion shows internal vascularity (black arrow)

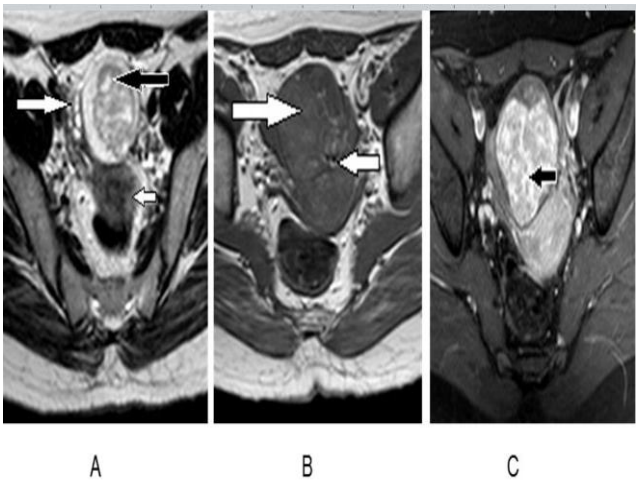

Figure 3A, 3B, 3C-
(A) MRI pelvis-T2W axial section large, well-circumscribed heterogenous signal intensity mass (black arrow) in the left adnexa, right ovary (large white arrow) and uterus (small white arrow) are normal.
(B) MRI- T1W axial section, the left adnexal mass shows isointense signal intensity (large arrow) and multiple signal voids (small arrow).
(C) Contrast enhanced MRI mass shows intense contrast enhancement (black arrow).

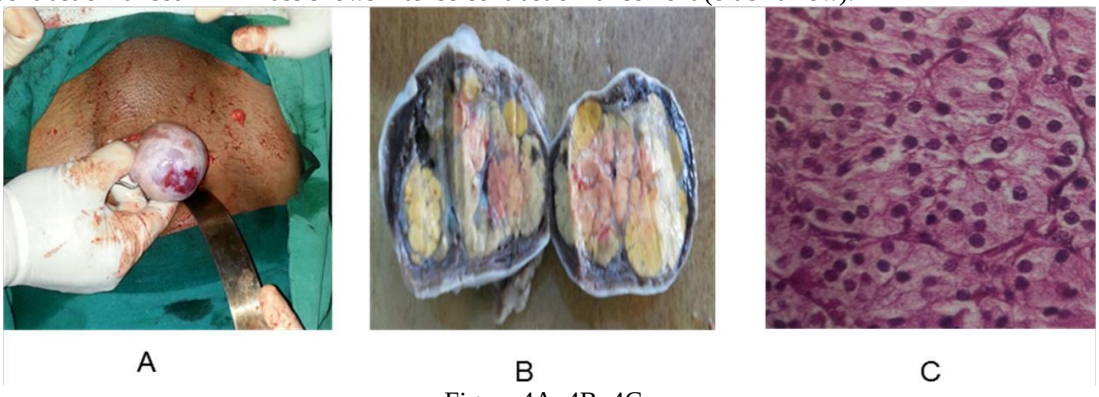

Figure 4A, 4B, 4C-
(A) Intraoperative highly vascular large capsulated solid greywhite mass lesion.
(B) The cut-section of the specimen shows a yellow nodular appearance
(C) Histopathology of the tumor showing vacuolated clear cells suggestive of NOS subtype of steroid cell tumor.