

EP16.77 Rapid Dual Maternal and Fetal Virilization Revealing a Mischaracterized Steroid Cell Tumor in Pregnancy

Kylie R. Roslin BS, Christina Attia MD, Chitra Gotluru MD, Megan Piacquadio MD, Edna Kathleen Porter MD, MBA, Joseph Pearson MD, Wafa Khadraoui MD, Anna Sfakianaki MD, Rodrigo Ruano MD, PhD.
University of Miami Miller School of Medicine, Department of Obstetrics & Gynecology, Miami, FL

Introduction

- ❖ Virilization or male secondary sex characteristics in a genotypically XX patient
 - ❖ Most common etiologies: ovarian/adrenal pathology, pituitary dysfunction, or exogenous androgens.
- ❖ Ovarian sex cord tumors (OSCTs) account for 2–8% of ovarian masses and are typically hormonally active.
- ❖ Steroid cell tumors (SCTs) are rare (~0.1% of ovarian tumors), may co-secrete androgens and cortisol, and carry 25–40% malignancy risk.
- ❖ Dual maternal and fetal virilization is exceptionally rare, posing a significant diagnostic challenge.

Case Presentation

- ❖ 30-year-old G1P0 with primary infertility and obesity by embryo transfer with PGT-confirmed XX embryo.
 - ❖ Pre-conception workup noted a 6-cm pelvic mass, presumed to be pedunculated leiomyoma.
- ❖ At 15 weeks noted new onset acne, voice deepening, hirsutism, clitoromegaly, hypertension, hyperemesis, and cervical shortening.
- ❖ At 22 weeks elevation of all androgens critical transaminitis (ALT >1,000 U/L) and proteinuria (Table 1)
- ❖ MRI initially mischaracterized the mass as leiomyoma; repeat MRI showed 10.8×9.7×7.9 cm mass likely ovarian (Figure 1)
- ❖ Fetal ultrasound showed male-appearing external genitalia in XX fetus — concurrent with fetal virilization (Figure 2)
- ❖ **At 25w1d underwent laparoscopic left salpingo-oophorectomy and pathology showed a Stage IA steroid cell tumor, NOS (Figure 3)**
 - ❖ Rapid postoperative normalization with testosterone down to 79 ng/dL and transaminases by POD 6
- ❖ At 29w3d, she had PPROM with intermitted vaginal bleeding
- ❖ At 32w had vaginal delivery of 1.71 kg XX neonate with virilized phallus; APGAR 7/8/9
 - ❖ Neonatal labs were normal and neonatal pelvic MRI confirmed uterus and no testes
 - ❖ Neonate was discharged after 4-week NICU stay.

Table 1. Key Laboratory Values

Hormone	Normal Range	Pre-op	Post-op
Total Testosterone	< 50 ng/dL (pregnancy)	2,351 ng/dL	79 ng/dL
DHEA-S	35–430 µg/dL	Markedly elevated	89 µg/dL
Androstenedione	0.5–3.4 ng/mL	Markedly elevated	Normalized
Estradiol	< 3,000 pg/mL (2nd tri)	> 15,000 pg/mL	Normalized
Estriol (uE3)	2–12 ng/mL (2nd tri)	1.49 ng/mL (↓)	N/A
ALT	7–56 U/L	> 1,000 U/L	< 40 U/L (day 6)
AST	10–40 U/L	> 1,000 U/L	Normalized

Figures



Figure 1: T2-weighted MRI showing heterogeneous 10.8×9.7×7.9 cm left adnexal mass

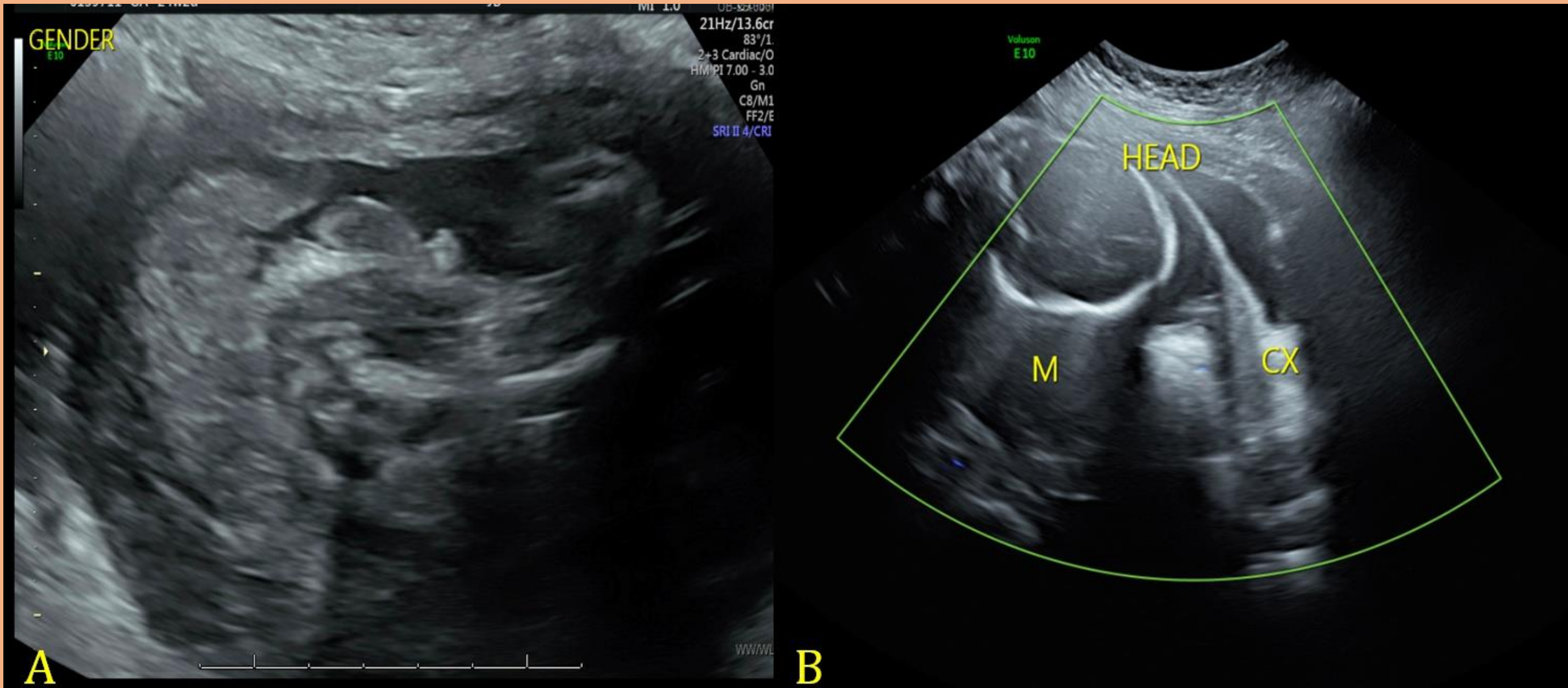


Figure 2: A. Fetal ultrasound demonstrating virilized external genitalia (XX karyotype) B. Maternal pelvis demonstrating the adnexal mass

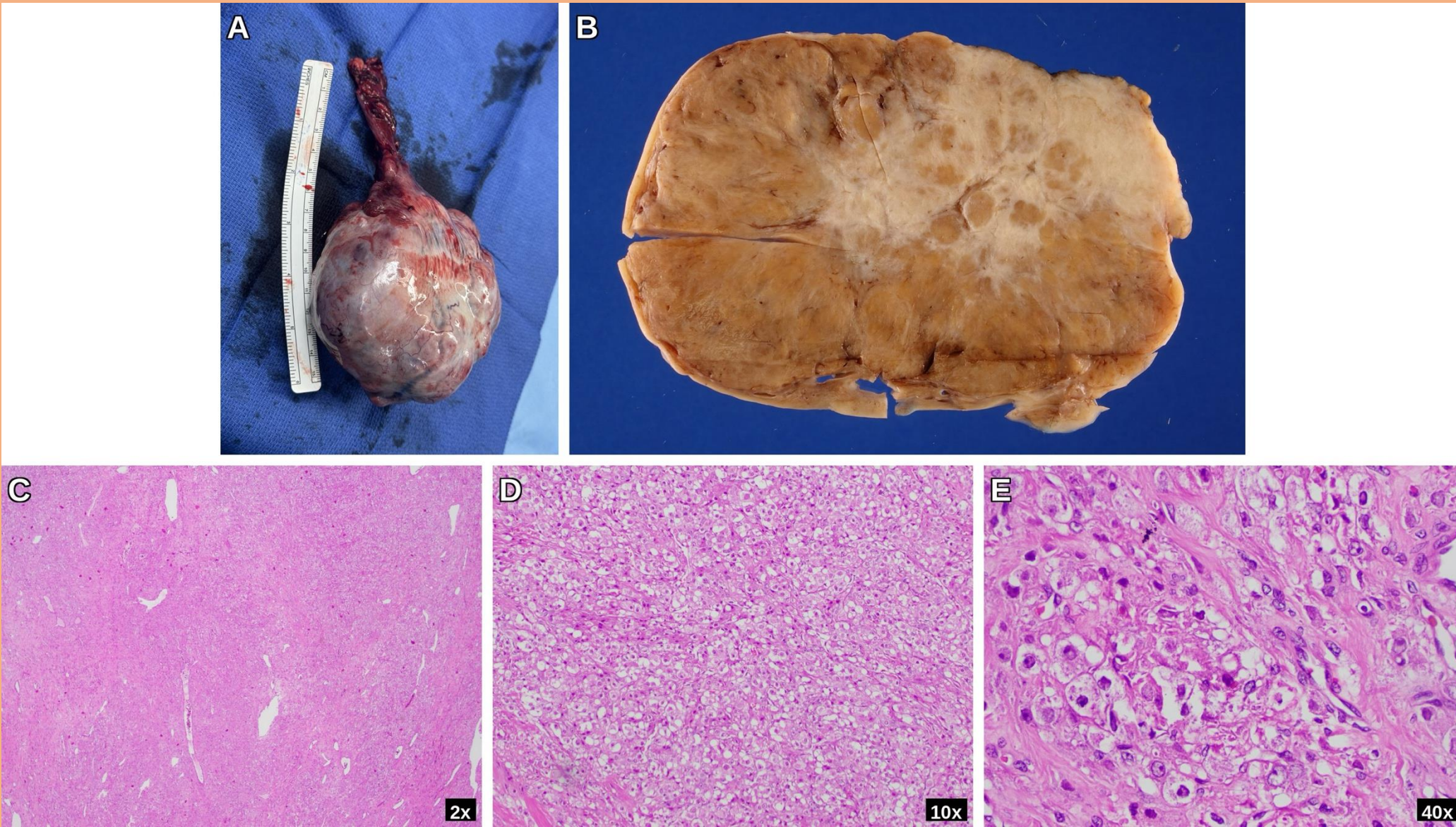


Figure 3A–B: Gross specimen (10.5×10×9 cm) and H&E histopathology (2× and 40×) C–D: IHC positive for SF-1, calretinin, inhibin; negative for CK, CD56, desmin

Discussion

- ❖ Placental aromatase and sex hormone binding globulin normally protect against hyperandrogenism in pregnancy
 - ❖ Fetal virilization signals this barrier was overwhelmed
 - ❖ Suspect testosterone exposure occurred during genital differentiation (7–12 weeks)
 - ❖ Prenatal androgen excess carries long-term offspring risks: metabolic syndrome, cardiovascular disease, autism spectrum disorder, ADHD
- ❖ Suppressed estriol (1.49 ng/mL) likely reflects functional aromatase saturation from massive androgen overload, not a genetic CYP19A1 mutation (carrier screening negative)
- ❖ Atypically elevated DHEA-S (classically adrenal) may reflect SCT origin from ectopic adrenal rests retaining SULT2A1 enzyme activity. However, magnitude substantially exceeds prior SCT reports
- ❖ Imaging mischaracterization (leiomyoma vs. ovarian mass) highlights limitations of morphology alone in hormonally active adnexal lesions
- ❖ Multiorgan involvement (transaminitis, hypertension, psychiatric worsening) reflects severe hypercortisolism from excess steroid hormone production
- ❖ Recurrence in Stage IA SCTs is rare (6.1%) but can occur up to 15 years post-resection
 - ❖ Long-term surveillance is mandatory

Conclusion

- ❖ This case represents a rare instance of dual maternal and fetal virilization secondary to a steroid cell tumor in pregnancy.
- ❖ Physiologic hormonal shifts, altered protein binding, and fetoplacental steroid metabolism can obscure pathologic androgen excess in pregnancy
- ❖ Timely multidisciplinary evaluation and surgical excision are essential when a structurally concerning adnexal mass coexists with severe gestational hyperandrogenism
- ❖ Delayed intervention risks irreversible fetal virilization and progressive maternal organ dysfunction.

References

