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Jordanian Girl with Juvenile Granulose Cell Tumor Presented as Precocious Puberty: A Case Report

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Abstract

Objective: Our objective is to explain precocious puberty, type, etiology, investigation, how to differentiate between central and peripheral precocious puberty, and focus on one of the rarest causes of precocious puberty.

Case report: In our case of precocious puberty, a two-year-old female presented with bilateral breast enlargement, vaginal secretion, pubic hair, and abdominal swelling. An abdominal and pelvic CT shows a very large ovarian mass. Asalpino-oophorectomy was carried out with regression of symptoms and signs and improvement in laboratory exams. The biopsy showed Juvenile Granulosa Cell Tumors (JGCT).

Discussion: Due to an increase in the levels of estradiol and no increase in the gonadotropins (LH, FSH), advanced bone age and height age are compatible with the chronic age of 2 years old, so we think about peripheral precocious puberty. Due to abdominal swelling and patient age (small age), we suspect malignancy, so we did a CT scan and sonography of the pelvis.

Conclusion: Every patient with signs and symptoms of precocious puberty must do all the investigation that leads to diagnosis, especially when there is a red flag like in our patient (very small age).

Keywords: Precocious puberty • Juvenile granulosa cell tumors • Estradiol • Gonadotropins

Introduction

Precocious Puberty (PP) is the premature development of pubertal changes that begins before 8 years in girls and before 9 years in boys [1]. Precocious puberty may be categorized as central or peripheral Precocious Puberty [2]. Diagnostic criteria for PP include:

- Early appearance of 2dary sexual characteristics.
- An acceleration of growth velocity.
- Rapid bone maturation resulting in the early fusion of the growth plates, potentially responsible for the adult height deficit in these individuals [3].

The main etiologies of Central precocious puberty are idiopathic, organic brain tumors (Astrocytoma, optic glioma), congenital anomalies (arachnoid cyst, cerebral dysgenesis), CNS insult (infection, head trauma). While, Peripheral Precocious Puberty caused by gonadal source (ovarian or Leydig cell tumor, ovarian cyst), Adrenal source (Congenital adrenal hyperplasia, Adrenal tumors), Human chorionic gonadotropin producing tumors (Choriocarcinoma, hepatoma, teratoma), and others (Primary hypothyroidism, Iatrogenic).

The pattern of pubertal development in peripheral PP may be asynchronous, unlike in normal puberty or CPP, for example, with menarche occurring at an early stage of breast development. Autonomous ovarian cysts may present with vaginal bleeding and breast development. Non–germ cell gonadal tumors are very uncommon in pediatrics and occur predominantly in the ovary. Epithelial carcinomas (usually an adult tumor), Sertoli-Leydig cell tumors, and granulosa cell tumors may occur in children. Carcinomas account for about one-third of ovarian tumors in females younger than 20 years of age; most of these occur in older teens and are of the serous or mucinous subtype. granulosa cell tumors and Sertoli- Leydig cell tumors produce hormones that can cause virilization, feminization, or precocious puberty, depending on the pubertal stage and the balance between Sertoli cells (estrogen production) and Leydig cells (androgen production). The diagnostic evaluation usually focuses on the chief complaint of inappropriate sex steroid effect and includes hormone measurements, which reflect gonadotropin independent sex steroid production. Appropriate imaging also is performed to rule out a functioning gonadal tumour. Surgery usually is curative. No effective therapy for the non-resectable disease has been found.

Case Report

A two-years-old girl presented to the endocrine clinic at Queen Rania Children Hospital with progressive bilateral breast enlargement, vaginal secretion, and pubic hair development for 2 months. Her history was unremarkable. Her general examination and vitals were normal. Tanners staging was Breast stage 3, Pubic hair stage 3. There was no axillary hair. Cardiovascular, pulmonary, and neurological examinations were normal. A solitary well defined abdominal mass firm to hard, mobile was palpable in the left lower abdomen. Her bone age was 5 years, height age 4 years (100cm), and chronological age 2 years. Her investigations showed normal hemogram, liver, renal, and thyroid functions. Hormonal profile: luteinizing hormone (LH), <0.1mIU/ml, follicle-stimulating hormone (FSH) <0.1 mIU/

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ml, Testosterone 2.5 ng/dl, Estradiol 237.1 pg/ml, Dehydroepiandrosterone Sulfate (DHEAS) 11.9 µg/dl, 17-hydroxy progesterone 0.89 ng/ml. Tumor markers were within normal levels: beta- HCG was less than 0.1mIU/ml and alpha-fetoprotein 2.9 ng/ml.

CT abdomen and pelvis with contrast showed an 18 × 16 × 10 cm abdominopelvic mass with no visualization of left ovary suggestion of a left ovarian mass (Figure 1). The patient underwent a left salpino-oophorectomy and a resection of the big mass (Figure 2). The findings of the histology were compatible with granulosa cell tumor, juvenile type stage IA according to the International Federation of Gynecology and Obstetrics (FIGO) system (Figure 3). During her follow-up, there was a regression of the pubertal signs, the serum estradiol level had dramatically declined to 5 pg/ml, CT abdomen and pelvis with contrast showed no pathological findings after one month of surgery.

Discussion

Ovarian tumours in children are uncommon, with an incidence of about 25 in 100,000 children's hospital admissions. Granulosa cell tumors account for 2-3% of all ovarian tumours [4], two subtypes of GCT have been described based primarily on clinical behaviour and histopathological characteristics, the juvenile, and the adult form, but the juvenile variety is the most common before 20 years of age [5]. They must be considered in any girl with lower abdominal pain, an abdominal mass or precocious puberty. In our case, she presented with progressive bilateral breast enlargement, vaginal secretion, pubic hair, and abdominal distension.

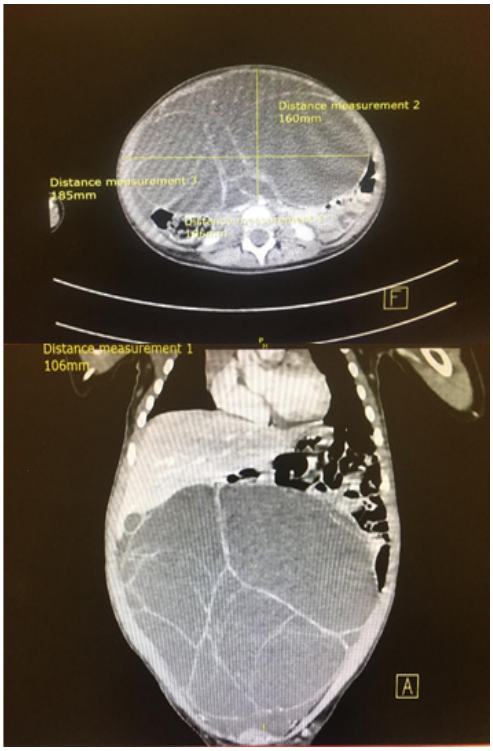

Figure 1. CT abdomen and pelvis CT with contrast abdominopelvic mass.

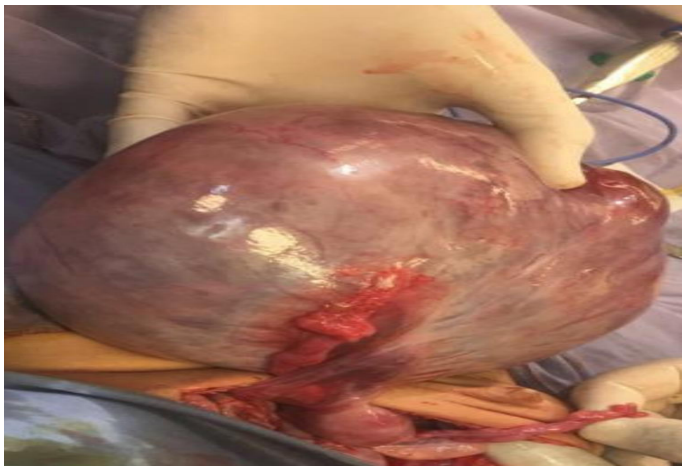

Figure 2. A resection of the big mass.

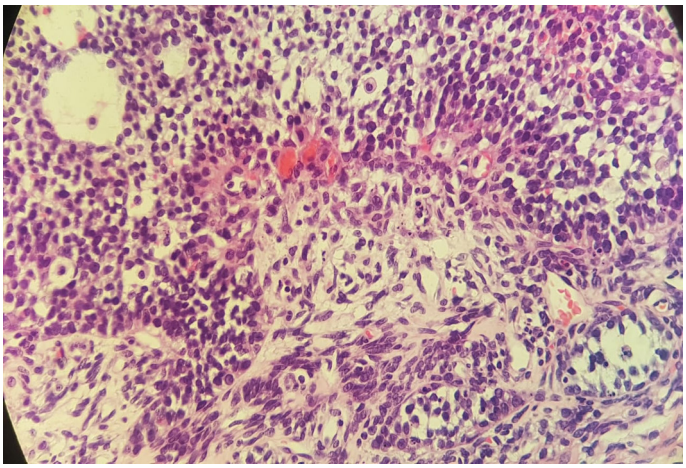

Figure 3. Juvenile type stage IA according to the International Federation of Gynecology and Obstetrics (FIGO) system.

Regarding the laboratory tests, we observed an increase in the levels of estradiol and there is no increase of the gonadotropins (LH, FSH). Due to tumor-derived estradiol, she had advanced bone age and height age compatible with the chronic age of 2 years old in our case. The image test must include an abdominal and pelvic US, and Magnetic Resonance Imaging (MRI) or CT. Even though it can show up as a solid mass or as a cystic mass. They should exclude primary gonadal disorders, adrenal gland, and exogenous secretion of gonadotropin by a tumor, or an autonomous ovarian cyst [6]. In our case, CT scan and sonography of the pelvis showed large multiseptate mass seen extending from the epigastric area to the pelvis displacing the small bowel posteriorly. Surgical resection with a unilateral salpingo-oophorectomy is typically the only required therapy and carries a good prognosis; particularly in the case of juvenile granulosa cell tumors stage IA [7]. Pubertal regression should ensue.

Conclusion

When the signs and symptoms of precocious puberty appear in children at the age of fewer than six years in both sexes, it is considered one of the red flags, and the doctor must do the essential tests to determine the main cause that led to precocious puberty. We must differentiate between central and peripheral precocious puberty. Treatment depends on the cause. In our case, the cause was juvenile granulose cell tumors, which consider one of the rarest causes, and the treatment was surgical removal of the tumor and follow-up in the endocrine clinic.

References

1. Luigi Garibaldi and Wassim Chemaitilly. "Disorders of Pubertal Development." *Nelson Text Pediatr* 2 (2020): 2656-2862.
2. Sasha R. Howard, Nicolas Roux, Juliane Leger and Jean-Claude Carel, et al. "Puberty and Its Disorders." *Brook's Clin Pediatr Endocrinol* 7(2020): 235-288.
3. Nakamoto, Jones. "Pediatric Practice Endocrinology." *Endocrinol* 2 (2010): 256-298.
4. Alhabib, Majed. "A Four-Year-Old Girl with Ovarian Tumor Presented with Precocious Pseudo Puberty." *J Diabetes Metab Disord Control Case* 3 (2016): 96-97.

5. Mahin Hashemipour, Mohammad Hassan Moaddab, Masoud Nazem and Parvin Mahzouni, et al. "Granulosa Cell Tumor in a Six-Year-Old Girl Presented as Precocious Puberty." *J Res Med Sci* 15 (2010): 240-242.

6. Mahtab Ordooei, Mojgan Karimi-Zarchi, Golnaz Malekzadeh and Mansour Moghimi, et al. "Investigation of the Cause of Precocious Puberty in an 8-Year-Old Girl Ended up in Juvenile Granulosa and Theca Cell Tumor of the Ovary." *J Cancer Ther* 2013 (2013): 1391-1394.

7. Hamillour Boussouf and Boussouf Nggada. "Ovarian Juvenile Granulosa Cell Tumour in Childhood: About Two Cases." *EC Paediatr* 5 (2018): 459-464.

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