A rare cause of hirsutism

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Clinical Presentation

Presenting complaints and past medical history

50 yrs old lady, Deepening & hoarseness of voice since 18 months - 2yrs
Excess facial hair, coarse & thick hair on chin, chest & back – 6 months or so
Non specific Lt sided chest pains
Excess hair since 1 ½ yrs, shaving regularly, tried laser treatment privately, on & off lower abdominal pains since 6 – 8 months. Loss of appetite & lost 1 stone in 6 weeks, Ex smoker, No regular medications. No history suggestive of Cushing`s syndrome or thyroid problems. Endometrial ablation for menorrhagia – 2008
No significant family history.

Examination

Wt 80 Kg, BP- 130/80, Mild hoarseness of voice
Evidence of shaved hair on face, chin, upper chest & back, some over abdomen mostly terminal hair; Modified Ferriman Gallwey scale > 15 (Normal < 6-8).
No striae/neck hump/moon face/easy bruising signs, No temporal hair recession
No muscular or psychological changes, On systemic examination- no abnormality
PA- No palpable mass.

Differential Diagnosis

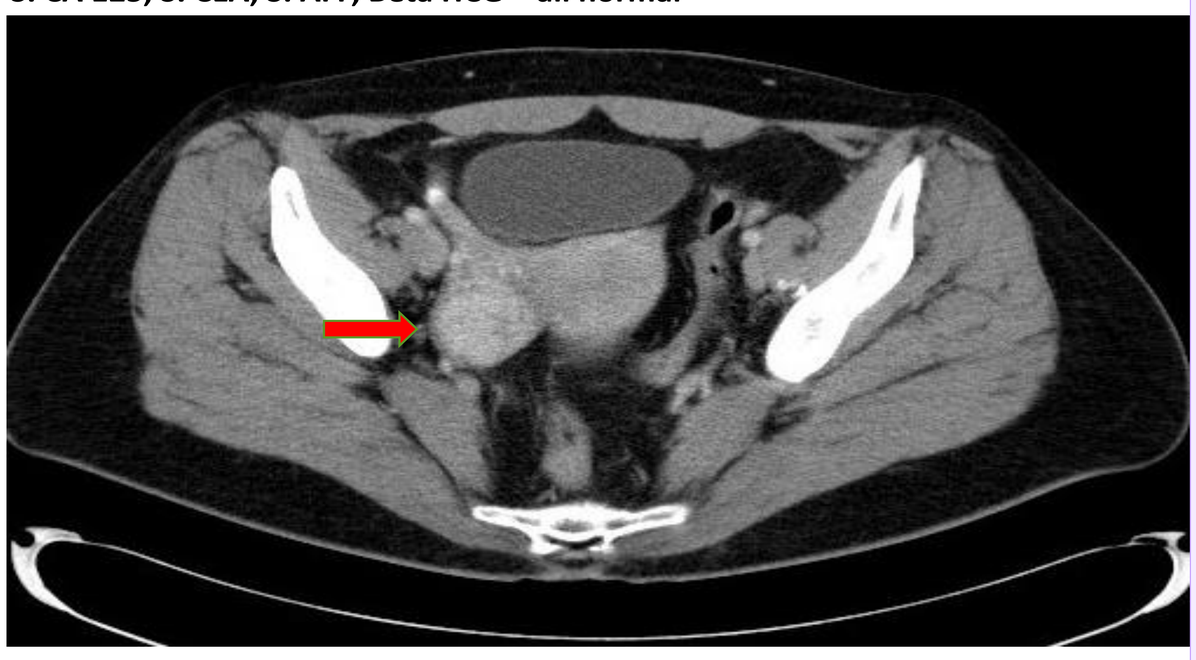
Middle aged lady with hirsuitism & features of hyperandrogenism
Ovarian or adrenal tumor
Cushing`s syndrome
Drug induced – exogenous hormones
Idiopathic
PCOS, CAH, Porphyria unlikely here.

Investigations and Management

U/Es- creat. 91, e GFR-68, HbA1c- 37, TFTs- Normal
LFTs- normal, rest routine bloods- NAD
S. Testosterone – 20.1 (0-4.3 nmol/L),
SHBG- 61 (19- 117), DHEA – 2.7,
Androstenedione – 7.5 (0-6.0)
S. Free androgen index- 33 (0- 15)
FSH- 7.4, LH-7.4, S. Oestradiol (17 Beta)- 385 pmol/L,
S. CA 125, S. CEA, S. AFP, Beta HCG – all normal
Requested CT abdomen while waiting for results & OBGY Consult.

OBGY review

No clitoromegaly, Endometrial ablation in past, Fullness in Right Adnexa with mild tenderness, Cervix healthy, rest – NAD
Advised scans & tumour markers.
S. CA 125, S. CEA, S. AFP, Beta HCG – all normal



CT Abdomen & pelvis

Normal adrenals, Right ovarian mass- 3.5 x 3.4 cm, predominantly solid, small cystic component, Probably functioning ovarian tumour, NO Mets

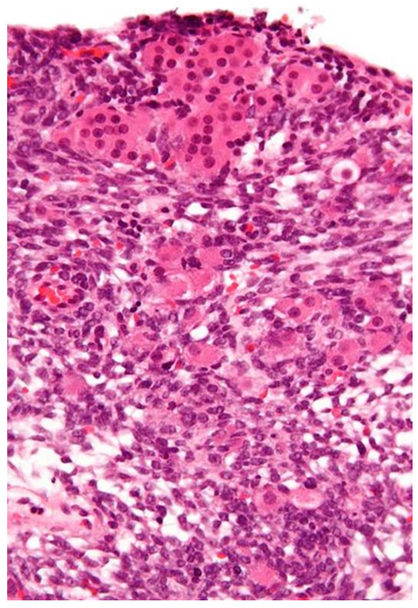
Treatment : Bilateral salpingo-oophorectomy

Rt. Ovarian mass- confirmed as,well differentiated Sertoli Leydig cell tumor, no serosa involvement, confined to body, no spillage (Stage Ia), Lt ovary normal
No further treatment required.

Post-OP Follow-up

Voice - mild improvement
Reduction in hair growth but mature hair persisted
S. Testosterone - < 0.45 nmol/L ,S. Free androgen index – 0.9
Vaniqua cream (Eflornithine) or laser on medical grounds after D/W GP
Symptomatic treatment for hot flushes.

Sertoli-Leydig Cell Tumours



The Leydig cells have abundant eosinophilic or light pink cytoplasm. The Sertoli cells have a pale/clear cytoplasm.

Leydig cells are named after German anatomist Franz Leydig, discovered in 1850.
Leydig cells secrete androgens testosterone, androstenedione and dehydroepiandrosterone (DHEA),when stimulated by LH

Sertoli cells are named after Enrico Sertoli Italian Physiologist discovered them in 1865.
Sertoli cells- Nurse cells for sperm production.
Secrete various chemicals like- AMH, Inhibin, androgen binding proteins, activin, estradiol, transferrin and various immunoregulatory molecules

Sertoli-Leydig Cell Tumours

Sex cord stromal tumours , Rare ovarian tumors (< 0.5%)
Mostly unilateral, testicular structure
1/3rd produce androgen, Virilisation according to androgen quantity
Mostly 2nd or 3rd decade, Rarely malignant
20 % show heterologus element eg. Bone,cartilage,muscle, glands, teeth etc.

Clinical presentation and Management of Sertoli-Leydig Cell Tumours

Clinical features

Non functional tumors- abdominal pains, swelling, avg. size 16 cm at presentation
Androgen secreting tumors – features of virilisation- oligomenorrhea, amenorrhea, breast atrophy, hirsuitism, deepening of voice, acne, male type baldness, clitorial enlargement
Renin – refractory hypertension & hypokalemia

Management

Surgery is the mainstay (TAH + BSO)
Depends on patients age & childbearing
Adjuvant chemo. - Only in Ic onwards, poorly differentiated, tumors with heterologous elements or metastatic tumors
NCCN recommends platinum based therapy, No standard regimen
BEP is most commonly used(Bleomycin, Cisplatin & etoposide).

Follow-Up

Physical examination & testosterone levels – every 3-4 months for first 2 years
Then every 6 months for next 3 years
Inhibin & AFP if initially elevated
US abdomen or MRI depending on symptoms & markers

Summary

Sertoli Leydig cell tumors are rare, mostly benign
1/3rd women present with virilisation
Diagnosis is by histology at surgical excision
Surgery is the mainstay of treatment, Platinum based chemo. as adjuvant

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