

Laparoscopy decision for precocious pseudopuberty of ovarian origin

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Summary

Development of autonomous functional ovarian follicular cysts results in precocious pseudopuberty. Conservative approach is the first choice for these patients, and the authors present two cases of precocious pseudopuberty due to autonomous ovarian cysts to discuss the decision of surgery.

Key words: Autonomous functional ovarian follicular cysts; Precocious pseudopuberty.

Introduction

Girls before adolescence may have follicular cysts as a normal response of the ovary to GnRH secretion. It is usually self-limited and non-functional in most cases. Ovarian cysts are prevalent in 2% to 5% of prepubertal girls, and of those, 5%, cysts may enlarge and continue to produce estrogen, resulting in precocious pseudopuberty of one in 400 (0.25%)[1]. Conservative approach is the first choice for these patients, and the authors present two cases of precocious pseudopuberty due to autonomous ovarian cysts to discuss the decision of surgery.

Case Report

Case 1

A 5.0-year-old girl, presents with breast development (Tanner stage II) with areola pigmentation and increased vaginal discharge. The estradiol level was 127.9 pg/ml and LH < 0.11 U/L. The GnRH stimulation test showed suppressed LH and FSH response. Abdominal and pelvic ultrasonography revealed a 3.2×2.0×2.7-cm hypoechoic cystic of the left ovary. Bone age was advanced by 6.1 years. One month later, the girl experienced remission of the autonomous ovarian cyst. No signs of McCune-Albright syndrome (MAS)-related café-au-lait skin pigmentation were identified on physical examination, and radionuclide bone scan did not reveal any abnormalities. Up to now, aged 6.3 years, the patient has experienced no further relapse of the ovarian cyst.

Case 2

A 2.3-year-old girl, presents with signs of vaginal bleeding and breast development (Tanner stage III) with areola pigmentation and axillary and pubic hair (Tanner stage II). No skin lesions characteristic of MAS showed, and radionuclide bone scan did not reveal any abnormalities. The estradiol level was 30.5 pg/ml, LH <

0.11 U/L. Serum gonadotropin responses to GnRH stimulation were prepubertal. The initial abdomen and pelvic ultrasonography revealed a 1.7-cm hyperechoic in 2.1 cm hypoechoic cystic of the right ovary, with presence of a definite endometrial echo. Bone age was advanced by 3.3 years. She continued to experience repeated episodes of vaginal bleeding for two months under conservative observation. Repeated pelvic ultrasound revealed a 4.5×4.0×3.3-cm echo-free right ovarian cyst with a 3.0×2.8×2.4-cm inhomogeneous echo in the cyst cavity, and the uterus had a prominent endometrium. CT showed the presence of a 4.2-cm right ovarian cyst with a solid component and no other abdominal or pelvic abnormalities were noted. Repeated hormonal analysis revealed an elevated level of estradiol 310.4 pg/ml and CA19-9 65.1 U/ml. To exclude malignant tumor, laparoscopic cystectomy was performed (Figure 1), pathological test confirmed the diagnosis of ovarian follicular cyst. Up to now with 15 months' follow-up, there is no relapse and she still undergoes out-patient follow-ups.

Discussion

Precocious pseudopuberty is rare and gonadotropin-independent. Development of autonomous functional ovarian follicular cysts, germ cell tumor, or MAS result in

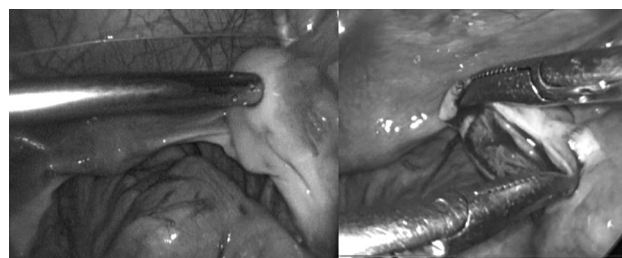


Figure 1. — Laparoscopic view of autonomous ovarian cyst.

precocious pseudopuberty of ovarian origin, because of peripheral estrogen production.

Autonomous functional ovarian follicular cysts can develop at any age and might regress after a few weeks or months [2, 3]. Precocious pseudopuberty caused by autonomous ovarian cysts may have elevated estrogen levels, but not always (due to regression of the cyst), and both basal and GnRH-stimulated gonadotropin concentrations are low. Most autonomous ovarian cysts can be managed conservatively as spontaneous remission generally occurred [3]. However if malignancy cannot be excluded, or when there might be ovarian torsion or hemorrhage, surgery should be proceeded.

Papanikolaou *et al.* [4] have reviewed articles published in English, from 1980 to 2015 about ovarian autonomous cyst related pseudopuberty, which identified 11 of 26 (42%) cases presenting with recurrences, ranging from one to four episodes. Recurrences were more likely to occur in those managed conservatively, and occurred in nine of the 13 girls who did not undergo surgery. This may due to relapses of autonomous ovarian cysts will result in prolonged or repeated estrogen exposure which can precipitate early maturation of the hypothalamic-pituitary-gonadal axis, resulting in progression from precocious pseudopuberty to central precocious puberty, and may require GnRH analogue therapy.

Differential diagnosis should exclude MAS and germ cell tumor. As there might be further episodes of autonomous cyst formation, ultrasound monitoring at regular intervals

and long-term follow up should be performed in these patients. Papanikolaou *et al.* [4] also reported time at recurrence was mentioned in 19 episodes and ranged from one to 24 months, in which three patients were eventually given GnRH agonist because of progression to central precocious puberty, one patient was identified as MAS. Recurrence might be an early presentation of MAS, and diagnosis of MAS requires a prolonged clinical follow up with repeated skeletal radiographs [5].

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