# A Rare Case of Ovarian Juvenile Granulosa Cell Tumor in an Infant with Isosexual Pseudo Puberty

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## Introduction:

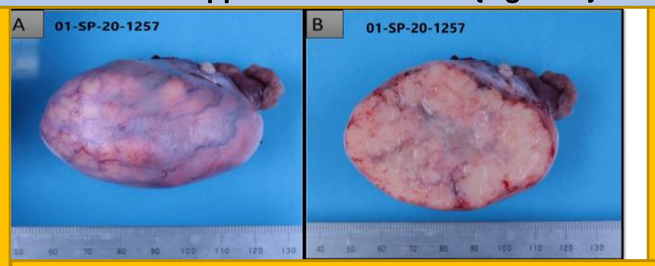
Ovarian neoplasms are infrequent in childhood, with an incidence of only 1–5% (1). They can be classified as epithelial, germ cell, or stromal. Within the stromal category, the most common tumor is the granulosa cell tumor (GCT).

The juvenile subtype accounts for 5% of these cases. In infants, less than 1-year JGCT is extremely rare, with very few reported cases in the literature .

## Case Presentation:

- A 9-month-old girl born at term presented with a day history of bloody vaginal secretions and 2 months bilateral breast development
- Her length and weight were in the normal range [75cm (1.66 SD) and 10.4 kg (1.85 SD), respectively]. She had no skin hyperpigmentation skeletal abnormalities or organomegaly .
- Breast development corresponded to Tanner's stage 2 and fine pubic hair was evident.
- No neurological abnormalities were detected
- Hormonal workup in {table 1}
- An abdominopelvic ultrasound showed well-defined round-shaped heterogeneous hypoechoic lesion measuring 5.4 x 4.0 x 5.4 cm with solid appearance {Figure 1}.
- Bone age was advanced by (+4 SD) {Figure 1}.
- MRI of the abdomen and pelvis confirmed a large solid lesion in the left-sided adnexa, measuring 5.4 x 4.12 x 5.39 cm (volume of 62.84 ml) . Chest computed tomography was normal.

### Gross appearance of Tumor {Figure 2}




## Intervention:

- An exploratory laparotomy via a midline incision was performed.
- An ovarian mass was detected and resected and left total salpingo-oophorectomy was done.
- The tumor was ovoid, encapsulated with smooth-surface measuring 6.5 x 4 x 6.5 cm and weighing 75 grams {figure 2}
- Histopathology revealed a granulosa cell tumor compatible with a juvenile type of JGCT. stage 1a, according to the International Federation of Gynecology and Obstetrics (FIGO) classification.
- No chemotherapy or radiotherapy was required.
- 1 week and 1 month after hospital discharge, hormonal testing revealed a prepubertal level of E2 and a decrease of AMH and inhibin levels (Table 2).
- Breast development regressed significantly within 6 months{Table 2}.
- After 12 months of follow-up, the patient had no recurrence.

### Table 1

| Test | Value | Normal |
|---|---|---|
| Estradiol | 1,091.0 pmol/L | 0.0-132.0 |
| FSH | <0.3 IU/L | 1.2-12.5 |
| LH | <0.3 IU/L | 0.3-2.5 |
| AMH | >1714.0 pmol/L | 0.1-38.6 |
| TSH | 2.77 mIU/L | 0.70-8.40 |
| Testo Level | 2.2 nmol/L | 0.0-2.2 |
| Beta HCG | 0.9 IU/L | 0.0-5.0 |
| Alpha Fetoprotein | 6 kIU/L | 1-33 |
| S. Inhibin A | 385 pg/ml | <4.7 |
| S. Inhibin B | 3805 pg/ml | <111 |

### Table 2

| Test | Value 1 week after the surgery | Value 1 month after the surgery | Normal Range |
|---|---|---|---|
| Estradiol | 62 pmol/L (Normal) | < 55 pmol/L (Normal) | 0.0-132.0 |
| AMH | 9 pmol/L (Normal) | 9 pmol/L (Normal) | 0.1-38.6 |
| Beta HCG | 0.8 IU/L (Normal) | < 0.6 IU/L (Normal) | 0.0-5.0 |
| Alpha Fetoprotein | 4 kIU/L (Normal) | 4 kIU/L (Normal) | 1-33 |

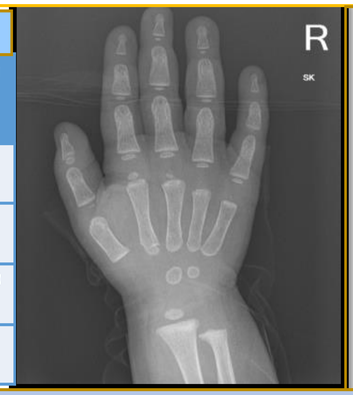



**Figure 1. Advanced bone age (+4 SD) according to Greulich and Pyle female standard.**

## Discussion:

- Our patient presented with secondary sexual characteristics that occurred as early as 7 months of age and vaginal bleeding at 9 months of age.
- Evidence of rapid growth has been reported in few cases with similar conditions. However, our patient had a length SD of 1.66 with markedly advanced bone age (+4 SD). This clearly demonstrated a marked effect of high sex steroid (E2) on skeletal maturation very early in life.
- In our patient, inhibin level was significantly elevated pre-operatively and decreased to normal after surgical resection of the tumor. Granulosa cells secrete mainly inhibin B, which makes its determination useful as a tumor marker in the diagnosis and monitoring. The AMH is also, co-secreted by granulosa cells. Its normalization occurred one week after surgery and appears to be a good biomarker for tumor monitoring
- Most cases, as in our infant, are unilateral and limited to the ovary at diagnosis, placing them in FIGO stage 1A.Recurrence is rare and related to the stage at diagnosis.
- Cases of recurrence have been reported up to 3 years after the initial surgery, so tumor surveillance is important.
- Hormone levels, most commonly serum inhibin concentration, should return to normal postoperatively and can be used to assess response to treatment and monitor for recurrence and spread

## Conclusions

JGCT is a rare ovarian neoplasm in infants. The prognosis is favourable in patients who have only ovarian involvement when treated with early surgical removal.