Juvenile Granulosa Cell Tumour as a Rare Cause of Isosexual Precocious Puberty in A child: Case Report and Review of the Literature
Amer O. Al Ali, Mohammed Abdu A Soeid, Ghadah M Gosadi, Mohammed A Faqehi, Nabil T. Mikhail, Nasir A.M. Al Jurayyan

1Consultant Pediatric Endocrinologist, 2Senior Pediatric Resident, 3Consultant Pediatrician, 4Professor and Chief Histopathologist, King Fahad Central Hospital, Jizan Saudi Arabia
5Professor and Senior Consultant Endocrinologist, College of Medicine and King Khalid University Hospital, King Saud University, Riyadh, Saudi Arabia

Abstract: Juvenile granulosa cell tumour is a rare cause of isosexual precocious puberty in children. Early diagnosis and surgical removal of the tumour is essential since the delay in treatment can result in malignant transformation. A 22-month-old girl presented with isosexual precocious puberty. Huge pelviabdominal mass involving the right side, which was firm, smooth surface, mobile, not tender, not pulsatile measuring 9x12 cm which was confirmed by computed-tomography (CT) of the abdomen. Laparotomy revealed a unilateral involvement, with successful removal of the tumour. Histopathology reported a juvenile granulosa cell tumour. Postoperatively, she experienced cassation of vaginal bleeding and her breast size regressed. She has a good prognosis.

Keywords: Juvenile granulosa cell tumor, precocious puberty, isosexual, ovarian tumour.

INTRODUCTION
Granulosa cell tumour (GCT) of the ovary is a rare neoplasm that originates from sex-cord stromal cells and relatively imprevalent which includes 1.5% of the whole of the ovarian neoplasm, and 6% of its malignant tumours. Only 5% of GCT cases are observed before puberty that in regard to most of their special pathologic are divided under the Juvenile granulosa cell tumour (JGC) [1-11].

In this report, we present a 22-month-old girl with juvenile granulosa cell tumour of the right ovary, who presented with isosexual precocious puberty with a brief review of the literature.

CASE SUMMARY
A 22-month-old girl who presented with history of slowly progressive breast enlargement since the age of 4 months and the appearance of pubic hair at 18 months of age. Also, she had vaginal bleeding at 20 months of age. Two weeks prior to her presentation to the emergency room (ER), she started to have abdominal pain with distension and low grade fever. There were no symptoms or signs suggestive of increased intra-cranial pressure hypo or hypermetabolic, nor head trauma or irradiation. She was not on any medication. Apart from history of premature the larche in the mother, past medical history and systemic review of systems were unremarkable. Physical examination showed nodysmorphic feature, nor hypo or hyperpigmentation. Thyroid was not palpable.

Her height was 85 cm, 50th percentile, and weight of 14 kg, 95th percentile, and head circumference of 47 cm, 25th percentile. Clinical signs of isosexual precocity, figure 1. Abdomen was distended with a huge pelvi-abdominal mass involving the right side, which was firm with smooth surface not pulsatile and not tender measuring 12 x 9.0 cm. No bony deformities or cafe-au-lait spots. Head computed tomography (CT) was normal, and abdominal CT scan, figure 2 showed a large solid mass with heterogenous pattern with septi and areas of cystic change. No evidence of vascular invasion. Both adrenal glands were normal.
A diagnosis of isosexual precocious puberty was made. Laparotomy and right ovary was resected. It was encapsulated without any capsule breach. The left ovary was normal. Uterus was normal in size and endometrial cavity was expanded and filled with fluidly component. Fine needle aspiration showed granulosa cell tumour, juvenile type which was confirmed by histological examination, figure 3. Tumor marker including beta HCG and alpha-fetoprotein were negative.
Laboratory investigation revealed low hemoglobin of 6.9, WBC 23, and platelets 354. Liver, renal function and thyroid function were normal. Serum estradiol levels were slightly increased at 18.3 pmol/L. Normal serum LS 0.4 iu/L, and FSH 0.115 iu/L. Prolactin was high at 424 miu/L (normal 21-30). Postoperative course was uneventful, her breast size regressed and menstrual bleeding stopped.

**DISCUSSION**

Ovarian tumors are uncommon during childhood and constitute rare cause of precocious puberty. Among the most common types, epithelial cell tumors (70%), germ cell tumors (20%) and sex-cord-stromal tumors (10%). Granulosa cell tumors (GCT) represent 1-2%. GCT is classified into adult and juvenile types. JGCT represent about 55% of childhood ovarian tumors, and usually present in the first two decades of life. The most frequent presentation in the prepopulated girls is precocity. Clinical manifestation may also include hyperandrogenicity, pleural effusion, ascites, or surgical abdomen, as in our patient. Secondary amenorrhea, virilization, abdominal pain, or abdominal mass may be the presenting symptoms in post-pubertal girls [1-11].

Recognition of the symptoms, signs and abnormal hormone production, and consideration of such tumors in the differential diagnosis can allow early identification and timely surgical management and, hence, an excellent outcome [11-13].

Tumor staging is done with International Federation of Gynecology and Obstetrics (FIGO) system. Tumors are usually encapsulated with solid or cystic components. Extracapsular invasion is unilateral and are FIGO stage 1, with sporadic origin [14-16].

Surgery remains the mainstay of therapy with favorable prognosis. However, adjuvant radiotherapy or chemotherapy with cisplatin based regimen may be required in tumours with advanced stage or has high mitotic rate [4, 17-22].

The prognosis is usually excellent in the early stages. Five-year survival rate are 90-95% for FIGO stage 1 tumors. Also, age less than 10 years presence of precocious puberty, and FOXL2 expression are associated with good prognosis [23-27].

**ACKNOWLEDGEMENT:**
The authors would like to thank Miss France Solomon for typing the manuscript and extend their thanks and appreciations to Miss Hadeel N. Al Jurayyan for her help and assistance in preparing this manuscript.

**REFERENCES:**


