# MALIGNANT JUVENILE GRANULOSA CELL TUMOR OF THE OVARY

***R.Garg*, S.Agarwala, M.Srinivas, V. Bhatnagar**
***Department of Pediatric Surgery,***
***All India Institute of Medical Sciences, New Delhi-110029, India***

## BACKGROUND

**Malignant juvenile granulosa cell tumor (JGCT) is a sex cord stromal tumor of ovary and constitutes around 1-10% of all ovarian malignancies in children. Most of the patients present with precocious puberty and vaginal bleeding and few patients present with galactorrhoea.**

## AIMS

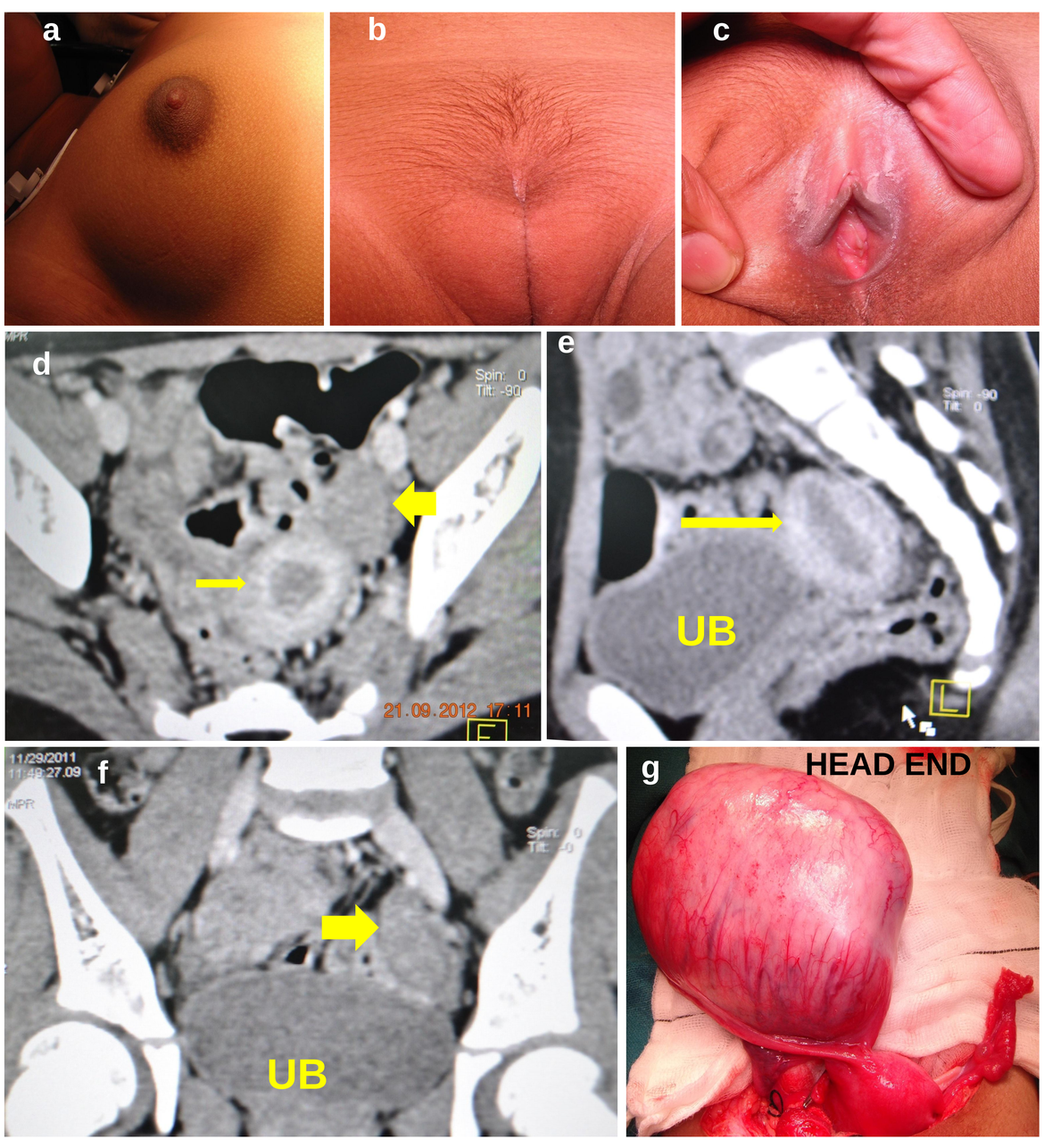
**To assess the presentation, diagnosis, treatment and the outcome of Juvenile Granulosa Cell Tumor of the Ovary in children**

## MATERIALS AND METHODS

**All children less than 12 years and registered in the Pediatric Solid Tumor Clinic of our hospital from 2005-11 who were diagnosed to have JGCT were retrospectively studied for the presentation, diagnostic investigations, management and outcome.**

## PATIENT DETAILS

- **Total No. of 6 patients**
- **Age range of 12-141 months (mean 70.8 months; median 36 months)**
- **Common presenting features:**
  - **Abdominal mass: 4(66%)**
  - **Vaginal bleeding: 4(66%)**
  - **Breast enlargement: 4 (66%).**
- **Other presenting symptoms:**
  - **Premature appearance of pubic hair: 2(33%)**
  - **Pain abdomen: 2(33%)**
  - **Abdominal distention, obstructive symptoms of bladder and bowel, bilateral inguinal swellings: 1each (16.5%)**




## TREATMENT AND OUTCOME

- **Basis of diagnosis:**
  - **Clinical features of precocious puberty**
  - **Raised serum estradiol (Range 162-710pg/ml) levels**
  - **Ovarian mass on imaging (Ultrasound/ CT scan)**
  - **Histological confirmation after resection.**
- **Treatment done:**
  - **All patients underwent salpingo-oophorectomy on the affected side**
    - ✓ **one laparoscopic**
    - ✓ **5 laparotomy and resection**
  - **All staged as POG stage 1 tumors**
  - **None received adjuvant chemotherapy**
- **Patient Outcome and follow-up**
  - **Followed up with clinical examination, serum inhibin levels and serial ultrasonography 3 monthly for 1 year then 6 monthly for 2 years**
  - **After 3 years- clinical examination and ultrasonography yearly**
  - **The symptoms of isosexual precocious puberty regressed in all, except persistence of some pubic hair**
  - **There was no recurrence during a follow up of ranging from 3-72 months.**

## DISCUSSION AND REVIEW

**Stromal cell tumors of the ovary constitute around 15% of malignant ovarian tumors in children. Both granulosa-theca cell tumors and Sertoli-Leydig cell tumors are rare but their importance lies in their propensity to cause precocious puberty. Most of these tumors present in their early stage because of early appearance of secondary sexual characteristics. Most patients respond to complete surgical excision. Platinum based chemotherapy is reserved only for advanced cases. Presenting as ovarian torsion is a rare occurrence. Inhibin B levels are required for follow up.**

**Multiple case reports have been published over the last 10 years with the age range from newborn to 12 years. Completeness of resection has been suggested as the most important prognostic criteria. Those localized to the ovary have responded well to surgery alone, while those who had extension outside ovary or tumor rupture need cisplatinum based chemotherapy.**

**LEGENDS**

- **Figure: a, b and c shows clinical findings of Gynaecomastia, Pubic hair growth and Clitoromegaly.**
- **Figure d,e and f: CECT scans showing the mass from left ovary (thick arrows) and the thickened uterine wall with hyperplastic endometrium (thin arrows). UB—Urinary Bladder**
- **Figure g shows the operative specimen (different patient) with ovarian mass from the right ovary, uterus, intact left fallopian tube and left ovary**

## CONCLUSION

**Juvenile granulosa cell tumors of the ovary presents with varied features of iso-sexual precocious puberty with or without palpable abdominal mass. After complete excision, they have very good prognosis with complete regression of features of isosexual precocious puberty and no recurrence without any chemotherapy.**