

# Neonatal Ovary tumor

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# Clinical History

- Prenatal diagnosis (27 WG) macrosomia, clitoromegaly, labial hypertrophy and 2cm cystic left ovary mass
- Delivery (41 WG) by caesarean section

# Clinical data

- External genitalia : virilized aspect with clitoral enlargement, hypertrophy of labia majora
- No inguinal gonads were noted.
- Pelvic and abdominal ultrasound scan (US) showed a uterus and two apparently normal ovaries.

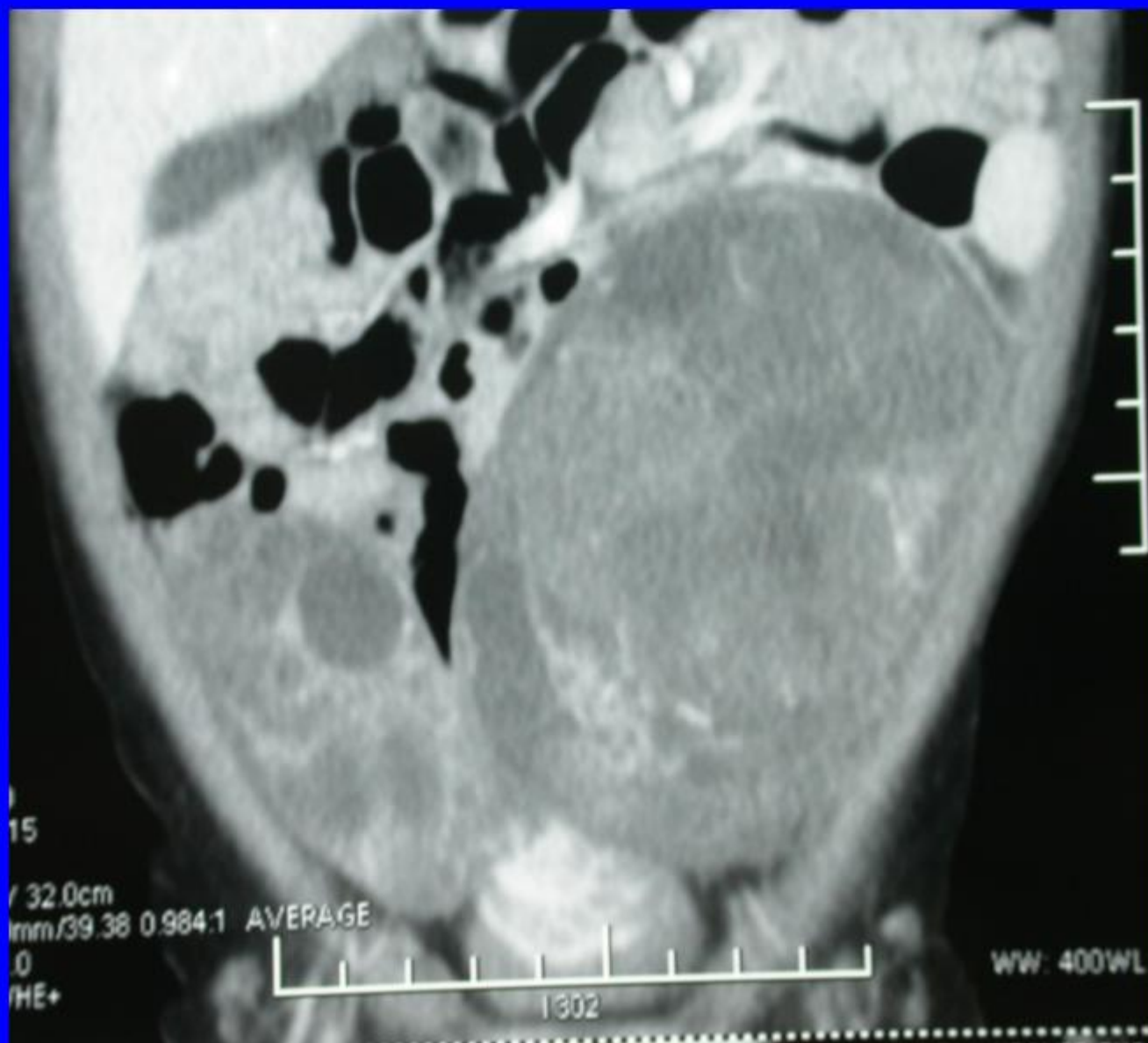
# *1<sup>st</sup> period*

## *( birth - 5months )*

hCG stimulation was done for endocrine diagnosis

- 2 injections induced: isosexual precocious puberty, breast development, uterine bleeding, pubic hair development
- Abdominal mass appears within 10 days.
- Abdominopelvic CT scan showed two enlarged ovaries: **left** 11cm; right 6cm

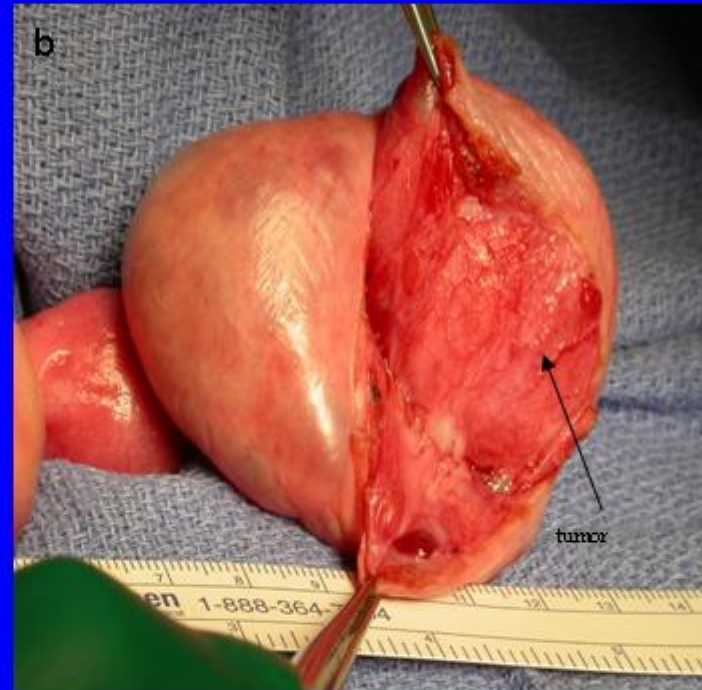
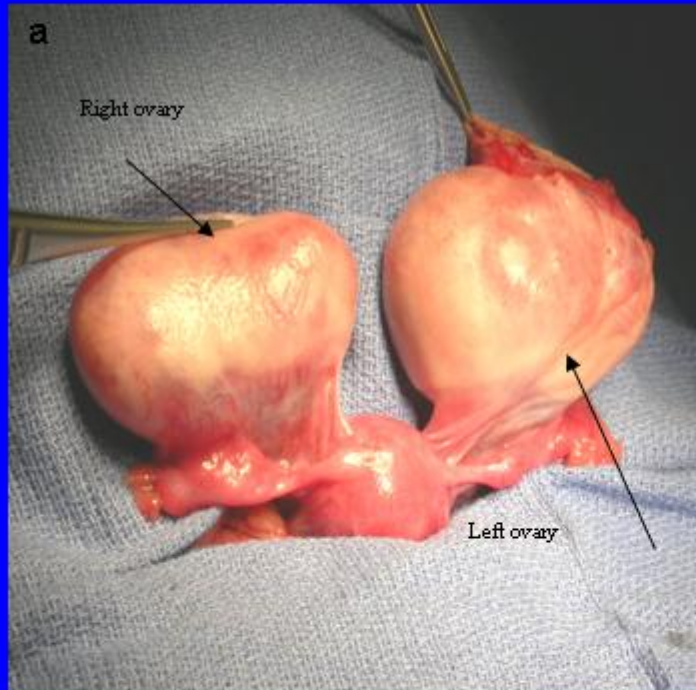
# CT-Scan



# Biology

- increased testosterone (37ng/ml) and Estrogene (760pg/ml) levels.
- The levels of AFP and beta-HCG were within normal
- Dosages of IGF1 (39ng/ml), TSH and T4 were normal for age

# Surgery



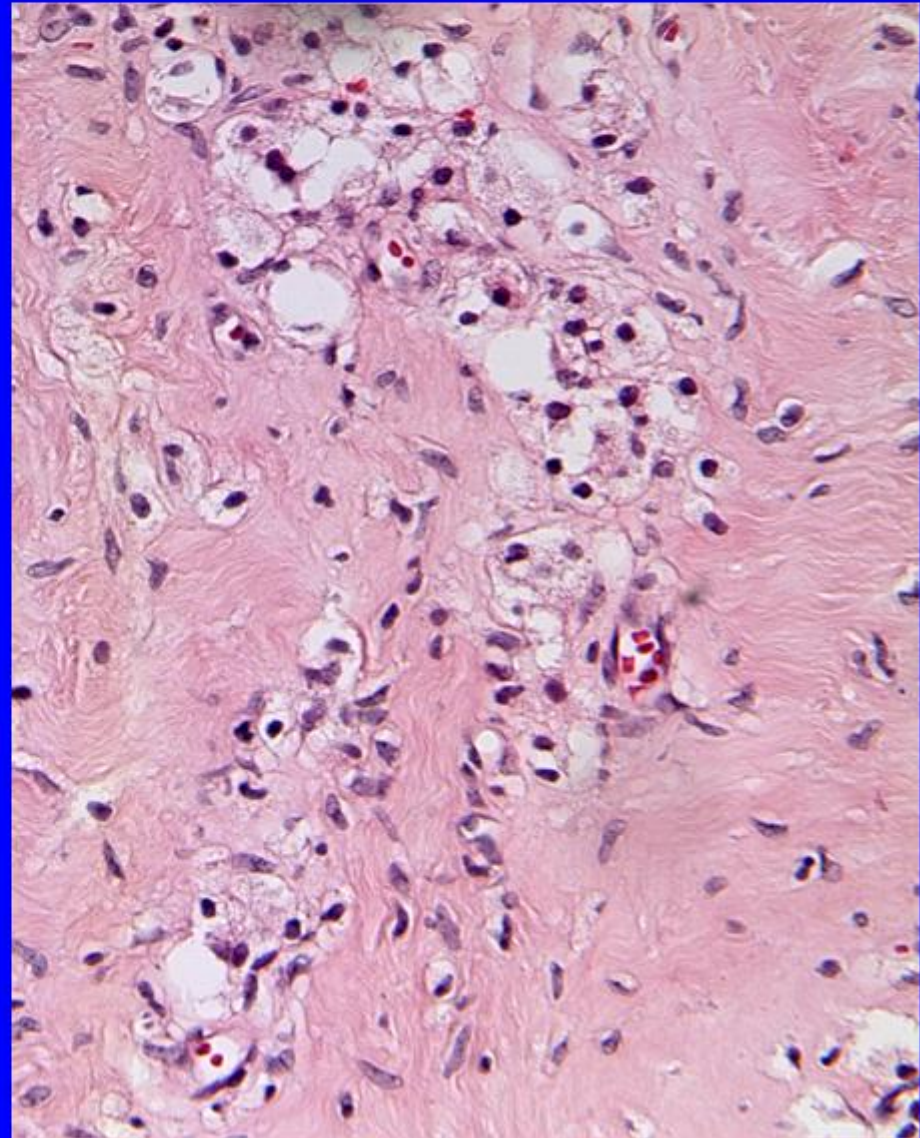
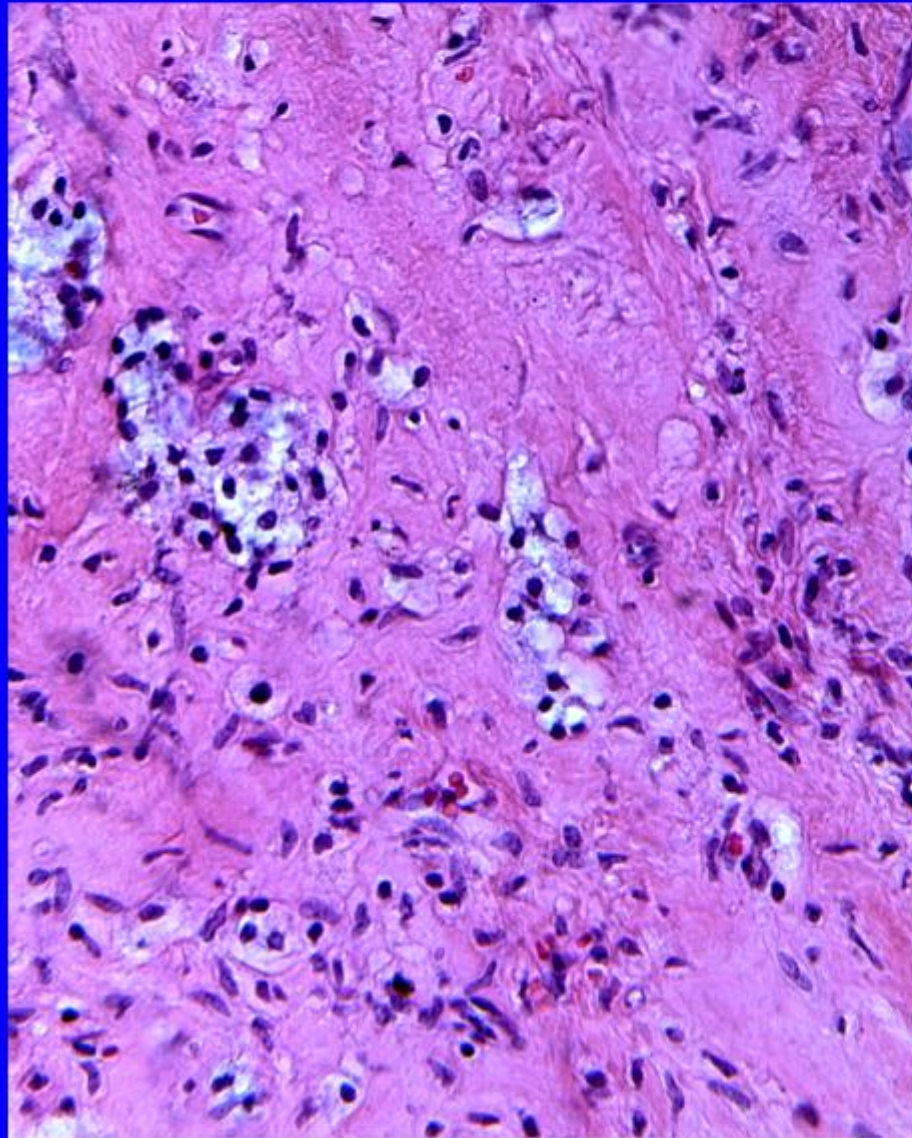
# Macroscoscopy: left ovary



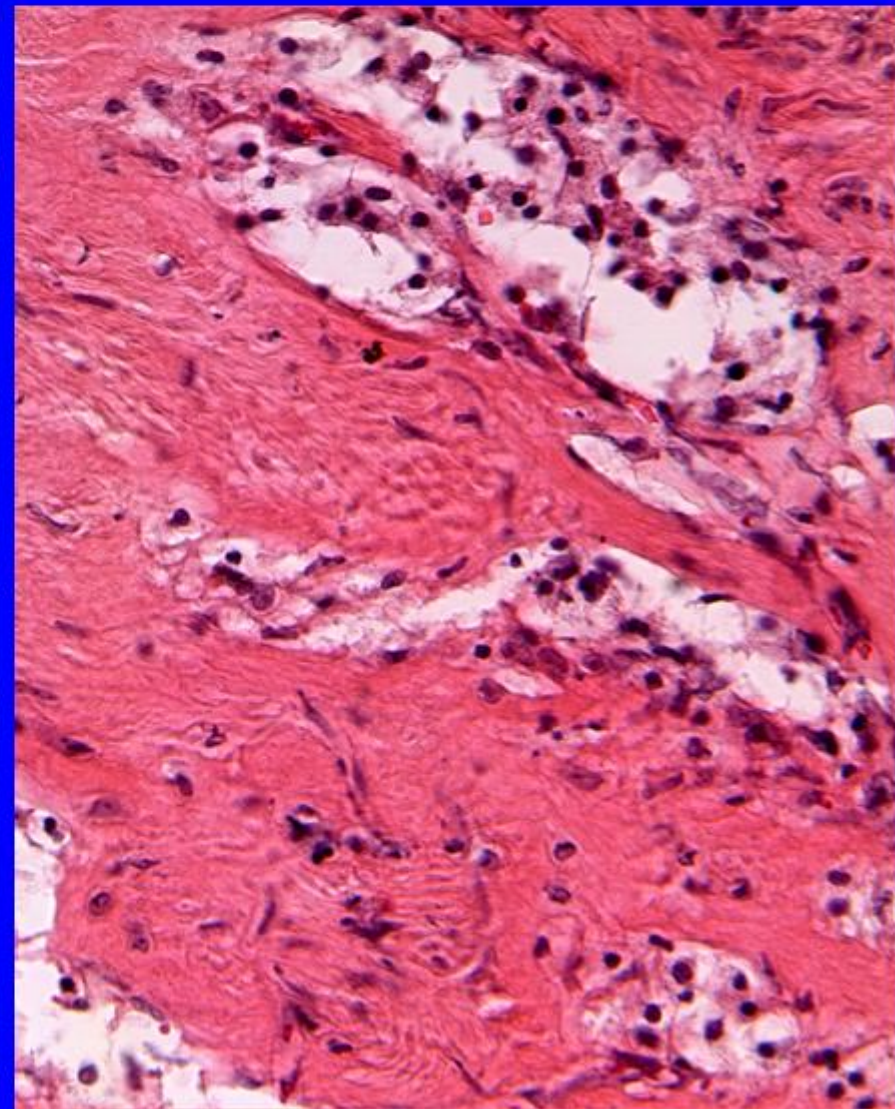
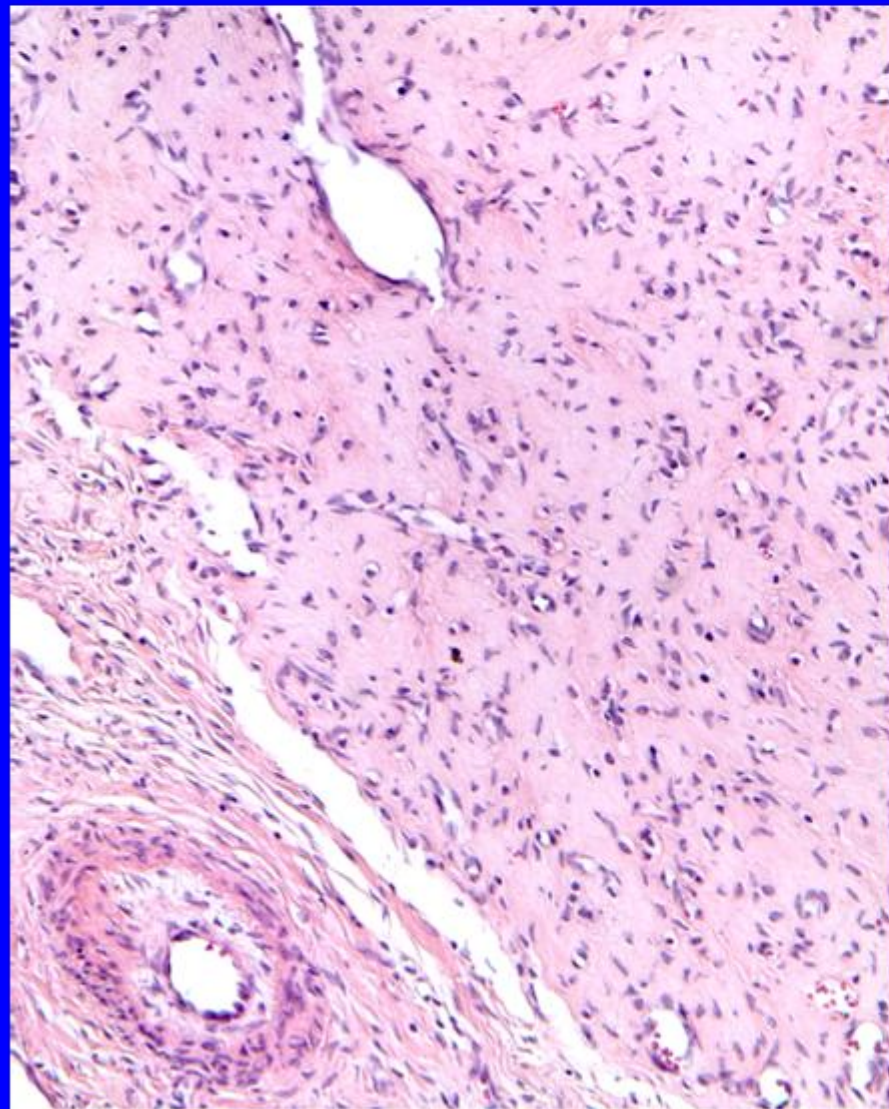
# Histopathology

Frozen

Fixed



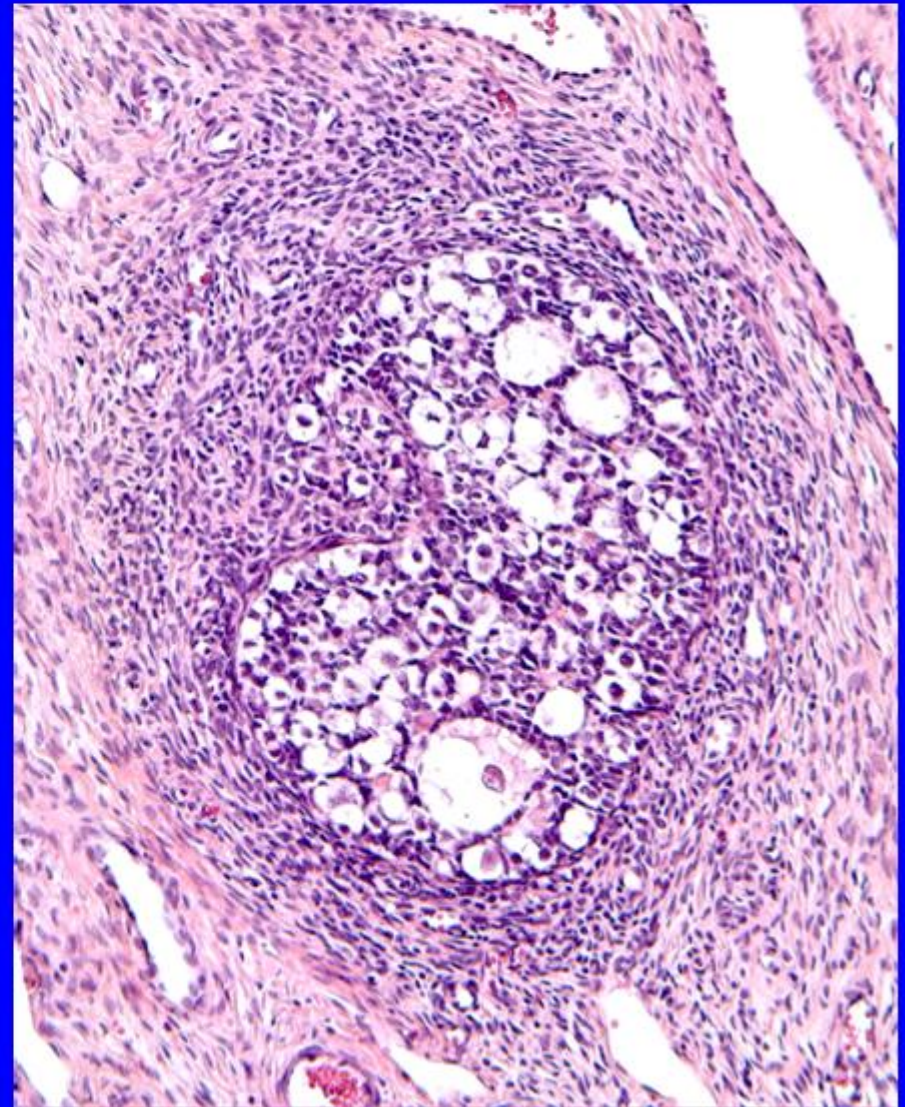
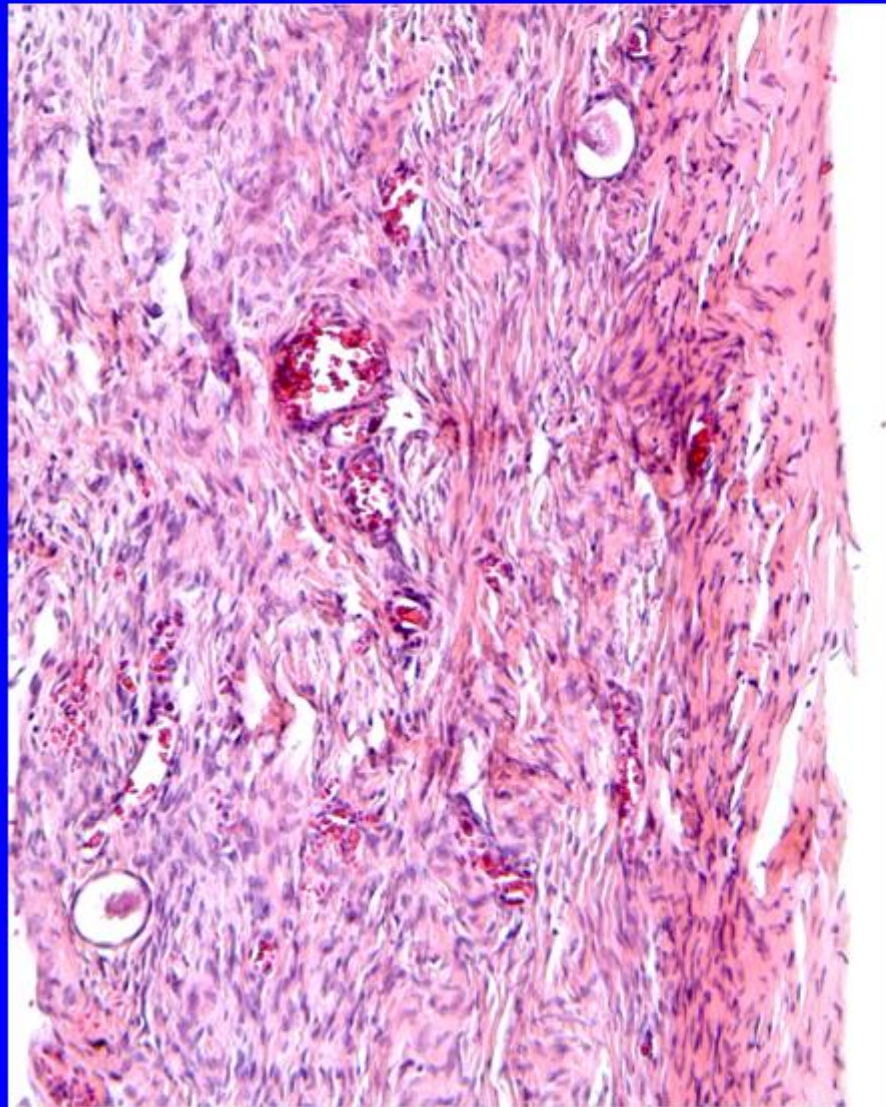
# Sex cord tumor: fibrothecoma with sertoli cells



# Ovary

Rare follicles

Abnormal follicles

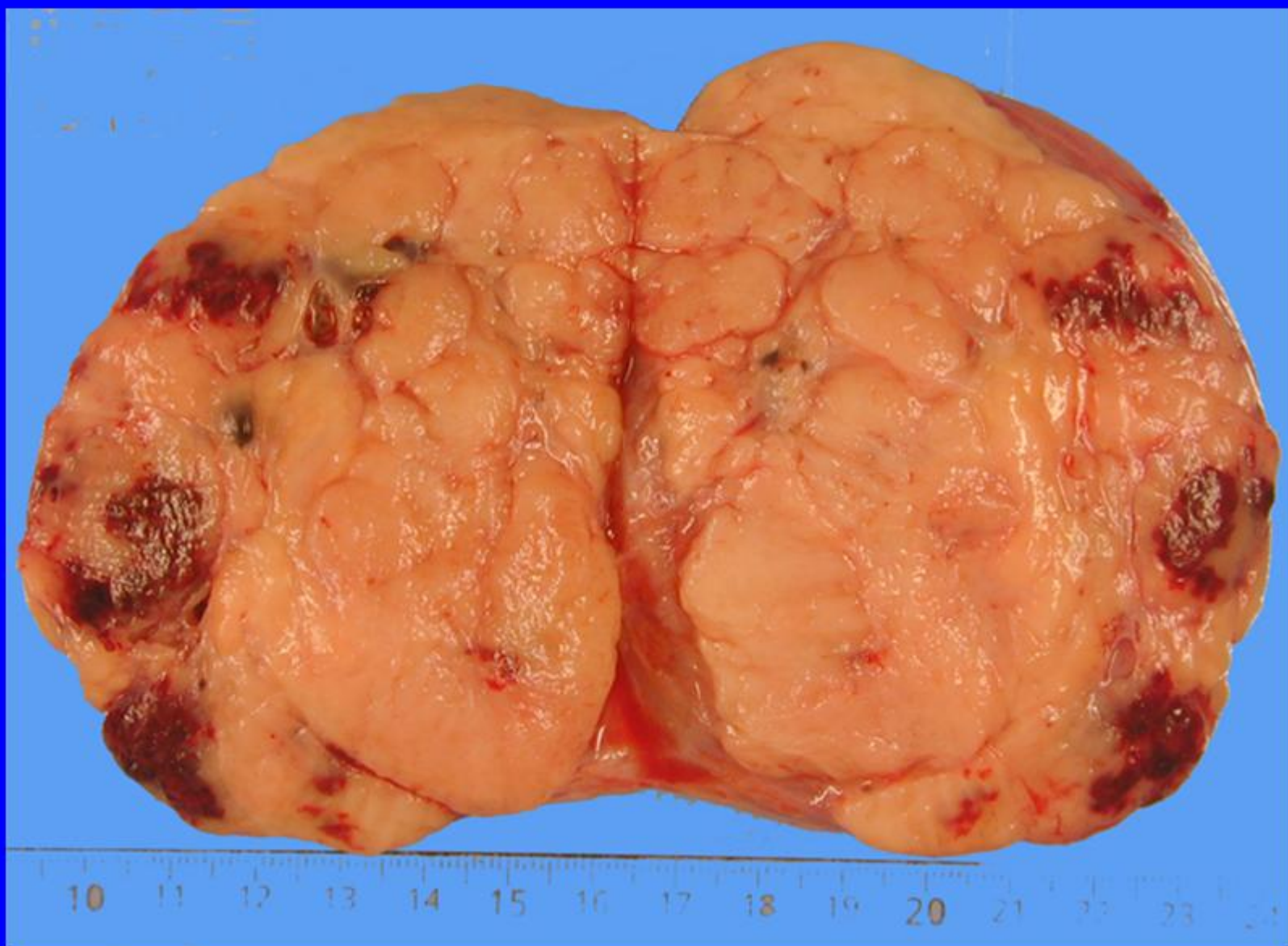


## *2d period*

### *(relapse 6 - 7.5 months )*

- 6 weeks later, age 6 months
- Breast development and relapse: uterine bleeding, associated with another abdominal mass.
- Estradiol elevated (297pg/ml) but low testosterone (1.32ng/ml).
- CT scan : **left** ovarian tumor 9cm, right ovary 5.7cm multicystic

# left salpingo-oophorectomy



## *3d period*

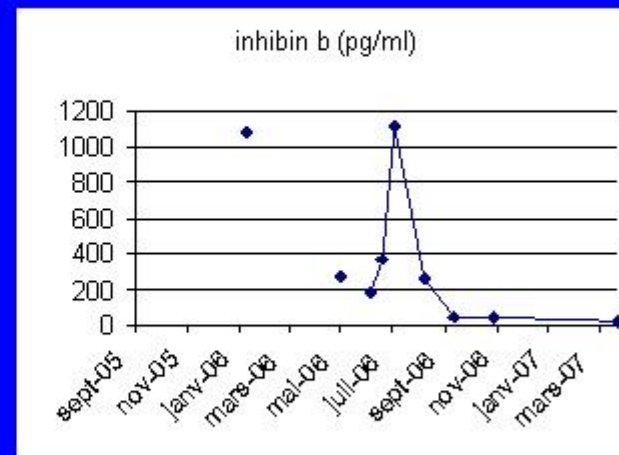
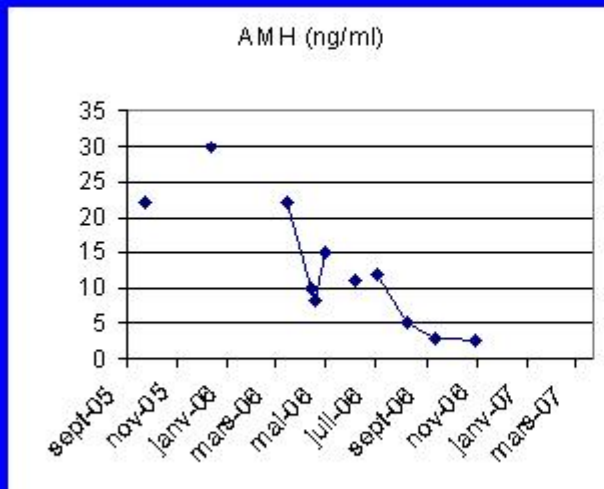
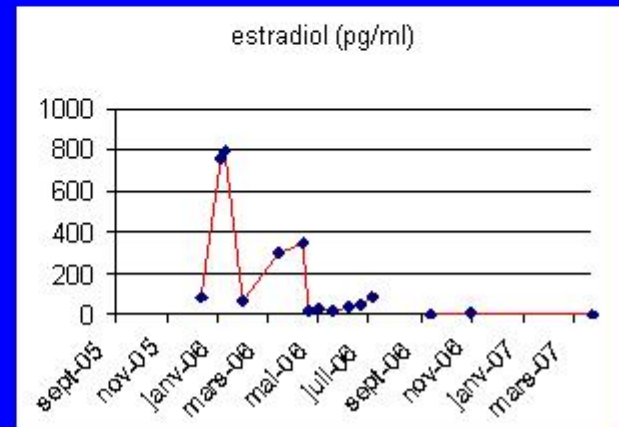
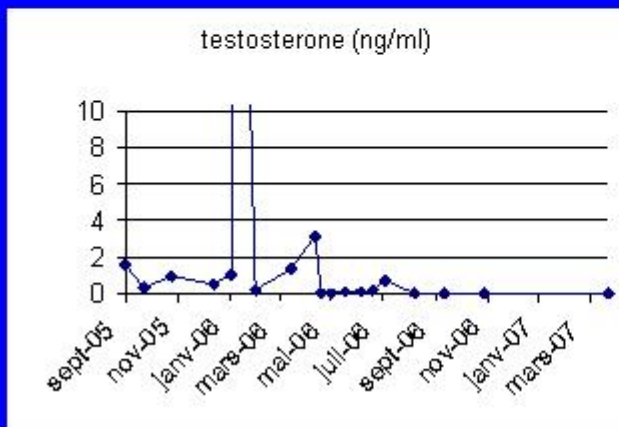
### *( 7.5 - 19 months )*

- Inhibin B, E2 and AMH levels rose until respectively 1110pg/ml, 88pg/ml and 12ng/ml at 10 months old (2.5 months after the second surgery).
- A diagnosis of a **right** granulosa cell tumor was done and a right oophorectomy was proposed to parents who refused further surgery but agreed with a clinical, biological and ultrasound scan follow-up

# Follow-up

- Within two months, a progressive normalization of hormonal levels associated with a regression of the clinical signs of precocious puberty was observed.
- With a follow-up  $> 1$  year, E2, testosterone, AMH and inhibin B are still within a normal range

# Biochemical follow-up



# Discussion 1

- Juvenile Granulosa cell tumor is the most frequent endocrine tumor of the ovary in childhood
- It originates from the granulosa cells of ovarian follicle.
- The main steroid secretion is estradiol which induces isosexual precocious puberty in prepubertal girls, or menstrual anomalies in pubertal girls

## Discussion 2

- Sclerohyalines areas may coexist with juvenile granulosa cell tumors and mimick a fibrothecoma
- They are unilateral
- Bilaterality may be induced by hormonal stimulation or TSH hypersecretion ( the beta chain is common to FSH & TSH)

# Acknowledgement

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Thank you for your attention

