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Case Report

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A four-year-old girl with ovarian tumor presented with precocious pseudo puberty

Abstract

Precocious puberty in girls is generally defined as appearance of secondary sexual characteristics before eight years of age. Precocious puberty is divided into central precocious puberty and precocious pseudo puberty (peripheral).[1] Central precocious puberty (gonadotropin-dependent), which involves the premature activation of hypothalamic-pituitary-gonadal axis. Precocious pseudo puberty (gonadotropin-independent) is caused by activity of sex steroid hormones independently from the activation of pituitary-gonadotropin axis.1 The commonest cause for precocious pseudo puberty is functional ovarian cyst. Ovarian masses are generally considered rare in the premenarchal age group.[2] Ovarian tumors of premenarchal girls generally originate from the germ cell line.[2] The most common presentation of these tumors in children is precocious pseudo puberty.[3] In This report we describe a 4year-old girl with isosexual precocity as premature thelarche and vaginal bleeding. CT of pelvis showed right ovarian mass. The pathology report of the excised mass was consistent with Juvenile Granulosa Cell Tumors (JGCT).

Keywords: ovarian tumor, granulose cell tumor, precocious puberty, children, gonadotropin-dependent, premature thelarche, hypothalamic-pituitary-gonadal, sex steroid hormones

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Case report

A 4year old healthy girl presented to the pediatric emergency department (PED) with a 5months history of breast enlargement and pubic hair. One day prior to her presentation to PED, she developed vaginal bleeding with mild lower abdominal pain. There was no history of meningitis, cranial irradiation, head trauma, seizures, medication intake, headache, visual problem or behavior al change. There was no evidence of local trauma or sexual abuse. Family history was negative for precocious puberty, endocrine, autoimmune and malignancy disorders.

On physical examination she looked well, conscious, alert and oriented. Vital signs included a temperature 37°C, respiratory rate 28breaths/minute, and heart rate 110, not dysmorphic. Her weight was: 16.1kg (75th) percentile and her was height 104cm (75th), Breast tanner stage IV, pubic hair tanner III. No palpable mass in abdominal exam, local genital exam within normal. There were no café-au-lait spots, or bony deformity. The remainder of her physical examination was unremarkable. Laboratory findings were consistent with pseudo precocious puberty: LH and FSH pre and post GnRH stimulation test: less than 0.1IU/l. Estradiol 264pmol/l (high). Testosterone (total): less than 0.04nmol/l.

Other laboratory investigations: Serum sodium 139mmol/l, potassium 4.2mmol/l. thyroid function test, TSH: 6.9mIU/l, free T4:18.9pmol/l. 17-hydroxy progesterone 0.5nmol/l. cortisol 151nmol/l, ACTH 10.8pmol/l. prolactin 341mIU/l.

Tumor markers were within normal levels: beta HCG was less than 0.1IU/l, LDH: 209U/l and alfafeto protein 1.7ug/l. Bone age was advanced: At a chronological age of 4years, it was 7years. Ultrasonography of pelvis showed: predominantly solid mass lesion in the right adnexal region most likely arising from the right ovary measuring 5.6 x 3.8 x 6.0cm volume approximately 66ml. Uterus enlarged for the age of the patient with endometrial thickening. CT abdomen and pelvis with contrast showed: smoothly marginated solid soft tissue mass involving the right ovary, shows contrast enhancement and tumor vessels. Mass is displacing the surrounding bowel loops, abutting the urinary bladder, anterior abdominal wall and the psoas muscle. There was no evidence of free fluid or lymphadenopathy in abdomen or pelvis. A diagnosis of precocious pseudopuberty due to estrogen-producing ovarian tumor was reached. She underwent Laparotomy, a right oophorectomy was done and the mass was sent for histopathological examination as shown in Figures 1–3. The pathology reports of the excised mass were consistent with Juvenile Granulosa Cell Tumors (JGCT).

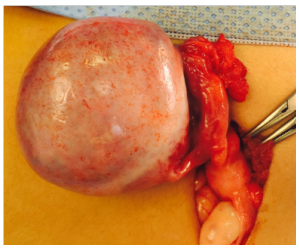

Figure 1 Laparotomy revealed right ovarian tumor.

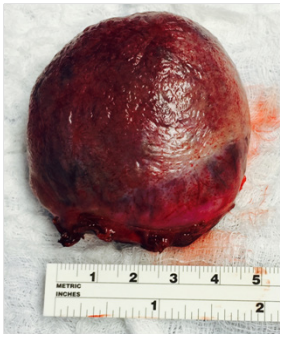

Figure 2 The size of the ovarian tumor is 4.5 x 6cm.

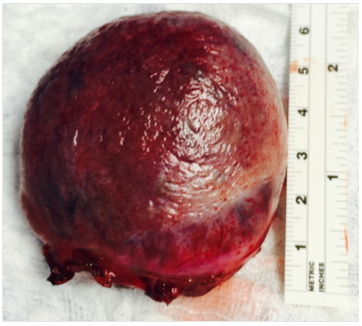

Figure 3 The size of the ovarian tumor is 4.5 x 6cm.

Discussion

Ovarian tumors are uncommon during childhood and are rare causes of precocity.[4] Ovarian sex cord-stromal tumors are neoplasms containing granulosa cells, sertoli cells, theca cells, leydig cells, and fibroblasts of gonadal stromal origin. Granulosa cell tumors (GCT) represent about 2% of all ovarian tumors and fall under ovarian sex cord–stromal tumors. There are two types: adult granulosa cell tumor (95% of cases) and juvenile granulosa cell tumor (5% of cases).[5,6] Isosexual precocity occurs in 70-80percent of prepubertal girls with GCT.[7] However, only 1% of all cases of sexual precocity in prepubertal girls are due to granulosa and theca cell tumors. This hormonally active ovarian neoplasm is an estrogen-producing tumor which may lead to its early detection. The presence of undetectable basal serum FSH and LH levels had a specificity of 95% in girls with gonadotropin- independent precocious pseudo puberty.[5] In this condition, There will be high level of sex hormones and suppressed gonadotropins as in our case. Tumor markers, whether positive or negative are not conclusive in all cases but, useful for postoperative surveillance. Bone age usually advanced due to tumor-derived estradiol as in our case. Ultrasonography is the investigation of choice.[8] On CT and Ultrasonography, they most typically appear as large, multilocular masses, with either thin or thick septations, as well as solid components, predominantly solid or large multisystem mass.[5] Ovarian tumors are usually capsulated and extra capsular invasion is rare.[5] In our case, the sonography and CT scan of pelvis showed a solid ovarian mass. Unilateral oophorectomy is the first-choice of therapy. Early diagnosis of JGCT is important, as adjuvant treatment may not be necessary if the tumor is localized to ovaries.[7,5] Tumor staging is by International Federation of Gynecology and Obstetrics (FIGO) system. Most tumors at FIGO stage IA have a favorable prognosis, whereas those at higher stages have a less favorable outcome.[5] Five-year survival rates are 90-95% for FIGO stage I tumors and 25-50% for advanced stages.[5]

Because most of these lesions are benign, ovarian-preserving operations should be performed whenever feasible.[8] Ovarian masses, although rare in children, must be included in the differential diagnosis of all girls who present with abdominal pain, swelling or precocious puberty.

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None.

Conflict of interest

Author declares that there is no conflict of interest.

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