Abdominal computed tomography showed a solitary ovarian cyst measuring 85×62 mm with clear boundaries and without solid component (1A). Giant ovarian cyst disappeared naturally after adjustment of glucocorticoid therapy (1B).

The patient had demonstrated clitoral enlargement at 2 years of age and was brought to a local hospital by her parents. Thereafter, she was treated with hydrocortisone under a diagnosis of congenital adrenal hyperplasia due to 21-hydroxylase deficiency (21-OHD).

She had menarche at 12 years old and menstruation had been regular thereafter. However, she developed oligomenorrhea at 22 years old. She consulted our hospital in 2006, at the age of 28, when she married. Her ACTH level was 422 pg/mL, 17-hydroxyprogesterone (17-OHP) 304 ng/mL, testosterone (T) 1.27 ng/mL in the morning after fasting overnight, and was taking hydrocortisone 40 mg/day. An ovarian cyst measuring 85×62 mm was observed on abdominal computed tomography in July 2006. At that time, 17-OHP was 2.5 ng/mL, T 0.09 ng/mL in the early morning after an overnight fast, while taking hydrocortisone 20 mg in the morning and 10 mg in the evening, and prednisolone 2 mg in the evening. We were planning the excision of the ovarian cyst, but it had disappeared on ultrasonography by November 2006. Thereafter, she became pregnant naturally and gave birth to a female infant by caesarean section in February 2008.

It has been reported that the pregnancy rate in a female with 21-OHD is low because of their anatomic abnormalities of genitalia or gonad, and abnormal gonadotropin dynamics with an excess of 17-OHP and androgens may occur. Ovulatory rate in females with 21-OHD relate to 17-OHP and T, and appropriate steroid management is necessary for patients who hope to become pregnant (1). At physiological doses, hydrocortisone prevents adrenal insufficiency but does not suppress ACTH or androgen production. Thus, it is sometimes quite difficult to reduce the excess androgen without giving excess glucocorticoid (2).
case was treated with prednisolone in order to suppress the ACTH level in the morning. As a result, menstruation became regular and the giant ovarian cyst disappeared naturally.

Giant ovarian cyst may occur in association with menstrual irregularity, and oral combined contraceptives usually cause an ovarian cyst to regress (3). Plasma progesterone shows a high level during the follicular phase of the menstrual cycle in women with 21-OHD, and these patients seem to accept a biological ‘mini pill’ (4) and giant functional ovarian cyst would be unlikely to occur in women with 21-OHD. The giant ovarian cyst in the present case showed spontaneous regression after combined treatment with prednisolone. Hyperandrogenism may have contributed to the development of this giant ovarian cyst.

References


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