Sertoli-Leydig Cell Tumor as a rare cause of hirsutism in a young adolescent

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Nothing to disclose

Introduction

In girls the abrupt onset and rapid progression of hirsutism as well as additional signs of virilization are suggestive for an androgen secreting tumor from the adrenal gland or the ovary. There are three types of ovarian tumors that can cause hirsutism and virilization: Sertoli-Leydig Cell Tumor (SLCT), Lipoid Cell Tumor and Hilus Cell Tumor. SLCT's account only for 1 % of all ovarian neoplasia's and occur more commonly in the second or third decade.

Objective and hypothesis

To report the hormonal and biological profile of a SCLT in a young adolescent. Ovarian tumor markers screening as well as FDG -PET scanning might be helpful in diagnosing ovarian malignancy in case of normal ultrasound imaging.

Case Presentation

A 13 7/12 year old girl with unexplained hyperandrogenemia. Since menarche at 12 10/12 years, rapid hair grow over her chest, abdomen, back, arms and legs. Recently new hair growth occurred at the chin and upper lip. Menses were irregular and prolonged. No use of medication, dermal products or nutritional supplement. Family history was negative for consanguinity, infertility, rare tumors or multinodular goiter

Physical Examination

Weight: 63.2 kg, Height: 168.2 cm, BMI: 22.6 kg/m²
BP : 113/80 mmHg
A3P6M4
Voice deepening, Muscular build, Slight acne
Cliteromegaly
Excessive hair growth on the face, trunk and legs
(fig 1-3) Ferryman Galway score : 26

Hormonal analysis

testosterone : 425 ng/dL (RIA), **222** ng/dL (LC MSMS)
17OHProg : 4,6 ng/ml, Androsteendione : 4,35 ng/ml
cortisol :12 µg/dl, DHEAS : 297 µg/dl
AMH : 17.4 mcg/L, LH 7 U/L, FSH 4.9 U/L
estradiol : 40 ng/ml
Alfa fetoprotein : **268** mcg/L, Inh B and bhCG : normal
ACTH testing: normal cortisol and androgen response

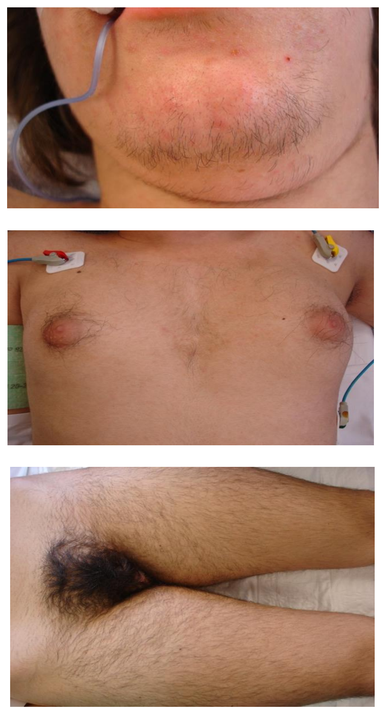

Figure 1-3:
Excessive hair growth

Genetic studies

Karyotype : 46, XX
Dicer1gene mutation screening: negative

Imaging

Pelvic US: normal sized ovaries, small cysts
CT adrenals: normal
FDG PET CT scan: oval mass with sharp borders with high peripheral FDG uptake in the right ovary
MRI: right ovarian mass with high intensity at T1& T2

Histology

Macroscopy: Solid ovarian mass, size 4 x 3 x 2,5 cm.
Microscopy: Tubular structures, with slightly atypical cells, including clusters of Leydig cells without atypia.
Histopathological and histochemical studies confirmed a **Sertoli-Leydig Cell Tumor (SLCT)**

Management

Unilateral right oophorectomy was performed laparoscopically. Serum AFP level was undetectable after tumor resection and serum testosterone declined to normal levels.

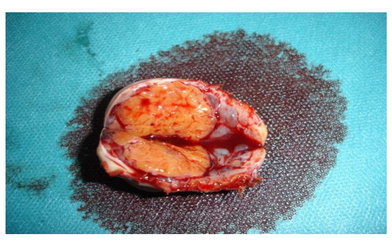

Figure 4: Yellow oval mass

Discussion

Adrenal causes of hyperandrogenemia were excluded by imaging and hormonal testing. The high/normal gonadotropin levels argue against the exogenous administration or cutaneous contact with testosterone. On the other hand, normal gonadotropin levels have been observed in female adolescents with testosterone producing tumors and are explained by a low degree of aromatization to estradiol. Diagnosing androgen secreting ovarian tumors can be a diagnostic challenge, since they are uncommon and difficult to detect at imaging and/or by venous sampling. An ovarian mass was detected by FDG PET CT scanning, avoiding venous sampling.

Conclusion

- Ovarian malignancy should be suspected in case of severe clinical features of hyperandrogenism and a very high (> 200 ng/dl) serum testosterone in hirsute adolescent girls.
- Screening for ovarian tumor markers as well as FDG-PET scanning are helpful in diagnosing ovarian malignancy.

References: Rohini Danya, Somanath Padhi, Renu G'Boy Varghese, C. S. Sertoli-Leydig Cell Tumor of ovary-a diagnostic dilemma, *Journal of Clinical and Diagnostic Research. 2014 Mar, Vol-8(3): 127-129*

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Gonads & DSD
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