**Scholars Journal of Medical Case Reports** **ISSN 2347-6559 (Online)**
Sch J Med Case Rep 2016; 4(8):568-571 **ISSN 2347-9507 (Print)**

(An International Publisher for Academic and Scientific Resources) DOI: 10.36347/sjmcr.2016.v04i08.002

# 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[1], Mohammed Abdu A Soeid[1], Ghadah M Gosadi[2], Mohammed A Faqehi[3], Nabil T. Mikhail[4], Nasir A.M. Al Jurayyan[5]**

[1]Consultant Pediatric Endocrinologist, [2]Senior Pediatric Resident, [3]Consultant Pediatrician, [4]Professor and Chief Histopathologist, King Fahad Central Hospital, Jizan Saudi Arabia

[5]Professor and Senior Consultant Endocrinologist, College of Medicine and King Khalid University Hospital, King Saud University, Riyadh, Saudi Arabia

***Corresponding author**
Nasir A.M. Al Jurayyan
Email: njurayyan@gmail.com

**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

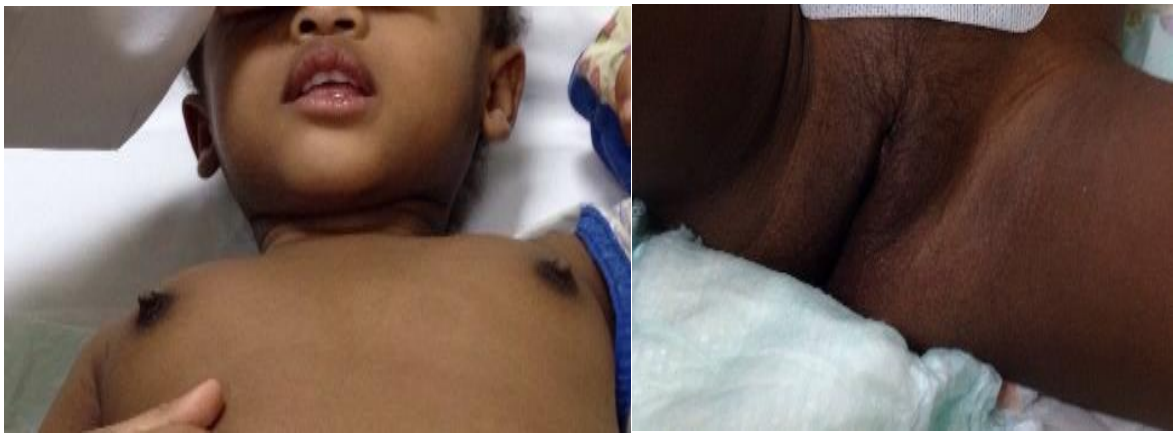
Granulosa cell tumour (GCT) of the ovary is a rare neoplasm that originates from sex-cord stromal cells and relatively imprevalent which includes 1.5% of the whole of the ovarian neoplasm, and 6% of its malignant tumours. Only 5% of GCT cases are observed before puberty that in regard to most of their 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, 50$^{th}$ percentile, and weight of 14 kg, 95$^{th}$ percentile, and head circumference of 47 cm, 25$^{th}$ 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$^{10}$ x 9.0 cm. No bony deformities or c*afe-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) (B)

**Fig 1: Photographs of the patient showing signs of isosexual precocious puberty; (A)Breast and (B) pubic hair**

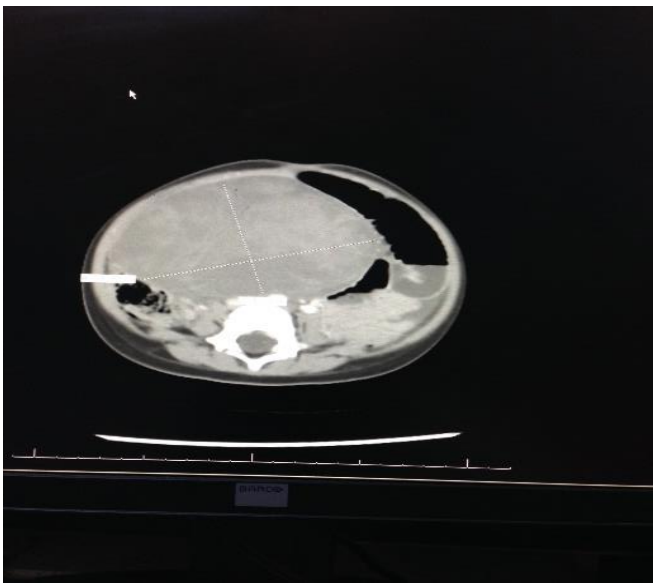
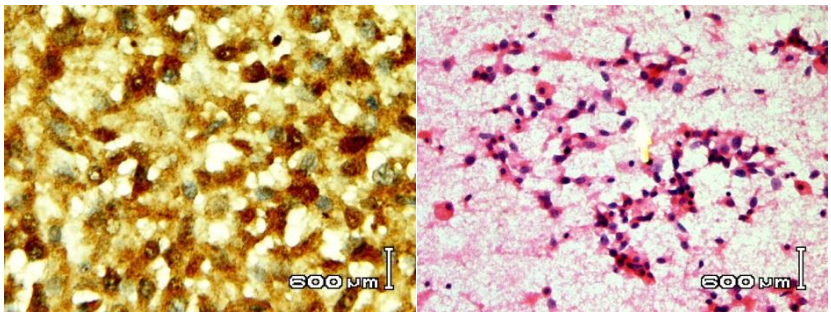


**Fig 2: Abdominal computed Tomography (CT) scan, showing a heterogeneous large solid mass with cystic changes**.

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.



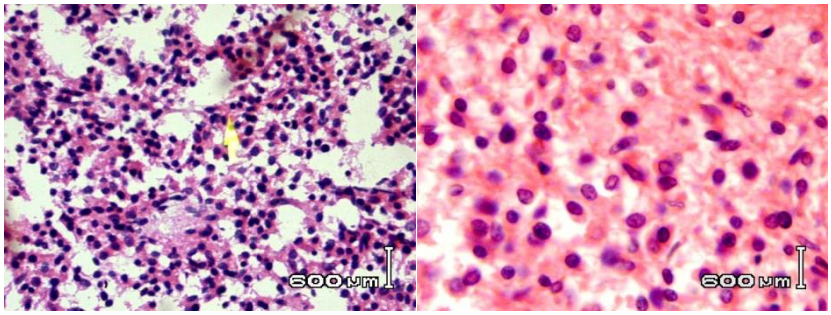



**Fig 3: Pathological protomicrograph slides showing cellular tumor with irregular, variable sized follicles (frozen section, and haematoxylin and eosin stain)**

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 serums LH 0.4 iu/L, and FSH 0.115 iu/L. Prolactin was high at 424 miu/L (normal 21-30). Postoperative course was uneventual, 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 tumours 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 are 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:

1. Biscotti CV, kennedy AW; Ovarian Juvenile Granulosa Cell Tumours. Adolesc Pediatr Gynecol 1990; 3(1): 15-19.
2. Schumer ST, CAnnistra SA; Granulosa Cell Tumor of the Ovary. J Clin Oncol 2003; 21: 1180-9.
3. Pectasides D, Pectasides E, Psyrri A; Granulosa Cell Tumor of the Ovar Cancer. Treat Rev 2008; 34: 1-12.
4. Stuart GC, Dawson LM; Update on Granulosa Cell Tumours of the Ovary. Curr OpinObstetGynec 2003; 15: 33-7.
5. Plantaz D, Flamant F, Vassal G, Chappuis JP, Baranzelli MC, Bouffet E, *et al.;* Granulosa Cell Tumours of the Ovary in Children and Adolescents. Multicenter retrospective study in 40 patients aged 7 months to 22 years. Arch Fr Pediatr. 1992; 49:793-8.
6. Lee CT, Tung YC, Tsai WY; Etiology and Clinical Features of Isosexual Precocious Puberty in Taiwanese Girls. Twenty-three years' experience in national Taiwan University Hospital. J Pediatr Endo crinol Metab. 2009; 22:947-53.
7. Nomelini RS, Micheletti AM, Adad SJ, Murta EF; Androgenic Juvenile Granulosa Cell Tumor of the Ovary with Cystic Presentation: A Case Report. Eur J Gynecol Oncol 2007:28:236-8.
8. Patel SS, Carriel KS, Carr BR; Virilization Persists in an Isomer with Androgenic Secreting Granulosa Cell Tumor. FertilSteril 2009; 91:933-935.
9. Gui H, Keefe KA, Kohler MF, Chan JK; Juvenile Granulosa Cell Tumor of the Ovary Associated with Tuberous Sclerosis. GynecolOncol 2006; 102: 118-120.
10. Brisigotti M, Fabbretti G, Pesce F, Gatti B, Cohen A, Parenti G *et al.;* Congenital Bilateral Juvenile Granulosa Cell Tumor of the Ovary in Leprec

haunism: A Case report. Pediatr Pathol 1992; 13:549-558.
11. Haroon NN, Agarwal G, Pandey R, Dabadghao P; Juvenile Granulosa Cell Tumor presenting as Isosexual Precocious Puberty: A Case Report and Review of Literature. Indian J EndocrinolMetab, 2013; 17 (1):157-159.
12. Ko SF, Wan YL, NG SH, Lee T.Y, Lin J.W, Chen W.J *et al.;* Adult Ovarian Granulosa Cell Tumors: Spectrum of Sonographic and CT Findings with Pathologic Cerrelation. AJR AM J Roentgenol 1999; 172:1227-1233.
13. Morikawa K, Hatabu H, Togashi K, Kataoka ML, Mori T, Konishi J; Granulosa. Cell Tumor of the Ovary: MR Findings. J Comput Assist Tomogr 1997; 21:1001-1004.
14. Mayr D, Kaltz-Wittmer C, Arbogast S, Amann G, Aust DE, Diebold J; Characteristic Pattern of Genetic Aberration in Ovarian Granulosa Cell Tumors. Mod Pathol 2002; 15:951-7.
15. Young RH, Dickersin GR, Scully RE; Juvenile Granulosa Cell Tumor of the Ovary: A Clinicopathological Analysis of 125 Cases. Am J SurgPathol 1984:8:575-90.
16. Sivasankaran S, Itan P, Ayensu-CAker L, Sanchez J, Egler R.A, Anderson M.L et al.; Juvenile Granulosa Cell Ovarian Tumor: A Case Report and Review of Literature. J Pediatr Adolesc Gynecol 2009; 22(5) e114-117.
17. Grabie CM, Kwailkowsin M, Valentowicz M, Grezlikowska V; Current View on treatment of the Ovarian Granulosa Cell Tumors, Ginekologia 2008; 79(1):42-46.
18. Suh-Burgmann E; Long-term Outcomes Following Conservative Surgery for Borderline Tumor of the Ovary: A Large Population-Based Study. Gynecologic Oncology 2006 ;( 0313):841-7.
19. Sekkate S, Kairouani M, Serji B, Tazi A, Mrabti H, Boutayeb S, Errihani H; Ovarian Granulosa Cell Tumours: A retrospective Study of 27 Cases and Review of Literature. World J SurgOnco 2013; 11:142.
20. Colombo N, proma G, Zanagnolo V, Insinga A; Management of Ovarian Stromal Cell Tumours. J. ClinOnco 2007; 25(20):2944-51.
21. Koukourakis GV, Kouloulias VE, Koukourakis MJ; Granulosa Cell Tumor of the Ovary: Tumor Review. Integrative Cancer Therapies 2008; 7(3):204-2015.
22. Calaminus G, Wessalowski R, Harms A, Gobel V; Juvenile Granulosa Cell Tumors of the Ovary in Children and adolescents: Results from 33 Patients registered in a prospective Cooperative Study. Gynecol Oncol 1997; 65(3):447-52.
23. Wu L, Zhang W, Li L; Prognostic Factors in Granulosa Cell Tumor of the Ovary. Zhanghua Fu Chan KeZaZhi 2000, 35 (11):673-6.
24. Chan JK, Zhang M, Kaleb V, Loizzi V, Benjamin J, Vasilev S et al.; Prognostic Factors Responsible for Survival in Sex Cord Stromal Tumors of the Ovary: A Multivariable Analysis. GynecolOncol 2007, 104:396-400.
25. Khosia D, Dimri K, Randey AK, Mabajan B, Trehan R; Ovarian Granulosa Cell Tumor: Clinical Features, Treatment, Outcome, and prognostic Factors. North Am J med 2014; 6(3):133-38.
26. Kanthan R, Senger JL, Kanthan S; The Multifaceted Granulosa Cell Tumor Myths and realities: A Review. ISRN ObstetGynecol 2012;8(8):25-8
27. Kalfa N, Philibert P, Patte E; Juvenile Granulosa Cell Tumor: Clinical and Molecular Expression. Gynecol Obstet Fertil 2009; 37:33-44.