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). 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. The commonest cause for precocious pseudo puberty is functional ovarian cyst. Ovarian masses are generally considered rare in the premenarchal age group. Ovarian tumors of premenarchal girls generally originate from the germ cell line. The most common presentation of these tumors in children is precocious pseudo puberty. In this report we describe a 4-year-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

Case report

A 4-year-old healthy girl presented to the pediatric emergency department (PED) with a 5-months 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 behavioral change. 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 28 breaths/minute, and heart rate 110, not dysmorphic. Her weight was: 16.1 kg (75th) percentile and her height 104 cm (75th). Breast Tanner stage IV, pubic hair Tanner III. No palpable mass 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 report of the excised mass was consistent with Juvenile Granulosa Cell Tumors (JGCT).

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

Ovarian tumors are uncommon during childhood and are rare causes of precocity. 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). Isosexual precocity occurs in 70-80 percent of prepubertal girls with GCT. 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. 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. 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. Ovarian tumors are usually capsule and extra capsular invasion is rare. 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. 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. Five-year survival rates are 90-95% for FIGO stage I tumors and 25-50% for advanced stages.

Because most of these lesions are benign, ovarian-preserving operations should be performed whenever feasible. 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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Conflict of interest

Author declares that there is no conflict of interest.

References