

Maternal ovarian luteoma causing complete virilization of a female fetus

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Background:

Maternal ovarian luteoma is a rare condition that may cause virilization of female fetuses.

Case Presentation:

We present the case of a baby born at 29 weeks GA to a 20 yo mother with history of 2 spontaneous abortions and no live births. Physical exam after birth revealed ambiguous genitalia, with a stretched phallic length of 1 cm with hypospadias on the ventral surface of phallus and complete fusion of rugated labioscrotal folds. (Figure 1) No palpable gonads in the folds or inguinal canal. Mother denied any medications during pregnancy. Mother was noted to have a low pitch voice and hirsutism. She reported that 3 months into her pregnancy, she noticed the appearance of facial hair and increased clitoral size. No family history of ambiguous genitalia. Baby had a 46XX female karyotype and negative SRY. A pelvic ultrasound showed normal uterus and ovaries. Baby's testosterone at birth was 71.6 ng/dL and at 2 weeks of age it was 32.6 ng/dL. Her 17 hydroxyprogesterone was high initially and later normalized. Her androstenedione was normal. Right after delivery, maternal testosterone was markedly elevated at 1867 ng/dL. Normal estradiol and DHEAS. Maternal pelvic US and MRI showed multiple leiomyomas and an oval shaped solid mass 31 X 41 mm within right adnexa with a cystic 19 X 24 mm lesion medial to the mass suggestive of a tumor. The mother was temporarily lost to follow-up. She returned for follow up two months after delivery. At that time her repeat testosterone was 13.7 ng/dL. Her hirsutism had improved and clitoral size had returned to normal.



Figure 1

Discussion:

We present the case of a female baby with complete virilization of the genitalia due to a presumed maternal ovarian luteoma. There are only other 3 cases reported, 2 of them twins, that presented with complete virilization. (1, 2) Other reported cases have partial virilization. The incidence of hyperandrogenism during pregnancy is low. Hyperandrogenism and virilization during pregnancy are usually the result of a disorder arising during gestation, as hyperandrogenism in a non-pregnant female frequently causes infertility.

Fetal conditions causing virilization, such as congenital adrenal hyperplasia, are more common than maternal disorders. Two of the most frequent causes of hyperandrogenism during gestation are luteomas and theca-lutein cysts of the ovary. (3) A luteoma is a rare ovarian mass that appears during gestation and regresses and disappears after delivery. About 25% of them secrete androgens. Many luteomas may be subclinical. Diagnosis is made only when the luteomas are large or cause virilization.(4) Maternal virilizing symptoms and signs may be present in approximately 35% of reported cases of pregnancy luteoma, and around 80% of female infants born to virilized mothers are virilized.(5) The degree of fetal virilization depends on the timing of the increased maternal androgen production and severity. Early gestation androgen exposure is needed for severe virilization, as in our case.

References:

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